Challenging and Rare Cases in Urology
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Forewords

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Atypical phenotype of human species is also reflected in the presentation of diseases afflicting human beings. Two similar diseases may not have the same presentation, and the same management protocol may not have the same outcome in two similar patients. Despite tremendous growth of evidence-based medicine (EBM), the practice of medicine is still an art and could never become mathematics. For cracking every complex diagnostic code, the physician adds his/her own style of practice and the synthetic sixth sense to the knowledge base and the available investigative information. Rich synthetic sense inbuilt by skills helps to rightly interpret and carry forward the learning from one situation to other similar situations. Still every clinician in his/her lifetime encounters a number of situations when he/she has no experience to fall back on and in such situations, search of published literature and interaction with the colleagues at the conferences comes handy to learn from the experience of colleagues across the world.

Experience of a surgeon is a long span, crescendo bridge between formal medical education and the practice of surgery making training of surgeon a hard task master. The need for a lifetime learning commitment makes the journey challenging and exciting. The entire medicine is a continuum of What, Why and How?

Dr Gupta’s book is about “How?”—How he has managed complex urological problems. It is an account of his long and illustrious career enriched with vast experience in the practice and education of urology gained at two premier institutions of India. This complete library of cases speaks volumes about his dedication to the specialty. He needs to be complimented for meticulous preservation of the case material and apply his photographic skills for the long-term preparation of the manuscript.

I strongly believe this material would turn into a great reference book and necessitate its possession on the shelves of the postgraduates and the practicing urologists alike. It will offer an opportunity to learn straight from the real-life experience of an intellectual giant. I hope Dr Gupta will extend this work in yet another project of establishing an online library including the experts at the national and international levels.

It is my privilege to present this book for the larger benefit of our patients.

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Dr NP Gupta deserves to be complimented for the vast wealth of academic material, which he has carefully collected over a period of four decades. Such an accomplishment has been possible because of Dr Gupta's long stint at two major tertiary care hospitals of the country. This book covers almost all the topics of urology. The language is simple and lucid and has relevant bibliography. Some of the cases have already been published in reputed and indexed journals and form the part of the existing literature.

While browsing through the book, I was most impressed with the quality of the images of various anomalies and diseases, which one may not see in a lifetime. I was reminded of residency days in urology, when our mentors used to recommend us the textbook, UROLOGICAL-RADIOLOGY by Emmett and Witten. Such images leave a lasting impression on young minds and help them to arrive at accurate diagnosis.

The chapters are unique in the sense, that we rarely see cases in the early stages of various diseases and what we see is mostly the disease of sequela, which have been deftly managed.

The management of difficult situations has been done with the evolving technologies and minimally invasive operative procedures, such as Endourology, Laparoscopic and Robotic Accesses. This book also reflects how the specialty of urology has changed with time.

This book is also a reflection of Dr Gupta's love for photography and his academics of the highest standard. It would be a useful guide for the younger practicing and trainee urologists.

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During my long career in urology for last 38 years, I had the opportunity to work in two major healthcare institutions of India—All India Institute of Medical Sciences, New Delhi and Medanta—The Medicity Hospital, Gurgaon, Haryana. These being tertiary care referral hospitals, I came across several challenging and rare cases of urology which an urologist may see only once in his life time. My hobby of photography was of great help in keeping the records and follow-up of these cases. I used to keep my camera with me in the hospital; and whenever I came across these interesting cases, I tried to capture the clinical and investigative images.

The specialty of urology has had several advances during the last four decades. These advances are reflected in the modality of investigations and management of these cases, which changed depending on the year in which they were treated. However, the basic concept and philosophy behind the management process remained the same. This book is a collection of cases of last 38 years. Therefore, some old images may not be of good quality and some links may be missing for which the reader is advised to refer to standard textbooks in urology. Some of these cases have already been published in the literature for ready reference.

This book has been conceptualized with the idea to share my experience to the next generation of urologists and postgraduates in order to enhance their knowledge and approach in the management of these cases. This book is an additional supplement to the existing books in urology. It emphasizes the management of a case according to the experience of the treating surgeon, availability of diagnostic and therapeutic facilities at that time, the level of healthcare and affordability of the patients who are paying from their own pockets. These factors influence the process of management in many parts of the world particularly in developing countries.

I hope my collection will prompt the next generation urologists and practitioners to keep records of their patients and document them, thus, sharing the clinical experience which can enlighten others for the benefit of humanity.

Narmada Prasad Gupta
I dedicate this book to my parents, late Shri Mohanlal Gupta and my mother Late Smt Sato Bai, who brought me to this world and inspired me to become a doctor. I am also grateful to my teachers Dr OP Mishra, Professor and Head, Department of Surgery, Government Medical College, Jabalpur, Madhya Pradesh, India, who taught me the basics of surgery and Late Dr Sarindar Man Singh, Professor and Head, Department of Urology, All India Institute of Medical Sciences, New Delhi, for imbuing in me honesty, punctuality and sincerity in life. I am also grateful to Dr Naresh Trehan, for providing me the opportunity to join Medanta—The Medicity Hospital and for his vision, dedication and zeal to do more for our profession.

My sincere thanks to my colleagues in Urology—Dr SN Wadhwa, Dr PN Dogra, Dr Ashok Hemal, Dr Amlesh Seth, Dr Rajeev Kumar, Dr Rajiv Yadav and also to more than 120 MCh Urology students and fellows who have worked with me and were associated in the management of these cases during the last 38 years.

I am grateful to Dr Mahendra Bhandari, Formerly, Director & Professor and Head, Department of Urology and Renal Transplantation, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow, Uttar Pradesh, India, and currently Director, Robotic Surgery Research and Education, Vattikuti Urology Institute, Henry Ford Hospital, Detroit, USA, and Dr SK Sharma, Formerly, Director & Professor and Head, Department of Urology, Postgraduate Institute of Medical Education and Research, Chandigarh, India, for writing the Forewords. I had privilege of knowing and working with them since 1975.


I also thank Dr Rajan Duggal, for his help with the pathology figures. My special thanks to Dr Anand Kumar, Dr Emmanuel Akpo and my secretary Abhishek Singhal, for their help in preparation of this manuscript.

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Last but not least, I am thankful to my wife Dr Mrs Usha Gupta, who sacrificed many things in life for my career and allowed me to fulfill my dreams.
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1.1 ANTENATAL PELVIURETERIC JUNCTION OBSTRUCTION

**INTRODUCTION**

With routine use of prenatal ultrasonography, hydronephrosis is the most commonly diagnosed condition antenatally. The complete natural history of this condition is still not known. There has been much concern for the unilateral antenatally diagnosed pelviureteric junction (ADPUJ) obstruction due to the studies which note the vulnerability of developing kidneys. If the function of the affected kidney is well preserved, then conservative treatment with regular follow-up is recommended, otherwise surgery should be done.

**CLINICAL PRESENTATION**

A 28-weeks pregnant lady had prenatal ultrasound which revealed dilatation of the pelvicalyceal system of the right kidney (Fig. 1.1.1).

![Prenatal ultrasound showing hydronephrosis in the right kidney](image)

*Fig. 1.1.1:* Prenatal ultrasound showing hydronephrosis in the right kidney
Prenatal ultrasound has become routine during pregnancy. A large number of children are diagnosed with prenatal hydronephrosis. It is important to differentiate between pelviureteric junction (PUJ) obstruction and posterior urethral valves. After birth, these children need further evaluation to decide for conservative treatment or surgery.

**BIBLIOGRAPHY**

1.2 BILATERAL PELVIURETERIC JUNCTION OBSTRUCTION

**INTRODUCTION**

Bilateral ureteropelvic junction (UPJ) obstruction is rare. Traditionally, surgical management is by staged pyeloplasty to minimize the morbidity associated with performing procedures concurrently. With the advent of minimal invasive approaches concurrent surgical management can more readily be performed, laparoscopically or robotically.

**CLINICAL CASES**

**Case 1**

A 42-year-old male presented with right flank pain. Intravenous urogram (Fig. 1.2.1) revealed bilateral UPJ obstruction. Diuretic renogram confirmed the obstruction. Patient underwent bilateral pyeloplasty.

![Fig. 1.2.1: Case 1—Intravenous urogram showing bilateral PUJ obstruction](image)

**Case 2**

A 39-year-old male presented with right flank pain. Intravenous urogram (Fig. 1.2.2) revealed right UPJ obstruction with hydronephrosis and left dilatation of pelvis with normal calyx. Dynamic renal scan confirmed obstruction on the right side but pattern III on left side. Right pyeloplasty was done and followed by left pyeloplasty.
Case 3

A 45-year-old female presented with dull abdominal pain. Intravenous urogram (Fig. 1.2.3) revealed bilateral pelviureteric junction (PUJ) obstruction. Bilateral pyeloplasty was done in the one sitting.
**COMMENTS**

Bilateral PUJ obstruction can occur in about 10% of cases of PUJ obstruction, can be diagnosed synchronously, or later in life. Concurrent bilateral pyeloplasty can be done safely.

**BIBLIOGRAPHY**


1.3 CIRCUMCAVAL URETER

- **INTRODUCTION**

  It is commonly referred to as circumcaval or retrocaval ureter. More appropriate term may be preureteral vena cava. It is due to a congenital abnormality in development of the vena cava where the right ureter courses posterior to the inferior vena cava and partially encircles it. Although the lesion is congenital, patients usually present in the third to fourth decade of life.

- **EMBRYOLOGY**

  It results from persistence of the subcardinal venous system that anomalously forms the inferior vena cava (IVC) (Fig. 1.3.1).

  ![Fig. 1.3.1: Embryology of circumcaval ureter](image)

- **INCIDENCE**

  The incidence of circumcaval ureter is 1 in 1100. It commonly presents in the third to fourth decade of life, being more common in females and mostly on the right side.
**CLINICAL PRESENTATION**

Abdominal and flank pain, recurrent urinary tract infection, and hypertension.

**Types of Circumcaval Ureter**

*Type I*

Ureter with an S-shaped, fish hook, or Shepherd’s crook appearance, more hydronephrosis and more symptomatic (Fig. 1.3.2A).

*Type II*

A less angulated “sickle-shaped” ureteral deformity is classified as type II, less hydronephrosis and less symptomatic (Fig. 1.3.2B).

**IMAGING**

Intravenous pyelogram (IVP) or contrast enhanced computed tomography (CECT) with urogram (Figs 1.3.3 to 1.3.8).

**TREATMENT**

The classical treatment for retrocaval ureter consists of separating the ureter, re-anastomosing its stumps and replacing the ureter in its usual position while maintaining its patency. Pyeloureterostomy or ureteroureterostomy can be done and the approach may be open laparoscopic transabdominal/retroperitoneoscopic/robotic (Fig. 1.3.9).
Fig. 1.3.3: Intravenous pyelogram (IVP) showing hydroureteronephrosis (HDUN) in upper ureter

Fig. 1.3.4: Contrast enhanced computed tomography (CECT) showing reverse J picture

Fig. 1.3.5: Contrast enhanced computed tomography (CECT) showing reverse J picture
Fig. 1.3.6: Computed tomography (CT) Urogram showing reverse J picture

Fig. 1.3.7: Non Contrast CT showing hydroureteronephrosis (HDUN) with thickening around vena cava

Fig. 1.3.8: Contrast enhanced computed tomography (CECT) showing hydroureteronephrosis (HDUN) with enhanced mass lesion around vena cava

Fig. 1.3.9: Intraoperative picture showing caval division and circumcaval ureter
1.4 CROSSED RENAL FUSED ECTOPIA

INTRODUCTION

Crossed fused ectopia of the kidneys is the most common fusion abnormality of the urinary tract after horseshoe kidney. In this abnormality, both kidneys are present on the same side of the body (in most cases fused, with two separate ureters arising from the two kidneys). The crossed kidney has an aberrant vascular supply. The ureter arising from the crossed over kidney travels back to the opposite side and enters the bladder. Although crossed renal ectopia is usually clinically silent, they are sometimes responsible for infection and urinary stones and may be associated with a high incidence of ureteropelvic junction obstruction, vesicoureteral reflux and renal multicystic dysplasia.

CLINICAL PRESENTATION

A 26-year-old male presented with complaints of hematuria off and on for 6 months duration, and pain in right lower abdomen and periumbilical region for 4 months duration.

No association with bowel or bladder symptoms, no history of blood in stool or altered bowel habits and no history of weight loss.

GENERAL EXAMINATION

Pulse: 80/min, Blood pressure: 124/86 mm Hg, Per abdominal examination: Soft, non-tender, no lump palpable.

INVESTIGATIONS

- CBC and RFT: Normal, Urine R/M-normal—WBC: 6–10/hpf, RBC 3+, Urine C/S: Sterile, 24 hrs Urine—albumin: 71 mg%, creatinine: 31 mg/dL, calcium: 2 mg/dL, po4: 7 mg%
- Kidney, Ureter, Bladder (KUB) X-ray (Fig. 1.4.1): 2 × 2.5 cm stone seen in lower abdomen
- IVP (Fig.1.4.2): Left kidney—placed on right side with a stone in the left renal pelvis, Pelvicalyceal system (PCS) dilated on left side. Rotation of left kidney noted. Urinary bladder normal
- CECT (Figs 1.4.3 and 1.4.4): 2.5 × 2.2 cm stone in extrarenal pelvis of crossed fused ectopic left kidney. The left kidney at L3–5 location in right paraspinale location and fused with lower pole of right kidney. Left ureter crossing midline towards the left side with drainage at the normal left vesicoureteric junction (VUJ).
Fig. 1.4.1: Plain X-ray of the kidney, ureter, bladder (KUB) region showing radio-opaque shadow in right lumbar vision

Fig. 1.4.2: Intravenous pyelogram showing left crossed renal ectopia with calculus

Fig. 1.4.3: Noncontrast computed tomography (NCCT) of the abdomen
Fig. 1.4.4: Contrast enhanced computed tomography (CECT) of the abdomen

**TREATMENT**

Open pyelolithotomy was done.

**COMMENTS**

Crossed fused ectopia of kidney associated with stones can be managed by open surgery, and recently by percutaneous nephrolithotomy (PCNL) or laparoscopic-assisted PCNL.

**BIBLIOGRAPHY**

1.5 RETROILIAC URETER WITH CONTRALATERAL TRANSVERSE MALROTATION OF KIDNEY

**INTRODUCTION**

Retroiliac ureter is a well recognized, although extremely rare, congenital anomaly. It can be associated with other congenital anomalies.

**CLINICAL PRESENTATION**

An 18-year-old male presented with the complaints of nonradiating central abdominal pain, intermittent moderate fever and diarrhea for 3 months duration. He also had nausea, anorexia and weight loss. He noticed a lump in the lower abdomen. Abdominal examination revealed a 10 × 6 cm sausage shaped, smooth, firm, non-tender, midline, intra-abdominal lump in the hypogastrium and extending into both iliac region. There were no associated congenital anomalies.

**INVESTIGATIONS**

Hemoglobin (Hb): 6.7 gm, Blood urea: 72 mg/dL, Serum creatinine: 2.6 mg/dL. Creatinine clearance was 20 mL/minute. Urine examination revealed pus cells and urine culture was positive for *Escherichia coli*. Plain X-ray KUB region revealed a radiopaque stone opposite L4 vertebra on left side. Intravenous urogram revealed a left hydronephrotic kidney opposite L4-S1 with nonvisualized right kidney. Ultrasound revealed a transversely oriented left kidney with moderate hydronephrosis that crossed the midline. The right kidney was in normal position with hydroureteronephrosis. A dynamic renal scan confirmed a non-functioning right kidney with transverse poorly functioning left kidney. A voiding cystourethrogram revealed normal capacity bladder with grade IV reflux on right side. Cystoplasty revealed normal left and gaping right ureteric orifice. A left retrograde ureteropyelogram revealed ureteropelvic junction obstruction with transverse kidney.

**TREATMENT**

Abdominal exploration revealed left kidney was malrotated transversely with a dilated pelvis cephalad to the kidney. The calculus was extracted through vertical pyelotomy and nephrostomy was done. A nephrostogram (Fig. 1.5.1) done on 14th day revealed no drainage. Re-exploration was done. Right kidney was found hydronephrotic with dilated ureter coursed posterior to the iliac artery. Right nephrectomy was done and ureter was divided 3 cm above the iliac vessels. The distal half of the ureter freed and brought anterior to the iliac artery and crossed ureterocalyceal anastomosis was done with the left inferior calyx after excision of a disc of the parenchyma. A stent and nephrostomy was kept. Postoperative nephrostogram (Fig. 1.5.2) revealed a good drainage with patent anastomosis.
FOLLOW-UP

After 3 months, postoperative cystogram (Fig. 1.5.3) revealed right refluxing ureter. Right ureteral advancement procedure was done for the vesicoureteral (VU) reflux. His renal function improved and was normal for a 5-year follow-up.
COMMENTS

A retroiliac ureter is very rare and preoperative diagnosis is difficult. By crossed ureteral re-implantation single functioning kidney can we salvaged by use of contralateral ureter of nonfunctioning kidney.

BIBLIOGRAPHY

1.6 DOUBLE MOIETY WITH PELVIURETERIC JUNCTION OBSTRUCTION LOWER MOIETY

**INTRODUCTION**

Ureteropelvic junction (UPJ) obstruction may also occur in the upper or lower, usually lower half of a duplicated collecting system.

**CLINICAL CASES**

**Case 1**

A 26-year-old female presented with vague pain in abdomen. Ultrasound (Fig.1.6.1) revealed double moiety with pelviureteric junction (PUJ) obstruction inferior moiety. Intravenous urogram (Fig.1.6.2) and CECT (Fig.1.6.3) showing double moiety with PUJ obstruction inferior moiety. Treatment was done by right pyeloplasty of the inferior moiety.

![Ultrasound showing double moiety with PUJ obstruction inferior moiety](image-url)

**Fig. 1.6.1:** Case 1—Ultrasound showing double moiety with PUJ obstruction inferior moiety
Fig. 1.6.2: Case 1—Intravenous pyelogram (IVP) showing double moiety with PUJ obstruction inferior moiety

Fig. 1.6.3: Case 1—Contrast enhanced computed tomography (CECT) showing double moiety with PUJ obstruction inferior moiety

Case 2
A 32-year-old female presented with pain in upper abdomen. Intravenous pyelogram (IVP) (Fig. 1.6.4) revealed double moiety with PUJ obstruction of inferior moiety. Right pyeloplasty of the inferior moiety was done.
Case 3

A 32-year-old female presented with right flank pain. Contrast enhanced computed tomography (Fig. 1.6.5) revealed double moiety with obstruction of inferior moiety. Dynamic renal scan revealed very poorly functioning inferior moiety. Right lower pole nephrectomy was done.
COMMENTS

Careful preoperative evaluation of patients with ureteropelvic junction obstruction will usually identify segmental obstruction in a duplicated system. Treatment should be individualized based on site of obstruction and degree of function remaining in the affected segment.

BIBLIOGRAPHY

1.7 ECTOPIC PELVIC KIDNEY

INTRODUCTION

An ectopic kidney is a birth defect in which a kidney is located below, above, or on the opposite side of its usual position. About one in 900 people has an ectopic kidney. Abnormal positions are described as pelvic, iliac, abdominal, thoracic and crossed, with pelvic location being the most common. An ectopic kidney may not cause any symptom and may function normally, even though it is not in its usual position. No treatment for an ectopic kidney is needed if urinary function is normal and no blockage of the urinary tract is present. Common complications of ectopic kidney are infection, stone formation and obstruction.

CLINICAL PRESENTATION

A 32-year-old male presented with lower abdominal pain of 6 months duration. On examination, there were no positive findings.

INVESTIGATIONS

Plain X-ray KUB region (Fig. 1.7.1) revealed no radiopaque shadow. Intravenous urogram (Figs 1.7.2A and B) revealed left ectopic kidney with hydronephrosis, with normal functioning right kidney.

Fig. 1.7.1: Plain X-ray KUB region
TREATMENT

Left calicovesicostomy was done. Patient did well after surgery.

COMMENTS

Pelvic ectopic kidney with hydronephrosis is an extremely rare condition. Due to proximity of the kidney with the bladder, pelvio/calicovesicostomy can be done with good outcome.

BIBLIOGRAPHY

## CONGENITAL ANOMALIES OF KIDNEY AND URETER

### 1.8 ECTOPIC URETER

**INTRODUCTION**

An ectopic ureter is one which is not opening at the normal location at the angle of the trigone. Eighty percent of ectopic ureter occurs with ureteral duplication; however, single system ectopic ureter can also occur. Ectopic ureter is more common in females. Ectopic ureter can be unilateral or bilateral. Ectopic ureter may open into the urinary tract or genital tract following an ectopic pathway. Opening of ectopic ureter from the vesical neck distally can cause symptoms of obstruction; reflux and in females, incontinence. It can be associated with other congenital anomalies.

**CASE 1**

**Clinical Presentation**

A 14-year-old female presented with incontinence of urine since birth. She was voiding normally along with one to two pads leakage of urine.

**Investigations**

Intravenous urogram (Fig. 1.8.1) revealed right double collecting system with dilated superior moiety. Antegrade pyelogram (Figs 1.8.2A and B) through superior moiety was done which revealed dilated superior moiety and ureter with ectopic opening of ureter.

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**Fig. 1.8.1:** Case 1—Intravenous urogram
Treatment
Superior moiety nephroureterectomy was done. As a result, her incontinence stopped.

CASE 2
Clinical Presentation
A 38-year-old female presented with incontinence of urine few drops along with normal voiding.

Investigations
Intravenous urogram (Fig. 1.8.3) revealed right double moiety with poorly functioning dilated superior moiety with ectopic opening.

Treatment
Right superior moiety nephroureterectomy was done which stopped her incontinence.
CASE 3

Clinical Presentation

A 15-year-old female presented with history of paradoxical incontinence since birth. She had ectopic anus for which cut back operation was done at birth. Local examination showed patulous urethral orifice with two separate vaginal introitus leading to separate 5 cm deep vagina. There was one cervix and one uterus on each side (uterus didelphus).

Investigations

Intravenous urogram showed normal upper tract. Voiding cystourethrogram (Fig. 1.8.4) showed grade III left vesicoureteral reflux with a wide open bladder neck and contrast in both vagina. Cystoscopy revealed an absent trigone, both ureteric orifices at the bladder neck at 3 and 9 O’clock position. The length of the urethra was 3 cm.

Treatment

Cohen’s bilateral transtrigonal ureteroneocystostomy with Young-Dees-Leadbetter bladder neck reconstruction and Marshall-Marchetti-Krantz urethral fixation was done. Postoperative recovery was uneventful and continence was achieved.
Ectopic ureter usually occurs with double collecting system but can also occur in a single system and can be unilateral or bilateral. If symptomatic, needs surgical treatment.

**BIBLIOGRAPHY**

1.9 EQUIVOCAL PELVIURETERIC JUNCTION OBSTRUCTION

**INTRODUCTION**

Intravenous urogram is the most common investigation for evaluation of pelviureteric junction (PUJ) obstruction. Classic urographic features of equivocal PUJ obstruction are dilatation of renal pelvis, minimum or no calyceal dilation and minimum filling of ureter. Diuresis renogram will differentiate whether functional obstruction is present or not. In the absence of obstruction, these patients can be kept under observation and unnecessary surgery can be avoided.

**CLINICAL PRESENTATION**

A 35-year-old male presented with right flank pain off and on for a duration of 6 months. Examination was normal.

**INVESTIGATIONS**

Intravenous urogram (Fig. 1.9.1) revealed right dilatation of the renal pelvis, with minimum dilatation of the calyces and the ureter was partially filled. Diuresis renogram revealed Type 3A pattern.

![Fig. 1.9.1: Intravenous pyelogram (IVP) showing features of equivocal PUJ obstruction—dilated pelvis, cupping of calyx maintained and partial filling of ureter](image-url)
**TREATMENT**

He was kept under follow-up. He was asymptomatic there was no further deterioration of renal function.

**COMMENTS**

It is important to identify cases of hydronephrosis without functional obstruction. They can be followed-up periodically, and if asymptomatic and without any complications, then surgery can be avoided.

**BIBLIOGRAPHY**

1.10 EXTRARENAL PELVIS AND CALYCES

INTRODUCTION

Extrarenal calyces (wherein the calyces and renal pelvis lie outside the renal parenchyma) is one of the rare anomaly of the collecting system. It may develop due to a disparity resulting from slow development of the metanephric tissue or to a relatively rapid development of the ureteric bud. This anomaly may be associated with other anomalies of the urogenital system. Extrarenal pelvis and calyces usually do not produce symptoms, but failure to drain normally may lead to stasis, infection and calculi formation. Surgery is reserved for those patients in whom infection or obstruction is present.

CLINICAL CASES

Case 1

A 28-year-old male presented with vague pain in the left flank. Intravenous urogram (Fig. 1.10.1) revealed left malrotated ectopic kidney with extrarenal pelvis. Dynamic renal scan revealed no obstruction. He was kept under observation.

Fig. 1.10.1: Case 1—Intravenous urogram

Case 2

A 35-year-old female presented with left flank pain. Intravenous urogram (Fig. 1.10.2) revealed bilateral extrarenal pelvis with left PUJ obstruction. Dynamic renal scan revealed obstruction on the left side. She underwent left pyeloplasty.
Case 3

A 32-year-old male presented with vague pain in the abdomen. Intravenous urogram revealed a right double collecting system with an extrarenal pelvis of the inferior moiety (Fig. 1.10.3). Dynamic renal scan revealed no obstruction. He was kept under observation.
**COMMENTS**

Extrarenal pelvis and calyces can be incidental findings on imaging. If asymptomatic, can be observed, otherwise; surgery is required.

**BIBLIOGRAPHY**

1.11 INCOMPLETE DUPLICATION OF URETER

**INTRODUCTION**

Ureteral duplication is the most common ureteral anomaly. It can be complete duplication, where second ureter opens in the bladder, urethra or ectopic position. In incomplete duplication, there is bifid ureter. In many cases, it can be an accidental finding causing no functional disturbances, otherwise it can be associated with upper tract stasis, obstruction and reflux. There is a higher incidence of duplicated ureter in cases with urinary infection. It is more common in females.

**CLINICAL PRESENTATION**

A 35-year-old female presented with vague pain in the abdomen.

**INVESTIGATIONS**

Intravenous urogram (Fig.1.11.1) revealed a left incomplete duplication of ureter. Urine examination was normal.

![Intravenous urogram](image)

**TREATMENT**

She was counseled and was advised for observation.
**COMMENTS**

Duplication of ureter can be detected on imaging. Asymptomatic cases without obstruction can be kept on follow-up.

**BIBLIOGRAPHY**

1.12 MEGAPELVIS

**INTRODUCTION**

Megapelvis means a big renal pelvis. It is a condition in which there is congenital dilation of the renal pelvis due to smooth muscle aplasia. This causes hydronephrosis also. Renal pelvis is the area of kidney where urine is collected before passing through ureter. When urine gets backed up, the renal pelvis gets dilated. It can occur in adults as well as infants.

**CLINICAL PRESENTATION**

A 20-year-old female presented with bilateral dull flank pain for 6 months duration. No urinary symptoms, no operative intervention in the past and no physical findings on examination.

**INVESTIGATIONS**

Urine R/M: Normal, Urine C/S: Sterile, Renal function test: Normal. Ultrasound (Fig. 1.12.1): showing dilatation of pelvis of both kidneys. Plain X-ray (Fig. 1.12.2): No radiopaque shadow. IVP (Fig. 1.12.3): showing dilatation of both pelvis and calyces but cupping is maintained. Dynamic Renal Scan (Fig. 1.12.4): showing normal function of both kidneys with normal drainage. MRI T1 images (Fig. 1.12.5), MRI T2 images (Fig.1.12.6) and MRI T2 coronal images (Fig.1.12.7) showing dilated pelvis and calyces with no obstruction.

Fig. 1.12.1: Ultrasound
Fig. 1.12.2: Plain X-ray

Fig. 1.12.3: Intravenous pyelogram

Fig. 1.12.4: Dynamic renal scan
Fig. 1.12.5: MRI T1 images

Fig. 1.12.6: MRI T2 images
Fig. 1.12.7: MRI T2 coronal images

**MANAGEMENT**

The patient was counseled for conservative treatment and follow-up.

**COMMENTS**

Megapelvis is a very rare condition and less reference about it is available in the literature. If there is no obstruction in the dynamic renal scan (DRS), patients can be kept under follow-up.
1.13 PRIMARY MEGAURETER

**INTRODUCTION**

Megaureter is a ureter that exceeds the upper limits of normal size. Any ureter greater than 7 mm in diameter is defined as a megaureter based on measurements in fetuses greater than 30 weeks gestation and children younger than 12 years.

Primary megaureter results from a functional or anatomical abnormality involving the ureterovesical junction. Megaureter can be obstructive or nonobstructive and can be refluxing or nonrefluxing.

**TREATMENT**

Treatment is decided on the basis of obstruction and associated complications like stones, infection, etc.

**CLINICAL PRESENTATION**

Nine cases are presented with variable symptoms and their imaging results (Figs 1.13.1 to 1.13.9).

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**Fig. 1.13.1:** Case 1—Intravenous pyelogram: rat tail appearance of megaureter  
**Fig. 1.13.2:** Case 2—Intravenous pyelogram: left megaureter
Fig. 1.13.3: Case 3—Intravenous pyelogram: right megaureter

Fig. 1.13.4: Case 4—Intravenous pyelogram: right megaureter

Fig. 1.13.5: Case 5—Intravenous pyelogram: bilateral megaureter
Fig. 1.13.6: Case 6—Micturating cystourethrogram (MCU): Bilateral refluxing megaureter

Fig. 1.13.7: Case 7—Intravenous pyelogram: single ectopic kidney with megaureter

Fig. 1.13.8: Case 8—Intravenous pyelogram: single pelvic ectopic kidney with megaureter
COMMENTS

Most cases of primary megaureter resolve spontaneously or improve without loss of function or development of symptoms. Careful observation allows surgery to be delayed beyond the neonatal period in most patients. Long-term follow-up is recommended because symptoms can develop years later. Washout pattern and age at presentation are statistically significant predictors of spontaneous resolution.

BIBLIOGRAPHY

1.14 PARAPELVIC CYST

**INTRODUCTION**

Parapelvic cysts of the kidneys are simple renal cysts, which are adjacent to the renal pelvis or the renal sinus. They are usually derived from lymphatic vessels or from renal parenchyma. Normally, there are no complaints. Parapelvic cysts can compress the pyelocalyceal system or ureter, cause flank pain or lead to other symptoms of urinary obstruction. Renal ultrasound should be sufficient for the diagnosis; in doubtful situations an intravenous pyelography, computed tomography (CT) of abdomen or renal scintigraphy can rule out urinary obstruction.

**CLINICAL PRESENTATION**

A 30-year-old female presented with left hypochondrium and flank discomfort, post-meal fullness. Her history suggested no urinary symptoms, normal bowel functions and no operative intervention in the past.

**EXAMINATION**

No palpable mass or organomegaly, no lymphadenopathy and no pedal edema.

**INVESTIGATION**

Hemogram, RFT, LFT: Normal. Urine R/M: Normal; Urine culture: Sterile, UGI endoscopy: Normal. Barium meal and follow through: Normal; USG: Normal right kidney, 5 × 6 cm parapelvic cyst in left kidney. CECT revealed parapelvic cyst (Fig.1.14.1).

![Fig. 1.14.1: Parapelvic cyst revealed by contrast enhanced computed tomography (CECT)](image)

**TREATMENT**

Retroperitoneoscopic cyst deroofing was done.
COMMENTS

Ultrasonography and CT scan are the main diagnostic methods. Enhanced CT is extremely helpful in differential diagnosis of hydronephrosis. Surgical management is suitable for big cysts, lumbar pain, hematuria, hypertension and other complications.

BIBLIOGRAPHY

1.15 PELVIURETERIC JUNCTION OBSTRUCTION
WITH ECTOPIC KIDNEY

INTRODUCTION
Pelvic ectopic kidney can be associated with congenital PUJ obstruction with hydronephrosis. Due to close proximation with the bladder, the surgical treatment options are calycovesical or pyelovesical anastomosis which provide excellent drainage.

CLINICAL PRESENTATION
A 22-year-old male presented with periumbilical pain for 2 years, no operative intervention and no bowel/bladder symptoms.

INVESTIGATIONS
Urine routine and culture, and renal function were normal. Plain X-ray KUB region (Fig. 1.15.1): No radiopaque calculus; Intravenous urogram (Fig. 1.15.2) showed normal right kidney and ectopic left kidney. Intravenous urogram with delayed films (Fig. 1.15.3) show left hydronephrosis.
TREATMENT

Pyelovesicostomy was performed.

COMMENTS

Pyelovesicostomy is a good surgical option for patients with PUJ obstruction with hydronephrosis in ectopic pelvic kidney.

BIBLIOGRAPHY

1.16 PERSISTENT UMBILICAL ARTERY CAUSING URETERAL OBSTRUCTION

INTRODUCTION

Hyams, in 1929, first described vascular obstruction of the distal ureter and illustrated the anomalous routes of the umbilical arteries. Since then only few cases have been described in the literature. The diagnosis is usually made at laparotomy, frequently following previous failed attempts at endourological management.

CLINICAL PRESENTATION

A 32-year-old female presented with the history of recurrent left flank pain of 1 year duration. She had no urinary complaints, was married for 3 years and was investigated for infertility.

INVESTIGATIONS

Intravenous urogram (Fig. 1.16.1) revealed moderate hydroureteronephrosis on the left side, with ureter seen 1 cm distal to sacroiliac joint. Magnetic resonance (MR) urogram (Fig. 1.16.2) revealed left hydroureteronephrosis. Left retrograde pyelogram (Fig. 1.16.3) and drainage film (Fig. 1.16.4) revealed hydroureteronephrosis with sudden cut of lower ureter and was hook shaped. Ureteric catheter could not be negotiated beyond 8 cm.

Fig. 1.16.1: Intravenous urogram

Fig. 1.16.2: Magnetic resonance urogram
TREATMENT

On exploration, a 4-mm vessel was seen crossing over the ureter at the site of narrowing of the ureter. The ureter was divided and ureteral reimplantation was done.
**FOLLOW-UP**

Postoperative intravenous urogram (Fig. 1.16.5) after 3 months showed nonobstructive drainage on left side.

**COMMENTS**

Persistent umbilical artery causing ureteral obstruction is a very rare condition and should be kept as a differential diagnosis in lower ureteric obstruction.

**BIBLIOGRAPHY**

1.17 PELVIURETERIC JUNCTION OBSTRUCTION WITH CALCULI

INTRODUCTION

Pelviureteric junction obstruction can be associated with renal calculi. The etiology for calculus is due to stasis of urine and can be associated with metabolic abnormalities. These stones are usually round or facetted and can be multiple. Management is removal of calculi along with obstruction by endopyelotomy or more commonly performed pyeloplasty.

CLINICAL CASES

Case 1

A 45-year-old female presented with left flank pain. Plain X-ray KUB region (Fig. 1.17.1) revealed multiple radiopaque round shadows in the left renal area. Intravenous pyelogram (Fig. 1.17.2) revealed left pelviureteric junction obstruction with hydronephrosis with calculi. Left Anderson-Hynes pyeloplasty with removal of calculi was done. Figure 1.17.3 shows multiple calculi with excised pelvis. She recovered well.

Fig. 1.17.1: Case 1—Plain X-ray KUB region
Case 2

A 28-year-old male presented with left flank pain. Plain X-ray KUB region (Fig. 1.17.4) revealed three radiopaque round shadows in the left renal area. Intravenous pyelogram (Fig. 1.17.5) revealed left pelviureteric junction obstruction with hydronephrosis with calculi. Left robotic Anderson-Hynes pyeloplasty with removal of calculi was done. The patient recovered well.
COMMENTS

Concurrent stone extraction and pelviureteric junction obstruction (PUJO) repair can be successful. Operative time is longer than in patients with isolated PUJO repair, but this is to be expected as there is an additional procedure.

BIBLIOGRAPHY

1.18 PELVIURETERIC JUNCTION OBSTRUCTION
WITH MALROTATED KIDNEY

**INTRODUCTION**

Pelviureteric junction (PUJ) obstruction associated with malrotated kidney is very rare. In such cases, dismembered pyeloplasty poses technical difficulties.

**CLINICAL CASES**

**Case 1**

A 28-year-old male presented with vague abdominal pain. Plain X-ray of KUB region revealed no radiopaque shadows. Intravenous urogram (Fig. 1.18.1) revealed bilateral malrotated kidneys with hydronephrosis. Dynamic renal scan revealed bilateral partially obstructed pattern. Bilateral dismembered open pyeloplasty was done.

![Case 1—Intravenous urogram](image)
Case 2

A 35-year-male presented with left flank pain. Plain X-ray of KUB region revealed no radiopaque shadows. Intravenous urogram (Fig. 1.18.2) revealed left malrotated kidney with hydronephrosis with right ectopic pelvic kidney.

![Fig. 1.18.2: Case 2—Intravenous urogram](image)

**COMMENTS**

PUJ obstruction with malrotated kidney is very rare and only few cases are reported. The surgical outcome of pyeloplasty may not be good.

**BIBLIOGRAPHY**

1.19 PELVIURETERIC JUNCTION OBSTRUCTION WITH MEGAPELVIS

**INTRODUCTION**

Patients with extrarenal pelvis with pelviureteric junction obstruction can have a large renal pelvis. The obstruction can be anywhere in the ureter and above that pelvis can be dilated.

**CLINICAL CASES**

**Case 1**

A 24-year-old male presented with right flank pain. Plain X-ray KUB region revealed no radiopaque shadow. Intravenous urogram (Fig. 1.19.1) revealed right hydronephrosis with long dilated pelvis with obstruction at the L4 level. Right dismembered pyeloplasty with ureterization of pelvis was done.

![Fig. 1.19.1: Case 1—Intravenous urogram](image)
**Case 2**

A 28-year-old male presented with right flank pain. Plain X-ray KUB region revealed no radiopaque shadow. Intravenous urogram (Fig. 1.19.2) revealed right hydronephrosis with long hugely dilated pelvis with obstruction at the L4 level. Right dismembered pyeloplasty with ureterization of pelvis was done.

![Intravenous urogram](image)

**Fig. 1.19.2:** Case 2—Intravenous urogram

**COMMENTS**

Pelviureteric junction obstruction with large megapelvis is a rare congenital anomaly and can be managed surgically by excision of large pelvis.

**BIBLIOGRAPHY**

1.20 PELVIURETERIC JUNCTION OBSTRUCTION WITH SINGLE KIDNEY

**INTRODUCTION**

Pelviureteric junction (PUJ) obstruction is associated with other anomalies. Unilateral agenesis of the kidney can be associated in almost 5% of children with PUJ obstruction.

**CLINICAL PRESENTATION**

A 35-year-old male presented with left flank pain. Abdominal examination was normal. Blood and urine examination was normal. Intravenous urogram (Fig. 1.20.1) revealed single kidney with PUJ obstruction. Cystoscopy revealed right hemitrigone with absence of ureteric orifice. Right retrograde pyelography (RGP) confirmed PUJ obstruction. Left open pyeloplasty was done.

![Intravenous urogram](image)

**Fig. 1.20.1: Intravenous urogram**

**COMMENTS**

Congenital absence of kidney can be associated with pelviureteric junction obstruction and management is same as in the presence of two kidneys and prognosis is good.

**BIBLIOGRAPHY**

1.21 PELVIC ECTOPIC KIDNEY

**INTRODUCTION**

When a fully mature kidney is not in its normal location, it is known as renal ectopia. Pelvic kidney is the most common type of renal ectopia. The ectopic pelvis kidney is opposite the sacrum and below the aortic bifurcation. The contralateral kidney is usually normal. This can be associated with other congenital anomalies, the common ones are genital anomalies. Imaging by intravenous pyelogram, ultrasound and CT scan can diagnose ectopic kidney. The kidney function can be normal. These abnormally positioned kidneys are often clinically asymptomatic. However, they are prone to urinary infection, stone formation and trauma. It can be associated with obstruction and needs surgical correction.

**CLINICAL PRESENTATION**

A 32-year-old male presented with vague abdominal pain.

**INVESTIGATIONS**

Intravenous urogram revealed normal right kidney with ectopic pelvic kidney with mild hydronephrosis (Fig. 1.21.1). Dynamic renal scan revealed normal function and drainage of the ectopic kidney.

![Intravenous urogram](image-url)

**Fig. 1.21.1:** Intravenous urogram
**TREATMENT**

He was advised conservative treatment.

**COMMENTS**

Pelvic ectopic kidney can be an incidental finding on imaging. If symptomatic, surgery is recommended.

**BIBLIOGRAPHY**

1.22 URETEROCELE

INTRODUCTION

A ureterocele is cystic dilatation of the intravesical submucosal ureter. The size may vary from a small bulge of 1–2 cm to a size which can fill complete bladder. This swollen area can block urine flow. Ureterocele occurs in about one in 500 to one in 4,000 people. It is more common in Caucasians. Ureterocele is equally common in left- and right-side ureters.

Most common symptoms are abdominal pain, back pain, possibly only on one side, blood in the urine, burning pain while urinating (dysuria), flank pain, lump (mass) in the abdomen that can be felt, ureterocele tissue falls down (prolapse) through the female urethra and into the vagina, urinary incontinence, urinary tract infection.

During the last 20 years, the surgical approach to ureterocele has evolved from major open surgery to minimally invasive endoscopic puncture. The author believes that the endoscopic approach decreases the need for open surgical procedures.

CLINICAL CASES

Case 1

A 36-year-old female presented with left flank pain. Plain X-ray (Fig. 1.22.1) revealed a round radiopaque shadow in the pelvis. Intravenous urogram (Fig. 1.22.2) revealed left ureterocele with a calculus with mild

Fig. 1.22.1: Case 1—Plain X-ray pelvis
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hydroureteronephrosis. Ultrasound (Fig. 1.22.3) revealed left ureterocele with a calculus. Cystopanendoscopy revealed left ureterocele. A smiling incision was made by holmium laser and stone was also fragmented with the laser.

Fig. 1.22.2: Case 1—Intravenous urogram showing ureterocele with calculus

Fig. 1.22.3: Case 1—Ultrasound showing left ureterocele with calculus

Case 2

Another case with similar presentation. Intravenous urogram (Fig. 1.22.4) revealed a large left ureterocele (Cobra head appearance) with hydroureteronephrosis (HDUN). Endoscopic incision was done.
Case 2—Intravenous urogram showing cobra head appearance of ureterocele

Case 3
Another case with left flank pain. Intravenous urogram revealed bilateral double collecting system with left lower moiety ureterocele (Fig. 1.22.5). Common sheath ureteric reimplantation was done.

Fig. 1.22.5: Case 3—Intravenous urogram showing left ureterocele
Case 4

A 56-year-old presented with vague abdominal pain. CECT abdomen revealed a mass lesion near left ureteric orifice (Fig. 1.22.6). Cystoscopy revealed left ureterocele (Fig. 1.22.7) which was ballooning with efflux of urine. Endoscopic incision was made.

Fig. 1.22.6: Case 4—CECT showing a mass lesion at the base of bladder

Fig. 1.22.7: Case 4—Cystoscopic view showing ureterocele
Case 5

A 6-year-old male child presented with a single episode of periumbilical, moderate, non-colicky, nonradiating pain associated with vomiting; 6 months ago. He had no associated bowel, bladder symptoms. No similar episodes in past. No history of urinary tract infection (UTI), hematuria/lithuria/poor stream-operative intervention. Urine and blood investigations were normal. Ultrasound (Figs 1.22.8A and B) revealed right double moiety with dilatation of upper moiety ureter with ureterocele. Intravenous urogram (Figs 1.22.9A and B) revealed the same findings. Dynamic renal scan (Figs 1.22.10A and B) revealed poor function in the right upper moiety. CECT scan (Figs 1.22.11A to C) reconfirmed the findings of ultrasound. Excision of ureterocele with common sheath ureteroneocystostomy of both ureters was done. Patient recovered well. Postoperative intravenous urogram (Fig. 1.22.12) revealed improved renal function in right superior moiety with normal ureters.

Figs 1.22.8A and B: Case 5—Ultrasound
Figs 1.22.9A and B: Case 5—Intravenous urogram
Figs 1.22.10A and B: Case 5—Dynamic renal scan
Figs 1.22.11A to C: Case 5—Contrast enhanced computed tomography (CECT) scan
COMMENTS

Ureterocele can be an incidental finding on imaging and can occur in single ureter or in the upper moiety of a duplicated ureter. If asymptomatic, does not require any treatment. If associated with ureteric obstruction, mostly can be managed endoscopically or by ureteric reimplantation.

BIBLIOGRAPHY

1.23 VESICOURETERAL REFLUX

**INTRODUCTION**

Vesicoureteral reflux (VUR) is the retrograde flow of urine from the bladder into the ureter and towards the kidney due to abnormal functioning of vesicoureteric junction. This junction usually acts like a one-way valve, allowing urine flow from the ureter into the bladder and closing during micturition, preventing back flow. Most review articles suggest a frequency of around 1% in healthy children. It can be primary or secondary to high vesical pressure and obstructive uropathy. It is commonly associated with recurrent urinary tract infection. It is diagnosed by cystography and radionuclide scanning. Reflux can be graded from I to V. For grade I and II, conservative treatment is recommended whereas higher grade can be managed by endoscopic injection or by antireflux surgery.

**CLINICAL PRESENTATION**

A 15-year-old female presented with history of recurrent urinary tract infection (UTI) and frequency of urination. Her creatinine was 3.2 mg/dL. Ultrasound revealed bilateral hydroureteronephrosis. Micturating cystourethrogram (MCU) revealed small capacity bladder with bilateral grade V bilateral vesicoureteral reflux (Fig. 1.23.1). Initially, she was kept on continuous drainage for improvement of renal function.

*Fig. 1.23.1: Micturating cystourethrogram showing bilateral vesicoureteral reflux*
COMMENTS

Vesicoureteral reflux is a common cause for recurrent UTI and can be primary or secondary. Sometimes it can compromise the renal function. It should be managed appropriately to prevent renal damage by medically or surgically according the grade of vesicoureteral reflux.

BIBLIOGRAPHY

2.1 CHORDEE WITHOUT HYPOSPADIAS

INTRODUCTION

Chordee is the congenital curvature of the penis. Usually, it is associated with hypospadias. “Chordee without hypospadias” is rare. This is also known as “congenital short urethra” or “congenital ventral curvature of the penis”. Chordee without hypospadias is classified as: (A) Cutaneous chordee; (B) Fibrous chordee; (C) Corporocavernosal chordee; (D) Urethral tethering with hypoplastic urethra; (E) Congenital short. Surgery has to be modified according to the classification.

CLINICAL CASES

Case 1

A 24-year-old male presented with abnormal curvature of the penis. On examination, he had prepuce in normal position with ventral chordee (Figs 2.1.1A and B). The penile skin was not tethered. On exploration, urethra was found short. Division of the urethra and dorsal preputial graft urethroplasty was done in single stage.

Fig. 2.1.1A and B: Case 1—Clinical pictures
Case 2

A 6-month-old child was brought by parents for abnormal curvature of penis. On examination, dorsal hood of the prepuce was well developed (Fig. 2.1.2). The meatus was of normal size and normal position. Dorsal chordee was present. The parents were counseled and advised for surgery at the age of 1 year.

Fig. 2.1.2: Case 2—Clinical picture

COMMENTS

Chordee without hypospadias is a rare condition. According to etiology and classification, surgery should be planned. Surgery can be done in one or two stages.

BIBLIOGRAPHY

2.2 COMPLETE DUPLICATION OF URETHRA

**INTRODUCTION**

Duplication of urethra is an uncommon anomaly. Most duplications occur in the same sagittal plane, on top of the other. In the dorsal variety, the normal urethra is in the ventral position and the channel normally ends in a normal meatus of the glans. The accessory (abnormal) channel opens on the shaft in an epispadic position anywhere from the glans to the base of the penis. Often, dorsal chordee is present and foreskin may be infused dorsally.

Complete duplication of the urethra with a single bladder is an exceptional finding. The anomalous urethral canal originates from the bladder separately, runs parallel and usually dorsal to the normally situated urethra, and opens on the dorsum of the penis. Conservative therapy is the treatment of choice, and that surgery should be reserved for incontinent patients only. Because the normally positioned urethra has a normal bladder neck and sphincter mechanism, excision of the abnormal, epispadiac urethra will usually cure any incontinence affecting the patient.

**CLINICAL PRESENTATION**

A 25-year-old male presented with abnormal opening in the penis and incontinence of the urine. Examination revealed normal urethral meatus with abnormal opening dorsally (Fig. 2.2.1). A ureteric catheter could be passed in the abnormal urethra. Cystoscopy was done and cystoscope could be passed in both urethra (Fig. 2.2.2). Excision of the accessory dorsal urethra was done.

![Fig. 2.2.1: Clinical picture](image)
Fig. 2.2.2: Cystoscope in both urethra

**COMMENTS**

Complete duplication of urethra is a very rare anomaly, but can be managed successfully.

**BIBLIOGRAPHY**

2.3 PENILE DUPLICATION

**INTRODUCTION**

Diphallia, penile duplication (PD), diphallic terata, or diphallasparatus, is a rare medical condition in which a male infant is born with two penises. It has a spectrum from a small accessory penis to a complete duplication. The first reported case was by Johannes Jacob Wecker in 1609. Its occurrence is one in 5,500,000 men in the United States.

When diphallia is present, it is usually accompanied by other congenital anomalies such as renal, vertebral, hindgut or anorectal duplication. There is also a higher risk of spina bifida. Treatment must be individualized to attain a satisfactory functional and cosmetic result.

**CLINICAL PRESENTATION**

A 5-year-old male presented with history of double penis. There were no urinary complaints.

**EXAMINATION**

Examination revealed two well-developed, equal size penises (Fig. 2.3.1). There was no other congenital anomaly. The parents were counseled, but they refused for any treatment.

![Fig. 2.3.1: Clinical picture showing double penis](image)
**COMMENTS**

Complete duplication of the penis is a very rare condition and should be managed according to merit.

**BIBLIOGRAPHY**

INTRODUCTION

Bladder exstrophy, cloacal exstrophy and epispadias are variants of exstrophy-epispadias complex. The etiology of this complex has been attributed to the failure of the cloacal membrane to be reinforced by in-growth of mesoderm. The incidence of bladder exstrophy is between one in 10,000 to one in 50,000 live births. Usually these birth defects are noticed immediately after birth and managed accordingly. It is rare to see an adult presenting with exstrophy of bladder but in India, due to unawareness, poverty and lack of medical care, the patients present late in life. Due to prolonged exposure of bladder mucosa, malignant changes can occur and bladder loses its pliability and patient may require cystectomy.

The primary objectives of the management are: (1) Secure abdominal wall closure; (2) Urinary continence with preservation of renal function and; (3) Reconstruction of functional and cosmetically acceptable external genitalia.

CLINICAL CASES

Case 1

A 20-year-old male presented with a defect in lower abdominal wall and genitalia, continuous leak of urine from same site since birth. He was operated for right inguinal hernia at the age of 6 years. He had no other surgical intervention. Examination revealed exposed bladder with epispadias (Fig. 2.4.1).

Fig. 2.4.1: Case 1—Clinical picture
Investigations

Plain X-ray of the pelvis revealed widening of the symphysis pubis and soft tissue shadow of the penis (Fig. 2.4.2). Intravenous urogram revealed normal upper tracts and no contrast in the bladder (Fig. 2.4.3). Bladder biopsy showed squamous dysplasia.

Fig. 2.4.2: Case 1—Plain X-ray of the pelvis

Fig. 2.4.3: Case 1—Intravenous urogram
**Treatment**

In view of changes in the bladder, cystectomy and continent diversion was done followed by correction of the epispadias.

**Case 2**

A 26-year-old male presented with similar complaints as in case 1 (Fig. 2.4.4) and same treatment was done.

**Fig. 2.4.4: Case 2—Clinical picture**

**COMMENTS**

Bladder exstrophy in adult males is very rare. Due to unawareness and poverty, such patients present with it in adult age in developing countries. Due to dysplastic changes in the exposed mucosa, the risk for malignancy increases, and bladder closure with continence chances are less. In view of this, cystectomy and continent diversion is advisable.

**BIBLIOGRAPHY**

2.5 EPISPADIAS

INTRODUCTION

Epispadias is a rare defect that is present at birth (congenital). It varies from a glandular defect in a covered penis to the penopubic type with incontinence, to the complete variety associated with bladder extrophy.

The causes of epispadias are not known. It may occur because the pubic bone does not develop properly. Epispadias can occur with bladder extrophy. In this rare birth defect, the bladder is inside out and sticks through the abdomen wall. Epispadias can also occur with other birth defects. Epispadias occurs in one out of every 117,000 newborn boys and one in 484,000 newborn girls. The condition is usually diagnosed at birth or soon afterwards.

Males usually have a short, wide penis with an abnormal curve. The urethra usually opens on the top or side of the penis instead of the tip. However, the urethra may be open along the whole length of the penis.

Signs include abnormal opening from the bladder neck to the area above the normal urethra opening, backward flow of urine into the kidney (reflux nephropathy), urinary incontinence, urinary tract infections and widened pubic bone.

Patients who have more than a mild case of epispadias will need surgery. Leakage of urine (incontinence) can often be repaired at the same time. However, second surgery may be needed in some cases.

CLINICAL CASES

Case 1

A 21-year-old male presented with abnormal urethral opening over the dorsum of the penis. He was continent with no operative intervention in past. Examination revealed short, wide penis with abnormal opening of the urethra at the dorsum of the penis (Fig. 2.5.1). There was no chordee. Double layered urethroplasty was done.

Case 2

A 24-year-old male presented with abnormal urethral opening over the dorsum of the penis. Examination revealed similar finding as in Case 1 (Fig. 2.5.2). Urethral reconstruction was done.
Case 3

A 26-year-old male presented with ectopia vesicae with epispadias since birth. First stage closure of ectopia vesicae was done during childhood. He presented with short penis and abnormal curvature towards abdomen during erection. He also had stress incontinence. Examination revealed short, wide penis with ventral chordee (Figs 2.5.3 and 2.5.4). Young-Dees-Leadbetter bladder neck reconstruction and urethral reconstruction was done. Figure 2.5.5 shows clinical picture after surgery.
Follow-up

His incontinence improved and had straight erect penis, but size remained short.

COMMENTS

Incomplete epispidias has good outcome after surgery, whereas in the complete variety of epispidias, the outcome may not be satisfactory.

BIBLIOGRAPHY

INTRODUCTION

Urethral duplication is a rare congenital anomaly with less than 200 cases reported. It predominantly occurs in males and is nearly always diagnosed in childhood or adolescence. It is defined as a complete second passage from the bladder to the dorsum of the penis or as an accessory pathway that ends blindly on the dorsal or ventral surface.

Incomplete duplication is more common than complete duplication.

CLINICAL CASES

Case 1

A 26-year-old male presented with the history of abnormal opening at the base of the penis. Examination revealed abnormal opening at the base of the penis with normal urethral meatus (Fig. 2.6.1). The abnormal opening was blind ending as ureteric catheter could not be passed. Micturating cystourethrogram revealed normal ventral urethra and blind ending dorsal urethra (Fig. 2.6.2).

Treatment

Surgical excision of the blind ending urethra was done.

Fig. 2.6.1: Case 1—Clinical picture
Case 2

A 62-year-old male presented with lower urinary tract symptoms (LUTS) and investigations confirmed benign enlargement of prostate. Examination revealed double meatal opening (Fig. 2.6.3). Ventral one was the normal urethra and dorsal one was ectopic blind ending urethra.
**Treatment**

Patients was counseled that no treatment is required.

**COMMENTS**

Incomplete duplication of urethra is a rare anomaly. It can be diagnosed easily and can be managed surgically.

**BIBLIOGRAPHY**

INTRODUCTION

Megalourethra is a rare congenital penile anomaly. It is caused by an embryogenic failure of the mesenchyme to differentiate into corpora cavernosa or corpus spongiosum. Two types of megalourethra have been described. The first type, scaphoid megalourethra, in which the corpora spongiosum is absent but corpora cavernosa is well developed. During voiding, the penis swells up locally to become boat shaped with a dorsal chordee. The second type, fusiform megalourethra, in which corpora cavernosa are absent and the spongiosum is deficient or absent.

CLINICAL CASES

Case 1

A 3-month-old male child, born at full term normal delivery. He was the first child and his mother had noticed a large size penis which swelled during voiding. Local examination of the penis revealed that the ventral side of penis was flattened and the skin was wrinkled and redundant (Fig. 2.7.1). Micturating cystourethrogram (MCU) (Fig. 2.7.2) and retrograde urethrogram (Fig. 2.7.3) revealed distal urethra filled with contrast medium, causing a localized scaphoid appearance.

Treatment

Imbricated urethral tube repair was done.
Case 2

A newborn, premature, male child presented with diffuse swelling of the penis (Fig. 2.7.4). Unfortunately, child could not survive due to other problems.

Fig. 2.7.4: Case 2—Clinical picture

**Comments**

Megalourethra is a rare congenital condition and can be managed surgically.

**Bibliography**

2.8 POSTERIOR URETHRAL VALVE

INTRODUCTION

Posterior urethral valve is an obstructing membrane that radiates in a distal direction towards the membranous urethra anteriorly. This is classical type II valve which is the most common variety.

Posterior urethral valve (PUV) can present in a variety of ways depending on the degree of obstruction and presenting symptoms are age dependent. In newborns, it can present with retention of urine, urinary tract infections (UTI) and deranged renal function. In later age group, it can present with lower urinary tract symptoms.

Ultrasound examination reveals bladder hypertrophy and dilatation of upper urinary tracts. Micturating cystogram suggests the diagnosis which is confirmed by endoscopy and the treatment of valves is done endoscopically. Diathermy or laser incision of valves is recommended. In patients with deranged renal function, initial diversion of urine is recommended followed by valve treatment. Follow-up is important to check upper tract deterioration or infection, and for bladder hypertrophy which is known as valve bladder.

CASE 1

Clinical Presentation

A 7-year-old boy, asymptomatic up to the age of 5 years, presented with history of incontinence for the past 2 years both during day and night. Normal voiding in between, normal bowel habits and no history of bowel incontinence. General and neurological examination was normal.

Investigations

Blood urea (BU): 26 mg/dL, creatinine: 0.6 mg/dL, Na: 141 mEq/dL, K: 4.4 mEq/dL urine microscopy: Normal; Urine culture: Sterile. Ultrasound revealed normal upper tracts with thick walled bladder with significant residual urine. Micturating cystourethrogram (MCU) (Fig. 2.8.1) revealed large capacity bladder with dilated posterior urethra with secondary bladder neck hypertrophy. No vesicoureteral reflux (VUR) noted.

Treatment

Cystoscopy confirmed the presence of type 2 posterior urethral valves. Laser incision of the valve was done at 5 and 7 o'clock position. The patient improved postoperatively and is under follow-up.
CASE 2

A 10-year-old boy presented with recurrent UTI. Micturating cystourethrogram revealed features of posterior urethral valve with vesicoureteric reflux (Fig. 2.8.2). Endoscopic incision of valves was done.
**COMMENTS**

Posterior urethral valve is a common problem in children. Any male child presenting with lower urinary tract symptoms and infection should be investigated for the presence of posterior urethral valves. If found, should be treated and followed up lifelong.

**BIBLIOGRAPHY**

2.9 PARAMEATAL CYST

INTRODUCTION

Parameatal cyst may rarely form along with median raphe. These cysts result from occlusion of the paraurethral ducts and are usually diagnosed after eversion of the prepuce. These cysts can be treated by simple excision.

CLINICAL PRESENTATION

A 9-year-old male presented with abnormal swelling protruding out from the meatus (Fig. 2.9.1). Examination revealed about 7-8 mm cystic lesion at the meatus. Surgical excision was done. Histopathology revealed a cyst lined by simple cuboidal to columnar epithelium.

Fig. 2.9.1: Clinical picture

COMMENTS

Parameatal cyst is a rare condition and can be managed surgically.

BIBLIOGRAPHY

2.10 PARAURETERIC BLADDER DIVERTICULUM

**INTRODUCTION**

Bladder diverticula are protrusions of the mucosal and submucosal layers of the bladder through the muscular wall of the bladder. Congenital or true diverticula involve all layers of the structure, including muscularis propria and adventitia. Acquired or false diverticula do not involve muscular layers or adventitia.

**CLINICAL PRESENTATION**

A 15-year-old male presented with difficulty in passing urine. Clinical examination was normal.

**INVESTIGATIONS**

Intravenous urogram revealed left paraureteric diverticulum with normal functioning kidney (Fig. 2.10.1). Micturating cystourethrogram (MCU) confirmed diverticulum with no reflux (Fig. 2.10.2). Cystopanendoscopy confirmed the presence of paraureteric diverticulum on left side.

**TREATMENT**

Diverticulectomy was done. Patient’s symptoms improved.

![Intravenous urogram](image)
**COMMENTS**

Bladder diverticulum can be congenital or acquired due to lower urinary tract obstruction. Congenital diverticulum is asymptomatic, and can be kept under observation. Bladder diverticulum with obstruction needs endoscopic management by laser incisions.

**BIBLIOGRAPHY**

2.11 UNDESCENDED TESTIS

INTRODUCTION

Undescended testicle occurs when one or both testicles fail to move into the scrotum before birth. Generally, testicles descend by the time the child is 9 months old. Undescended testicles are fairly common in infants who are born early (premature infants). It occurs in about 3–4% of full-term infants.

Absence of testis in the scrotum can be due to undescended testis, congenital absence of the testis, ectopic testis and also retractile testis. Testicles that do not descend by the time the child is 1 year old should be evaluated. Studies suggest that surgery should be done by this age. Surgery can reduce the chances of permanent damage to the testicles, which can lead to fertility problems later in life. Bringing the testicle into the scrotum can improve sperm production and increases the odds of good fertility. It also allows the health care provider to examine the testicle to detect cancer early.

There are usually no symptoms, except that the testicle cannot be found in the scrotum (this may be described as an empty scrotum).

An examination confirms that one or both of the testicles are not in the scrotum. Imaging tests, such as an ultrasound or CT scan/MRI may be done for localization of the testis. Diagnostic laparoscopy is advisable which can confirm the presence of testis and spermatic vessels.

TREATMENT

Usually the testicle will descend into the scrotum without treatment during the child’s first year of life. If this does not occur, the child may get hormone injections (beta-hCG or testosterone) to try to bring the testicle into the scrotum.

Surgery (orchidopexy) to bring the testicle into the scrotum is the main treatment. Having orchidopexy early may prevent damage to the testicles that can cause infertility.

CLINICAL PRESENTATION

A 9-year-old male presented with absence of testis in the right side of the scrotum since birth. On examination, scrotum was empty on the right side (Fig. 2.11.1). Left testis was normal.
INVESTIGATIONS

Ultrasound revealed right testis at the right internal inguinal ring.

TREATMENT

Diagnostic laparoscopy confirmed the presence of spermatic vessels entering into inguinal ring. Mobilization of the spermatic vessels and right orchidopexy was done. Testis could be brought up to upper part of the scrotum.

COMMENTS

It is important that at the time of birth, presence of testis in the scrotum should be documented. If absent, periodic review is important and by the age of 12 months, investigations and treatment should be done for proper spermatogenesis and prevention of cancer.

BIBLIOGRAPHY

2.12 PENOSCROTAL TRANSPOSITION

INTRODUCTION

Penoscrotal transposition (PST) is a rare anomaly resulting in a partial or complete positional exchange between the penis and the scrotum. Its prevalence at birth is less than one in 10,000 and it is associated with hypospadias and chordee. It is due to a defect in caudal migration of the inchoate scrotum during the intrauterine life. Surgical repair of PST relies on the creation of rotational flaps to mobilize the scrotum, or on transposing the penis to its normal anatomical location.

CLINICAL PRESENTATION

A 50-year-old male presented with abnormal position of scrotum. He was married and had no problem during intercourse. On examination, partial penoscrotal transposition was noted (Fig. 2.12.1). The external genitalia were normal. He was counseled for his condition and no surgical treatment was done.

COMMENTS

Penoscrotal transposition is a rare condition and according to deformity, surgical correction can be done.

BIBLIOGRAPHY

2.13 WEBBED PENIS

**INTRODUCTION**

Webbed penis is a condition in which scrotal skin extends onto the ventrum of the penis. When this is congenital, underlying urethra and scrotum are normal. It can be acquired if excessive skin from ventral side is removed. Webbed penis is usually asymptomatic, but if the cosmetic appearance is not acceptable then surgical correction can be done.

**CLINICAL PRESENTATION**

A 28-year-old male presented with the complaints of covered penis under scrotum. On examination, small penis was found covered by large scrotum ventrally (Fig. 2.13.1). Patient was counseled for management but he decided against any surgical management.

![Fig. 2.13.1: Clinical picture](image)

**COMMENTS**

Webbed penis is a rare condition and should be managed according to merit.

**BIBLIOGRAPHY**

3.1 ADRENAL PHEOCHROMOCYTOMA

INTRODUCTION

Pheochromocytoma is a rare tumor of the adrenal gland that can release high levels of epinephrine and norepinephrine. Pheochromocytoma is a tumor of catecholamine-secreting cells which are mainly present in adrenal medulla, but they are found in other areas of the body also. Pheochromocytoma has three classic symptoms: headache, sweating and heart palpitations in association with markedly elevated blood pressure. Biochemical investigations of elevated catecholamines in 24-hour urinary samples confirm the diagnosis. Imaging like contrast enhanced computed tomography (CECT) and magnetic resonance imaging (MRI) localize the tumor. Surgical excision is the treatment of choice and can be done by open surgery, laparoscopically or robotically.

CLINICAL PRESENTATION

A 35-year-old male presented with complaints of palpitations for 1 year as well as giddiness for 1 year. He has history of hypertension for 1 year and was on 3 different antihypertensive drugs. He was a known case of diabetes, for which he was taking insulin.

INVESTIGATIONS


CT scan: $4.8 \times 6.8$ cm mass lesion left adrenal with cortical necrosis (Fig. 3.1.1), MRI: $4.6 \times 6.7$ cm left adrenal mass lesion-T2 weighted—bright bulb appearance (Fig. 3.1.2).
Fig. 3.1.1: Computed tomography scan

Fig. 3.1.2: Magnetic resonance imaging
TREATMENT

Left-sided robotic adrenalectomy was done and a 7 × 7 cm left adrenal tumor was removed.

HISTOPATHOLOGY

Pheochromocytoma with no capsular and vascular invasion; margins were negative.

FOLLOW-UP

Normal blood pressure, MIBG and urinary catecholamine were normal.

COMMENTS

Pheochromocytoma has a classical presentation and should be suspected and treated as it is a correctable cause of hypertension.

BIBLIOGRAPHY

3.2 ADRENAL ADENOMA

**INTRODUCTION**

Adrenal adenoma is a benign tumor of the glandular type (adenoma) in the adrenal gland. While some adrenal adenomas do not secrete hormones at all (nonfunctional, often diagnosed incidentally as incidentalomas), some secrete cortisol (causing Cushing’s syndrome), aldosterone (causing Conn’s syndrome) or androgens (causing hyperandrogenism). It commonly occurs in adults, but it can be found in persons of any age. Adrenal cortical adenomas are not considered to have the potential for malignant transformation. The treatment for a hormonally active (functional) adrenal tumor is surgery. Nonfunctional adrenal cortical adenomas are not premalignant, and surgical excision is not indicated.

**CLINICAL PRESENTATION**

A 53-year-old male presented with abdominal discomfort off and on for 1 year. He had a history of hypertension for last 8 years and a history of abdominal discomfort for which he was investigated in 2002. He took anti tubercular treatment for 9 months for the same.

**EXAMINATION**

Pulse rate: 84/min, regular rhythm, good volume; BP: 160/96 mm Hg (supine), 156/94 mm Hg (supine); Per abdominal examination—Normal.

**INVESTIGATIONS**

Urine R/M: Normal; Urine C/S: Sterile; Renal function tests: within normal limits, Serum Na: 135 mEq/dL, Serum K: 3.5 mEq/dL; 24 hour urinary VMA: 12 mg (Normal: 1.5–6.5); 24 hour urinary catecholamines: 225 µg (0–275); 24 hour urinary metanephrine and normetanephrine: 0.6 mg (Normal: 0–0.9); serum cortisol (8 am): 17 µg/dL (normal—up to 24 µg/dL); Overnight dexamethasone suppression test: Negative; Serum cortisol (8 am): 7.47 µg/dL, (4 pm): 5.63 µg/dL.

Ultrasonography abdomen: right adrenal mass—2 × 2.5 cm (Fig. 3.2.1); Contrast enhanced computed tomography (CECT): enhancing right adrenal mass (Figs 3.2.2 and Figs 3.2.3A and B); Magnetic resonance imaging (MRI): right adrenal mass hypoechoic both in T1 and T2 (Figs 3.2.4 and 3.2.5).
Fig. 3.2.1: Ultrasound

Fig. 3.2.2: Noncontrast computed tomography (NCCT) scan
**TREATMENT**

Laparoscopic adrenalectomy was done.

**HISTOPATHOLOGY**

Adrenal cortical adenoma.
COMMENTS

Adrenal adenoma can be detected incidentally during imaging or as a part of investigation for hypertension. They can be functional or nonfunctional. Functional tumor can be removed surgically by laparoscopy or robotically.

BIBLIOGRAPHY

3.3 ADRENAL CORTICAL ADENOMA MIMICKING PHEOCHROMOCYTOMA

**INTRODUCTION**

Adrenal cortical adenoma can have biochemical evidence of raised catecholamines and can mimic pheochromocytoma. The clinical findings may be mediated by the presence of neuroendocrine features in these tumors.

**CLINICAL PRESENTATION**

A 28-year-old male presented with right upper abdominal discomfort off and on for 2 months, headache for 2 months and no urinary complaints.

**PAST HISTORY**

Hypertension for 2 months (not on regular medication) and history of occasional palpitation for 2 months.

**FAMILY HISTORY**

Father, mother—history of hypertension for 10 years.

**EXAMINATION**

Pulse rate: 84/min, regular rhythm, good volume; BP: 160/96 mm Hg (supine), 156/94 mm Hg (supine); Per Adomen: Normal; Per rectum: grade 1 prostate, smooth, firm, nontender.

**INVESTIGATIONS**

Urine R/M: Normal; Urine C/S: Sterile; RFT: within normal limits, Serum Na: 135 mEq/dL, Serum K: 3.5 mEq/dL; CECT: adrenal mass 5 × 7 cm (Fig. 3.3.1); MRI T1: hypoechoic adrenal mass (Fig. 3.3.2), MRI T2: bright bulb appearance (Figs 3.3.3A and B).
Fig. 3.3.1: Computed tomography scan

Fig. 3.3.2: MRI T1 images
TREATMENT
Laparoscopic right adrenalectomy was done.

HISTOPATHOLOGY
Histopathology showed adrenal cortical adenoma.

COMMENTs
If features of pheochromocytoma are present then these cases should be prepared and operated as pheochromocytoma. Histopathology will clarify the nature of the tumor.

BIBLIOGRAPHY
3.4 ADRENAL CYST

**INTRODUCTION**

Adrenal cysts are rare lesions that usually present as an incidental finding during surgery or at the time of autopsy. The cysts are usually small, seldom exceeding 10 cm in diameter, and are generally asymptomatic. In cases which are symptomatic, radiographic examination is the most important tool in establishing a correct preoperative diagnosis. Of the four main categories, endothelial cysts represent the most common type of adrenal cyst. Observation for asymptomatic cases and simple surgical enucleation of the cyst with preservation of the remaining adrenal gland is the treatment of choice in other cases.

**CLINICAL PRESENTATION**

A 65-year-old male presented with vague abdominal pain.

**INVESTIGATIONS**

Ultrasound revealed a right renal cyst. Contrast enhanced computed tomography (CECT) (Fig. 3.4.1) and MRI revealed a hypoechoic right renal cyst. Biochemical investigations including catecholamines were normal.

![Contrast enhanced computed tomography (CECT) scan](Image)

*Fig. 3.4.1: Contrast enhanced computed tomography (CECT) scan*
**TREATMENT**

Counseled and agreed for no surgical intervention.

**COMMENTS**

Adrenal cyst can be detected during imaging of abdomen for abdominal pain. If symptomatic, surgical excision is advised.

**BIBLIOGRAPHY**

3.5 CONN’S SYNDROME

■ INTRODUCTION

Primary aldosteronism is usually diagnosed after a battery of biochemical tests and adrenal imaging. A case of hypertension with persistent hypokalemia needs investigation for Conn’s syndrome. If an adrenal tumor is identified, surgery is indicated.

■ CLINICAL PRESENTATION

A 53-year-old female presented with weakness for 3–4 months duration. There is no history of hypertension.

■ INVESTIGATIONS

Investigations revealed persistent low serum potassium—2.8 mEq/dL. Magnetic resonance imaging revealed small tumor in left adrenal gland (Fig. 3.5.1). Urinary catecholamines were normal.

![MRI showing left adrenal tumor](image)

Fig. 3.5.1: MRI showing left adrenal tumor

■ TREATMENT

Laparoscopic left adrenalectomy was done.

■ HISTOPATHOLOGY

Histopathology confirmed cortical adenoma.

■ FOLLOW-UP

Serum potassium level returned to normal range. Symptoms improved.

■ COMMENTS

Cortical adenoma with Conn’s syndrome is a surgically correctable condition.

■ BIBLIOGRAPHY

Adrenal Gland

3.6 CASTLEMAN’S DISEASE MIMICKING ADRENAL TUMOR

INTRODUCTION
Castleman’s disease also known as giant lymph node hyperplasia and angiofollicular lymph node hyperplasia, is a disease of lymph nodes and related tissues. Castleman’s disease is a rare disorder characterized by benign proliferation of lymphoid tissues. Most cases occur as mediastinal masses, although extrathoracic involvement including nodal and extranodal locations has been reported. The pararenal location of localized disease may sometimes be mistaken for adrenal or renal tumor.

There are two forms of Castleman’s disease: localized and multicentric. Castleman’s disease can also be classified on the basis of how the lymph node tissue appears on histopathology. These are called microscopic subtypes. These subtypes are hyaline vascular, plasma cell and mixed subtypes.

CLINICAL PRESENTATION
A 25-year-old man underwent abdominal ultrasonography for vague upper gastrointestinal tract symptoms and was found to have a 6–8 cm mass in the right suprarenal area.

INVESTIGATIONS
Computed tomography (CT) revealed a calcified lesion in the region of the adrenal gland, and the latter was not separately identifiable (Figs 3.6.1 and 3.6.2). Pulse rate and blood pressure were normal. Renal function tests, serum cortisol, electrolytes and urinary catecholamines were all normal. Preliminary diagnosis was nonfunctioning adrenal tumor.

Fig. 3.6.1: Contrast enhanced computed tomography (CECT) abdomen—transverse films
**TREATMENT**

The patient underwent exploration through the bed of the 11th rib via an extrapleural transperitoneal approach. There were two lobulated masses in the suprarenal area which were carefully dissected and separated from the inferior vena cava and excised.

Histopathological analysis demonstrated Castleman’s disease (hyaline vascular type) and the adrenal gland itself was histologically unremarkable (Fig. 3.6.3).
COMMENTS

Castleman’s disease, if present in adrenal area can be misdiagnosed for adrenal tumors.

BIBLIOGRAPHY

3.7 MULTIPLE PHEOCHROMOCYTOMA

INTRODUCTION

In 15–20% of the cases pheochromocytoma localizes in extra-adrenal sites, and in about 15% of all cases it seems to be multiple. Ectopic, associated with familial syndromes and multiple pheochromocytomas are not uncommon and although recovery in surgically treated patients is excellent, lifelong follow-up is necessary.

CLINICAL PRESENTATION

A 42-year-old male presented with paroxysmal hypertension of 1 year duration.

EXAMINATION

Blood pressure: 160/100 mm Hg. Abdominal examination: no lump palpable.

INVESTIGATIONS

Investigations revealed raised 24-hour urinary catecholamines. Contrast enhanced computed tomography (CECT) abdomen revealed bilateral adrenal tumors (Figs 3.7.1A and B).

TREATMENT

Abdominal exploration with a large midline incision was done. Bilateral adrenalectomy was done. On the left side, tumor could not be separated from the kidney, so left kidney was also removed with left pheochromocytoma. After removal of tumors, his blood pressure was still raised. Further abdominal exploration revealed another tumor at the organ of Zuckerkandl, which was
Adrenal Gland

excised following which blood pressure returned to normal. Figures 3.7.2A and B show removed specimen of three tumors with cut section. Histopathology confirmed pheochromocytoma in all the three tumors.

**FOLLOW-UP**

Blood pressure returned to normal with normal catecholamine levels postoperatively.

**COMMENTS**

With better imaging facilities, multiple pheochromocytomas can be diagnosed preoperatively. Otherwise after removal of pheochromocytoma, if blood pressure do not drop then search for extra-adrenal pheochromocytoma is necessary.

**BIBLIOGRAPHY**

3.8 PHEOCHROMOCYTOMA INVOLVING RIGHT RENAL ARTERY

**INTRODUCTION**

Pheochromocytoma can be associated with renal artery stenosis or if pheochromocytoma is large and close to renal hilum it can involve renal artery.

**CLINICAL PRESENTATION**

A 43-year-old male presented with hypertension for 3 years duration. Clinical examination was normal. 24 hours catecholamines were raised.

**INVESTIGATIONS**

Contrast enhanced computed tomography (CECT) revealed enhancing right adrenal tumor which encroached around right renal pedicle (Figs 3.8.1A and B).

**TREATMENT**

Right adrenalectomy was done along with right nephrectomy as the tumor could not be separated from the renal pedicle. Intraoperative picture shows the tumor (Fig. 3.8.2). Specimen picture shows the adrenal tumor with the right kidney (Fig. 3.8.3).

**FOLLOW-UP**

Blood pressure was normal with normal catecholamine levels.

**COMMENTS**

Large adrenal tumors can involve renal artery and it is difficult to separate them. In such a situation, nephrectomy may be required along with removal of pheochromocytoma.

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**Figs 3.8.1A and B:** CECT with CT angiography showing right adrenal tumor encroaching right renal pedicle
Fig. 3.8.2: Intraoperative picture showing adrenal tumor

Fig. 3.8.3: Operative specimen of the adrenal tumor and right kidney

BIBLIOGRAPHY

3.9 PHEOCHROMOCYTOMA WITH CALCIFICATION

**INTRODUCTION**

Pheochromocytoma can be associated with calcification. The significance of calcification is not known; however, surgical removal of pheochromocytoma controls hypertension.

**CASE 1**

**Clinical Presentation**

A 45-year-old male presented with hypertension. Investigations revealed raised 24-hour urinary catecholamine levels.

**Investigations**

Contrast enhanced computed tomography shows right adrenal tumor with calcification (Fig. 3.9.1).

**Treatment**

Right adrenalectomy was done.

**Histopathology**

Histopathology confirmed pheochromocytoma.

![Fig. 3.9.1: Case 1—CECT showing right adrenal tumor with calcification](image)
CASE 2

Clinical Presentation

A 39-year-old female presented with hypertension.

Investigations

Ultrasound revealed left adrenal tumor with calcification (Fig. 3.9.2). Contrast enhanced computed tomography also confirmed enhancing adrenal tumor with calcification (Fig. 3.9.3). 24-hour urinary catecholamines were raised.

Fig. 3.9.2: Case 2—Ultrasound showing left adrenal tumor with calcification

Fig. 3.9.3: Case 2—CECT showing left adrenal tumor with calcification
Treatment
Left adrenalectomy was done.

Histopathology
Histopathology confirmed pheochromocytoma.

COMMENTS
Pheochromocytoma with calcification needs the same treatment.

BIBLIOGRAPHY
3.10 RECURRENT PHEOCHROMOCYTOMA

**INTRODUCTION**

Recurrence following adrenalectomy for a pheochromocytoma is rare and is seen in 6.5–8.5% of cases. Recurrence of pheochromocytoma may result from the development of a metachronous primary tumor in the retained adrenal or extra-adrenal paraganglionic tissue, intraoperative disruption of the original tumor with secondary implantation and the development of metastatic disease.

**CLINICAL PRESENTATION**

A 22-year-old unmarried female presented with persistent headache, palpitation, sweating and hypertension since the age of 12 years. She was evaluated outside for adrenal pheochromocytoma. Right adrenalectomy was done in 2005. It was a difficult surgery and associated with significant bleeding for which she was re-explored. Symptoms persisted post surgery.

**EXAMINATION**

Pulse rate: 108/min, Blood pressure: 110/70 mm Hg (sitting), 118/78 mm Hg (supine).

Abdominal examination—hypertrophic scar of previous surgery noted in the right subcostal and thoracic region (Fig. 3.10.1).

**INVESTIGATIONS**

24-hour urinary catecholamines: 46 µg (normal upto 30 µg). Contrast enhanced computed tomography of the abdomen revealed a mass in the region of adrenal with clips of previous surgery, and mass extending to interaortocaval region with right small kidney (Fig. 3.10.2). MRI revealed enhancing tumor in T2 images in interaortocaval region (Figs 3.10.3 to 3.10.5). MIBG scan revealed enhancing tumor in upper abdomen in aortocaval region (Fig. 3.10.6).

**TREATMENT**

Mercedes Benz incision with excision of scar (Fig. 3.10.7), right nephrectomy with excision of tumor, infrahilar followed by suprahilar dissection (Figs 3.10.8A to C). Excised tumor with right kidney (Fig. 3.10.9).

**HISTOPATHOLOGY**

Polygonal cells suggestive of pheochromocytoma, margins were free.

**FOLLOW-UP**

Blood pressure was normal, 24-hour urinary catecholamines were normal.
Fig. 3.10.1: Previous surgery scar

Fig. 3.10.2: Contrast enhanced computed tomography of the abdomen
Fig. 3.10.3: MRI abdomen—sagittal sections

Fig. 3.10.4: MRI abdomen—coronal section
Fig. 3.10.5: MRI abdomen—sagittal section

Fig. 3.10.6: MIBG scan

Fig. 3.10.7: Incision

Figs 3.10.8A to C: Intraoperative pictures: (A) Showing tumor (B) Dissection of vessels (C) After removal of tumor
The recurrent pheochromocytomas are not malignant in nature, but they have considerable malignant tendency and should undergo intensive surveillance and regular examinations, especially for the primary or recurrent extra-adrenal pheochromocytomas.

**BIBLIOGRAPHY**

4.1 ANGIOMYOLIPOMA

**INTRODUCTION**

Angiomyolipoma (AML) is the most common benign tumor of the kidney. It is composed of fat, smooth muscle and abnormal blood vessels. AML can be diagnosed by identifying the intratumoral fat component, which shows negative attenuation on noncontrast computed tomography (CT) scans. First-line therapy includes renal preserving surgery or angioembolization (RAE) both with good outcomes.

**CASE 1**

**Clinical Presentation**

A 28-year-old female presented with sudden onset of pain in the epigastrium and left loin. It was not associated with dysuria, hematuria or lithuria, weight loss, anorexia or bowel symptoms.

No history of diabetes mellitus (DM), hypertension (HT), neurologic symptoms or surgical intervention in past. No family history of renal disease.

**Examination**

Face showed features of tuberous sclerosis (Fig. 4.1.1), Blood pressure (BP): 130/80 mm Hg, no pallor, edema or lymphadenopathy. Bilateral palpable kidneys on abdominal examination.

**Investigations**

Hemoglobin: 12 mg/dL, Urea: 20 mg/dL, Creatinine: 1.2 mg/dL, Urine R/M: NAD, Urine culture: Sterile. Contrast enhanced computed tomography (CECT) showing bilateral angiomyolipoma (Fig. 4.1.2); CECT head showed subependymal nodules suggestive of tuberous sclerosis; Renal dynamic scan split GFR: right kidney—68%, left kidney—32%.
Fig. 4.1.1: Case 1—Clinical picture

Fig. 4.1.2: Case 1—Contrast enhanced computed tomography (CECT) of the abdomen
**Treatment**

Patient was counseled and treatment options discussed.

**CASE 2**

A 69-year-old male, follow-up case of post-transurethral resection of the prostate (TURP), complaints of pain left flank. Abdominal examination was normal. Intravenous pyelogram (IVP) showed parapelvic mass with lateral displacement of pelvicalyceal system (Fig. 4.1.3). CECT confirmed angiomyolipoma involving parapelvic region and displacing pelvicalyceal system (PCS) (Figs 4.1.4A and B).

![Fig. 4.1.3: Case 2—Intravenous urogram](image1)

![Figs 4.1.4A and B: Case 2—Contrast enhanced computed tomography (CECT) of the abdomen](image2)
**CASE 3**

Another case, CECT showing bilateral AML with left small kidney (Fig. 4.1.5).

![Image of CECT showing bilateral AML with left small kidney](image)

**Fig. 4.1.5:** Case 3—Contrast enhanced computed tomography (CECT) of the abdomen

**CASE 4**

Another case, CECT showing left AML with small kidney (Figs 4.1.6A and B).
Angiomyolipoma can be diagnosed by typical features on imaging. Management depends upon the symptoms and size of the tumor.

**BIBLIOGRAPHY**

INTRODUCTION

Tumors of renal pelvis and ureter are rare but clinically important neoplasms. Urothelial tumors are the commonest followed by squamous cell carcinoma and adenocarcinoma. High-grade and unusual morphology as well as advanced tumor stage are the frequent findings in pelvicalyceal urothelial carcinomas. In addition, divergent morphology could be present and represent advanced tumor stage.

CLINICAL PRESENTATION

A 65-year-old male presented with history of hypertension for 10 years, type II diabetes mellitus for 25 years, chronic obstructive pulmonary disease (COPD) for 10 years and chronic renal failure (CRF) for 2 years. He had cardiac arrest in March 2013, due to hyperkalemia. He was revived and treated outside. During evaluation, ultrasonography (USG) was done which revealed left renal mass. A noncontrast MRI was done due to raised creatinine.

EXAMINATION

He was conscious, oriented in time, place and person. No pallor, cyanosis, icterus or lymphadenopathy. Pulse rate: 80/min, BP: 120/70 mm Hg, respiratory rate (RR): 22/min. Chest: Bilateral rhonchi. Per abdominal examination: soft, bowel sounds present. No organomegaly, no tenderness.

INVESTIGATIONS


Magnetic resonance imaging revealed left renal mass lesion involving adjacent descending colon and perinephric fat (size 5.8 × 5.5 × 7.8 cm) (Figs 4.2.1A to C). Dynamic renal scan revealed bilateral poor function with total glomerular filtration rate (GFR) of 62 mL/min (Fig. 4.2.2).

TREATMENT

Explored through 11th rib bed incision; findings—left renal mass 6 × 6 cm size, involving distal pancreas, spleen, greater curvature of stomach and descending colon, adherent to the diaphragm. Left radical nephrectomy with partial gastrectomy, splenectomy, distal pancreatectomy and descending colonic end to end resection anastomosis was done. (Figs 4.2.3A to C). Histopathology revealed high grade urothelial carcinoma with divergent differentiation with squamous and glandular components (Figs 4.2.4A to C). Pathological stage was pT4 pNx pMx. Perineural invasion was present. Tumor invades adjacent organs, i.e. stomach, pancreas, large intestine, spleen and left adrenal gland. Resection margins were free of tumor. Advanced diabetic changes in native renal parenchyma were noted.
Figs 4.2.1A to C: Magnetic resonance imaging (MRI)
Fig. 4.2.2: Dynamic renal scan

Figs 4.2.3A to C: Operative specimen
Postoperative period was uneventful. Serum creatinine rose up to 3.82 mg/dL. Postoperative dynamic renal scan revealed GFR—25.31 mL/min, right kidney normal in size, shape and location. Perfusion and cortical uptake mildly reduced impaired glomerular function with fair drainage. In view of CRF, adjuvant chemotherapy could not be given. In one year follow up, patient is doing well.

**COMMENTS**

Clinical presentation, diagnosis and management of tumors of renal pelvis are challenging and finally it is histopathology that clinches the diagnosis. These tumors are more aggressive and need multimodality treatment.

**BIBLIOGRAPHY**

4.3 HYDATID CYST OF KIDNEY

**INTRODUCTION**

Hydatid disease (HD) is caused by the cestode *Echinococcus granulosus* mainly involving the liver and the lung. Kidney involvement is rare in hydatid disease and constitutes only 2–4% of all cases.

The treatment of hydatid cyst of the kidney is surgical. Renal preserving surgery, hydatid cystectomy plus pericystectomy are done in most cases. Nephrectomy is reserved for destroyed kidneys resulting from aged cysts opening into the excretory cavities and complicated by renal infection. Pretreatment with albendazole alone or in combination with praziquantel is very important as the cyst material becomes non-antigenic, cyst tension is reduced and thus reducing the risk of spillage. Postoperatively, albendazole has shown to reduce the risk of implantation of scolices.

**CLINICAL PRESENTATION**

A 32-year-old female presented with complaints of lump right flank for 6 months and right flank pain for 4 months. Lump was of insidious onset, gradually increasing in size. No history of hematuria, lithuria, trauma, no comorbid illnesses.

**EXAMINATION**

Pulse: 80/min, BP: 124/86 mm Hg, Chest/Cardiovascular system: No abnormality detected (NAD). Abdominal examination revealed firm 10 × 15 cm nontender mass in right lumbar region, well-defined margins, moving with respiration, bimanually palpable. Dull on percussion.

**INVESTIGATIONS**


Ultrasoundography of the abdomen (Fig. 4.3.1): left kidney—normal, enlarged right kidney with cyst showing daughter cysts. Intravenous urogram (Fig. 4.3.2): right superior calyx visualized, middle and inferior calyx not seen, LK: Normal; CECT abdomen (Figs 4.3.3A and B): left kidney—normal, right kidney: large multicystic lesion 11.3 × 9.8 cm indenting superior pole. IgG-Echinococcus: 9.2 (Normal: 8.00)—borderline.
Fig. 4.3.1: Ultrasound

Fig. 4.3.2: Intravenous urogram—supine and prone
Figs 4.3.3A and B: Contrast enhanced computed tomography (CECT) scan
**TREATMENT**

Right retrograde pyelography (RGP) and stenting, and right open partial nephrectomy were done.

**COMMENTS**

Hydatid disease of the kidney is very rare and imaging helps in the diagnosis. Renal conservative surgery is possible if the disease is localized to on pole of the kidney.

**BIBLIOGRAPHY**

4.4 INCIDENTAL LARGE RENAL TUMORS

**INTRODUCTION**

Presentation of renal tumors is very variable. With the availability of imaging, about 50% of these tumors are detected incidentally. These tumors are small and have good prognosis. The classical triad of presentation of renal tumors pain, hematuria and abdominal lump is seen in about 20% cases. There are cases which present with large renal tumors without any symptoms and in absence of imaging, are diagnosed late.

**CLINICAL CASES**

**Case 1**

A 35-year-old male presented with abdominal swelling. On examination, a large left renal tumor was palpable. Contrast enhanced computed tomography (CECT) revealed a large left renal tumor with variegated appearance (Fig. 4.4.1). Left radical nephrectomy was done. Histopathology confirmed renal cell carcinoma (RCC).

![Case 1—Contrast enhanced computed tomography (CECT) scan](image)
Case 2

A 39-year-old male presented with abdominal swelling. On examination, a large right renal tumor was palpable. CECT revealed a large right renal tumor with variegated appearance and also displacing vena cava and aorta (Figs 4.4.2A and B). Right radical nephrectomy was done. Histopathology confirmed RCC.

Figs 4.4.2A and B: Case 2—Contrast enhanced computed tomography (CECT) scan

 COMMENTS

Due to lack of awareness and absence of imaging facilities, cases of renal tumors can present late with large in size. However, with change of time, big size tumors are seen less and less.

 BIBLIOGRAPHY

4.5 INCIDENTAL RENAL CELL CARCINOMA WITH ADRENAL HYPERPLASIA

INTRODUCTION

Renal cell carcinoma (RCC) can be associated with adrenal hyperplasia, ipsilateral or contralateral. It has to be differentiated with metastasis from RCC. Functional study of adrenal gland should be done before surgery. Adrenalectomy can be combined with radical nephrectomy.

CLINICAL PRESENTATION

A 58-year-old male, known diabetic on insulin, known case of hypertension, asthmatic; on medications and clinically asymptomatic; incidentally detected to have 6 × 5 cm right renal mass on ultrasound. Abdominal examination revealed no tenderness or palpable lump.

INVESTIGATIONS

Contrast enhanced computed tomography abdomen revealed 6 × 5 cm hypodense exophytic mass from posterolateral aspect of mid pole of right kidney with heterogeneous enhancement. Right adrenal gland was enlarged with 3 × 2 cm hypodense oval mass. Renal vein and inferior vena cava (IVC) were free of tumor (Fig. 4.5.1).

Fig. 4.5.1: Contrast enhanced computed tomography (CECT) of the abdomen
**TREATMENT**

Patient underwent right open radical nephrectomy with adrenalectomy (Figs 4.5.2A and B). Post operative period was uneventful.

**HISTOPATHOLOGY**

Histopathology revealed papillary carcinoma with large areas of necrosis, Adrenal gland hyperplasia. Margins and hilar lymph node—free of tumor.

**COMMENTS**

Imaging of adrenal gland is important in cases in RCC. RCC may be associated with adrenal tumor or secondaries.

**BIBLIOGRAPHY**

4.6 LEFT RENAL CELL CARCINOMA WITH IVC THROMBUS

**INTRODUCTION**

Renal cell carcinoma (RCC) has propensity for thrombus invasion of renal vein and inferior vena cava (IVC). 10% of patients with RCC have tumor thrombus involving the renal vein or vena cava, and 1% have tumor thrombus extending into the right atrium. It is more common on right side but can also occur with left RCC.

**CLINICAL PRESENTATION**

A 70-year-old male presented with history of weight loss for 4 months. There was no history of anorexia, no urinary symptoms louse, no history of bone pain, headache or chest discomfort. He had past history of fistula in ano with fistulectomy done 15 years back.

**EXAMINATION**

Conscious, oriented, no pallor/cyanosis/clubbing/icterus/lymphadenopathy/raised jugular venous pressure (JVP); afebrile; Pulse rate: 78/min; BP: 110/70 mm Hg; Chest: Bilaterally clear; Per abdominal examination: soft, no organomegaly, no tenderness; umbilical hernia present; central nervous system (CNS): WNL.

**INVESTIGATIONS**

Serum creatinine: 1.5 mg/dL, USG: detected left upper pole renal mass. MRI upper abdomen (Figs 4.6.1A and B): large renal mass involving the superior and mid pole of the kidney extending into the left perinephric fat without disruption of Gerota’s fascia. Tumoral thrombosis of left renal vein and IVC. No significant retroperitoneal lymphadenopathy. T3b N0M0. CT scan thorax: no significant diagnostic abnormality identified at present in the region studied. Color Doppler ultrasound abdomen: left upper and midpolar renal mass with left renal vein thrombus. Thrombus in infrahepatic and hepatic segments of IVC reaching upto sinoatrial junction.

Figs 4.6.1A and B: Magnetic resonance imaging (MRI) of the abdomen
TREATMENT

Left radical nephrectomy with splenectomy, distal pancreatectomy with IVC thrombus removal done under general anesthesia (Fig. 4.6.2–4.6.6). Tumor involved left mesocolon, distal pancreas and adhesions with spleen. Neovascularity in retroperitoneum over aorta and IVC. Dilated left renal vein with infrahepatic IVC thrombus.

Fig. 4.6.2: Intraoperative picture showing thrombus in inferior vena cava (IVC)

Fig. 4.6.3: Intraoperative picture showing removal of thrombus from inferior vena cava
Fig. 4.6.4: Intraoperative picture showing inferior vena cava after removal of thrombus

Fig. 4.6.5: Intraoperative picture showing inferior vena cava after suturing

Fig. 4.6.6: Operative specimen showing left kidney with spleen, colon and thrombus in renal vein
**HISTOPATHOLOGY**

Histopathology revealed left clear cell carcinoma, Fuhrman grade 2 with IVC thrombus.

**FOLLOW-UP**

Patient was well for 2 years. Then he started complaining weakness. CECT abdomen revealed recurrence near right adrenal. He was advised surgery but he refused. He was given molecular targeted therapy with Tab. sunitinib, to which he responded and is still alive.

**COMMENTS**

IVC thrombus with left RCC is very rare. The surgery is more difficult in comparison to IVC thrombus with right RCC

**BIBLIOGRAPHY**

4.7 MUCINOUS ADENOCARCINOMA OF RENAL PELVIS

INTRODUCTION
Primary mucinous adenocarcinoma comprises less than 1% renal pelvic tumors (< 70 cases reported). No sex preponderance and it affects patients in the 6-7th decade of life. Possible etiology is stones (most common), pyelonephritis, congenital malformations, parasitic infections, dietary and environmental factors. Prognosis is poor, 1-year survival is 75% and 5-year survival is 21%. Metastatic adenocarcinoma should be ruled out before labeling primary adenocarcinoma. Metastatic mucinous renal adenocarcinoma can be from breast, gastrointestinal (GI) tract, pancreas, mostly multifocal, widespread nonrenal metastases and occasional large solitary metastasis.

CLINICAL PRESENTATION
A 62-year-old male presented with complaints of swelling and pain in left upper abdomen for 3 months duration. No urinary complaints. Known case of hypertension and coronary artery disease (CAD) since 1999. No history of renal calculi.

EXAMINATION
Per abdominal examination revealed large swelling, tense cystic involving left half of abdomen which moved with respiration (Fig. 4.7.1).

Fig. 4.7.1: Clinical picture of the abdomen
INVESTIGATIONS

Urine R/M: NAD, renal function test (RFT): WNL, Chest X-ray: NAD.

Ultrasonography (KUB) revealed large cystic mass left kidney (Figs 4.7.2A and B), CECT revealed left large renal cyst with thick septae (Figs 4.7.3A to C), USG-guided fine needle aspiration cytology (FNAC): mucoid material with collagenous content.

Upper gastrointestinal (UGI) endoscopy: NAD; Carcinoembryoninoic antigen (CEA): 2 ng/mL; Colonoscopy: NAD.

Figs 4.7.2A and B: Ultrasound

Fig. 4.7.3A: Contrast enhanced computed tomography (CECT) of the abdomen
Exploration revealed a large cystic tumor of the kidney (Fig. 4.7.4). Left radical nephrectomy was done. Cut open specimen revealed large mucinous material (Figs 4.7.5 and 4.7.6).

**TREATMENT**

Figs 4.7.3B and C: Contrast enhanced computed tomography (CECT) of the abdomen
Fig. 4.7.4: Intraoperative pictures

Fig. 4.7.5: Specimen
Primary renal pelvic mucin secreting adenocarcinoma.

Primary mucinous adenocarcinoma of renal pelvis is very rare.

BIBLIOGRAPHY

4.8 PRIMARY MALIGNANT NEUROEPITHELIAL TUMORS OF THE KIDNEY

**INTRODUCTION**

Primary malignant neuroepithelial tumors of the kidney (PNET) comprise a group of primitive, highly malignant neoplasms that histologically and clinically are not well characterized. Primitive neuroectodermal tumor (PNET) is a malignant small cell neoplasm of neural crest origin. It is a very rare tumor representing about 1% of all sarcomas. Renal PNET was first reported by Mor and colleagues. Renal PNET is a highly aggressive malignant neoplasm and is more aggressive than PNET arising from other sites.

Primitive neuroectodermal tumor is treated with a combination of radical nephrectomy and chemotherapy, which includes vincristine, doxorubicin, cyclophosphamide, etoposide and ifosfamide. In cases where there is an incomplete resection or positive margin or recurrence of the tumor, radiation is recommended.

**CLINICAL PRESENTATION**

A 45-year-old male presented with history of pain and swelling of the left upper abdomen. There was no history of hematuria but had significant loss of appetite. On abdominal examination, there was a left renal tumor firm to hard in consistency with a smooth surface. Urine examination was normal.

**INVESTIGATIONS**

Plain X-ray KUB region revealed a large mass left upper abdomen displacing descending colon (Fig. 4.8.1). Ultrasound revealed an 8 × 8 cm mass in front of the splenic vein. CECT revealed a necrotic tumor arising from the superior pole of the left kidney with involvement of spleen (Figs 4.8.2 and 4.8.3).
**Fig. 4.8.1:** Plain X-ray of the kidney, ureter and bladder (KUB) region

**Figs 4.8.2A to C:** Computed tomography (CT) with oral contrast
**TREATMENT**

Angioinfarction followed by left radical nephrectomy along with en bloc removal of spleen and distal pancreas was done (Fig. 4.8.4).

**HISTOPATHOLOGY**

Left renal tumor consisted of small round cells without tubular and globular differentiation. The pancreas and spleen were involved. Tumor emboli were present in the small renal arteries but the lymph nodes were not involved.
**FOLLOW-UP**

Patient was advised adjuvant chemotherapy but he refused. Follow-up CT revealed local recurrence after 14 months. Adjuvant combined chemotherapy (etoposide, ifosfamide and cisplatin) was then given.

**COMMENTS**

Primary malignant neuroepithelial tumor of kidney is a rare tumor and is diagnosed on histopathology. These tumors are more angioinvasive and are also chemosensitive. Adjuvant chemotherapy is recommended. Prognosis is poor.

**BIBLIOGRAPHY**

4.9 RENAL CELL CARCINOMA—PARTIAL NEPHRECTOMY

**INTRODUCTION**

Nephron-sparing partial nephrectomy is used when the tumor is small (<4 cm in diameter) or when the patient has other medical diseases such as diabetes or hypertension. The partial nephrectomy involves the removal of the tumor only, sparing the rest of the kidney, Gerota's fascia and the regional lymph nodes. This allows for more renal preservation as compared to the radical nephrectomy, and this can have positive long term health benefits.

**CLINICAL PRESENTATION**

A 40-year-old female, known hypertensive, on ultrasound examination detected to have right lower renal mass. Contrast enhanced computed tomography (CECT) revealed enhancing lower renal tumor of 3 cm size (Figs 4.9.1A and B); lower pole partial nephrectomy was done. Figure 4.9.2 shows tumor specimen.

![Figs 4.9.1A and B: Contrast enhanced computed tomography (CECT) scan](image-url)
**FOLLOW-UP**

After follow-up of 5 years, patient has both kidney functioning normally and no recurrence of tumor.

**COMMENTS**

Nephron-sparing surgery (NSS) is becoming the standard of care for renal cell carcinoma less than 4 cm in size, as an elective procedure with contralateral normal functioning kidney. If the renal function is compromised or there is only single functioning kidney, then NSS can be done for larger tumor also. Partial nephrectomy can be done by open surgery, laparoscopically or robotically.

**BIBLIOGRAPHY**

4.10 SARCOMATOID RENAL CELL CARCINOMA

Introduction

Sarcomatoid renal cell carcinoma (SRCC) is defined in the 2004 World Health Organization (WHO) classification of renal tumors as—that histologic type of RCC which contains foci of high-grade malignant spindle cells. Thus, sarcomatoid RCC is a biphasic lesion with both mesenchymal (sarcomatous) and epithelial (carcinoma) elements. It has been found to have an increased proliferative activity and is locally aggressive, has high metastatic potential, and is associated with poor prognosis.

In patients with the disease, reported median survival durations from the time of diagnosis are 3.8–6.8 months, when no treatment is given. The 5-year cancer-specific survival rates for patients with SRCC are 15–27%, compared with 79% for patients with clear cell renal cell carcinoma (CCRCC) showing no sarcomatoid change. Surgical resection alone does not change the prognosis of these patients. Most patients (80%) with SRCC who undergo nephrectomy for clinically localized disease subsequently present with metastatic disease. Patients who are treated with aggressive surgical resection and high-dose interleukin 2 (IL-2) therapy have a significantly improved survival compared to patients treated with any other form of immunotherapy or with surgery alone.

Clinical Presentation

A 45-year-old male presented with left side abdominal swelling for 6 months. On examination, a large hard palpable tumor involving left upper abdomen, moving with respiration (Fig. 4.10.1).
INVESTIGATIONS

Ultrasound examination (Fig. 4.10.2) revealed large left renal tumor with mixed echogenicity, CECT abdomen revealed large left tumor with enhancement and variegated appearance (Figs 4.10.3A to C).

**Fig. 4.10.2:** Ultrasound (left kidney)

**Figs 4.10.3A to C:** Contrast enhanced computed tomography (CECT) of the abdomen
TREATMENT
Left open radical nephrectomy was done.

HISTOPATHOLOGY
Renal cell carcinoma with sarcomatoid changes.

COMMENTS
Sarcomatoid RCC is an aggressive tumor and needs adjuvant treatment following radical surgery.

BIBLIOGRAPHY
4.11 CYSTIC RENAL CELL CARCINOMA

INTRODUCTION

Unilocular and multilocular cystic renal cell carcinoma (CRCC) is a rare entity, comprising approximately 1–2% of all renal tumors, and their true biologic behavior is not well-known. It has a variable imaging pattern, the Bosniak category ranging from IIF to IV. As multilocular CRCC lesions increase in complexity on images (higher Bosniak category), there is a corresponding increase in the volume of malignant cells lining the tumor and an increase in the presence of vascularized fibrous tissue. The patient has an excellent prognosis and because of better prognosis of CRCC, a conservative surgical approach would be the treatment of choice whenever technically feasible.

CLINICAL PRESENTATION

A 52-year-old male presented with left flank pain for 6 months duration. There was no history of hematuria. On examination, left kidney was palpable, firm to soft in consistency. Ultrasound revealed a cystic mass with increased echogenicity. Magnetic resonance imaging (MRI) revealed a lower pole tumor enhancing in T2 images (Figs 4.11.1A and B). Left radical nephrectomy was done (Fig. 4.11.2A). Cut section (Fig. 4.11.2B) revealed a large lower pole tumor with necrosis. Histopathology confirmed CRCC.

Fig. 4.11.1A: Magnetic resonance imaging (MRI)
Cystic renal cell carcinoma is a variant of RCC and should be suspected in higher Bosniak type picture on imaging.

**BIBLIOGRAPHY**

**INTRODUCTION**

There are a variety of causes of adrenal pseudotumors on CT scan, including upper pole renal mass, gastric diverticulum, prominent splenic lobulation, pancreatic mass, hepatic mass and periaortic varices. Sometimes, it is very difficult to differentiate upper pole renal tumor with adrenal tumor, till histopathology confirms the diagnosis.

**CLINICAL PRESENTATION**

A 45-year-old male with noninsulin-dependent diabetes mellitus (NIDDM) presented with dull pain in abdomen, malaise for 8 months duration.

**EXAMINATION**

Examination revealed normal abdomen, no lump palpable, BP: 140/86 mm Hg (supine), 130/78 mm Hg (sitting).

**INVESTIGATIONS**

Blood urea: 14 mg/dL, Serum creatinine: 1.4 mg/dL, 24-hour urine catecholamines: 27 µg (upto 30 µg is normal), Urine vanillyl mandelic acid (VMA): 40.6 mg/dL (normal: 15 mg/dL), Dehydroepiandrosterone (DHEAS) level was normal and post-dexamethasone (DEXA) suppression was normal.

Ultrasound revealed a mass arising from upper pole of left kidney. CECT scan revealed large left adrenal mass 13.2 × 12.2 × 11 cm at upper pole of left kidney displacing renal vessels and spleen. MRI revealed a 13 × 11 × 12 cm heterogeneous mass in left lumbar and hypochondrium with infiltration of superior and mid pole of left kidney with involvement of hilum of kidney.

**Figs 4.12.1A and B: Magnetic resonance imaging (MRI)**
(Figs 4.12.1A and B) and no lymphadenopathy. Metaiodobenzylguanidine (MIBG) scan revealed I-131 concentrating in left adrenal region.

**DIAGNOSIS**

Left adrenal tumor involving kidney.

**TREATMENT**

Left robotic adrenalectomy with nephrectomy was done.

**HISTOPATHOLOGY**

Gross: 11.5 × 11.5 × 11 cm of tumor arising from upper pole of kidney (Figs 4.12.2A and B) which measured 10.5 × 5.5 × 5 cm, no capsular or perirenal fat invasion, clear cell RCC, Fuhrman grade 2. Immunohistochemistry (IHC): negative for chromogranin, inhibin, calretinin and melan-A, IHC: positive for cytokeratin.

**COMMENTS**

A tumor arising in the upper pole of the kidney may be difficult to differentiate from an adrenal tumor. Metabolic workup for adrenal functions should be done. During surgery, all precautions should be taken, as for adrenal surgery.

**BIBLIOGRAPHY**

INTRODUCTION

Presence of cholelithiasis can be an incidental finding along with RCC and is diagnosed during imaging. In right-sided tumor, both operations can be done together.

CLINICAL PRESENTATION

A 52-year-old male presented with pain in abdomen of 6 months duration. On investigations, ultrasound revealed cholelithiasis and right renal mass. CECT confirmed cholelithiasis and right renal tumor (Figs 4.13.1A to C).

Figs 4.13.1A to C: Contrast enhanced computed tomography (CECT) scan
**TREATMENT**

Laparoscopic right radical nephrectomy and cholecystectomy was done (Fig. 4.13.2). Patient tolerated the procedure well.

![Specimen](image)

*Fig. 4.13.2: Specimen*

**COMMENTS**

Combined surgery can be done for localized RCC and uncomplicated cholelithiasis.
INTRODUCTION

Malignant hypercalcemia can occur in patients with cancer as part of a paraneoplastic syndrome. Paraneoplastic manifestations of renal cell carcinoma (RCC), including hypercalcemia, polycythemia, hepatic dysfunction, amyloidosis, fever and weight loss are present in up to 20% of the patients. Treatment of hypercalcemia associated with RCC includes restoration of volume status, saline diuresis and use of intravenous (IV) bisphosphonates such as zoledronate or pamidronate. Approximately 50% of patients who undergo nephrectomy and/or tumor debulking will revert to normocalcemia.

CLINICAL PRESENTATION

A 50-year-old male had incidental detection of right renal stone and left renal mass. He had no comorbid illness. Past history was insignificant.

EXAMINATION

A tender mass was palpable in left hypochondrium reaching up to left lumbar region.

INVESTIGATIONS

Hemoglobin: 13.7 g/dL, TLC: 7,000/cumm, Urea: 25 mg/dL, Serum creatinine: 1.9 mg/dL, Serum calcium: 16.8 mg/dL, Serum phosphate: 1.8 mg/dL, Serum albumin: 4.3 mg/dL, Serum alkaline phosphatase: 280 (normal <240), Serum PTH: 2.30 pg/mL (Normal: 15–68.30 pg/mL), 24-hour urinary Calcium: 0.58 gm/day (Normal: 0.1–0.4), Phosphate: 0.656 gm/day (Normal: 0.4–1.3), Uric acid: 0.668 gm/day (Normal: 0.6–0.7), Bone scan and sestamibi scan for parathyroid were normal.

Management of hypercalcemia: Forced saline diuresis 150–200 mL normal saline (NS)/hour along with injection lasix; 40 mg after every 2 L of saline was given. For 2 days, zoledronic acid 4 mg was given. After 4 days of aggressive serum calcium came down to 9.9 mg/dL, serum phosphate to 2.2 mg/dL and serum parathyroid hormone (PTH) increased to 6.7 pg/mL (15–68 mL).

Plain X-ray KUB region (Fig. 4.14.1) showed right renal calculi, intravenous urogram (Fig. 4.14.2) showed normal functioning kidneys with splaying of calyces on left side, CECT scan (Figs 4.14.3A and B) showed enhancing tumor of the left kidney.
Fig. 4.14.1: Plain X-ray of the kidney, ureter and bladder (KUB) region

Fig. 4.14.2: Intravenous urogram
Figs 4.14.3A and B: Contrast enhanced computed tomography (CECT) scan
TREATMENT

Patient underwent laparoscopic left radical nephrectomy. Intraoperatively a large renal mass, $15 \times 8$ cm was seen (Figs 4.14.4A and B).

On postoperative day 3, patient noticed tingling and numbness of extremities. Trousseau and Chvostek’s sign for hypocalcemia was negative, serum Calcium: 7.9 mg/dL, serum phosphate being: 1.9 mg/dL. The patient was managed with calcium supplementation.

HISTOPATHOLOGY

Clear cell carcinoma: Fuhrman’s grade 2, tumor extending into renal vein, margins free.

FOLLOW-UP

On follow-up, serum calcium was: 10.6 mg/dL, at 1 month but he had complaints of cough, for which X-ray chest (Fig. 4.16.5) revealed multiple metastasis, CECT chest revealed multiple metastasis (Fig. 4.16.6).
RCC with hypercalcemia has more aggressive nature and is prone for metastasis. It is important to have more close follow-up and adjuvant treatment.

**BIBLIOGRAPHY**
4.15 RENAL CELL CARCINOMA WITH INFERIOR VENA CAVA THROMBUS

INTRODUCTION
Renal cell carcinoma (RCC) has propensity for thrombus invasion of renal vein and inferior vena cava (IVC). 10% of patients with RCC have tumor thrombus involving the renal vein or vena cava, and 1% have tumor thrombus extending into the right atrium. It is more common on right side but can also occur with left RCC.

CLINICAL PRESENTATION
A 56-year-old male police constable presented with history of hematuria, gross, painless, episodic since January 2004; loss of breath, generalized weakness for 4 months and right flank pain, dull mild continuous for 1 week, loss of appetite, loss of weight (not measured) for 4 months.

No history of fever, bone pains, vomiting or headache. No history of hypertension, diabetes mellitus, tuberculosis, coronary artery disease or surgery. History of jaundice 10 years ago. No history of blood transfusion.

EXAMINATION
Pulse: 70/min, BP: 150/80 mm Hg, afebrile, per abdominal examination: right flank mass 12 × 10 cm hard, nodular and bimanually palpable. No free fluid. No other organomegaly. External genitalia: right grade III varicocele. Left grade II varicocele. Bilateral testes: normal.

INVESTIGATIONS
Hemoglobin: 9.7 g/dL, TLC: 10,500/cumm, Blood sugar (r): 80 mg/dL, Urea: 45.2 mg/dL, Creatinine: 2.2 mg/dL, Na: 141.0 mEq/L, K: 3.8 mEq/L, Calcium: 8.9 mg/dL, Phosphate: 3.9 mg/dL, Uric acid: 6.4 mg/dL, LFT—bilirubin (total): 0.8 mg/dL, Total protein: 9.1 mg/dL, Albumin: 3.0 mg/dL, Globulin: 6.1 mg/dL, SGOT/SGPT 18.4/17.0 IU, Alkaline phosphatase: 487 IU. Ultrasound (Figs 4.15.1A and B) and CECT showed enhancing tumor in right kidney with IVC thrombus (Fig. 4.15.2). MRI shows right renal tumor with supradiaphragmatic thrombus IVC (Figs 4.15.3A and B).

TREATMENT
Right radical nephrectomy with removal of inferior vena cava (IVC) thrombus was done.
Kidney Tumors

Fig. 4.15.1A and B: Ultrasound

Fig. 4.15.2: Contrast enhanced computed tomography (CECT) scan
Renal cell carcinoma with inferior vena cava thrombus is a challenging problem. The level of thrombus is to be identified by proper imaging and treatment is to be planned accordingly.

**BIBLIOGRAPHY**

INTRODUCTION

Metastatic renal cell carcinoma (mRCC) is the spread of the primary renal cell carcinoma from the kidney to other organs. 25–30% of the cases have this metastatic spread by the time they are diagnosed with RCC. This high proportion is explained by the fact that clinical signs are generally mild until the disease progresses to a more severe state. The most common sites for metastasis are the lymph nodes, lungs, bones, liver and brain.

CASE 1: RCC WITH BONE AND TESTICULAR METASTASIS

Clinical Presentation

A 55-year-old male had fracture of right femur in 1990, for which he underwent nailing (Fig. 4.16.1). At the time of surgery, biopsy was taken which revealed metastatic RCC.

Examination

No palpable lump in abdomen. Scrotal examination revealed left testicular tumor.

Investigations

His intravenous urogram revealed lower polar space occupying lesion (Fig. 4.16.2). CECT scan revealed left renal tumor (Fig. 4.16.3).

Fig. 4.16.1: Case 1—X-ray of the right femur
He underwent left radical nephrectomy and left high inguinal orchidectomy.

**Histopathology**

Histopathology confirmed clear cell carcinoma of the kidney and metastatic tumor in left testis.
Follow-Up

He was given adjuvant interferon therapy as no other adjuvant therapy available at that time.

CASE 2: RCC WITH BONE AND LUNG METASTASIS

Clinical Presentation

A 65-year-old male presented with left flank pain along with difficulty in walking for 3 months duration. There was no history of hematuria.

Examination

Abdominal examination revealed no palpable lump. There was tenderness over left side of pelvic bone and compression sign was positive.

Investigations

His blood and urine examination were normal. Serum creatinine was 1.5 mg/dL. Ultrasound revealed left lower pole renal tumor. CECT chest revealed bilateral pulmonary metastasis (Fig. 4.16.4). CECT abdomen revealed enhancing tumor of left lower pole of the kidney and osteolytic lesion in left pelvic bone (Fig. 4.16.5). PET-CT revealed Fluorodeoxyglucose avid lesion in the chest, left iliac bone and left kidney (Fig. 4.16.6).

Fig. 4.16.4: Case 2—Contrast enhanced computed tomography (CECT) of the chest
Fig. 4.16.5: Case 2—Contrast enhanced computed tomography (CECT) of the abdomen

Fig. 4.16.6: Case 2—Positron emission tomography and computed tomography (PET-CT) scan
**Treatment**

He underwent left adjuvant radical nephrectomy (Figs 4.16.7A and B).

**Histopathology**

Histopathology confirmed clear cell carcinoma of the kidney.

**Follow-Up**

Postoperative recovery was uneventful. He was started with tablet sunitib to which he responded well. He started walking well. He tolerated 6 cycles of sunitib. After 1 year, his condition deteriorated and he died.

**COMMENTS**

Adjuvant radical nephrectomy is recommended in cases presenting with RCC with metastasis followed by immunotherapy and recently by molecular targeted therapy.

**BIBLIOGRAPHY**

4.17 PYREXIA OF UNKNOWN ORIGIN AS PRESENTATION FOR RENAL CELL CARCINOMA WITH LUNG METASTASIS

**INTRODUCTION**

Among solid tumors, renal cell carcinoma is most commonly associated with pyrexia of unknown origin (PUO).

**CLINICAL PRESENTATION**

A 50-year-old female was being evaluated for fever of unknown origin for 2 months duration. On ultrasound examination, there was incidental detection of left renal tumor mass. 8 years back she underwent vaginal hysterectomy.

**INVESTIGATIONS**

Contrast enhanced computed tomography (CECT) of the chest revealed: multiple metastatic lesions in the lungs, enlarged nodes in the mediastinum (Fig. 4.17.1). CECT abdomen revealed left enhancing tumor of the kidney with involvement of descending colon (Figs 4.17.2A to D).

*Fig. 4.17.1: Contrast enhanced computed tomography (CECT) of the chest*
Figs 4.17.2A and B: Contrast enhanced computed tomography (CECT) of the abdomen
Figs 4.17.2C and D: Contrast enhanced computed tomography (CECT) of the abdomen
**TREATMENT**

Open left radical nephrectomy with segmental colectomy with colostomy was done (Fig. 4.17.3).

![Operative specimen showing kidney with colon](image)

**HISTOPATHOLOGY**

A 10 cm, clear cell renal cell carcinoma with sarcomatoid features, extending into perirenal tissue and intestinal wall. G3 pT3a. All the resection margins are free of malignancy. Adrenal gland is free of malignancy.

**ADJUVANT TREATMENT**

In view of sarcomatoid changes in histopathology, she was subjected to adjuvant chemotherapy. She died after three months.

**COMMENTS**

Pyrexia of unknown origin (PUO) is a known presentation of RCC and ultrasound can clinch the diagnosis.

**BIBLIOGRAPHY**

4.18 RENAL CELL CARCINOMA WITH RENAL CALCIFICATION

**INTRODUCTION**

RCC can present with calcification which can be focal or diffuse. Calcification can be associated with tumor necrosis. Calcified renal cell carcinoma appears to be a biologically distinct subgroup of renal parenchymal tumors.

**CLINICAL PRESENTATION**

A 51-year-old male presented with right flank pain.

**INVESTIGATIONS**

Plain X-ray KUB region revealed calcification in the renal area with enlargement of the kidney (Figs 4.18.1A and B). Contrast enhanced computed tomography (CECT) confirmed enhancing right renal tumor.

![Figs 4.18.1A and B: Plain X-ray of the kidney, ureter and bladder (KUB) region](image)

**TREATMENT**

Right radical nephrectomy was done. Histopathology confirmed RCC with calcification.

**COMMENTS**

RCC with calcification do not change prognosis and survival.

**BIBLIOGRAPHY**

4.19 RENAL CELL CARCINOMA WITH CALCULI

INTRODUCTION

Despite the high prevalence of renal calculi in North Indian population, there are few reports of the synchronous presentation of renal cancer with renal calculus. They were diagnosed as incidental renal cancers in the era of open surgery for renal calculi. With better imaging, they can be diagnosed preoperatively and management can be planned. In long standing cases of renal calculi, renal tumor should be suspected and CECT is recommended.

CASE 1

Clinical Presentation

A 35-year-old male presented with dull pain right flank for 3 months duration. No history of hematuria. No past history of renal calculi. History of hypertension since last 1 year.

Examination

Examination revealed no positive findings.

Investigations

Plain X-ray KUB region revealed radio opaque shadows in the renal area (Fig. 4.19.1). Intravenous urogram revealed normal functioning of both kidneys with stone in right kidney (Fig. 4.19.2). Ultrasound revealed right renal calculi

Fig. 4.19.1: Case 1—Plain X-ray of the kidney, ureter and bladder (KUB) region

Fig. 4.19.2: Case 1—Intravenous urogram
and a small mass lesion in lower pole of right kidney (Fig. 4.19.3). CECT scan revealed right lower pole enhancing tumor of 3 cm size with right renal calculus (Figs 4.19.4A to C).

![Case 1—Ultrasound abdomen](image)

**Fig. 4.19.3:** Case 1—Ultrasound abdomen
Figs 4.19.4A to C: Case 1—Contrast enhanced computed tomography (CECT) of the abdomen

**Treatment**

Right lower pole partial nephrectomy (Fig. 4.19.5) was done followed by extracorporeal shock wave lithotripsy (ESWL) for right renal calculi.

**Follow-Up**

No recurrence of tumor in 2 years follow-up.
CASE 2

Clinical Presentation

A 65-year-old male, diagnosed to have left staghorn renal calculi in 1994. In 1994, he was advised for surgery. He did not undergo surgery and lost to follow-up. No records available for same. In May 2010, he presented with left flank pain. There is history of coronary artery disease 1.5 years back, hypertension and chronic kidney disease (CKD). He had left percutaneous nephrostomy (PCN) for hydronephrosis, later on his PCN tube slipped out. Two years later, he presented with left flank pain.

Examination

No palpable lump in abdomen.

Investigations

Serum creatinine (on December 21, 2010): 2.2 mg/dL. Plain X-ray KUB region slowed left renal calculi (Fig. 4.19.6). Intravenous urogram revealed poorly functioning kidney with sparing of calyces with renal calculi (Figs 4.19.7A and B). Ultrasound done June 08, 2010 revealed left kidney—enlarged by a complex cystic lesion (6 × 12 cm), multiple septae in upper pole with wall calcifications with multiple calculi in it (Fig. 4.19.8). Diethylene triamine pentaacetic acid (DTPA) scan done on May 25, 2010 revealed right kidney
GFR: 12.08 mL/min, left kidney GFR: 8.57 mL/min, Total GFR: 20.66 mL/min (Fig. 4.19.9). MRI done on June 14, 2010: enlarged left kidney with a large lobulated cystic/hydronephrosis (HDN) collection involving upper and mid pole region, internal septations and multiple internal hypointense foci suggestive of calculi with possible patchy wall calcification, with a small doubt full ill-defined soft tissue focus apparent peripherally along inferolateral aspect of this collection or cystic lesion of unknown nature (Fig. 4.19.10).

**Fig. 4.19.6:** Case 2—Plain X-ray of the kidney, ureter and bladder (KUB) region

**Figs 4.19.7A and B:** Case 2—Intravenous urogram
Fig. 4.19.8: Case 2—Ultrasound abdomen

Fig. 4.19.9: Case 2—Dynamic renal scan
Treatment

Patient underwent left PCN under local anesthesia (LA) on December 16, 2010, approximately 1,000 mL hemorrhagic fluid was drained. Daily output from PCN was approximately 200 mL. Drain fluid for cytology: cells with marbled pleomorphism with high neocyttoplasmic ratio and hyperchromatic nuclei numerous scattered polymorphs and histiocytes. Positive for malignant cells (possibly adenocarcinoma).

He underwent left radical nephrectomy on December 22, 2010 (Fig. 4.19.11). Histopathology confirmed clear cell RCC.

Fig. 4.19.10: Case 2—Magnetic resonance imaging (MRI)
Renal cell carcinoma can be associated with renal calculi. In case of any doubt on ultrasound or intravenous pyelogram (IVP), CECT is recommended which can diagnose the tumor. Management can be according to the merit of the case.

**BIBLIOGRAPHY**

INTRODUCTION

Renal pseudotumors are lesions that mimic renal neoplasm on imaging but after surgery and histopathology are proven to be composed of normal or benign renal tissue. Attempts at characterizing renal pseudotumors have been difficult on imaging and methods like contrast enhanced ultrasound (CEUS), CECT are likely to help over non contrast studies. Renal pseudotumor can be classified as developmental, infectious, granulomatous or vascular (Table 4.20.1).

Table 4.20.1: Classification of renal pseudotumors

<table>
<thead>
<tr>
<th>Types of renal pseudotumors</th>
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<tbody>
<tr>
<td><strong>Developmental</strong></td>
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<tr>
<td>Prominent columns (septa) of Bertin</td>
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<tr>
<td>Persistent fetal lobulation</td>
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<tr>
<td>Dromedary hump</td>
</tr>
<tr>
<td>Splenorenal fusion</td>
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<tr>
<td>Cross-fused renal ectopia</td>
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<tr>
<td><strong>Infectious</strong></td>
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<tr>
<td>Abscess</td>
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<tr>
<td>Pyelonephritis</td>
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<tr>
<td>Scarred kidney</td>
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<tr>
<td><strong>Granulomatous</strong></td>
</tr>
<tr>
<td>Xanthogranulomatous pyelonephritis</td>
</tr>
<tr>
<td>Sarcoidosis</td>
</tr>
<tr>
<td>Malakoplakia</td>
</tr>
<tr>
<td>Tuberculosis</td>
</tr>
<tr>
<td><strong>Vascular</strong></td>
</tr>
<tr>
<td>Extramedullary hematopoiesis</td>
</tr>
<tr>
<td>Arteriovenous malformation</td>
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<tr>
<td>Renal pelvic hematomas</td>
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<tr>
<td>Anticoagulant-induced subcapsular hemorrhage</td>
</tr>
<tr>
<td><strong>Miscellaneous</strong></td>
</tr>
<tr>
<td>Regenerating nodule after reflux</td>
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<tr>
<td>IgG4 related disease</td>
</tr>
</tbody>
</table>

CLINICAL PRESENTATION

A 48-year-old female had a history of recurrent UTIs for last 18 years. She was married in 1981, had her first child birth in 1986. She had a history of fall during pregnancy and had pelvic tear. Delivery was normal. Pelvic floor repair was done in 1987 post delivery. Second child was born in 1995. It was a complicated forceps delivery. She developed urinary tract infection
(UTI) and urinary incontinence post delivery. In 1999, Marshall-Marchetti-Krantz (MMK) procedure was done which relieved her incontinence but UTI continued. In 2002, cystoscopy was done which revealed chronic cystitis. Repeated cystoscopies and urethral dilatations were done. In 2008, cystoscopy, bladder biopsy, bladder neck incision (BNI) and left retrograde pyelography procedures were done. Bladder biopsy was negative for malignancy and showed chronic cystitis. Ultrasound done in March 2012 was reported normal (Fig. 4.20.1).

In October 2012, she presented to us with weight loss and loss of appetite. Physical and abdominal examination was normal. Investigations revealed erythrocyte sedimentation rate (ESR) of 66 mm/1st hour, urine culture positive for *Escherichia coli*, antinuclear antibodies (ANA) positive and low serum thyroid-stimulating hormone (TSH). CECT chest was normal. CECT abdomen revealed a poorly enhancing partly calcified mass in lower pole of left kidney involving both cortex and medulla and distorting the pelvicalyceal system; suggestive of infective etiology possibly tubercular (Fig. 4.20.2). A further renal core biopsy was done which revealed chronic inflammation with no evidence of malignancy. A DTPA scan showed a GFR of 26.3 mL/sec for left kidney and 56.7 mL/sec for right kidney.

She was advised long term suppressive antibiotics, multivitamin supplements and cranberry tablets. However, no improvement was noted in next 2 months. She was then started on antitubercular treatment which had to be stopped after 1 week due to excessive vomiting and dehydration.

Fig. 4.20.1: Ultrasonography (USG) of the abdomen showing left kidney with arrows pointing to calcification in left lower pole
INVESTIGATIONS

Serum TSH was found to be low, serum adrenocorticotropic hormone (ACTH) was also low, IgG subclass 4 was elevated. She was started on thyroxin and prednisolone.

Repeat CT scan (Fig. 4.20.3) revealed no interval change with previous CECT. A poorly enhancing in homogeneous partly calcified lesion was noted in the lower half of the left kidney involving both cortex and medulla (renal shape maintained); distorting the pelvicalyceal system as well as smaller hypodense lesion in the interpolar cortex of the right kidney posterolaterally remained unchanged. Few prominent retroperitoneal lymph nodes were noted as before.

Positron emission tomography-computed tomography (PET-CT) (Fig. 4.20.4) revealed hyper metabolic large inhomogeneously enhancing mass lesion in the lower half of cortex and medulla of the left kidney with areas of nodular calcification within, extending into the renal sinus and abutting the posterior paraspinal muscles with no obvious infiltration. Hypermetabolic small wedge shaped cortical based hyperdense area in the interpolar region in the posterior cortex of the right kidney. Hypermetabolic mediastinal and retroperitoneal lymphadenopathy. Metabolically indeterminate few randomly distributed nodular opacities in both the lung fields.

![Contrast enhanced computed tomography (CECT) of the abdomen](image_url)
Fig. 4.20.3: Contrast enhanced computed tomography (CECT) of the abdomen

Fig. 4.20.4: Positron emission tomography-computed tomography (PET-CT) scan
Robot assisted laparoscopic left nephrectomy was done on March 21, 2013. Colon was adherent to the left lower pole.

Kidney specimen (Figs 4.20.5 and 4.20.6), cut section showed tumor in lower half of the kidney.
HISTOPATHOLOGY

Multiple section studied from the tumor reveal an inflammatory pseudotumor composed of storiform fibrosis, moderate to marked lymphoplasmacytic infiltrate with formation of lymphoid follicles. Admixed with the inflammatory cells were scattered fibroblastic cells. Majority of the vessels entrapped in this inflammatory lesion revealed obliterative phlebitis. This inflammatory process involved renal sinus and pelvicalyceal system. In addition, areas of calcification were also noted. There was no evidence of malignancy.

The sclerosing areas and fibroblastic areas were vimentin and smooth muscle actin (SMA) positive and anaplastic lymphoma kinase (ALK) negative.

Impression: Inflammatory sclerosing pseudotumor.

COMMENTS

In view of ANA positivity and this tumor morphology, possibility of IgG4 related sclerosing disease needs consideration.

She was discharged on thyroxin 50 μg and prednisolone 5 mg daily along with multivitamins.

On follow-up at 3 months after the surgery a weight gain of 13 kg was noted and the appetite had returned to normal. ESR was down to 30 mg/1st hour. No urinary tract infection was noted in the intervening period. PET-CT was normal (Fig. 4.20.9).

BIBLIOGRAPHY

4.21 RENAL TUMOR WITH RENAL CYST

- **INTRODUCTION**
  
  Renal cell carcinoma can be associated with renal cyst which can be simple cyst or associated with adult polycystic kidney disease (APKD). Renal cyst can also have cystic carcinoma.

- **CASE 1**

  **Clinical Presentation**

  An 80-year-old male presented with vague abdominal pain.

- **Investigations**

  Ultrasound revealed right lower pole tumor and left renal cysts. CECT confirmed enhancing right lower polar tumor with left large simple renal cyst at upper pole and small cyst at lower pole (Figs 4.21.1A to C).

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**Figs 4.21.1A to C:** Case 1—Contrast enhanced computed tomography (CECT) of the abdomen
**Treatment**

Right lower pole partial nephrectomy and left upper pole cyst excision and deroofing of lower pole cyst was done simultaneously. Figure 4.21.2 shows specimen of partial nephrectomy and excised cyst. Procedure went uneventful.

![Fig. 4.21.2: Case 1—Specimen of kidney and cyst](image)

**Follow-Up**

Magnetic resonance imaging revealed normal functioning right upper half of kidney and normal functioning left kidney.

**CASE 2**

**Clinical Presentation**

A 55-year-old male presented with right side abdominal pain for 6 months duration.

**Investigations**

Ultrasound revealed right lower pole renal cysts and left side tumor in lower pole of the kidney. CECT (Figs 4.21.3A and B) confirmed enhancing left lower polar tumor and small cyst upper pole with right large simple renal cyst at lower pole.
Figs 4.21.3A and B: Case 2—Contrast enhanced computed tomography (CECT) of the abdomen

**Treatment**

Left lower pole partial nephrectomy (Figs 4.21.4A to C) and right lower pole cyst deroofing was done simultaneously. Figure 4.21.5 shows specimen of partial nephrectomy. Procedure went uneventful.
Figs 4.21.4A to C: Case 2—Intraoperative picture of partial nephrectomy
RCC can be associated with simple renal cyst and it can be an incidental finding. Simultaneous management of both should be planned.

**BIBLIOGRAPHY**

4.22 SPLENIC TUMOR MIMICKING RENAL CELL CARCINOMA

**INTRODUCTION**

There are a variety of causes of tumors near adrenal gland on CT scan, including upper pole renal mass, gastric diverticulum, prominent splenic lobulation, pancreatic mass, hepatic mass and periaortal varices. Discrimination of origin of these tumors from neighborhood organs is difficult preoperatively.

**CLINICAL PRESENTATION**

A 30-year-old young lady presented history of recurrent right flank pain for the last 6 years. Episodes of continuous, dull, localized pain were accompanied by frequency of urination along with fever for which she was investigated and treated in Nepal. NCCT done outside showed a left renal calcific mass (Figs 4.22.1 to 4.22.3). She gave history of multiple interventions in the form of repeated Double J (DJ) stent placement on the right side with symptomatic relief in pain during these 6 years. She also gave history of being without right sided DJ stent for varying duration.

There is no history of pain on left side of abdomen or flank. No intervention was ever done on left side kidney or ureter. No history of bowel symptoms. No history of diabetes mellitus, tuberculosis or hypertension, no weight loss. No history of contact with pets.

Six months back, she had raised blood urea and creatinine. Ultrasonography revealed bilateral enlarged kidneys (right 12.5 × 7.4 cm, left side 12.5 × 6.7 cm) with hydronephrosis, cortical thinning and loss of corticomedullary differentiation. No stone or mass lesion was seen. Right sided percutaneous nephrostomy was done. The creatinine came down to 1.9 mg/dL. CECT abdomen and CT nephrostogram was done on November 12, 2010 (Fig. 4.22.4). After that right DJ stenting was done and nephrostomy catheter was removed and patient was referred for further management.

**EXAMINATION**

She was conscious, oriented, no pallor, cyanosis, clubbing, icterus, lymphadenopathy or raised jugular venous pressure (JVP). Afebrile, Pulse: 80/min, RR: 22/min, BP: 110/70 mm Hg. Per abdominal examination: soft, Bowel sounds: normal, no organomegaly, no tenderness. Right PCN scar present. CNS examination was normal.

**INVESTIGATIONS**

Blood urea: 40 mg/dL, Serum creatinine: 1.70 mg/dL. DTPA renal scan revealed right obstructed kidney with maintained cortical function and nonvisualized left kidney (nonfunctioning).
**Kidney Tumors**

**Fig. 4.22.1:** Noncontrast computed tomography (NCCT) scan of the abdomen

**Fig. 4.22.2:** Contrast enhanced computed tomography (CECT) of the abdomen
Fig. 4.22.3: Contrast enhanced computed tomography (CECT) of the abdomen—coronal

Fig. 4.22.4: Nephrostogram
TREATMENT

Exploratory laparotomy by midline incision was done on February 9, 2011. Large hard mass from the spleen adherent to left lateral segment of liver, splenic flexure of colon, distal pancreas, greater curvature of stomach and diaphragm. Multiple calcific hard nodules present in omentum.

En bloc resection of spleen, wedge resection of segment III of liver, wedge resection of greater curvature of stomach, segmental resection of splenic flexure of colon, distal pancreatectomy with colocolic anastomosis was done (Figs 4.22.5 to 4.22.7). Mesenteric lymph node biopsy was done. Left nephrectomy with end to side (left ureter to the side of right ureter) uretero-ureteral anastomosis was done. DJ stent placed in both the ureters across the anastomosis.

She tolerated the procedure well. She had increased serum creatinine and drain output in the postoperative period which settled after right percutaneous nephrostomy placement.
Fig. 4.22.6: Operated specimen—spleen cut open

Fig. 4.22.7: Operated specimen—left kidney
**FOLLOW-UP**

Patient recovered and was discharged on 10th postoperative day. Nephrostomy catheter was clamped and removed after 4 weeks. Blood urea was 44 mg/dL and Serum creatinine: 1.9 mg/dL, DJ stent was removed after 3 months. On 2 years follow-up, she was asymptomatic with normal renal parameters.

**FINAL DIAGNOSIS**

Small dysplastic, nonfunctioning left kidney, right mid ureteric stricture with hydroureteronephrosis, mild CRF and splenic tumor with calcification.

**COMMENTS**

Due to nonvisualization of left kidney, it was difficult to diagnose this tumor preoperatively. During surgery, it was diagnosed as splenic tumor with small left kidney. There was a right ureteric stricture; therefore, decision was taken intraoperatively to perform left to right ureteroureterostomy.

**BIBLIOGRAPHY**

4.23 INCIDENTAL DETECTION OF TRANSITIONAL CELL CARCINOMA OF THE RENAL PELVIS

**INTRODUCTION**

Transitional cell carcinoma (also known as urothelial carcinoma) is the most common type of malignant tumor of the renal pelvis and ureter, although it is relatively rare in the population. This type of tumor is often found in multiple areas (synchronous lesions) of the upper urinary tract.

Transitional cell carcinoma is more common in men than women, and patients who smoke or have certain chemical exposures may also be at higher risk for this type of cancer.

For low grade, smaller cancers, minimally invasive endoscopic management whenever possible is advisable. For larger tumors that require kidney preservation, percutaneous approaches to the kidney may be used. For moderate- to high-grade cancers, removal of the entire kidney, ureter and a small cuff of bladder where the ureter enters the bladder is the treatment of choice (nephroureterectomy).

**CLINICAL PRESENTATION**

A 59-year-old male presented with an incidental detection of left lower pole mass lesion on ultrasound. There was no history of hematuria or flank pain.

**PAST HISTORY**

Patient is a chronic smoker and known case of hypertension.

**EXAMINATION**

No abdominal lump palpable on examination.

**INVESTIGATIONS**

Creatinine: 1.2 mg/dL, total leukocyte count (TLC): 13,400/cumm, Urine cytology (3 samples): negative, Urine R/M revealed pus cells: 20/hpf, Urine C/S: *Candida albicans*.

X-ray chest (Fig. 4.23.1): no abnormality detected, noncontrast computed tomography (NCCT) of the abdomen (Fig. 4.23.2) revealed soft tissue mass lesion in the pelvic region, CECT abdomen showed poorly enhancing tumor pelvis and lower calyx (Figs 4.23.3A to C), CT angiogram revealed poorly enhancing tumor pelvis and lower calyx and cyst lower pole of the kidney (Figs 4.23.4A and B), positron emission tomography and computed tomography (PET-CT) revealed fluorodeoxyglucose (FDG) avid lesion in the pelvis and lower calyx (Figs 4.23.5A and B).
**Fig. 4.23.1:** X-ray chest

**Fig. 4.23.2:** Noncontrast computed tomography (NCCT) of the abdomen
**Figs 4.23.3A to C:** Contrast enhanced computed tomography (CECT) of the abdomen

**Figs 4.23.4A and B:** Computed tomography (CT) angiogram
Figs 4.23.5A and B: Positron emission tomography and computed tomography (PET-CT)
**TREATMENT**

Cystoscopy, left retrograde pyelography and left robotic radical nephroureterectomy with para-aortic lymphadenectomy was done on January 31, 2013. Postoperative recovery was uneventful. Nephroureterectomy specimen (Fig 4.23.6A) and cut open specimen (Fig. 4.23.6B) showed tumor involving lower half of the kidney.

![Fig 4.23.6A and B: Nephroureterectomy specimen (A) and cut open specimen (B)](image)

**HISTOPATHOLOGY**

High grade urothelial carcinoma of the left kidney, 0/8 lymph nodes, resection margins free of tumor, pT3a pNo pM0.

**ADJUVANT CHEMOTHERAPY**

In view of high grade and stage of the tumor, four cycles of adjuvant chemotherapy with gemcitabin and paclitaxel were given.

**FOLLOW-UP**

Patient is asymptomatic and had no recurrence in 6 months follow-up.
COMMENTS

Tumors of renal pelvis and ureter have varied presentation and can be confused with RCC. A high index of suspicion is required and diagnosis can be confirmed by ureteroscopy and biopsy. High grade tumor needs radical surgery with lymphadenectomy and adjuvant chemotherapy.

BIBLIOGRAPHY

4.24 VON HIPPEL-LINDAU DISEASE

**INTRODUCTION**

Von Hippel-Lindau (VHL) disease is an autosomal dominant disease with features of hemangioblastomas and cysts involving the pancreas, liver and kidneys; and have a propensity to develop renal cell carcinoma and pheochromocytoma. Hemangioblastomas are most common in the cerebellum and retina. The disease frequency is 1 in 30,000–40,000.

**CLINICAL PRESENTATION**

A 50-year-old male presented with inability to sleep and lie down, backache, urgency and urge incontinence and multiple episodes of gross hematuria. He also had difficulty in walking and imbalance. He had family history of renal cyst and brain tumor. Figure 4.24.1 shows his family tree.

**PAST HISTORY**

He had history of gross hematuria in May 2012. Ultrasound done in May 2012, revealed multiple bilateral renal cyst with mass lesion in left kidney. A CT/IVP revealed multiple pancreatic cysts, left kidney.

**EXAMINATION**

Right kidney enlarged and palpable, neurological deficit in both lower limbs.

**INVESTIGATIONS**

Hemoglobin: 13.7 gm/dL, TLC: 6.22 × 10^3, Urea: 37 mg/dL, Creatinine: 1.2 mg/dL. NCCT and CECT contrast enhanced computed tomography revealed multiple bilateral complex renal cortical cysts, large heterogeneous enhancing mass involving the upper and middle pole of the right kidney with
loss of fat plane with liver, tumor thrombus extending into the right renal vein. Another heterogeneously enhancing mass in interpolar region of left kidney. Multiple pancreatic cysts with a heterogeneously enhancing lesion in the tail of the pancreas likely representing complex cyst versus neoplastic change. Homogeneously enhancing intramedullary lesion in the conus medullaris. In view of the history of von hippel-lindau disease it is likely a hemangioblastoma. Multiple prominent retroperitoneal lymph nodes (Figs 4.24.2 to 4.24.6A and B).

**Magnetic Resonance Imaging of the Brain and Spine**

Magnetic resonance imaging revealed large well-defined circumscribed cystic mass lesion in the posterior fossa involving the cerebellar vermis; and right cerebellar hemisphere intensely enhancing mural nodule with mild perilesional edema causing mass effect over the brainstem, and anterior displacement of the fourth ventricle and upstream prominence of the supratentorial ventricular system.

Multiple, at least four other intensely enhancing nodular lesions are also seen in the posterior fossa involving the bilateral (L>R) cerebellar hemispheres. These findings are suggestive of multiple hemangioblastomas (Von Hippel-Lindau syndrome).

Screening MRI of the cervical spine revealed subtle intramedullary hyperintensity involving the cervical spinal cord at C3–C4 level on T2 weight (T2W) images.

![Noncontrast computed tomography (NCCT) of the abdomen showing enlarged kidneys with right renal calcification](image)
Fig. 4.24.3: Contrast enhanced computed tomography (CECT) of the abdomen—arterial phase

Fig. 4.24.4: Contrast enhanced computed tomography (CECT) of the abdomen—venous phase
Fig. 4.24.5: Contrast enhanced computed tomography (CECT) of the abdomen—delayed phase

Figs 4.24.6A and B: Contrast enhanced computed tomography (CECT) of the abdomen—coronal view
**Dynamic Renal Scan**

Dynamic renal scan revealed overall subnormal GFR. Left non obstructed kidney with normal cortical function. Faintly visualized right kidney with grossly impaired cortical function. No comment is possible on drainage pattern in view of poor function. GFR: right kidney—16.6 mL/min, left kidney—55.2 mL/min.

**TREATMENT**

Patient was counseled for treatment, he decided for regular observation.

**COMMENTS**

There is an established correlation between RCC and Von Hippel-Lindau disease. The disease is hereditary and the VHL gene can be identified on genetic testing. Patients of VHL need detailed evaluation and management.

**BIBLIOGRAPHY**

5.1 BILATERAL TRANSITIONAL CELL CARCINOMA OF THE UPPER TRACT

**INTRODUCTION**

Upper tract transitional cell carcinoma (TCC) constitutes about 5% of urothelial malignancies. Although it is associated with “field change” and is frequently multifocal (44%), synchronous bilateral involvement is rare, constituting only 1% of all upper tract TCC. No specific guidelines for management have so far been formulated, given the rarity of the disease. One series of 10 cases with bilateral synchronous TCC reported all 7 cases with compromised renal function and all patients died within 36 months of diagnosis, despite radical or conservative surgery combined with maintenance dialysis.

There are several treatment options for managing localized upper tract TCC with nephroureterectomy and cuff of bladder excision being the standard treatment which can be done by open, laparoscopic or robotic approach. In patients with severe comorbidities precluding extensive surgery or patients with renal insufficiency, solitary kidney or synchronous bilateral disease endoscopic management through antegrade or retrograde access provides an acceptable alternative without rendering the patient anephric.

**CLINICAL PRESENTATION**

A 57-year-old male, known chronic smoker and diabetic, presented with complaints of painless hematuria with clots for 1 month duration. There was no history of flank pain and dysuria. Past history of pituitary gland enlargement in 1995, which was diagnosed as pituitary tuberculosis and treated with antitubercular therapy (ATT) for 6 months.
EXAMINATION

No palpable lump in abdomen. Digital rectal examination revealed prostate to be normal, firm, smooth and mucosa was free.

INVESTIGATIONS

Urine cytology: Positive for malignant cells; Hemoglobin (Hb): 11.1 g/dL; Blood urea: 43 mg/dL, Serum creatinine: 2.1 mg/dL, Liver function test, serum cortisol and thyroid function test were within normal limits.

Ultrasound KUB revealed 3.75 × 2.26 cm mass in right kidney with normal pelvicalyceal system (PCS). Another mass of 1.9 × 2.8 cm was found in left ureter extending to left vesicoureteric junction. Left sided moderate hydroureteronephrosis was detected (Figs 5.1.1 to 5.1.3).

Magnetic resonance imaging (MRI) abdomen revealed 3.7 × 2.2 cm mass in right renal pelvis inferiorly. Right pelvicalyceal system (PCS) was normal (Fig. 5.1.4). Left kidney was small in size with perinephric stranding. There was moderate dilatation of left PCS and ureter with a 3.1 × 2.7 cm mass lesion in distal left ureter involving left vesicoureteric (VU) junction (Fig. 5.1.5). Fat planes were maintained between the mass and prostate. Chest X-ray was normal. Renal dynamic scan: bilateral renal parenchymal dysfunction without any obstruction—total glomerular filtration rate (GFR): 27.58 mL/min. Right kidney function was 89% (GFR: 24.53 mL/min) and left kidney function was 11% (GFR: 3.05 mL/min) (Figs 5.1.6 and 5.1.7).

Fig. 5.1.1: Ultrasound of the right kidney showing tumor
Fig. 5.1.2: Ultrasound of the left kidney showing hydroureteronephrosis (HDUN)

Fig. 5.1.3: Ultrasound of the bladder showing tumor left lower ureter
**Fig. 5.1.4:** Magnetic resonance imaging (MRI) hydroureteronephrosis (HDUN) left and tumor lower one-third ureter

**Fig. 5.1.5:** Magnetic resonance imaging (MRI) of the lower abdomen showing tumor left ureter
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Upper Tract Urothelial Tumor

Fig. 5.1.6: Dynamic renal scan showing poorly functioning left kidney and normal functioning right kidney

CYSTOSCOPY

Tumor was protruding from the left ureteric orifice. Right ureteric orifice and bladder walls were normal. Biopsy was taken from the left vesicoureteric junction. Right retrograde pyelography (RGP) revealed tumor over lateral wall of the pelvis and upper ureter extending into the inferior calyx (Fig. 5.1.8). Laser resection and fulguration was done and double J (DJ) stent was placed (Fig. 5.1.9). Injection mitomycin C (40 mg) was instilled intravesically after 4 hours of surgery in Trendelenburg position.

Histopathology: Papillary lesion with features of papillary neoplasm of low malignant potential (PUNLMP) on both right pelvis and left ureter. Biopsy from bladder trigone did not reveal any tumor.
**DIAGNOSIS**

Final diagnosis was right renal and inferior calyceal tumor with good functioning kidney and left multiple ureteric tumors involving lower ureteric orifice with poorly functioning kidney.

**TREATMENT**

Right percutaneous nephroscopy and tumor resection was done. An infracostal, superior calyceal puncture was made and tumor was resected and its base fulgurated with holmium laser. Incomplete removal of tumor was done due to bleeding. Injection mitomycin C, 40 mg was administered through the percutaneous nephrostomy after 4 hours.

Robotic left nephroureterectomy with lymphadenectomy and excision of the cuff of the bladder was done (Fig. 5.1.10).

*Histopathology:* Lower end of left ureter showed low grade papillary urothelial carcinoma. The resected end of bladder cuff was free of tumor. Kidney showed features of chronic pyelonephritis. Renal hilar lymph node showed three lymph nodes and all were free of tumor.

After 1 week, relook right nephroscopy, resection of the remaining tumor and laser fulguration of the base was done.
Fig. 5.1.8: Right tumor specimen

Fig. 5.1.9: Nephroureterectomy specimen

Fig. 5.1.10: Retrograde pyelogram (RGP) after 3 months
**POSTOPERATIVE PARAMETERS**

Postoperative blood parameters were—Hb: 11.5 g/dL, blood urea: 42 mg/dL and serum creatinine: 1.8 mg/dL.
The patient was discharged and was administered 6 doses of mitomycin C weekly through the nephrostomy on outpatient basis. Cystoscopy and nephroscopy at 6 weeks were normal. Nephrostomy catheter was removed.

**FOLLOW-UP**

At 6 months—he was asymptomatic. Serum creatinine was 2.2 mg/dL. Urine cytology was negative. He was on regular follow-up every 3 months for the 1st year and 6 monthly for the next 3 years. There was no recurrence of the tumor and renal functions were normal.

**COMMENTS**

Endourologic techniques through retrograde and percutaneous access for the right side along with left robotic nephroureterectomy and cuff of bladder excision provided excellent perioperative outcomes in this patient.

**BIBLIOGRAPHY**

5.2 SMALL SUPERIOR CALYCEAL TUMOR

**INTRODUCTION**

Transitional cell carcinoma (TCC) of the upper urinary tract is a common malignancy affecting the genitourinary tract. The common presentation is hematuria. They are detected by imaging like ultrasound, intravenous urography, computed tomography (CT) urography and rarely by magnetic resonance (MR) urography. They can be multifocal; therefore, careful evaluation of complete urothelial system should be done.

Management is by nephroureterectomy; however, in single kidney or in chronic kidney disease (CKD), renal preserving surgery can be done by endourological procedures.

Prognosis is good in early cases. In high-grade advanced cases, adjuvant chemotherapy is required and prognosis is poor.

**CLINICAL PRESENTATION**

A 56-year-old male presented with one single episode of hematuria 1 week back. There was no history of flank pain or dysuria.

**EXAMINATION**

Examination was normal. Prostate grade I, firm, smooth and mucosa was free.

**INVESTIGATIONS**

Urine (routine/microscopic) revealed red blood cells (RBC) ++, no pus cells, urine cytology was negative. Ultrasound revealed a tumor in left upper pole of the kidney. Contrast enhanced computed tomography (CECT) abdomen revealed an enhancing lesion in left superior calyx (Figs 5.2.1A to C). Right kidney was normal.
Figs 5.2.1A to C: Contrast enhanced computed tomography (CECT) of the abdomen

Fig. 5.2.2: Left retrograde pyelogram
**TREATMENT**

Left retrograde pyelogram revealed a filling defect in superior calyx (Fig. 5.2.2). Left nephroureterectomy was done. Kidney specimen revealed a small tumor in left superior calyx (Figs 5.2.3A and B).

![Nephroureterectomy specimens](image)

**Figs 5.2.3A and B: Nephroureterectomy specimens**

**HISTOPATHOLOGY**

Transitional cell carcinoma of low grade in left superior calyx was seen. The other parts of the kidney was normal.

**FOLLOW-UP**

Patient was asymptomatic on follow-up for 3 years.

**COMMENTS**

Hematuria in adults should be investigated in detail to identify the cause. TCC renal pelvicalyceal system if diagnosed early has got good prognosis.

**BIBLIOGRAPHY**

5.3 TRANSITIONAL CELL CARCINOMA OF THE RENAL PELVIS

INTRODUCTION

Transitional cell carcinoma (TCC) of the renal pelvis is uncommon compared to renal cell carcinoma, and can be challenging to identify on routine imaging. Renal pelvis tumors are more common in males, and are typically diagnosed between 60 and 70 years of age. Imaging depends on the morphology: papillary tumors appear as soft tissue density filling defects whereas nonpapillary or infiltrating tumors are harder to detect as they are often sessile. The choice of treatment is surgical consisting of a nephroureterectomy with excision of cuff of bladder. In low stage tumors and especially in patients with bilateral tumors or solitary kidneys, renal sparing surgery may be attempted endoscopically (percutaneous or transurethral approach). High grade and high stage tumors need adjuvant chemotherapy.

CLINICAL PRESENTATION

A 64-year-old male presented with complaints of heaviness in abdomen and decreased urination for 15 days duration. No history of fever, hematuria, pyuria or dysuria. No history of diabetes mellitus.

EXAMINATION

Abdomen soft, no organomegaly, no tenderness.

INVESTIGATIONS

Serum creatinine was 1.3 mg/dL. Ultrasound revealed right mild hydronephrosis (HDN), Intravenous urogram revealed: right kidney showing filling defect in the renal pelvis and left kidney was normal (Fig. 5.3.1). CECT abdomen (on July 18, 2011): right hydroureteronephrosis (HDUN) with enhancing lesion in renal pelvis and upper ureter (Figs 5.3.2A to C). Diethylene triamine pentaacetic acid (DTPA) scan (on July 24, 2011): left kidney—normal in size, shape and location with mildly impaired parenchymal function and nonobstructed drainage pattern; right kidney—hydronephrotic, normal in size and location with severely impaired parenchymal function and likely obstructed drainage. GFR—right kidney: 12.6 mL/min (28.9%), left kidney: 31 mL/min (71.1%) (Fig. 5.3.3).
Upper Tract Urothelial Tumor

**Fig. 5.3.1:** Intravenous urogram

**Fig. 5.3.2A:** Contrast enhanced computed tomography (CECT) of the abdomen
Figs 5.3.2B and C: Contrast enhanced computed tomography (CECT) of the abdomen
Cystoscopy, right retrograde pyelography (RGP), ureteroscopy (URS) and ureteric stenting were done under general anesthesia (GA) on July 26, 2011. Cystoscopy revealed normal urethra and bladder. No tumor was detected. Right ureteric orifice was narrow. Guidewire followed by ureteric catheter was passed. Right RGP revealed a large filling defect in the renal pelvis. Ureter was narrow and did not admit the ureteroscope. Ureteric dilatation was done. Saline barbotage of renal pelvis was done and sample was sent for urine cytology. Urine cytology was positive for malignant cells.

Right robotic nephroureterectomy with cuff of bladder was done under GA on July 28, 2011.

Findings: Enlarged right kidney with dilated renal pelvis. No grossly enlarged lymph nodes. Lower ureter was normal (Figs 5.3.4A to C).

Histopathology

High grade papillary urothelial carcinoma with squamous differentiation; proximal ureter and pelvis; pathological stage: pT1pNxpMx; margins were free of tumor (bladder cuff). Lymphovascular invasion was not identified.
**FOLLOW-UP**

CECT abdomen post nephrectomy status revealed a recurrent soft tissue mass in the right renal fossa abutting the posterior crus of the diaphragm and extending into the right paravertebral space adjoining the L1 vertebra with nodular lesions, possibly metastatic (as not mentioned in the previous CT typed report) in bilateral adrenal glands. The left kidney, pelvicalyceal system, ureter and urinary bladder were normal. No local nodes were seen. PET-CT scan revealed a FDG avid soft tissue mass in right renal fossa closely abutting and possibly involving right crus of diaphragm, with a metabolically active right retrocrural lymph node.

Patient was given four cycles of injection gemcitabine $1,000 \text{ mg/m}^2$ intravenous (IV) on day 1 and day 8, injection carboplatin $AUC \ 5$ IV on day 1.

**COMMENTS**

A critical part of management of patients with TCCs is an awareness of the high rate of recurrence due to field effect on the urothelium. Adjuvant treatment is required for high grade invasive tumors as risk of metastasis is high.

**BIBLIOGRAPHY**

INTRODUCTION

Bilateral renal stones may occur in about 10% of the cases. In management of bilateral renal stones; more obstructed, symptomatic and infected side should be operated first. If one is contemplating nephrectomy on one side, then other kidney should be operated first. As per the site and size of stones, surgical intervention can be open surgery, percutaneous nephrolithotomy (PCNL) or ureteroscopy. Surgery can be performed simultaneously, or in two separate sittings.

Simultaneous bilateral PCNL has been demonstrated to be a well-tolerated, safe, cost-effective and expeditious treatment. The reduced total operative time, hospital stay, and total blood loss, along with the requirement for only one anesthesia, makes synchronous bilateral PCNL an attractive option for selected individuals.

CASE 1

Clinical Presentation

A 51-year-old male presented with complaints of bilateral flank pain for 2 years duration. The pain was dull, aching, intermittent and on and off with one severe episode in between. He also had one episode of hematuria.

Past History

Patient was a known hypertensive, for last 5–6 months and was not taking any medication for the same.
**Examination**

Pulse and blood pressure were normal. Abdomen was normal.

**Investigations**

Urine routine/microscopic (R/M): pH—6.5, Specific gravity—1.015, Protein—Nil, Sugar—Nil, Red blood cells urinary (RBC)—10–12/hpf, White blood cells (WBC)—70–80/hpf, Bacteria—positive, Cast—negative. Urine culture/sensitivity (C/S): Sterile. Blood urea: 36 mg/dL, Serum creatinine: 0.9 mg/dL. Serum calcium, uric acid and phosphate were normal. 24 hours urinary calcium, uric acid and phosphate were normal.

Ultrasound revealed: Left kidney—thin parenchyma, hydronephrotic with a calculus, Right kidney—hydronephrosis with multiple stones. Plain X-ray revealed multiple radiopaque shadows in both renal areas (Fig. 6.1.1). Intravenous urogram revealed bilateral renal stones with right hydronephrosis and non-excreting left system (Figs 6.1.2 and 6.1.3). Renal dynamic scan revealed glomerular filtration rate (GFR) revealed: Left kidney—7.6%, 3.9 mL/min, Right kidney—92.4%, 46 mL/min.
**Treatment**
Right PCNL was done followed by left nephrectomy.

**CASE 2**

**Clinical Presentation**
A 28-year-old male farmer presented with left flank pain for 1 year duration. There was no history of hematuria, pyuria, lithuria or urologic intervention in the past.

**Examination**
No organomegaly or any palpable lump, normal blood pressure

**Investigations**
Urine routine: pH—5.0, WBC—2/hpf, RBC—6/hpf. Urine culture: sterile, Renal function test: Urea—20 mg%, Creatinine—1.1 mg/dL. Plain X-ray revealed multiple radiopaque shadows in both renal areas (Fig. 6.1.4). Intravenous urogram revealed right hydronephrosis with left nonvisualized kidney (Fig. 6.1.5). Renal dynamic scan revealed GFR: Left kidney—5 mL/min, Right kidney—49 mL/min.

*Fig. 6.1.3: Case 1—Intravenous urogram delayed film*
Right PCNL was done in two stages followed by left nephrectomy.

**COMMENTS**

Management of bilateral renal calculi needs careful consideration for management in one sitting or in two sittings, technique for removal of calculi and conservative or ablative surgery.

**BIBLIOGRAPHY**

6.2 CROSSED RENAL ECTOPIA WITH CALCULI

**INTRODUCTION**

Renal ectopia is generally uncommon; its coexistence with nephrolithiasis is even rarer. Due to its variable presentation, it is usually discovered incidentally, especially while investigating patients for abdominal pain.

**CASE 1**

**Clinical Presentation**

A 50-year-old male presented with right iliac fossa pain for 1 year duration. He had history of diabetes mellitus for 5 years and hypertension for 3 years. There is past history of appendectomy via right paramedian approach.

**Examination**

Abdomen: right paramedian scar present, no organomegaly.

**Investigations**

Urine R/M and urine culture were normal. Renal function tests were normal. Plain X-ray kidney, ureter and bladder (KUB) region revealed a radiopaque shadow in right lumbar region (Fig. 6.2.1). Intravenous urogram revealed left to right crossed renal ectopia with hydronephrosis with renal calculi (Fig. 6.2.2). Noncontrast computed tomography (NCCT) abdomen revealed

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Fig. 6.2.1: Case 1—Plain X-ray kidney, ureter and bladder (KUB) region

Fig. 6.2.2: Case 1—Intravenous urogram
no kidney on the left renal area and fused kidney on right side with calculi (Fig. 6.2.3). Contrast enhanced computed tomography (CECT) abdomen revealed right fused kidney with hydronephrosis with calculi (Fig. 6.2.4).

**Fig. 6.2.3:** Case 1—Noncontrast computed tomography (NCCT) abdomen

**Fig. 6.2.4:** Case 1—Contrast enhanced computed tomography (CECT) abdomen

**Treatment**

He was explored and right open pyelolithotomy was done (Fig. 6.2.5). Postoperative plain X-ray revealed complete clearance of stones with double J (DJ) stenting (Fig. 6.2.6).
Fig. 6.2.5: Case 1—Picture of removed calculi

Fig. 6.2.6: Case 1—Postoperative plain X-ray
CASE 2

Clinical Presentation

A 12-year-old female presented with vague right flank pain for 6 months duration. No history of hematuria or urinary tract infection (UTI). Examination was normal.

Investigations

Urine R/M and urine culture were normal. Plain X-ray KUB region revealed radiopaque shadow in right renal area (Fig. 6.2.7). Intravenous urogram revealed left to right crossed renal ectopia with calculus in superior calyx of left moiety with normal functioning kidneys and no hydronephrosis (Figs 6.2.8A and B).

Fig. 6.2.7: Case 2—Plain X-ray kidney, ureter and bladder (KUB) region
Treatment

Two sittings of extracorporeal shock wave lithotripsy (ESWL) to the renal stone in superior calyx of left moiety was performed; stone completely cleared.

COMMENTS

Management of crossed renal ectopia with calculi is challenging because of altered anatomy. Careful planning is required for removal of stones without damage to other moiety. As per radiological findings, treatment can be planned by open surgery, PCNL or ESWL.

BIBLIOGRAPHY

6.3 EGGSHELL CALCIFICATION OF KIDNEY

**INTRODUCTION**

Eggshell calcification of kidney in case of ureteropelvic junction obstruction (UPJO) is an uncommon finding with only a few cases reported in literature. Although eggshell calcification is described with benign conditions, but surgical removal is advisable as 20% of the cases may develop malignancy.

**CLINICAL PRESENTATION**

A 30-year-old male presented with abdominal pain and palpable mass in the right lumbar region. He had no associated urinary or bowel symptoms. He had past history of pulmonary tuberculosis for which he was treated.

**EXAMINATION**

A 7 cm hard lump, nontender, bimanually palpable with restricted mobility was palpable in right lumbar and umbilical region.

**INVESTIGATIONS**

Hemogram was normal. Urine routine and urine culture were normal. Serum creatinine, serum calcium was normal. Urine for acid fast bacilli (AFB) and urine polymerase chain reaction (PCR) for acid fast bacilli was negative.

X-ray KUB region revealed an eggshell calcification in the right renal region (Fig. 6.3.1). Ultrasound and CECT revealed well-defined cystic kidney (Figs 6.3.2 and 6.3.3). Dynamic renal scan revealed right nonfunctioning kidney.

*Fig. 6.3.1: X-ray kidney, ureter and bladder (KUB) region*
Fig. 6.3.2: Contrast enhanced computed tomography (CECT)

Fig. 6.3.3: Ultrasound

**TREATMENT**

Right laparoscopic nephrectomy was done.

**HISTOPATHOLOGY**

Histopathology revealed fibrocollagenous cystic wall with hyalinization, calcification and ossification. No granulomatous inflammation or parasite was present.

**COMMENTS**

Eggshell calcification of the kidney is rare and should be factored in the differential diagnosis.
BIBLIOGRAPHY


6.4 ENDEMIC CALCULI

**INTRODUCTION**

The endemic bladder calculi have become rare in developed countries due to industrialization, improvement in nutrition and diet. These stones remain an important cause of morbidity in an endemic belt extending from the Middle East across India and Thailand to Indonesia, including North Africa. The incidence of endemic bladder calculi is high in children of lower socioeconomic status.

The composition of endemic bladder calculi includes most commonly ammonium acid urate alone, or in combination with calcium oxalate, but may also contain calcium phosphate. These stones form mainly due to dietary and nutritional deficiencies in these children, who are fed with a cereal based diet (low protein, high carbohydrate diet), lacking in animal proteins. Other predisposing risk factors are hot climate, chronic diarrhea or dehydration states, decreased animal milk intake (less than 25% of total protein intake: low phosphate, calcium), increased oxalate consumption (green leafy vegetables), nutritional deficiency of vitamin A, B₆ and magnesium, male sex (longer, tortuous urethra is a predisposing factor) and increased tissue turnover in children less than 10 years.

**Management**

Management depends on the size of the stone and can be managed by open or endourologic procedures.

**Prevention**

A mixed cereal diet supplemented with animal milk or animal proteins, along with correction of dehydration or diarrhea, and avoiding excess of oxalate consumption (green leafy vegetables) can decrease the incidence of endemic bladder stones.

**CLINICAL PRESENTATION**

A 4-year-old male presented with the history of fever, vomiting and diarrhea off and on since last 1 year. Patient was treated for enteric fever, and symptoms subsided. Four months later, patient developed hematuria and pyuria. Ultrasonography (USG) was done at a government hospital and patient was diagnosed to have bladder calculus and was started on medical treatment. Symptoms did not subside and patient had persistent pyuria and was referred. Patient was admitted with symptoms of decreased urine, altered sensorium, fever and anasarca. Bilateral percutaneous nephrostomy (PCN) and peritoneal dialysis was done. Symptoms improved. Bilateral PCN tubes were removed before discharge.
EXAMINATION

Facial puffiness was present. He had hypopigmented hair. No organomegaly, protruded abdomen. External genitalia was normal.

INVESTIGATIONS

Urine—Specific gravity: 1.005, pH: 7.5, Protein: 1+, Sugar: Nil, RBC: 8–10/hpf, WBC: 8–12/hpf, No crystals. Urine culture: Mixed growth. 24 hours urine analysis—Calcium: 0.51 g/L, Phosphate: 0.11 g/L, Uric acid: 0.18 mg/dL, Sodium: 30 mEq/L, Potassium: 8 mEq/L.

Plain X-ray KUB region (Fig. 6.4.1): vesical calculus with bilateral ureteric calculus. USG: bilateral kidneys—normal in size and echo texture, 2 cm calculus in urinary bladder. Two centimeter calculus seen at bilateral lower ureteric end. Calculus seen in lower pole of left kidney. Bilateral hydroureronephrosis (HDUN) with low level echoes seen within lumen suggestive of pyonephrosis. Left medullary nephrocalcinosis.

Intravenous urogram (Fig. 6.4.2): delayed excretion of contrast with clubbing of calyces with bilateral HDUN. NCCT abdomen (Fig. 6.4.3): vesical calculus with bilateral lower ureteric calculus with bilateral HDUN, multiple small calculi in lower pole of right kidney. 3 × 3 cm hypodense lesion with thick walls seen arising from lower pole of left kidney with 1.2 cm calculus within.

Noncontrast computed tomography of the head: sclerosis of cranial bones likely due to renal osteodystrophy.

Fig. 6.4.1: Plain X-ray kidney, ureter and bladder (KUB) region
Fig. 6.4.2: Intravenous urogram

Fig. 6.4.3: Noncontrast computed tomography (NCCT) abdomen
**TREATMENT**

The bladder and bilateral ureteric calculi were treated by open surgery. Left renal calculi were removed by PCNL.

**COMMENTS**

Endemic calculi are now rare and can be prevented by proper nutrition.

**BIBLIOGRAPHY**

INTRODUCTION

Horseshoe kidney is more prone to stone formation because of stasis of urine due to abnormal position. The management options are open surgery or percutaneous nephrolithotomy (PCNL). The outcome of extracorporeal shock wave lithotripsy (ESWL) for treatment is poor.

Percutaneous nephrolithotomy is becoming first-line management technique for staghorn stones and stones located in a horseshoe kidney. It can be challenging due to the altered anatomical relationship in the retroperitoneum, and usually requiring upper pole access and flexible nephroscopy due to the altered anatomical relationships of the fused renal units. PCNL in horseshoe kidneys is no more difficult than normal kidneys and does not carry a greater risk than reported for normal kidneys.

CASE 1

Clinical Presentation

A 35-year-old male presented with left flank pain. No past history of stone disease. Abdominal palpation was normal. Plain X-ray revealed radiopaque shadows in left renal area, the shape was abnormal and the lower end medially deviated (Fig. 6.5.1). Intravenous urogram confirmed horseshoe kidney with calculi in left moiety (Fig. 6.5.2).
**Treatment**

Left PCNL was done.

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**CASE 2**

**Clinical Presentation**

A 55-year-old male presented with history of vague pain in abdomen. Plain X-ray revealed bilateral large renal calculi with abnormal shape and lower ends were directed medially (Fig. 6.5.3). Metabolic evaluation was normal. Serum creatinine was 2 mg/dL. Dynamic renal scan revealed very poorly functioning right kidney and normal functioning left kidney.

![Case 2—Plain X-ray kidney, ureter and bladder (KUB) region](image)

**Treatment**

Open surgery was done. Right moiety nephrectomy with isthmectomy was done. Left side stones were removed.

**Follow-Up**

Patient is doing well after 15 years with serum creatinine of 2 mg/dL with no recurrence of stones.
CASE 3

Clinical Presentation

A 42-year-old male presented with history of left flank pain. Plain X-ray KUB region revealed bilateral renal calculi with medial rotation (Fig. 6.5.4). Intravenous urogram revealed horseshoe kidney with bilateral renal calculi (Fig. 6.5.5).

Treatment

Left PCNL followed by right PCNL.

COMMENTS

Management of horseshoe kidney with renal calculi is challenging and proper planning is necessary. PCNL is becoming the treatment of choice.

BIBLIOGRAPHY

Matrix stones are an uncommon form of urinary calculi, presenting a diagnostic and therapeutic dilemma to the practicing urologist. These radiolucent concretions are composed primarily of a noncrystalline mucoprotein matrix. Matrix calculi are more common in females. Urinary tract infection (UTI), usually with *Proteus* species or *Escherichia coli*, is a known predisposing factor for developing matrix calculi. The successful management of urinary matrix calculi depends on a high index of suspicion. They are best treated by percutaneous or surgical extraction, sterilization of the urine and maintenance of dilute urine. Extracorporeal shock wave lithotripsy is unsuccessful because of the gelatinous nature of the stone and lack of a crystalline structure.

**Clinical Presentation**

A 15-year-old male presented with left flank pain off and on since last 3 years. No history of hematuria, infection and fever.

**Examination**

Abdomen was normal. No lump palpable.

**Investigations**

Urine R/M examination revealed plenty of WBC and RBC. Urine C/S revealed *E. Coli* infection. Plain X-ray KUB region revealed multiple faceted radiopaque shadows in left renal area (Fig. 6.6.1). Intravenous urogram revealed normal
functioning right kidney. Left kidney was hydronephrotic with multiple renal calculi (Fig. 6.6.2).

**TREATMENT**

Left percutaneous nephrolithotomy was done by superior calyceal puncture. Stones along with jelly-like material was removed (Fig. 6.6.3). Nephrostomy catheter was kept. Relook nephroscopy and removal of remaining debris was done.

![Fig. 6.6.3: Picture of removed calculi and jelly like material](image)

**FOLLOW-UP**

Patient was given long term prophylactic antibiotics.

**COMMENTS**

Matrix stones are an uncommon form of urinary calculi and may be mistaken for tumors involving the renal collecting system, thereby presenting a diagnostic and therapeutic dilemma to the practising urologist. A high index of suspicion is required in diagnosing matrix calculi in addition to carefully selected radiographic imaging. Percutaneous removal is the primary treatment modality to render patients stone-free.

**BIBLIOGRAPHY**

6.7 MEGAURETER WITH CALCULI

INTRODUCTION

Adult and adolescent primary obstructive megaureter is a congenital abnormality that does not regress. The primary presentation of congenital megaureter in adults is rare. Complications such as stone formation and altered function of the affected kidney are common, and when associated with recurrent urinary tract infections, require surgical intervention. Urinary tract stones can form either within the dilated ureteral segment or in a part of the upper urinary tract proximal to the abnormal ureteral segment. Large stones can develop in the dilated portion of the ureter due to stasis of urine. Management of both megaureter and calculi can be done together.

CLINICAL PRESENTATION

A 55-year-old male presented with left flank pain for 5 months duration. There was no fever, hematuria or lithuria. He was a known diabetic taking oral hypoglycemic drugs for the same and was on medical management for benign prostatic hyperplasia (BPH). There was no history of tuberculosis (TB) or hypertension (HTN).

INVESTIGATIONS

Urine R/M: Normal; Urine C/S: Sterile. Renal function tests were within normal limits. Serum calcium, phosphate and uric acid were within normal limits. Ultrasound revealed multiple left renal calculi (Fig. 6.7.1). Plain X-ray KUB region revealed radiopaque shadows in left renal and left pelvis. Intravenous urogram revealed left hydroureteronephrosis with left megaureter with renal and lower end ureteric calculi (Fig. 6.7.2). Dynamic renal scan revealed delayed function and drainage from the left kidney (Fig. 6.7.3).

Micturating cystourethrogram (MCU): Bladder configuration and capacity was normal, posterior urethra was normal, no vesicoureteral reflux, insignificant post-void residual (PVR) urine.

Fig. 6.7.1: Ultrasound
TREATMENT

Left PCNL was done. Left renal stones were cleared. Three days later, open left ureterolithotomy with tailoring of ureter and ureteric reimplantation was done.
COMMENTS

Complicated megaureter with calculi needs surgical treatment for both stones and the megaureter.

BIBLIOGRAPHY


6.8 MILK OF CALCIUM IN KIDNEY

INTRODUCTION

Milk of calcium is a fine colloidal suspension of precipitated calcium salts (carbonate, phosphate and oxalate) and may be radiopaque or radiolucent. If radiopaque, it closely mimics renal calculus. Milk of calcium is usually asymptomatic and of little clinical significance. The etiology of milk of calcium in kidneys can be: (a) stasis; (b) urinary tract infection; (c) gravity; and (d) long-term physical immobility.

CLINICAL PRESENTATION

A 28-year-old female presented with right flank pain and there was no history of hematuria or urinary tract infection.

EXAMINATION

Abdominal examination was normal. No lump palpable.

INVESTIGATIONS

Urine R/M revealed RBCs and pus cells. Urine culture was positive for *E. Coli*. Plain X-ray in supine position revealed multiple radiopaque shadows in the kidney area (Fig. 6.8.1). Plain X-ray in standing position revealed radiopaque shadows with fluid levels (Fig. 6.8.2). Intravenous urogram revealed hydronephrosis with multiple calculi.

Fig. 6.8.1: Plain X-ray in supine position

Fig. 6.8.2: Plain X-ray in standing position
TREATMENT

Right PCNL was done. Multiple stones with jelly like material came out. Long term suppressive antibiotics were given.

COMMENTS

Milk of calcium in kidney can be confused with nephrocalcinosis or matrix calculi. Imaging by X-rays and ultrasound in standing position will help to differentiate the conditions. If symptomatic, PCNL is the treatment of choice.

BIBLIOGRAPHY

**INTRODUCTION**

In cases of ureteric calculi with non-functioning kidney, nephrectomy may be indicated. In such cases, nephroureterectomy should be done along with removal of calculi, otherwise patient will present with recurring UTI and symptoms due to residual ureteral stump.

**CLINICAL PRESENTATION**

A 36-year-old male presented with history of recurrent UTI since last 6 months with past history of left nephrectomy for calculus disease about 1 year back. No history of hematuria.

**EXAMINATION**

A left nephrectomy lumbar scar was present.

**INVESTIGATIONS**

Urine and blood examination was normal. NCCT scan revealed multiple left ureteric calculi in the line of left ureter (Fig. 6.9.1).

![Fig. 6.9.1: Noncontrast computed tomography (NCCT) scan](image)

**TREATMENT**

Left ureteroscopy and laser fragmentation and removal of all calculi were done.

**COMMENTS**

The ureter including the stones should be removed at the time of nephrectomy.

**BIBLIOGRAPHY**

INTRODUCTION

Prostatic parenchymal calculi are common, usually an incidental finding on morphological examinations. They are typically asymptomatic and may be present in association with normal glands, benign prostatic hyperplasia and prostate cancer.

CLINICAL PRESENTATION

A 59-year-old male presented with straining while micturition, frequency and dysuria for 1 year duration. Per rectal examination revealed enlarged prostate grade II, firm to hard, smooth and rectal mucosa free.

INVESTIGATIONS

Urine C/S was sterile. Plain X-ray pelvis revealed multiple, round small calculi in the prostatic region creating a horseshoe shape (Fig. 6.10.1). Ultrasound revealed normal kidneys, prostate—39 grams with multiple calculi with mixed echogenicity. Uroflowmetry revealed obstructed flow rate.
**TREATMENT**

Bipolar transurethral resection of prostate (TURP) with removal of calculi was done.

**FOLLOW-UP**

His urine flow improved and urine C/S was sterile.

**COMMENTS**

Patients with prostatic calculi can have associated infection and obstruction which needs surgical treatment.

**BIBLIOGRAPHY**

6.11 STEINSTRASSE FOLLOWING EXTRACORPOREAL SHOCK WAVE LITHOTRIPSY

**INTRODUCTION**

“Steinstrasse” is a German word which means “stone street”. It is a known possibility following extracorporeal shock wave lithotripsy (ESWL). Stone size and site, renal morphology and shock wave energy are the significant predictive factors controlling steinstrasse formation. It is associated with patient discomfort, infection or impaired renal function. The meticulous follow-up of patients with steinstrasse should prevent any loss of renal function. When there is obstruction and/or infection or renal damage, active treatment is indicated, of which ESWL and Percutaneous Nephrolithotomy (PCN) are the most effective, with ureteroscopy and open surgery being reserved for difficult cases.

**CLINICAL PRESENTATION**

A 15-year-old male presented with history of left renal calculus. In another hospital, left DJ stenting and ESWL was done. He developed high fever and was referred for further treatment.

**INVESTIGATIONS**

Plain X-ray KUB region revealed left renal calculus with DJ stent in situ (Fig. 6.11.1). Intravenous urogram revealed normal functioning kidney renal pelvic calculus (Fig. 6.11.2). Post ESWL, plain X-ray KUB region showed multiple calculi in the ureter and lower pole of the kidney (Figs 6.11.3A and B).

![Fig. 6.11.1: Plain X-ray kidney, ureter and bladder (KUB) region](image-url)
**TREATMENT**

Left PCN was done and broad spectrum antibiotics were given. His fever settled. Plain X-ray KUB region after PCN revealed few calculi in the kidney and ureter (Fig. 6.11.4). After that, left ureteroscopic removal of ureteric and renal calculi was done. Figure 6.11.5 shows plain X-ray of the KUB region after removal of calculi. PCN catheter was removed. Ureteric stent was removed after 4 weeks.
COMMENTS

The optimum selection of cases (aiming to pulverize the stones rather than fragment them) and accurate stone targeting are essential to minimize the development of steinstrasse.

BIBLIOGRAPHY

INTRODUCTION

Ureteric calculi are usually small, solitary and take shape of the ureter. If ureter is large and dilated, ureteric calculus can be giant and take round or other kind of shapes.

CLINICAL PRESENTATION

A 32-year-old male presented with right flank pain off and on since last 6 months.

EXAMINATION

Abdominal examination was normal.

INVESTIGATIONS

Plain X-ray KUB region revealed kyphoscoliosis with deviation to left side and a V-shaped radiopaque shadow at the level of L4 vertebra on the left side (Fig. 6.12.1). Intravenous urogram revealed right hydroureteronephrosis with dilated and tortuous ureter above the stone (Fig. 6.12.2). Computed tomography (CT) urogram noncontrast and contrast confirmed the same findings.
**TREATMENT**

Right ureterolithotomy was done. Patient recovered well.

**COMMENTS**

Ureteric calculus takes the shape of the ureter; therefore, they can be of abnormal size.

**BIBLIOGRAPHY**

6.13 URETERIC STRICTURE FOLLOWING URETEROSCOPY

**INTRODUCTION**

Ureteral mucosal injury is common but ureteric stricture is rarely formed. The etiology is ureteric perforation, ischemia and use of large size of ureteroscope and ureteral sheath. Prolonged duration of surgery also leads to ischemia. For prevention of ureteric injury; if ureter is narrow, ureteric dilatation should be done. Small size ureteroscope should be used and in case of any problem, staged ureteroscopy can be done.

**CLINICAL PRESENTATION**

A 35-year-old male presented with the history of left flank pain for 3 months duration.

**INVESTIGATIONS**

Plain X-ray KUB region revealed a small ureteric calculus in lower end of left ureter (Fig. 6.13.1). Intravenous urogram revealed mild hydroureteronephrosis (HDUN) on the left side (Fig. 6.13.2).

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**Fig. 6.13.1:** Plain X-ray kidney, ureter and bladder (KUB) region

**Fig. 6.13.2:** Intravenous urogram
TREATMENT

Patient underwent left ureteroscopy and stone removal. After 6 months, he presented to us with complaints of left flank pain. Intravenous urogram revealed left HDUN with delayed drainage (Fig. 6.13.3). Left ureterogram revealed stricture in left lower ureter. Thereafter, left ureteric reimplantation was done as treatment.

Fig. 6.13.3: Post ureteroscopy intravenous urogram

COMMENTS

Ureter is a very soft and delicate structure susceptible to iatrogenic injury during ureteroscopy.

BIBLIOGRAPHY

INTRODUCTION

Tuberculosis is a leading public health problem particularly in the developing nations of South East Asia. *Mycobacterium* is the causative agent of tuberculosis (TB), and genitourinary tuberculosis (GUTB) is a secondary manifestation either by hematogenous spread to the kidneys and epididymis, by reactivation of dormant bacilli or reinfection. There is a latent period of three to ten years for development of GUTB after primary infection. In 1937, Wildbolz coined the term “genitourinary tuberculosis” as this is a systemic disease and can involve multiple organs in the genitourinary system.

Renal involvement of TB starts with bacilluria, followed by ulceration at the apex of pyramid, which results into ulcerocavernous lesion, locked or lost calyx, pelviureteric junction (PUJ) stenosis, calcification which may lead to nonfunctioning kidney.

Ureteric lesions start with ureteritis, which may result into stricture formation or vesicoureteric reflux. The lower one third of the ureteric stricture is the most common form of manifestation. However, there can be involvement of middle and upper ureter. There can be single or multiple strictures in the ureter.

The bladder lesions start around ureteric orifice with inflammatory bullous edema, which later on can become golf-hole orifice. There can also be tubercular ulcers in the bladder. By healing, it results into fibrosis and small capacity bladder (thimble bladder), which can be associated with vesicoureteric obstruction or vesicoureteric reflux.

Tuberculosis can involve all parts of the genitourinary (GU) tract and is termed as global GUTB. The sequelae of TB is renal failure in 12% and hypertension in 4–12%. 5-year survival without surgery in 15–42% and 10-year survival with surgery in 50%.

The most common presenting symptoms in patients of genitourinary tuberculosis are irritative voiding symptoms and hematuria. A high index of suspicion is required for the diagnosis of tuberculosis, especially in patients with recurrent urinary tract infection. Diagnosis is made by nonspecific
investigations like acid sterile pyuria, raised erythrocyte sedimentation rate (ESR) or by direct investigations like presence of acid fast bacilli in urine smear and culture, urinary polymerase chain reaction (PCR) for TB and tissue diagnosis. The common radiological abnormalities seen were calcification, cortical scarring, calyceal cavitation and destruction, nonvisualized kidney, ureteric stricture or irregularity, and small capacity bladder. Intravenous urogram is the cornerstone in the diagnosis of GUTB. Ultrasound is commonly available and can detect cavities in the kidneys. However, three-dimensional spiral CT with urogram gives complete information. Percutaneous antegrade pyelogram is indicated in patients with percutaneous nephrostomy. Cystogram depicts changes in the bladder as well as associated with vesicoureteric reflux. Magnetic resonance imaging and renal angiogram have selective indications in selected cases.

Guidelines for management of renal TB are chemotherapy in stage I disease, conservative surgery in stage II, and ablative surgery in stage III. Drainage for hydronephrosis can be done by ureteric stenting or percutaneous nephrostomy in patients presenting with bilateral hydronephrosis/hydronephrosis of solitary kidney, renal insufficiency and septic complications need immediate surgical intervention and percutaneous nephrostomy can salvage renal function.

Percutaneous drainage of abscesses or localized collections can be done. Local treatment of the affected part can be by way of cavernotomy or calyceorrhaphy or partial nephrectomy or infundibuloplasty. Reconstruction of the upper urinary tract can be done by pyeloplasty or ureterocalicostomy. However, the results are poor.

Modern antitubercular chemotherapy remains the cornerstone of management of genitourinary tuberculosis. A short course combination chemotherapy is the standard of care, in which 4-drug regimen for two months and 2-drug regimen for 4 or 6 months is prescribed. Directly observed treatment short course or DOTS is acceptable, more effective with less complications. DOTS has become popular in India. In drug resistance TB, second line of treatment is used.

The role of surgery is complementary to antitubercular chemotherapy. Surgery is usually undertaken 4–8 weeks after instituting antitubercular treatment. Early obstructive lesions of the urinary tract can be managed initially with minimally invasive (endoscopic reconstructive) procedures, failing which, open surgery may be offered according to the merits of the case. Endoscopic procedures include urethral dilatation, bladder neck incision, internal urethrotomy, endopyelotomy, endoureterotomy and ureteric calibration, dilatation, and stenting. Reconstructive surgery in genitourinary tuberculosis is required in cases with a grossly distorted and dysfunctional anatomy that is unlikely to regress with chemotherapy alone. Reconstructive surgery consists of partial nephrectomy, pyeloplasty, augmentation cystoplasty with or without ileal replacement of ureter by use of various bowel segments.
Nephrectomy is recommended for nonfunctioning kidneys as it ensures removal of the infective pathology and decreases chances of resurgence of the disease. Nephroureterectomy is performed if there is associated ureteric pathology or vesicoureteric reflux. In some patients, who present with chronic renal failure, permanent nephrostomy may be required.

This chapter presents various aspects of genitourinary tuberculosis and various cases and features will be depicted.

**BIBLIOGRAPHY**

7.1 GENITOURINARY TUBERCULOSIS — AUGMENTATION CYSTOPLASTY

**INTRODUCTION**

Surgical management of tubercular thimble bladder is by augmentation cystoplasty to increase the capacity of the bladder. Any segment of bowel from the stomach to the sigmoid colon can be used. The choice of bowel for reconstruction is purely the surgeon’s prerogative—his skill, the ease, the mobility and length of mesentery (allowing bowel to reach the bladder neck without tension and maintaining an adequate blood supply). Ileum/ileocelec segment is most commonly used bowel segment. Sigmoid colon can be used by being opened along its antimesenteric border to form a capacious pouch. Gastrocystoplasty reduces the risk of acidosis and can be used in patients with compromised renal function but is associated with complications like hypochloremic alkalosis and ‘Hematuria-dysuria’ syndrome. Orthotopic neobladder reconstruction is a feasible option, suitable in cases of tubercular thimble bladder with a markedly reduced capacity (as little as 15 mL) where an augmentation alone may be associated with anastomotic narrowing or poor relief of symptoms.

**CASE 1**

**Clinical Presentation**

A 26-year-old male presented with frequency and dysuria. Radiological features revealed bilateral hydroureteronephrosis with small capacity bladder (Fig. 7.1.1). After anti-tuberculosis treatment (ATT), symptoms persisted and bladder was small in size.

**Treatment**

The patient underwent augmentation cystoplasty with ileocelec segment with reimplantation of ureter in the ileum. He did well after surgery. In follow-up for 10 years, he has no problems except slight mucus in the urine. Postoperative intravenous urogram (IVU) (Fig. 7.1.2) showed residual bilateral hydroureteronephrosis (HDUN) with augmented bladder.

**CASE 2**

**Clinical Presentation**

A 24-year-old male presented with symptoms of frequency and dysuria. Intravenous urogram (Fig. 7.1.3) revealed right HDUN with nonvisualized left kidney. After ATT, his symptoms persisted.

**Treatment**

He underwent left nephroureterectomy with augmentation cystoplasty. In follow-up, he did well. Intravenous urogram (Fig. 7.1.4) shows residual bilateral HDUN with augmented bladder and post-nephrectomy status left side.
COMMENTS

Long-term outcome is good in patients with augmentation cystoplasty. In cases with nonfunctioning kidneys, nephroureterectomy can be combined with augmentation cystoplasty.
7.2 GENITOURINARY TUBERCULOSIS—GASTRIC AUGMENTATION CYSTOPLASTY WITH TRANS-URETERO-URETERAL ANASTOMOSIS

INTRODUCTION

Genitourinary tuberculosis (GUTB) can be associated with compromised renal function. Use of ileum and colon in urinary tract with compromised renal function is not recommended due to higher incidence of metabolic complications. In such situations, stomach can be used. Transureteroureterostomy is an option for ureteral replacement in nonmalignant diseases, where other options are not available.

CLINICAL PRESENTATION

A 46-year-old male presented with frequency, dysuria and recurrent painless hematuria for 2 years.

PAST HISTORY

The patient had bilateral percutaneous nephrolithotomy (PCNL) in December 1999, followed by right PCNL for residual calculi in January 2001. He developed gross painless hematuria with frequency in February 2001. Diagnosed as GUTB, acid-fast bacillus (AFB) was positive in 2001. He had antituberculosis treatment (ATT) for 9 months. No history of HT/DM.

EXAMINATION

Examination was normal. He had bilateral PCN in situ with scar marks of bilateral PCNL.

INVESTIGATIONS

Urine R/M—Protein: Traces, WBC: 1–2/hpf, RBC: 2–4/hpf. Urine culture: Sterile, AFB microscopy: Negative, Blood Investigations—Hb: 8.0 gm%, TLC: 4,500/mm³, Urea: 70 mg%, Creatinine: 3.2 mg%. Plain X-ray KUB region (Fig. 7.2.1) revealed small residual calculus in the right kidney. Ultrasound (Figs 7.2.2A to C) revealed HDN left kidney with small capacity bladder. MR urogram (Fig. 7.2.3) revealed left HDUN up to lower ureter, right contracted hydronephrotic kidney with small capacity bladder. Bilateral PCN was done. Plain X-ray KUB region (Fig. 7.2.4) shows bilateral PCN tubes. Bilateral nephrostogram (Fig. 7.2.5) revealed bilateral HDUN with right lower ureteric stricture. Dynamic renal scan (Fig. 7.2.6) revealed right poorly functioning kidneys, right more than left. Micturating cystourethrogram (MCU) revealed small capacity bladder with bilateral vesicoureteral reflux (VUR), left dilated ureter and right lower ureteric stricture (Figs 7.2.7A and B).
**Fig. 7.2.1:** Plain X-ray KUB region

**Figs 7.2.2A to C:** Ultrasound
In view of compromised renal function and stricture of right lower ureter, gastrocystoplasty with transureteroureterostomy with joining of the right ureter to the left was done. The patient recovered well. Postoperative X-ray (Fig. 7.2.8) shows bilateral double J (DJ) stents in position. Micturating cystourethrogram (Fig. 7.2.9) shows reflux on left side with augmented bladder. Both nephrostomy catheters were removed. Patient had mild chronic renal failure (CRF) postoperatively but his symptoms improved.
**COMMENTS**

In patients with compromised renal function, gastric segment can be used for augmentation cystoplasty. In view of thick wall of the stomach, ureteric reimplantation is difficult. Due to stricture of the right ureter, transureteroureterostomy was done to salvage the situation.
INTRODUCTION

The sequel of tuberculosis is renal failure in advanced cases with involvement of both kidneys due to long standing infection and obstruction.

CLINICAL PRESENTATION

A 42-year-old male presented with history of frequency, dysuria and loss of appetite.

INVESTIGATIONS

Serum creatinine of 5.2 mg/dL. Noncontrast CT scan revealed bilateral hydroureteronephrosis with small capacity bladder (Fig. 7.3.1). Dynamic renal scan revealed bilateral poorly functioning kidneys with poor drainage (Fig. 7.3.2). Bilateral percutaneous nephrostomy (PCN) (Fig. 7.3.3) was done. On the right side, separate PCN was done on superior and inferior calyx for proper drainage.

Fig. 7.3.1: Noncontrast computed tomography scan

Fig. 7.3.2: Dynamic renal scan
**Fig. 7.3.3:** Plain X-ray after percutaneous nephrostomy

**TREATMENT**

As renal parameters did not improve, he was left with bilateral PCN.

**COMMENTS**

In some cases, renal function cannot be salvaged in spite of draining the kidney and the patients have to live with nephrostomy catheter.
7.4 GENITOURINARY TUBERCULOSIS WITH CONGENITAL SINGLE KIDNEY

■ INTRODUCTION

Genitourinary tuberculosis can be associated with congenital single kidney.

■ CLINICAL PRESENTATION

A 23-year-old male presented with symptoms of frequency of urination in 1980.

■ INVESTIGATIONS

Investigations revealed left congenital absent kidney with small capacity bladder with mild hydroureteronephrosis (HDUN). Cystoscopy revealed right hemitrigone with absent left ureteric orifice. After antituberculosis treatment (ATT) for 6 months, his symptoms persisted.

Intravenous urogram revealed normal functioning right kidney with mild HDUN with thimble bladder (Fig. 7.4.1).

![Fig. 7.4.1: Preoperative intravenous urogram](image1)

![Fig. 7.4.2: Follow-up intravenous urogram after 25 years](image2)
**TREATMENT**

Augmentation cystoplasty was done with ileocecal segment. His recovery was uneventful.

**FOLLOW-UP**

He was regularly coming for follow-up. His intravenous urogram in 2004, i.e. after 25 years revealed normally functioning right kidney with augmented bladder (Fig. 7.4.2).

**COMMENTS**

This case illustrates that augmentation cystoplasty has a good long-term outcome and can be done in single functioning kidney.
7.5 GENITOURINARY TUBERCULOSIS WITH REPLACEMENT OF URETER

**INTRODUCTION**

Ureteric stricture following tuberculosis is not uncommon. Management of ureteric stricture poses both a diagnostic dilemma as well as taxes the surgical skills of the reconstructive surgeon. If not properly managed, the kidney may be lost. For long ureteric strictures involving almost the whole length of ureter or upper ureteric strictures require ileal replacement of ureter.

**CLINICAL PRESENTATION**

A 45-year-old female presented with urinary frequency and dysuria and right flank pain.

**INVESTIGATIONS**

Intravenous urogram revealed nonvisualization of right kidney with small capacity bladder (Fig. 7.5.1). Urine examination for AFB was negative. Ultrasound revealed right HDUN. Right PCN was done. The output from right kidney was 600–700 mL. Right nephrostogram revealed mild HDUN with obstruction at the midureter level.

![Fig 7.5.1: Preoperative intravenous urogram](image1)

![Fig 7.5.2: Follow-up intravenous urogram](image2)
**TREATMENT**

Augmentation cystoplasty with ileal replacement of ureter was done.

**FOLLOW-UP**

Intravenous urogram revealed normal functioning right kidney with ileal ureter with augmentated bladder (Fig. 7.5.2). She is on regular follow-up for the last 15 years and is normal.

**COMMENTS**

Kidney function can be salvaged in mid and upper ureteric tubercular stricture ureter by ileal replacement of ureter along with augmentation cystoplasty.
7.6 GENITOURINARY TUBERCULOSIS WITH RENAL CALCIFICATION

**INTRODUCTION**

Presence of calcification in tubercular kidney is common and the incidence is increasing. Tubercular calcification can also be associated with renal calculi. The etiology of calcification is obscure, as there is no evidence to support different pathogenesis for tubercular calcification and for discrete renal calculi. The precipitating factors can be recumbency, high calcium intake, recurrent UTI, obstructive uropathy and hypercalciuria. The small calcification can be observed with anti tubercular treatment (ATT) whereas larger area of calcification should be excised and nonfunctioning kidneys with extensive calcification removed.

**CLINICAL PRESENTATION**

There is no specific clinical presentation in GUTB with calcification. Calcification is detected on imaging (Figs 7.6.1 to 7.6.5).

![Fig. 7.6.1: Plain X-ray showing small renal calcification](image1)

![Fig. 7.6.2: Plain X-ray showing diffuse cortical renal calcification](image2)
Fig. 7.6.3: Plain X-ray showing two small renal calcifications with a calculus

Fig. 7.6.4: Plain X-ray and intravenous urogram showing calcified nonfunctioning kidney—also known as putty kidney
**Fig. 7.6.5:** Plain X-ray showing renal calcifications with renal calculi

**COMMENTS**

Tubercular calcification can be associated with renal calculi and along with ATT needs surgical treatment. Tubercular calcification is to be differentiated from renal calculi which can be associated with GUTB.
7.7 GENITOURINARY TUBERCULOSIS WITH MULTIPLE ORGAN INVOLVEMENT

**INTRODUCTION**

Genitourinary tuberculosis is a systemic disease and can involve multiple sites in the urinary system.

**CLINICAL PRESENTATION**

A 26-year-old-male presented with difficulty in passing urine, frequency and dysuria.

**INVESTIGATIONS**

Investigations revealed raised erythrocyte sedimentation rate (ESR) and normal renal functions. Intravenous urogram revealed nonvisualization of right kidney with HDUN of left kidney with small capacity dumbbell-shaped urinary bladder (Figs 7.7.1A and B). Retrograde urethrogram revealed multiple strictures of proximal penile urethra (Fig. 7.7.2).

![Figs 7.7.1A and B: Intravenous urogram](image-url)
**TREATMENT**

After starting ATT, Johanson’s 1st stage urethroplasty was done for the stricture urethra. After 1 week, right nephroureterectomy with augmentation cystoplasty was done. The patient recovered well. In follow-up, 2nd stage urethroplasty was done.

**COMMENTS**

Genitourinary tuberculosis can involve kidney, ureter, bladder and urethra in the same patient. With proper medical and surgical treatment, kidney function can be salvaged.
7.8 GENITOURINARY TUBERCULOSIS WITH NONFUNCTIONING KIDNEY WITH THIMBLE BLADDER

**INTRODUCTION**

In genitourinary tuberculosis, kidney can become nonfunctioning due to involvement of kidney, ureter and bladder if diagnosed late. Nonfunctioning kidney can be due to ureteric obstruction or due to parenchymal involvement.

**CLINICAL PRESENTATION**

A 27-year-old male, presented with frequency of urination and dysuria.

**INVESTIGATIONS**

Investigations revealed raised ESR and acid sterile pyuria. Urine AFB was negative. Intravenous urogram revealed left nonvisualized kidney with HDUN of the right kidney with small bladder (Fig.7.8.1). Micturating cystourethrogram revealed small capacity bladder with right hydroureteronephrosis (Fig. 7.8.2).

![Fig. 7.8.1: Intravenous urogram](image1)

![Fig. 7.8.2: Micturating cystourethrogram (MCU)](image2)
**TREATMENT**

After ATT, he underwent left nephroureterectomy with augmentation cystoplasty. Nephrectomy specimen (Fig. 7.8.3) showed caseation with destruction of the parenchyma.

![Nephrectomy specimen](image)

**Fig. 7.8.3: Nephrectomy specimen**

**COMMENTS**

With the availability of newer ATT drugs, it is advisable to do nephrectomy to eradicate the source of infection to avoid resurgence of the disease.
7.9 GENITOURINARY TUBERCULOSIS—PARTIAL NEPHRECTOMY

**INTRODUCTION**

Genitourinary tuberculosis can involve only one pole of the kidney or one moiety in double collecting system. The remaining part of the kidney can be saved by partial nephrectomy.

**CLINICAL CASES**

**Case 1**

Intravenous urogram (Fig. 7.9.1) shows poorly functioning right upper pole of the kidney with normal functioning lower pole. The other kidney and bladder was normal. Right upper pole partial nephrectomy was done. Histopathology confirmed tubercular involvement of the kidney.

**Case 2**

Intravenous urogram shows bilateral double moiety with nonfunctioning right upper moiety. Right superior moiety nephroureterectomy was done. Follow-up intravenous urogram (Fig. 7.9.2) shows normally functioning right lower moiety.

**COMMENTS**

Remaining kidney can be salvaged by partial nephrectomy in GUTB.
7.10 TUBERCULAR URETERIC STRICTURE

**INTRODUCTION**

Following tubercular infection, the initial inflammatory response heals by fibrosis that leads to subsequent stricture formation. The most common site of stricture formation is the ureterovesical junction (UVJ). Stricture formation is also seen at the level of ureteropelvic junction and less commonly in the middle third of the ureter. Although rare, sometimes the entire length of ureter is involved. The length of stricture varies in length but UVJ strictures are usually less than 5 cm in length. The area of fibrosis is usually localized to the area of stricture only. Extensive fibrosis can shorten the length of the ureter, pulling up the orifice as a gaping hole—the so-called “golf hole”.

Early cases can be managed by endoscopic procedures like ureteral dilatation, double J stent, endoscopic incisions, etc. In established stricture, treatment is by ureteral substitution according to the function of the kidney and site of stricture. If associated with tubercular bladder, then augmentation cystoplasty with replacement of ureter is advisable.

**CLINICAL CASES**

**Case 1**

Retrograde ureterogram showed stricture at the level of pelviureteric junction managed by pyeloplasty (Fig. 7.10.1).

**Case 2**

Retrograde ureterogram shows stricture of middle ureter managed by regular ureteral dilatations (Fig. 7.10.2).

**Case 3**

Retrograde ureterogram shows stricture of lower ureter managed along with augmentation cystoplasty (Fig.7.10.3).
COMMENTS

Ureteric stricture should be identified early to prevent deterioration in renal function and should be managed adequately.
INTRODUCTION

Bladder tuberculosis generally results in patchy cystitis due to inflammation by the tubercle bacilli—*Mycobacterium tuberculosis*, of the urothelium. Resultant granulomatous inflammation, caseation necrosis, and final healing by fibrosis may lead to marked contracture of the urinary bladder within an year or so, the early disease being nonspecific. Two types of lesions in the tubercular bladder are common. The first type—which is more common, is when the bladder due to active infection has a reduced capacity of about 150–200 mL. The other type—true or structural bladder contracture, wherein the urinary bladder has permanently lost its capacity and has little or no value as a urinary reservoir. In the initial stages before cicatrization has taken place, the dome contracts but the trigone and bladder neck are relatively spared. The appearance of the bladder can be dumbbell shape. Tubercular bladder can be associated with vesicoureteric reflux and also obstruction at the vesicoureteric junction.

Antituberculosis treatment is often successful in preventing disease progression and restoring normal bladder function. But once the tubercular bladder shrinks to a very small size—tubercular thimble bladder—the process may no longer be reversible and corrective (reconstructive) surgery in the form of augmentation must be performed to prevent/arrest kidney damage. This entails enlarging the capacity of the small, contracted, irreparably damaged bladder that causes intolerable frequency.

CLINICAL CASES

Case 1

A 32-year-old female presented with frequency and dysuria. Urine for AFB was positive. Intravenous pyelogram (IVP) revealed normal upper tracts and cystogram revealed early changes in the bladder (Fig. 7.11.1). ATT was started and she improved completely. ATT was discontinued after 9 months.

Case 2

A 28-year-old male presented with frequency of urination and dysuria. Urine for AFB was positive. Cystogram revealed small rounded bladder with grade 1 right vesicoureteric reflux (VUR) (Fig. 7.11.2). ATT was started followed by augmented cystoplasty.

Case 3

This is another similar case presentation. Cystogram revealed small elongated bladder pulled to right side with VUR (Fig. 7.11.3). ATT was started followed by augmented cystoplasty.
Case 4
This is another similar presentation with frequency of urination every 2 hours. Cystogram revealed double bell shaped bladder with right grade 3 VUR (Fig. 7.11.4). ATT was started followed by augmented cystoplasty.

Case 5
A 32-year-old male presented with severe frequency of urination. Cystogram revealed small capacity bladder with left VUR (Fig. 7.11.5). ATT was started followed by augmented cystoplasty.

Case 6
A 45-year-old male presented with continuous passing of urine at both day and night. He used urine pot to collect the urine. Cystogram revealed thimble bladder of 5 mL capacity (Fig. 7.11.6). After ATT, due to very small bladder, cystectomy and orthotopic neobladder was done.
In GUTB, involvement of the bladder is very common and patients can present with a wide spectrum of bladder as shown in the above six cases. In early cases with bladder spasm, medical treatment alone can help, otherwise augmentation cystoplasty is required.
7.12 TUBERCULAR EPIDIDYMITIS

**INTRODUCTION**

Tuberculous/tubercular epididymitis is the most common kind of tuberculosis of male genitourinary tract. Usually, it is accompanied with tuberculosis of prostate, tuberculosis of seminal vesicle, tuberculosis of kidney, retrograde infection of prostate and vas deferens, or partial hematogenous infection. The key symptom of tuberculosis of epididymis is manifested as the inflammation and swelling of epididymis that would form epididymal induration, abscess, then sinus can be formed.

If chronic epididymitis does not resolve with antibiotics, then one should suspect TB and fine needle aspiration cytology (FNAC) can clinch the diagnosis. Management is by ATT and if it is nonresolving, then epididymectomy with or without orchidectomy can be done.

**CLINICAL PRESENTATION**

A 65-year-old male presented with a scrotal swelling on the left side of scrotum for 6 months duration. He had no past history of tuberculosis. On examination, he has enlarged epididymis, firm in consistency and could not be separated from the epididymis.

Figs 7.12.1A and B: Clinical pictures
## TREATMENT

He was treated with antibiotics but did not improve. The swelling increased in size, indurated and developed a discharging sinus (Figs 7.12.1A and B). Anti TB treatment was started. Swelling slightly decreased in size but sinus was persisting. Right epididymo-orchidectomy was done (Figs 7.12.2A and B). ATT was continued for 1 year. Histopathology confirmed TB epididymitis.

![Image of Epididymo-orchidectomy specimen](image)

**Figs 7.12.2 A and B:** Epididymo-orchidectomy specimen

## COMMENTS

Unresolving chronic epididymitis can be due to tuberculosis and should be suspected.
7.13 TUBERCULAR PROSTATITIS

## INTRODUCTION

Mycobacterial tuberculosis infection of prostate is rare and can only occur after pulmonary infection or miliary dissemination. Small (1–2 mm) caseating granulomas coalesce within the prostatic parenchyma, forming yellow nodules and streaks. Caseation and cavitation can be extensive. There are no specific presenting symptoms for tuberculosis. Usually patients present with lower urinary symptoms and are diagnosed on biopsy of prostate or in transurethral resection of prostate (TURP) chips. Management is by ATT.

## CLINICAL PRESENTATION

A 51-year-old man presented on 20th April 2013 with complaints of poor urinary stream for 3 months. Digital rectal examination revealed prostate grade I, firm prostate. There was a firm nodule on the right lobe of the prostate.

## INVESTIGATIONS

Urine R/M showed 5–7 pus cells/hpf; Urine C/S: Sterile, Uroflowmetry—MFR: 11.1 mL/s; AFR: 5.5 mL/s; VV: 315 mL, Ultrasound KUB showed grade I prostate with PVR of 24 cc, Serum PSA: 5.47 ng/mL. MRI (Figs 7.13.1A and B) showed a 31 cc prostate with hypointense peripheral nodules on the right side with type 2 and type 3 enhancement kinetics suggestive of mitotic pathology. Extracapsular extension and possible right neurovascular bundle involvement are suggested. No significant lymphadenopathy was seen.

Twelve Core transrectal ultrasound guided prostate (TRUS) biopsy was done.

![Magnetic resonance imaging (MRI)](Image)
**HISTOPATHOLOGY**

Histopathology slides (Figs 7.13.2A to C) of the sections of prostatic tissue cores from the right lobe showed granulomas comprising of epitheloid cells, lymphocytes, plasma cells, multinucleate and Langhans-type of giant cells. Foci of necrosis were seen. Sections of prostatic tissue core from left lobe showed a patchy moderate mixed inflammatory cell infiltrate. There was no evidence of malignancy in the sections examined. Special stain for fungus is negative. Ziehl-Neelsen (ZN) stain for AFB is negative. Impression: Necrotizing granulomatous inflammation.

*Fig. 7.13.2A: Prostate core biopsy showing lymphomononuclear infiltrate along with pale eosinophilic areas (H & E, 4X)*
Fig. 7.13.2B and C: Higher magnification of pale areas showing langhans giant cells and epitheliod cell granulomas (H & E, 40X)

■ TREATMENT

Patient was started on ATT along with alpha blockers. His symptoms improved.

■ FOLLOW-UP

Urinary symptoms improved. Prostate-specific antigen (PSA) returned to normal. Contrast enhanced MRI revealed decrease in size and localized necrotic lesion in the prostate. He was continued on ATT.

■ COMMENTS

Tubercular prostatitis is an incidental histological finding following transurethral resection of prostate (TURP) or needle biopsy of prostate. Anti-TB treatment is the treatment of choice.
8.1 NEUROGENIC BLADDER WITH SPHINCTERIC WEAKNESS

INTRODUCTION

Urinary incontinence secondary to spinal dysraphism in children is usually mixed type (>60%)—intrinsic sphincteric deficiency (ISD), detrusor hyperactivity and decreased compliance.

Medical Management: Alpha agonists for intrinsic sphincter deficiency—increase outlet resistance; anticholinergic agents—for detrusor hyperactivity; clean intermittent catheterization (CIC)—for high postvoid residual urine (PVRU).

Surgical intervention: Bladder neck suspension (Stamey/Raz/Burch/ Marshall-Marchetti-Krantz, MMK), sling cystourethropexy, bladder neck reconstruction (Kropp/Young-Dees-Leadbetter), artificial urinary sphincter, bladder neck submucosal injection, bladder neck closure with a continent catheterizable stoma and urinary diversion with a continent or incontinent stoma.

Advantages of fascial sling procedure: No foreign material is used unlike submucosal injection or artificial sphincter, improves intrinsic sphincteric deficiency unlike bladder neck suspension procedures, long-term success rate is from 50–70% to 80–90% in some series.

CLINICAL PRESENTATION

A 12-year-old male child, known case of meningomyelocele, was operated after 8 days of birth. He presented with urinary and fecal incontinence since birth. He was passing urine mostly in spurts at a frequency of 5–10 min. He also had incontinence on coughing and running and had complete nocturnal incontinence. He had fecal incontinent to liquid or semisolid stools.

He had full-term normal vaginal delivery at home. No history of motor, sensory or autonomic weakness. No history of previous recurrent urinary tract infections (UTIs), abdominal or flank pain, or urolithiasis. He achieved
normal milestones and was able to perform daily activities. He had weak erections. He had two siblings who were normal.

He was evaluated 5 years back and was found to have small capacity, hyperreflexic bladder with mixed (stress and urge) incontinence; he was put on anticholinergic drugs and advised to follow clean intermittent catheterization (CIC). He had partial relief of symptoms and effect waned off in 1–2 months. Intravesical botox injection was given 1 year back, after which he was fully continent for 2 months.

**EXAMINATION**

Transverse scar on lower back at L5-S1; perineal sensations—touch/pain—absent. Motor examination and knee/ankle jerks were normal, cremasteric reflex were normal, bilateral bulbocavernosus reflex weak. Per rectal examination revealed weak anal sphincter tone and voluntary contraction.

**INVESTIGATIONS**

Complete blood count (CBC), renal function test (RFT), liver function test (LFT), serum electrolytes and urine routine/microscopic (R/M) were normal; Urine culture: sterile; Ultrasound-upper tracts normal, mild irregular outline of urinary bladder, PVRU: 5 cc; Plain X-ray pelvis (Fig. 8.1.1): spina bifida S5, open posterior sacral segments; Intravenous urogram (Fig. 8.1.2): normal upper tracts and small bladder; micturating cystourethrogram (MCU) (Fig. 8.1.3): small capacity bladder, open bladder neck, no vesicoureteral reflux (VUR). Videourodynamics (Fig. 8.1.4) revealed: Bladder capacity—150 cc, PVRU—30 cc, bladder leak point pressure (BLPP)—60 cc H$_2$O, valsalva leak point pressure (VLPP)—30 cc H$_2$O.
Fig. 8.1.3: Micturating cystourethrogram

Fig. 8.1.4: Video urodynamics
**DIAGNOSIS**

Post-myelomeningocele small capacity, hyperreflexic bladder with urge and stress urinary incontinence, and internal and external sphincteric weakness.

**TREATMENT**

Cystoscopy, W-loop ileal augmentation cystoplasty and rectus sheath bladder neck sling were done. Figures 8.1.5 to 8.1.12 are showing the operative pictures.

![Fig. 8.1.5: Operative picture showing mobilization of bladder neck and urethra](image)

![Fig. 8.1.6: Opening of bladder](image)
Fig. 8.1.7: Isolation of 15–20 cm segment of ileum

Figs 8.1.8A and B: Detubularization of bowel segment

Figs 8.1.9A and B: Anastomosis of bowel to bladder
Figs 8.1.10A to C: Dissection of rectus sheath

Fig. 8.1.11: Fascial sheath in position

Fig. 8.1.12: Final picture
FOLLOW-UP

Patient developed full control of urine and was voiding well.

COMMENTS

Management of neurogenic bladder with sphincteric deficiency is challenging. If drug therapy fails, surgical treatment by augmentation cystoplasty and sphincter reconstruction is required.

BIBLIOGRAPHY

8.2 NEUROGENIC BLADDER WITH URETHRAL DIVERTICULUM

INTRODUCTION
Long term indwelling urethral catheter can form urethral diverticulum.

CLINICAL PRESENTATION
A 60-year-old male, a follow-up case of neurogenic bladder on continuous catheter drainage for many years, presented with Foley’s catheter in situ and difficulty in passing urine. He also had chronic constipation.

INVESTIGATIONS
Micturating cystourethrogram revealed a large diverticulum in the urethra with Foley’s catheter balloon in it (Fig. 8.2.1). X-ray also showed fecoliths in the abdomen.

Fig. 8.2.1: Micturating cystourethrogram

TREATMENT
Foley’s catheter was removed. After 4 weeks, diverticulum excision and reconstruction of the urethra was done.

COMMENTS
Urethral diverticulum can occur if Foley’s catheter balloon is inflated in the urethra. During Foley’s catheterization, one has to be careful to ensure Foley’s catheter in bladder by ensuring free flow of urine from the bladder.

BIBLIOGRAPHY
8.3 PINE TREE APPEARANCE WITH NEUROGENIC BLADDER

INTRODUCTION

Pine or Christmas tree is the name given to the neurogenic bladder due to its appearance. The shape of the bladder is highly abnormal with an elongated appearance, with the dome like the top of a Christmas tree. The associated bladder wall hypertrophy gives an outline which mimics the decorations that adorn a Christmas tree. It is typically seen in severe neurogenic bladder with increased sphincter tone (detrusor sphincter dyssynergia) due to suprasacral lesions (above S2-S4) or epiconal lesions (in and around S2-S4).

It is, however, not pathognomonic of a neurogenic bladder and can be seen in patients with lesions anywhere along the sacral reflex arc leading to poor detrusor compliance. Occasionally it is also seen in bladder neck obstruction of a non-neurogenic cause (Figs 8.3.1 to 8.3.3).

Fig. 8.3.1: Pine tree appearance in neurogenic bladder

Fig. 8.3.2: Pine tree appearance in neurogenic bladder

Fig. 8.3.3: Pine tree appearance in neurogenic bladder
**CLINICAL PRESENTATION**

Patient presented with history of neurologic defect with lower urinary tract symptoms. Urodynamic evaluation revealed detrusor hypertrophy. Cystogram and MCU revealed pine tree appearance (Fig. 8.3.4).

![Fig. 8.3.4: Pine tree bladder in voiding phase with open bladder neck](image)

**COMMENTS**

During evaluation and investigations for lower urinary tract symptoms, pine tree appearance helps to suggest diagnosis of neurogenic bladder.

**BIBLIOGRAPHY**

8.4 NEUROGENIC BLADDER WITH VESICOURETERAL REFLEX AND CHRONIC KIDNEY DISEASE

INTRODUCTION

Neurogenic bladder results in detrusor hyperreflexia with or without detrusor sphincter dyssynergia and hypo- or acontractility. These effects undermine safe, effective and controlled storage and voiding of urine and predispose to reflux nephropathy. Therefore, patients with neurogenic bladder are expected to have increased risk of renal failure.

CASE 1

Clinical Presentation

A 12-year-old male presented with lethargy, weakness, poor growth and recurrent UTI. His renal functions were deranged and he was on catheter for a long time. He had history of myelomeningocele for which he was operated during childhood.

Investigations

Serum creatinine was 4.3 mg/dL. MCU revealed grade IV bilateral vesicoureteral reflux (VUR) (Fig. 8.4.1).

Treatment

Supravesical urinary diversion was done by loop ureterostomy to improve his renal function and to make him catheter free. After improvement in renal function, augmentation cystoplasty with intermittent self-catheterization was done.

Fig. 8.4.1: Case 1—Micturating cystourethrogram
**Case 2**

Another case of neurogenic bladder with bilateral VUR (Fig. 8.4.2) with chronic kidney disease (CKD) was managed by intermittent self-catheterization and long-term suppressive antibiotics.

**Case 3**

Another case of neurogenic bladder with bilateral VUR (Fig. 8.4.3) with CKD required renal replacement therapy.

![Fig. 8.4.2: Case 2—Micturating cystourethrogram](image1)

![Fig. 8.4.3: Case 3—Micturating cystourethrogram](image2)

**COMMENTS**

Patients of neurogenic bladder develop chronic renal failure in long term. In management, it is important to prevent renal failure by low pressure bladder and control of infection.

**BIBLIOGRAPHY**

8.5 NEUROGENIC BLADDER WITH INCONTINENCE OF URINE

CASE PRESENTATION

A 40-year-old male presented with continuous leakage of urine. He had myelomeningocele for which he was operated during childhood. During all these years, he had incontinence of urine for which he managed with a local advise.

EXAMINATION

Clinical picture of the back (Fig. 8.5.1) showing operative scar. Clinical picture (Fig. 8.5.2) showing locally made urinary incontinence devise.

INVESTIGATIONS

X-ray spine showed a large defect in the spine (Fig. 8.5.3). MCU show bilateral vesicoureteric reflux with trabeculated bladder (Fig. 8.5.4).
TREATMENT

The patient opted to continue in the same way.

COMMENTS

In developing countries, due to limited resources and poverty, patients adjust and live their lives accordingly.

BIBLIOGRAPHY

9.1 BLADDER CANCER WITH PAROTID METASTASIS

■ INTRODUCTION

Transitional cell carcinoma (TCC) urinary bladder is the most common urothelial cancer. The common sites of metastasis are pelvic and paraaortic lymph nodes followed by liver, lungs, bones and adrenal. Head and neck metastasis are rare and only few cases are reported. It also indicates poor prognosis for the patient.

■ CLINICAL PRESENTATION

A 78-year-old male is presented here with complaints of hematuria since last 2 months. On examination, there is no positive finding.

■ INVESTIGATIONS

Bladder biopsy (outside slides for review): Infiltrating urothelial carcinoma and muscle included in the biopsy is involved. Contrast enhanced computed tomography (CECT) showed right hydronephrosis (HDN) with poorly functioning kidney with tumor bladder with extravesical extension (Figs 9.1.1 and 9.1.2).
TREATMENT

He underwent right open nephroureterectomy and radical cystectomy with ileal conduit on December 15, 2011. He tolerated the procedure well. His postoperative period was uneventful.

FOLLOW-UP

He was alright for 1 year with no symptoms. After that, he noticed a swelling on the right side of the face (Fig. 9.1.3). Fine-needle aspiration cytology (FNAC) was inconclusive. Fine-needle aspiration biopsy (FNAB) confirmed metastatic poorly differentiated TCC, supported by the immunohistochemical
findings. Positron emission tomography computed tomography (PET-CT) revealed fluorodeoxyglucose (FDG) avid lesion left parotid (Fig. 9.1.4). Bone scan revealed no other metastasis (Fig. 9.1.5). Local radiotherapy to the parotid region was given. At 1 year follow-up the patient is asymptomatic.

Fig. 9.1.3: Clinical picture

Fig. 9.1.4: Positron emission tomography computed tomography (PET-CT)
COMMENTS

Isolated metastasis to parotid from TCC urinary bladder is very rare. Treatment options are surgical excision, radiotherapy or chemotherapy. In view of age and extent of the disease, it was decided that he should undergo radiotherapy.

BIBLIOGRAPHY

9.2 BLADDER DYSPLASIA

INTRODUCTION

Urothelial dysplasia of the bladder is the precursor of urothelial carcinoma in situ (CIS) and invasive urothelial carcinoma of the bladder. Patients present with symptoms of frequency, dysuria and urgency. Many patients may be asymptomatic. Microscopic hematuria may be present.

Pioglitazone, an antidiabetic medication of thiazolidinedione group, is a peroxisome proliferator-activated receptor gamma (PPAR\(\gamma\)) agonist which is known to cause altered proliferation and differentiation in urothelial tissues. It is associated with an increased risk of bladder cancer possibly attributable to the PPAR\(\gamma\) activity. In the present case, the patient had a 1 year history of consuming pioglitazone, thereafter showing urothelial dysplasia on biopsy done for hematuria.

CLINICAL PRESENTATION

A 64-year-old male with a history of type 2 diabetes mellitus and hypertension for the past 10 years presented to the urology department with complaints of two episodes of hematuria with passing of clots 6 weeks back. Smoking history was negative. He was taking tablet pioglitazone 30 mg once daily for a period of 1 year till current visit. General and systemic examination was normal.

INVESTIGATIONS

Hematological and biochemical investigations were normal. CT urography showed mild urinary bladder wall thickening and irregularity along the base and posterior wall. Urine cytology reported superficial squamous cells with occasional lymphocytes. Malignant cells on urine cytology were absent. Cystoscopy revealed normal urethra and bilateral ureteric orifices with elevated bladder neck and velvety red tissue over trigone area (Fig. 9.2.1).

![Fig. 9.2.1: Cystoscopic picture showing diffused velvety bladder mucosa](image-url)
**TREATMENT**

Transurethral cold cup biopsies were obtained from trigone area followed by resection of bladder lesion. Patient was then discharged with the advice of discontinuing pioglitazone and was started on sitagliptin 100 mg once daily and metformin 1 g once daily.

**HISTOPATHOLOGY**

Histopathology revealed urothelial dysplasia (Figs 9.2.2A to C).

![Histopathology showing features of bladder dysplasia](image)

**FOLLOW-UP**

Check cystoscopy was done at 3 months follow-up. Cystoscopy showed a well-healed area over trigone and velvety edematous lesion above right ureteric orifice. Fulguration of bladder lesion was done and patient was kept on a 6-month follow-up. Sitagliptin and metformin were continued. On 1-year follow-up, patient was clinically asymptomatic with ultrasound kidney urinary bladder region reporting no abnormal findings.
**COMMENTS**

Patients of diabetes mellitus on pioglitazone should be informed for the risk of bladder dysplasia and bladder cancer following long-term uses.

**BIBLIOGRAPHY**

9.3 BLADDER PHEOCHROMOCYTOMA

INTRODUCTION

Pheochromocytoma of urinary bladder accounts for less than 0.05% of all bladder tumors and less than 1% of all the pheochromocytomas. Meta-iodobenzylguanidine (MIBG) scan is considered specific but CECT are also helpful as pheochromocytomas are usually hyper vascular. These tests are useful even in the patients where bladder pheochromocytoma is not considered as clinical diagnosis. Presence of clinical symptoms related to catecholamine hypersecretion points towards the diagnosis of pheochromocytoma although the differential diagnosis of carcinoma of bladder must be excluded.

TREATMENT

Transurethral resection is to be avoided which can precipitate catecholamine secretions and can be fatal. Partial cystectomy or total cystectomy is recommended.

CLINICAL PRESENTATION

A 59-year-old male, chronic smoker, hypertensive for 2 months—controlled on medication, presented with complaints of constipation and tenesmus for 1 year, sense of incomplete evacuation of bowel for 1 year and irritative voiding symptoms for 2 months. No history of hematuria, bleeding per rectum.

EXAMINATION

Pulse: 68 beats/min regular; Blood pressure—supine: 120/80 mm Hg, standing: 110/74 mm Hg; Chest/cardiovascular system (CVS)/abdomen: normal; Rectal examination: normal.

INVESTIGATIONS

Hemoglobin: 12.3 gm/dL; platelets: 327,000/cumm; hematocrit: 34.2%; urea: 50 mg/dL; serum creatinine: 1.5 mg/dL; liver function test (LFT): normal; stool for occult blood: negative; colonoscopy: normal; urine cytology: negative; ultrasound abdomen: right kidney—normal (Fig. 9.3.1), left kidney—mild hydronephrosis, (5.1 × 4.8 cm mass) posterior to urinary bladder indenting its wall, prostate: 17.7 grams. Cystoscopy, left retrograde pyelography and DJ stenting and biopsy of tumor was planned. Findings—large mass projecting into bladder over left side of base and posterior wall with intact mucosa and elevated left ureteric orifice. Cold cup biopsy was taken which lead to rise in blood pressure to 270/150 mm Hg, increasing the suspicion of bladder pheochromocytoma. The procedure was thus abandoned.
Further work up showed—24-hour urine metanephrines to be 189.58 μg/mg of creatinine (normal <155 μg/mg) and normetanephrines to be 6960.46 μg/mg of creatinine (normal < 256 μg/mg). Iodine-131 MIBG scan—increased concentration in region of left vesicoureteric junction even after catheterization, no other focal abnormal accumulation elsewhere in body. Magnetic resonance imaging (MRI)—5.5 × 4.5 cm T2-hyperintense mass left posterolateral wall of urinary bladder (Figs 9.3.2A and B), loss of fat planes
with left seminal vesicle, maintained with rectum and pelvic wall, no other adrenal or extraadrenal lesion.

**TREATMENT**

Robotic partial cystectomy and left ureteric reimplantation was done on July 8, 2008. Intraoperative findings—6 × 6 cm bladder mass over left posterolateral wall, left ureter could not be separated.

**HISTOPATHOLOGY**

Neuroendocrine tumor compatible with pheochromocytoma

**POSTOPERATIVE**

Patient recovered well, urinary catecholamines returned to normal.

**FOLLOW-UP**

After 1 year, he developed backache for which he was investigated and recurrence of tumor in the bones was found. Four cycles of radioisotope MIBG was given, which improved his symptoms and he is still surviving.

**COMMENTS**

Bladder pheochromocytoma is a rare tumor. Possibility should be kept in mind if patient has symptoms of high blood pressure during voiding or during bladder biopsy. Transurethral resection of bladder tumor (TURBT) should not be done. Surgical removal of the tumor should be done.

**BIBLIOGRAPHY**

9.4 CLOACAL ANOMALY WITH BLADDER TUMOR

INTRODUCTION

Cloacal malformations can be associated with wide spectrum of clinical conditions. Cloacal deformities occur due to poor development of urorectal septum which divides anterior urogenital sinus and a posterior gastrointestinal canal (Fig. 9.4.1). The literature reports that the incidence rate of cloacal malformations is approximately 1 per 20,000–25,000 live births. Cloacogenic tumors of anus, rectum and vulva are reported which are mainly adenocarcinoma. Squamous cell carcinoma of bladder with cloacal anomaly is very rare.

CLINICAL PRESENTATION

A 36-year-old female presented with urinary frequency, urgency and urge incontinence associated with low-grade fever. She had history of recurrent UTIs since childhood, received multiple courses of antibiotics. She was a known case of cloacal anomaly. She underwent cutback and anal dilatation at birth. She delivered two full term live issues by lower segment cesarean section (LSCS). Per abdominal examination revealed scar of LSCS. Local examination revealed a common channel opening present in the perineum.

Fig. 9.4.1: Cloacal anomaly
INVESTIGATIONS

Three-dimensional contrast enhanced computed tomography was done. The reconstructed images showed sacral deformity (Fig. 9.4.2) and cloacal anatomy (Fig. 9.4.3). Ultrasound revealed a large bladder tumor (Fig. 9.4.4). CT pelvis showed a large bladder tumor cystopanendoscopy (CPE) revealed large solid sessile growth arising from dome and anterior wall of urinary bladder, rectum opening just distal to bladder neck and two cervical openings.

Fig. 9.4.2: Three-dimensional (3D) CECT reconstructed images showing bony deformities

Fig. 9.4.3: Three-dimensional (3D) CECT reconstructed images—cloacal anatomy
posteriorly (Fig. 9.4.5). Cold cup biopsy from growth revealed moderately differentiated squamous cell carcinoma.

**TREATMENT**

On abdominal exploration, there was pouch of colon (approximately 15 cm) with large tumor involving anterior wall of bladder and infiltrating the pubic bones. The two uteri lay on either side of the bladder. Pelvic exenteration, bilateral pelvic lymph node dissection and pubectomy + wet colostomy were performed on October 24, 2007. Operative time was 6 hours with 1,200 cc blood loss and 2 units of blood transfusion.
**HISTOPATHOLOGY**

Histopathology confirmed moderately differentiated squamous cell carcinoma infiltrating transmurally up to serosa and pubic bone. Resected margins were free of tumor. Five pelvic lymph nodes out of nine were involved by the tumor.

She was given adjuvant chemotherapy methotrexate, vinblastine, Adriamycin and cisplatin (MVAC) four cycles.

**COMMENTS**

Cloacal anomaly with squamous cell carcinoma of the bladder is very rare. Usually advanced disease with poor prognosis.

**BIBLIOGRAPHY**

Introduction

Cystitis cystica glandularis is a rare benign condition of urinary bladder that can mimic bladder tumor. It is most commonly seen in the trigone area of the bladder. Clinically patients have irritative voiding symptoms and hematuria. Its transformation into adenocarcinoma is rare.

Treatment consists of removal of source of irritation and long-term antibiotic therapy for chronic urinary tract infections or transurethral resection (TUR) of cystitis cystica glandularis tissue. Surgical options reserved for patients who do not respond to conservative therapies.

Case 1

Clinical Presentation

A 37-year-old male presented with history of frequency of urination 7–8 times during the day and 1–2 times during the night. He was investigated and treated as chronic prostatitis for 2 years. Recently his frequency had increased on he had difficulty in passing urine. Physical examination was normal with normal sized prostate.

Investigations

Urine examination revealed 8–10 pus cells hpt and urine culture/sensitivity (C/S) revealed Escherichia coli infection. Urine cytology was negative for the malignant cells. Uroflowmetry revealed maximum flow rate of 7.2 mL/sec with voided volume of 371 mL. Ultrasound revealed left mild hydronephrosis (HDUN) and thickening of the base of the bladder. CECT revealed thickening of the base of the bladder with tumor near trigone with left HDUN (Figs 9.5.1A to C). Dynamic renal scan revealed right nonobstructed kidney with left HDUN with partially obstructed kidney with normal cortical function. Cystoscopy revealed (Figs 9.5.2A and B) bullous oedema over trigone with thickened mucosa and both ureteric orifices were obliterated.
Figs 9.5.1A to C: Case 1—Contrast enhanced computed tomography (CECT) abdomen

**Treatment**

Bipolar transurethral resection of the tumor was done over the trigone and both ureteric orifices (Fig. 9.5.3). Figure 9.5.4 is shows exposed left ureteric orifice after resection. Bilateral DJ stenting was done.

Figs 9.5.2A and B: Case 1—Cystoscopic view
Fig. 9.5.3: Case 1—Bipolar transurethral resection (TUR) of prostate

Fig. 9.5.4: Case 1—Left ureteric orifice after resection

Figs 9.5.5A and B: Case 1—cystoscopic view after 6 months
**Histopathology**

Histopathology confirmed the features of cystitis cystica glandularis.

**Follow-Up**

After 3 months, relook cystoscopy revealed healing tissue. Both DJ stents were removed. Cystoscopy after 6 months revealed healed bladder mucosa with scars and gaping left ureteric orifice (Figs 9.5.5A and B). On follow up, patient was voiding well since last 1 year and is on regular cystoscopy surveillance.

**CASE 2**

**Clinical Presentation**

A 41-year-old male presented with history of gross hematuria for 3–4 days. He had no past history of any significant disease. Physical examination was normal.

**Investigations**

Urine examination revealed plenty of red blood cells (RBCs). Urine cytology was negative for the malignant cells. Ultrasound revealed thickening of the base of the bladder. CECT (Figs 9.5.6A and B) revealed thickening of the base of the bladder with a tumor in the region of the trigone. Cystoscopy revealed bullous edema over trigone with thickened mucosa with tumor in the trigone (Fig. 9.5.7).

**Treatment**

Bipolar transurethral resection of the tumor was done over the trigone. Histopathology confirmed the features of cystitis cystica glandularis.
Follow-Up

Patient is voiding well with no hematuria. In 3-month follow-up, he developed recurrence of tumor with involvement of posterior urethra, trigone and both uretric orifices. Repeat TURBT was done, bilateral DJ stenting was done. Intravesical 6 cycles of hydrocortisone and heparin was given. He was advised for regular follow-up every 3 months.

COMMENTS

Cystitis cystica glandularis is rare. In younger patients with lower urinary tract symptoms (LUTS) and hematuria with bladder mass, possibility of cystitis cystica glandularis should be considered. Histopathology confirms the diagnosis. Cystoscopy surveillance is necessary as in long term, it can transform into a malignant disease.

BIBLIOGRAPHY

9.6 FOREIGN BODY—URINARY BLADDER

INTRODUCTION

Various types of foreign bodies have been found in the urinary bladder like electric wire, safety pin, hairclip, intrauterine device, gauze pieces, battery, leech, hairballs, etc. The causes include self introduction for sexual gratification, accidental migration through the urethra or iatrogenic introduction. Majority of patients present with lower abdominal pain, frequency, dysuria and hematuria. Sometimes, the foreign bodies may mimic carcinoma bladder or there can be stone formation over the foreign body.

The treatment can be endoscopic removal or removal by open surgery.

CASE 1

Clinical Presentation

A 45-year-old male presented with history of hematuria and painful micturition for 6 months duration. He had a past history of open surgery for removal of bladder calculus about 7 years back. On examination, there was lower abdominal scar and vague lump palpable in suprapubic region. Per rectal (PR) examination revealed grade I enlargement of prostate with a bimanually palpable mass.

Investigations

Urine examination revealed plenty of pus cells and RBCs. Urine culture was sterile. Urine cytology was negative. Plain X-ray revealed a vague shadow in the pelvis (Fig. 9.6.1). Intravenous urogram revealed normal upper tracts with a mass in the bladder (Fig. 9.6.2).

Fig. 9.6.1: Case 1—Plain X-ray of kidney, ureter, bladder (KUB) region
Urinary Bladder Diseases

Treatment

Cystoscopy revealed normal urethra, a huge mass occupying the complete bladder. Interlacing strands were found on the surface of the mass. Frozen section cold cup biopsy of the mass revealed cotton fiber. Suprapubic exploration revealed thick walled bladder. A surgical sponge measuring 7.5 × 6 × 4 cm was removed from the bladder (Fig. 9.6.3). Bladder biopsies taken which revealed chronic cystitis.

Fig. 9.6.2: Case 1—Intravenous Urogram

Fig. 9.6.3: Case 1—Picture of the sponge
CASE 2

Clinical Presentation

A 35-year-old male presented with frequency and dysuria. Past history was negative. Physical examination was normal.

Investigations

Urine and blood examination was normal. X-ray of kidney, ureter, bladder (KUB) region (Fig. 9.6.4) and pelvis (Fig. 9.6.5) revealed a vesical calculus over a wire type foreign body.

Fig. 9.6.4: Case 2—Plain X-ray of kidney, ureter, bladder (KUB) region
Treatment

Cystoscopy confirmed the presence of stone. Holmium laser lithotripsy was done and later on a wire was found which was removed endoscopically.

 COMMENTS

Foreign bodies in bladder are usually self introduced and iatrogenic bodies are uncommon. These should be suspected in patients with LUTS, and can be treated endoscopically or by open surgery.

 BIBLIOGRAPHY

9.7 PARAURETERIC DIVERTICULUM WITH TUMOR

**INTRODUCTION**

Carcinoma bladder can occur in bladder diverticulum. Due to thin wall of diverticulum, there are higher chances for local infiltration and spread. The management depends upon the grade and stage of the tumor. Paraureteric diverticulum is rare and can be associated with carcinoma bladder.

**CLINICAL PRESENTATION**

A 51-year-old male presented with history of hematuria for 1 month duration. Clinical examination was normal.

**INVESTIGATIONS**

Contrast enhanced computed tomography scan revealed a tumor on the left side of bladder and displacing seminal vesicle (Figs 9.7.1A and B). Cystoscopy revealed normal left ureteric orifice with a paraureteric diverticulum with tumor. Left retrograde ureterogram revealed medial deviation of ureter with hydroureteronephrosis (Figs 9.7.2A and B). Biopsy was taken from the parametric mass and bladder. Histopathology confirmed low-grade transitional cell in biopsy of the mass in the periureteric diverticulum carcinoma (TCC) and the bladder biopsy was normal.

![Fig 9.7.1A: Contrast enhanced computed tomography (CECT) scan](image-url)


**TREATMENT**

Patient was counseled for radical cystoprostatectomy, but he refused for a radical procedure. However, he agreed for conservative surgery. Robotic excision of diverticulum with seminal vesiculectomy with pelvic lymphadenectomy was done (Fig. 9.7.3). Operative specimen revealed large tumor confined to diverticulum (Figs 9.7.4A and B). Patient recovered well.
**FOLLOW-UP**

On regular follow-up with check cystoscopy, he was alright for 2 years. Then he did not come for follow-up for 4 years and lost to follow-up. He came back after 4 years with recurrent hematuria. CECT revealed large recurrence of tumor in left lateral wall with perivesical extension (Fig. 9.7.5). TURBT was done. Histopathology confirmed high-grade TCC. He was advised for adjuvant chemotherapy.
 COMMENTS

Bladder diverticular cancer is more aggressive and needs partial or radical cystectomy. In the present case, the local infiltration occurred due to conservative surgery. Aggressive follow up and adjuvant chemotherapy is necessary.

 BIBLIOGRAPHY

9.8 BLADDER DIVERTICULUM IN ADULTS

INTRODUCTION

Bladder diverticula are protrusions of the mucosal and submucosal layers of the bladder through the muscular wall of the bladder. Congenital or true diverticula involve all layers of the structure, including muscularis propria and adventitia. Acquired or false diverticula do not involve muscular layers or adventitia.

CASE 1

Clinical Presentation

A 15-year-old male is presented with difficulty in passing urine. Clinical examination was normal.

Investigations

Intravenous urogram revealed left paraureteric diverticulum with normal functioning kidney (Fig. 9.8.1). Micturating cysto-urethrogram (MCU) confirmed diverticulum with no reflux (Fig. 9.8.2). Cystopanendoscopy confirmed presence of paraureteric diverticulum on left side.

Fig. 9.8.1: Case 1—Intravenous urogram
Urinary Bladder Diseases

Treatment
Diverticulectomy was done. Patient improved of his symptoms.

CASE 2

Clinical Presentation
A 45-year-old male presented with straining to pass urine. Examination revealed normal size prostate.

Investigations
Ultrasound examination revealed thick-walled bladder with diverticulum right side with narrow opening (Fig. 9.8.3). Postvoid film revealed significant

Fig. 9.8.2: Case 1—Micturating cysto-urethrogram (MCU)

Fig. 9.8.3: Case 2—ultrasound
residual urine. Cystoscopy revealed normal urethra, high bladder neck and grade 1–2 trabeculations in the bladder with narrow opening diverticulum on the right side.

**TREATMENT**

Laser incisions Triradiate at 4, 8 and 12 o’clock positions were made at the diverticular neck and laser bladder neck incision (BNI) was done. Patient improved postoperatively.

**COMMENTS**

Bladder diverticulum can be congenital or acquired due to lower urinary tract obstruction. Congenital diverticulum is asymptomatic and can be kept under observation. Bladder diverticulum with obstruction needs endoscopic management by laser incisions.

**BIBLIOGRAPHY**

9.9 CARCINOMA URINARY BLADDER IN A 128-YEAR-OLD MAN

**INTRODUCTION**

Cancer of the urinary bladder can develop in any age group. The oldest person reported of carcinoma urinary bladder in literature is 97 years of age with a nested variant of TCC.

**CLINICAL PRESENTATION**

A 128-year-old man (Fig. 9.9.1) presented with painless hematuria for 2 months and lower urinary tract symptoms (LUTS) for 8 months. He was a known hypertensive on drugs. He was nondiabetic, nonsmoker and unmarried.

![Fig. 9.9.1: Clinical picture](image)

**INVESTIGATIONS**

Ultrasound revealed normal kidneys, multiple sessile echogenic soft tissue masses, largest measuring: $3.16 \times 2.76$ mm, prostate: 26 cc, post-void residual (PVR): 33.13 cc (Figs 9.9.2A and B). Cystopanendoscopy was done under spinal anesthesia which revealed stricture in proximal bulbar urethra. Optical internal urethrotomy (OIU) done. Multiple polypoidal tumors was seen urinary bladder.
TREATMENT

Transurethral resection of all tumors was done. Intravesical mitomycin C 40 mg was given within 4 hours of surgery. Postoperative period: normal was uneventful.

HISTOPATHOLOGY

Histopathology revealed high-grade papillary urothelial carcinoma with no deep muscle infiltration. Foley’s catheter was removed on 3rd day. Relook cystoscopy was done after 6 weeks. Cystoscopy and bladder biopsy revealed no tumor. Intravesical Bacillus Calmette-Guérin (BCG) 80 mg weekly for 6 weeks was given. Patient tolerated intravesical BCG therapy well.

COMMENTS

Age is no bar for carcinoma urinary bladder. Surgery can safely be performed at any age.

BIBLIOGRAPHY

9.10 CARCINOMA BLADDER WITH RENAL STONE

**INTRODUCTION**

Urinary stone disease and bladder cancer are two of the most commonly seen urologic diseases. As such, no correlation has been found between these two diseases, but patient may present with both diseases together co-incidentally.

**CLINICAL PRESENTATION**

A 55-year-old male presented with history of painless hematuria. Physical examination was normal.

**INVESTIGATIONS**

Urine and blood examination was normal. Plain X-ray of kidney, ureter and bladder (KUB) revealed radio opaque shadow in left renal area (Fig. 9.10.1). CECT abdomen revealed left renal stone, right normal kidney, tumor right lateral wall of bladder (Figs 9.10.2A to D). Cystoscopy revealed infiltrative tumor in right lateral wall.

![Fig. 9.10.1: Plain X-ray of kidney, ureter and bladder (KUB) region](image)
TREATMENT

Transurethral resection of bladder tumor was done. Histopathology revealed muscle invasive TCC urinary bladder. Patient was counseled for surgery. Open left pyelolithotomy (Fig. 9.10.3) and radical cystoprostatectomy with lymphadenectomy was done at the same time. Figure 9.10.4 shows renal stone and figure 9.10.5 shows radical cystectomy specimen with renal stone. Postoperative recovery was uneventful.
It may be a coincidence to have both carcinoma bladder and renal stone together. It is a challenging decision for management of carcinoma bladder and renal stone if present together. Both surgeries can be done together as decided in the above case.
10.1 BENIGN PROSTATIC HYPERPLASIA WITH BLADDER DIVERTICULUM

**INTRODUCTION**

Majority of bladder diverticula are acquired and are secondary to either bladder outlet obstruction or the upper motor type of neurogenic bladder. Acquired diverticula are often multiple and typically seen in older men (mirroring the incidence of obstruction of the bladder outlet). Treatment include relief of the obstruction and may or may not removal of the diverticulum. For narrow neck diverticulum, endoscopic incision of the bladder neck diverticulum can be done at the time of surgery for prostate enlargement. In some cases, bladder diverticulectomy may be required.

**CLINICAL PRESENTATION**

A 41-year-old male presented with irritative and obstructive lower urinary tract symptoms (LUTS) for past 6 months. There was no history of hematuria. No history of surgical intervention in the past. History of hypertension present for 2 years. Per rectal examination revealed grade 1 prostate, smooth, firm and nontender.

**INVESTIGATIONS**

Urine and blood examinations were normal. Ultrasound kidney, ureter and bladder (KUB) revealed a large diverticulum with narrow neck in right lateral wall with prostate enlargement (Fig. 10.1.1). Micturating cystourethrogram (MCU) revealed a large diverticulum in right lateral wall (Fig. 10.1.2).
Transurethral resection of the prostate (TURP) with four incisions were made at the neck of diverticulum. Postoperative recovery was uneventful. Postoperative ultrasonography (USG) revealed wide bladder diverticulum neck with minimum residual urine (Fig. 10.1.3). Uroflowmetry revealed peak flow rate of 30 mL/sec.
COMMENTS

Management of acquired diverticulum with benign prostatic hyperplasia (BPH) needs management of outlet obstruction with diverticular neck incision which can be done by holmium laser. Diverticulectomy can be done by open or laparoscopic or robotically in cases where symptoms persist with significant retention in the diverticulum.

BIBLIOGRAPHY

INTRODUCTION

Chronic renal failure (CRF) is a well-described complication of obstructive Benign Prostatic Hyperplasia (BPH). The degree of renal failure varies from a serum creatinine cutoff of 1.5–3.0 mg/dL. It is important to differentiate acute and chronic renal failure, and acute and chronic urinary retention as outcome will be different. Various combinations of chronic retention with large residual urine volumes (>300 mL), detrusor instability and decreased bladder compliance are associated with chronic renal failure. Ureterovesical junction obstruction from bladder remodeling in chronic urinary retention is the most commonly proposed mechanism for CRF. However, episodic acute urinary retention, urinary tract infections and secondary hypertension may also have a role. There will be significant improvement in renal function after prostate surgery, but the acuity of renal failure is generally not known.

CLINICAL PRESENTATION

A 55-year-old male presented with history of difficulty in passing urine for 3 months duration. He had no history of hematuria. Per rectal examination revealed grade 3 enlargement of prostate, firm, smooth and rectal mucosa free.

INVESTIGATIONS

Serum creatinine was 3.5 mg/dL. Prostate-specific antigen (PSA) was 2.5 ng/mL. Ultrasound revealed mild bilateral hydroureteronephrosis with prostate of 75 gm and large intravesical projection of the median lobe and significant post-void residual (PVR) urine (Fig. 10.2.1).

Fig. 10.2.1: Ultrasound
**TREATMENT**

Bipolar TURP was done and 55 gm of prostate was resected (Fig. 10.2.2). Postoperative recovery was uneventful. His serum creatinine returned to 1.5 mg/dL after 2 weeks and patient was voiding well.

![Resected transurethral resection of the prostate (TURP) chips](image)

**Fig. 10.2.2:** Resected transurethral resection of the prostate (TURP) chips

**COMMENTS**

Neglected enlargement of prostate can present with chronic renal failure. It may be associated with decompensation of bladder for which urodynamic evaluation is required. If serum creatinine levels are more than 3 mg/dL, then catheterization is required to bring down the creatinine level before surgery. Surgery is the treatment of choice. Patient should be followed up for serum creatinine level and bladder decompensation.

**BIBLIOGRAPHY**

10.3 CARCINOMA PROSTATE WITH FAMILY HISTORY

INTRODUCTION

Men with family history of prostate cancer have a significantly greater risk of developing prostate cancer than those with no such history. Risks are greatest for relatives of cases diagnosed when young and those with more than one relative affected. Familial aggregation of prostate cancer can be caused by genetic factors that are not inherited. Most studies suggest that risks in brothers are greater than in father-son relationships. Given the increased risk of the disease in men with a father or brother with early-onset disease (defined by an age of onset of <60 years, or with two affected first-degree relatives) screening these men should increase the yield and might prove to be a cost-effective strategy.

CLINICAL PRESENTATION

A 61-year-old male, had a family history of cancer prostate in his father, was regularly getting his PSA since last 10 years (Table 10.3.1). Initial PSA was 1.21 ng/mL which increased upto 3.30 ng/mL. Transrectal ultrasonography (TRUS) (on March 20, 2013) revealed—prostate 33 cc with a well-defined hypoechoic nodule in left peripheral zone (PZ) across mid portion of gland. TRUS guided 12 core prostatic biopsy was done which revealed adenocarcinoma of the prostate, Gleason 3+4. Magnetic resonance imaging (MRI) (on March 08, 2013) revealed enlarged prostate with altered signal intensity in left lobe in PZ with bright signal on diffusion-weighted imaging (DWI). Bone scan (on April 13, 2013) was negative for metastasis.

Robot-assisted radical prostatectomy (RARP) was done. Histopathology revealed acinar adenocarcinoma not otherwise specified (NOS); involving 5.74% of prostate. Gleason score was 3+4=7, bilateral disease, pathological stage: pT2c pN0. Right-sided nodes—4 lymph nodes isolated were free of malignancy. Left-sided nodes—3 nodes isolated were free of malignancy. Seminal vesicle was free of malignancy.
**Table 10.4.1: Prostate-specific antigen (PSA) levels**

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</tbody>
</table>

**FOLLOW-UP**

Patient was fully continent at 1 week after surgery. His PSA level was 0.01 ng/mL. He had partial erection at 3 months.

**COMMENTS**

Men with family history of prostatic carcinoma should have PSA level screening done regularly after the age of 40 years. The frequency of the PSA testing can be according to the level of PSA. If there is a rising trend, then prostatic biopsy is advisable.

**BIBLIOGRAPHY**

10.4 CARCINOMA PROSTATE WITH METASTASIS IN BLADDER

■ INTRODUCTION

Carcinoma prostate can have metastasis in urinary bladder but in very rare.

■ CLINICAL PRESENTATION

A 71-year-old male presented with hematuria for last 15 days duration.

■ PAST HISTORY

Patient presented with lower urinary tract symptoms in July 2009. Contrast enhanced computed tomography (CECT) scan revealed a bulge in the posterior margin of the prostate. TRUS guided prostatic biopsy revealed mucinous adenocarcinoma prostate, Gleason 4+5. His PSA was 3.2 ng/mL, carcinoembryonic antigen (CEA) was 5.2 IU. Sigmoidoscopy and bone scan revealed no tumor. MRI revealed right inferior pubic ramus suspicious for metastasis.

■ PAST TREATMENT

He was started injection leuprolide 3.75 mg monthly with bicalutamide (August 2009). He was given image-guided intensity-modulated radiotherapy (IG-IMRT) in November 2009. Therapy was stopped after nine cycles due to low platelet count. Cyber Knife treatment was given to complete radiotherapy. Patients was on regular follow-up since then. He had coronary artery disease for which coronary artery bypass surgery (CABG) was done in 2010.

■ EXAMINATION

Abdominal examination was normal. Per rectal examination revealed prostate hard nodular prostate with grade 1 enlargement.

■ INVESTIGATIONS

Computed tomography (CT) urogram revealed thickening of the left anterolateral wall of the bladder (Fig. 10.4.1). MRI prostate revealed mixed echogenicity in the prostate (Fig. 10.4.2). Cystoscopy revealed three tumors in the left anterolateral wall of the bladder, solid with thickened mucosa.
Transurethral resection of bladder tumor (TURBT) was done. All tumors were removed on March 19, 2013.

**HISTOPATHOLOGY**

Histopathology revealed highly atypical cells forming acinar structures and arranged in sheets, at places. These cells had high nucleus/cytoplasm (N/C) ratio, with brisk mitosis and prominent nucleoli. Section from deep biopsy revealed infiltration by the same tumor as described above; however, the deep muscle was free of tumor.

*Immunohistochemistry (IHC):* (1) PSA: diffusely positive; (2) Cytokeratin (CK) 7 and CK 20: positive in focal areas.

*Impression:* High-grade adenocarcinoma. The IHC profile is in favor of prostatic origin.
**FOLLOW-UP**

Patient again presented with acute retention of urine for which cystoscopy and TURP was done on March 28, 2013. Findings—grade I trilobar prostatic enlargement. Previous resection site in bladder was normal. Histopathology revealed stromal hyperplasia with prominent acute on chronic prostatitis. Focal areas of squamous metaplasia were seen. There was no evidence of malignancy in the sections evaluated.

His PSA increased to 4.02 ng/mL. Bone scan revealed metastasis in bones. Positron emission tomography and computed tomography (PET-CT) revealed fluorodeoxyglucose (FDG) avid increased activity in right 9th rib, pelvic and mediastinal lymph nodes and right lung. He was again started with injection zoladex and tablet bicalutamide. His PSA further increased to 8.95 ng/mL. He was advised for doxetacel-based systemic chemotherapy.

**COMMENTS**

Carcinoma of the prostate can involve bladder by direct infiltration which is usually at the base of the bladder. Isolated metastasis to bladder is rare. It is difficult to differentiate poorly differentiated cancer prostate and cancer bladder, but immunohistochemistry can help.

**BIBLIOGRAPHY**

10.5 CONCURRENT CARCINOMA PROSTATE AND PANCREATIC CANCER

■ INTRODUCTION

The incidence of multiple primary malignant neoplasm increases with age, reflecting an increase in overall cancer risk in older patients.

■ CLINICAL PRESENTATION

A 66-year-old male presented with raised PSA level to 24.70 ng/mL.

■ PAST HISTORY

Patient was a known case of moderately differentiated adenocarcinoma of the pancreas for which pancreaticoduodenectomy with partial gastrectomy was done on July 24, 2010. He developed metastasis in lungs in 2012. He was given chemotherapy in 2012. He had significant urinary complaints. Per rectal examination revealed prostate enlargement grade II, hard, nodular and rectal mucosa free.

■ INVESTIGATIONS

Magnetic resonance imaging of the prostate gland revealed hypoechogenicity in both lobes of prostate (Figs 10.5.1 and 10.5.2). Twelve core TRUS-guided biopsy of the prostate was done.

Fig. 10.5.1: Magnetic resonance imaging prostate—T2 images—sagittal section
Histopathology revealed acinar adenocarcinoma not overviews specified involving 71.5% of the cores. Gleason score being 3 + 4 and perineural invasion was seen.

**TREATMENT**

Hormonal treatment by injection leuprolide and tablet bicalutamide was started and he responded well. In follow-up, his PSA level returned to normal.

**COMMENTS**

There is no link between carcinoma of the prostate and carcinoma of the pancreas, and this is a chance existence due to higher incidence of cancers in elderly.

**BIBLIOGRAPHY**

10.6 GRANULOMATOUS PROSTATITIS

**INTRODUCTION**

A variety of granulomatous lesions of the prostate, with and without extensive infiltration by eosinophils have been described. Differing concepts of their pathogenesis and especially of their relation to allergic states have produced confusion and controversy. Granulomatous prostatitis may be specific or nonspecific. Nonspecific granulomatous prostatitis is densely cellular, granulomatous reactions extending throughout entire lobules.

**CLINICAL PRESENTATION**

A 45-year-old male presented with lower urinary tract symptoms. Digital rectal examination revealed prostate enlargement grade I, firm to hard and nontender.

**INVESTIGATIONS**

His PSA on April 05, 2013 was 3.04 ng/mL. MRI prostate revealed level diffuse T2 hypointensity with marked low apparent diffusion coefficient (ADC) values, high choline and type III curves in almost entire peripheral zone (PZ) of the prostate, highly suspicions of malignancy. Mild periprostatic fat

Figs 10.6.1A to C: Magnetic resonance imaging prostate
strandings was seen bilaterally and also around the neurovascular bundles, focal bulges in the left apex of the gland and basal PZ causing suspicion of infiltration of right seminal vesicle (Figs 10.6.1A to C). Twelve core TRUS-guided biopsy of prostate was done on May 06, 2013.

**HISTOPATHOLOGY**

Histopathology revealed dense mixed inflammatory cell infiltrate comprising of neutrophils, lymphocytes, plasma cells and eosinophils. Aggregates of epithelioid cells and multinucleate giant cells were also seen. No necrosis was seen. Ziehl-Neelsen (ZN) stain for acid-fast bacilli (AFB) was negative. Special stain for fungus was negative. There was no evidence of malignancy in the sections examined. Impression: Nonspecific granulomatous prostatitis.

**TREATMENT**

Patient was treated with alpha-blockers and anti-inflammatory drugs to which he responded.

**COMMENTS**

Patients present with features of chronic prostatitis and do not respond to treatment, need prostatic biopsy to clinch the correct diagnosis.

**BIBLIOGRAPHY**

10.7 LARGE PROSTATE WITH NO OBSTRUCTION

- **INTRODUCTION**
  The symptoms of BPH have no correlation with the size of prostate. Small prostate can give more symptoms and large prostate may be asymptomatic.

- **CLINICAL PRESENTATION**
  A 71-year-old male presented with frequency of micturition. Per rectal examination revealed grade III enlargement of prostate, firm, smooth and rectal mucosa free.

- **INVESTIGATIONS**
  Prostate-specific antigen was 1.5 mg/mL. Ultrasound revealed normal upper tracts (Fig. 10.7.1), prostate gland enlarged (110 mL) with intravesical projection (Fig. 10.7.2). Uroflowmetry revealed maximum flow rate of 27 mL/sec, average flow rate of 11.8 mL/sec, voided volume was 374 mL (Fig. 10.7.3).

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**Fig. 10.7.1:** Ultrasound kidneys

**Fig. 10.7.2:** Ultrasound bladder and prostate
As patient had no symptoms, no treatment was given. He was advised for regular follow-up once a year.

**COMMENTS**

Enlargement of prostate alone may not require treatment. These patients can be kept under observation.
Malakoplakia is an unusual chronic granulomatous inflammatory disorder that was originally described as affecting the bladder, but was subsequently found in other organs. Clinically, the patient presents with lower urinary tract symptoms. Digital rectal examination may reveal a hard nodule which increases the suspicion of malignancy. The ultrasonographic findings can also mimic prostatic adenocarcinoma. The majority of the cases have had proven previous episodes of *Escherichia coli* infection of the urinary tract, but bacilliform organisms have yet to be demonstrated in the diseased prostatic tissue.

**CLINICAL PRESENTATION**

A 59-year-old male presented with history of acute retention of urine which was relieved by Foley’s catheterization.

**EXAMINATION**

External genitalia were normal. Per rectal examination revealed grade II enlargement of prostate, hard and nodular on palpation and mucosa was free.

**INVESTIGATIONS**

The PSA level was found to be raised, (47.0 ng/mL) at the time of presentation. The PSA level reduced to 4.56 ng/mL after antibiotic treatment. The liver and renal function tests were found to be normal. The complete blood count, peripheral smear and coagulation profile were normal. Ultrasound reveals enlarged prostate (~66 gm) with fatty liver and gall bladder polyp.

Magnetic resonance imaging (MRI) showed diffuse T2 hypointensity in central and peripheral zone (PZ) with areas of restricted diffusion and low ADC. There is capsular breech and loss of fat planes with the pelvic side walls, neurovascular bundles and rectum on left side (Fig. 10.8.1).

Transrectal ultrasonography-guided 12 core biopsy was done.

**HISTOPATHOLOGY**

Microscopically, the cores showed dense mixed inflammatory cell infiltrate comprising of neutrophils, lymphocytes, plasma cells, histiocytes and foamy macrophages (Figs 10.8.2A and B). There was presence of Michaelis-Gutmann (M-G) bodies which are concentrically layered basophilic inclusions measuring 2–10 μm in diameter, and are thought to represent remnants of phagosomes mineralized by iron and calcium deposits (Fig. 10.8.2C). There was no evidence of malignancy in any of the cores studied.
Fig. 10.8.1: Magnetic resonance imaging

Fig. 10.8.2A: Prostate core biopsy showing inflammation completely replacing the glandular tissue (H & E, X 4x)

Fig. 10.8.2B: The inflammatory infiltrate is composed of lymphomononuclear cells and histiocytes (H & E, X 10x)

Fig. 10.8.2C: Histiocytic infiltrate in CD68 positive (CD68 IHC, X 40x)
Special stains were done on three blocks (B—left mid, H—right mid and I—right apex) which included periodic acid-Schiff (PAS) and von Kossa stains. For PAS stain 4 µm sections and for von Kossa 6 µm sections were collected on polylysine-coated slides. The slides were then subjected to dewaxing followed by standard procedures of staining for various stains. PAS stained positive the M-G bodies and von Kossa stained the calcium deposits.

Immunohistochemistry (IHC) was done using a panel of antibodies that included p63 (monoclonal, Biogenix, 4A4), Alpha-methylacyl-CoA racemase (AMACR) (Dako) and cluster of differentiation (CD) 68 (monoclonal, LEICA, S14H12, RTU-CD68). For IHC, 4 µm sections were collected on polylysine-coated slides and subjected to steps of dewaxing, antigen retrieval and blocking of nonspecific sites, followed by incubation with the respective primary antibody. Supersensitive polymer horseradish peroxidase (HRP) detection system (EnVision Flex) and 3,3-diaminobenzidine (DAB) chromogen (EnVision Flex) was used as the chromogenic substrate. Hematoxylin was used as counterstain. Appropriate positive and negative controls were incorporated in each run for all antibodies. p63 demonstrated preservation of basal layer of prostatic glands. AMACR were negative. CD68 highlighted the histiocytes.

**FOLLOW-UP**

Patient underwent bipolar TURP (Fig. 10.8.3). He started voiding well and was put on long-term suppressive antibiotics. On 3-month follow-up, it was asymptomatic with PSA level 1.08 ng/mL. Ultrasound revealed a residual prostate which measures 10 cc in volume, with no significant post void residual urine.

*Fig. 10.8.3: Transurethral resection of the prostate (TURP) chips*
**COMMENTS**

Malakoplakia is a difficult pathological diagnosis, and the prevalence of this disease may be underestimated. Clinical findings and imaging cannot distinguish between malignancy and malakoplakia. Biopsy is mandatory to clinch the correct diagnosis.

**BIBLIOGRAPHY**

10.9 METASTASIS LOWER URETER FROM CANCER PROSTATE

**INTRODUCTION**

In 1909, Stow reported the first case of true metastatic involvement of the ureter by a malignancy, a lymphosarcoma. A true metastasis to the ureter is defined as spread of a neoplasm to the ureteral wall, seldom invading the mucosa into the lumen probably with periureteral lymphatic involvement. To be a true metastasis, there must be no ureteral involvement of the tumor by direct extension or contiguity. Only a few ureteral metastases are diagnosed during life and up to 85% are asymptomatic. McLean and Towler reported 18 such cases among 10,223 consecutive autopsies and only one was from prostate adenocarcinoma.

The neoplasms more commonly metastatic to the ureter are from the breast, colon, lungs, stomach, uterus and lymphatic. Adenocarcinoma is the most frequent histopathology. Ureteral metastasis may occur at any age and at any level of the ureter. Multiple ureteral lesions are seen; 60% had bilateral involvement and 90% had metastases to other organs.

**CLINICAL PRESENTATION**

A 77-year-old male presented with left flank pain for 2 months. He underwent ureteroscopic biopsy with double J (DJ) stenting in another hospital. Histopathology revealed papilloma low grade transitional cell carcinoma (TCC). DJ stenting was done.

**PAST HISTORY**

Patient was known case of adenocarcinoma with Gleason score 4 + 3, with perineural spread and this was diagnosed in September 2007. He was treated with hormone therapy—maximal androgen blockade (MAB) (bicalutamide and bilateral orchidectomy) with TUR channeling in 2007. PSA level came down from 11.58 ng/mL to 0.43 ng/mL (on July 26, 2008).

Abdominal examination revealed no organomegaly. Digital rectal examination revealed grade 1 prostate, smooth, nontender, no nodularity and rectal mucosa free.

**INVESTIGATIONS**

Urine culture/sensitivity (C/S) was sterile. Urine cytology was negative. PSA level was 0.43 ng/mL. X-ray chest was normal. Ultrasound revealed normal kidneys, 17.8 gm prostate. CECT abdomen (Fig. 10.9.1A and B) revealed 2.3 \( \times 2 \times 1.7 \) cm mass in lower end of left ureter with no extension to the bladder no lesion in the left kidney and impaired excretion in right kidney.

*Cystoscopy*: Anterior urethra: normal, prostatic fossa: no growth. Right ureteric orifice: normal. Left ureteric orifice: normal. A bulge was seen superomedial to left ureteric orifice.
Left ureteric mass excision with cuff of bladder, with pelvic lymphadenectomy was done (Fig. 10.9.2). Figure 10.9.3 show excised specimen. Proximal (ureter) and distal margins (bladder) were sent for frozen section—margins were negative. Ureteroneocystostomy was done with 6 Fr DJ stent.
Gross examination revealed ulceroproliferative growth of 3.5 × 3 × 0.8 cm in left lower ureter. Histopathology revealed poorly differentiated adenocarcinoma with local infiltration in the muscle presently. PSA stain was positive, CK-7 and CK-20 were negative. Margins were proximal was negative, distal focal tumor infiltrate present. External iliac lymph nodes—1 out of 3 was positive. Postoperative recovery was uneventful. He was given hormonal treatment.

**COMMENTS**

Ureter can be involved by prostate carcinoma either by direct infiltration or by lymphatic spread. Although ureteral metastases from prostate carcinoma is rare, but it should be taken into consideration even when the PSA is normal before considering for double malignancy.

**BIBLIOGRAPHY**

INTRODUCTION

The French term “peau d’orange” means “orange peel skin,” or more literally, “skin of an orange.” *Peau d’orange* is caused by cutaneous lymphatic edema, which causes swelling. However, the infiltrated skin is tethered by the sweat ducts such that it cannot swell, leading to an appearance like orange skin. It is commonly seen in cases of breast cancer. Occasionally, the same phenomenon is seen over a chronic abscess.

CLINICAL PRESENTATION

A 74-year-old male presented with acute urinary retention. He had history of type 2 diabetes mellitus for 20–25 years.

EXAMINATION

Bladder was distended up to umbilicus. Per rectal examination revealed a hard, nodular prostate. Catheterization was done. Investigations revealed serum PSA level—7.31 ng/mL. TRUS-guided 12 core biopsy revealed adenocarcinoma, prostate, No otherwise specified (NOS) type, Gleason score being 5 + 4 = 9 with perineural and perivascular invasion. A total of 50% of the tissue submitted was involved by tumor. Infiltration into periprostatic muscle and fat was seen.

Fig. 10.10.1: Clinical picture
**TREATMENT**

Injection leuprolide 22.5 mg and tablet bicalutamide was started. Bone scan revealed multiple secondaries. MRI revealed features of Carcinoma prostate with extracapsular extension, involvement of urinary bladder, seminal vesicles, neurovascular bundles and rectum. There were nodular mass like lesions seen in the mesorectum and pararectal region with pelvic bony metastasis. There was bilateral hydroureter with left hydronephrosis. Multiple enlarged retroperitoneal, iliac, inguinal, perirectal and presacral lymph nodes were seen. Channeling TURP was done and patient started voiding well.

**FOLLOW-UP**

Patient was well for 1 year. After that, his PSA started rising. PET-CT scan revealed hypermetabolic left, supraclavicular, mediastinal, retroperitoneal, pelvic and inguinal lymphadenopathy with enhancing FDG avid nodules in prostate with hypermetabolic sclerotic bony lesions suggestive of metastatic carcinoma of the prostate. Docetaxel-based chemotherapy was given but he showed no response.

He presented with difficulty in passing urine and swelling around penis. On examination (Fig. 10.10.1), there was diffuse swelling over the suprapubic region, penis and scrotum with peau d’orange appearance. Trocar cystostomy was done to relieve his symptoms. He developed chronic renal failure and was put on hemodialysis. Finally, he died after 3 months.

**COMMENTS**

Peau d’orange appearance in prostate cancer can be seen in advanced stage with lymphatic obstruction.
**INTRODUCTION**

Hemorrhagic cystitis can occur 6 months to 10 years after pelvic radiation therapy with moderate to severe persistent rates of hematuria in 3–5% cases of radiotherapy for pelvic malignancies. Radiation-induced severe hemorrhagic cystitis is difficult to treat. Current treatment modalities for hemorrhagic cystitis include oral and intravenous agents, intravesical therapy and selective embolization of the hypogastric arteries. In extreme cases, surgery is required.

**CLINICAL PRESENTATION**

A 62-year-old man presented with intractable hematuria with clot retention on and off. He required frequent hospital admissions for bladder wash and blood transfusions.

**PAST HISTORY**

Patient was diagnosed within carcinoma prostate patient in 2006 for which he was treated with radiotherapy. He had obstructive symptoms for which TURP done in 2008. Post TURP, he developed stricture urethra with bladder neck obstruction for which laser optical internal urethrotomy (OIU) was done. He was on self-catheterization. In January 2013, he presented with retention of urine for which suprapubic catheterization (SPC) was done. Later, he underwent buccal mucosa urethroplasty after which he again developed retention of urine for which OIU was done. He subsequently developed urinary incontinence.

**EXAMINATION**

On examination, he had SPC in place. Per rectal examination revealed a flat prostate.

**INVESTIGATIONS**

Investigations revealed normal renal functions. Retrograde urethrogram (RGU) with MCU (Fig. 10.11.1) revealed stricture in bulbomembranous urethra with thick-walled bladder. CT urogram (Fig. 10.11.2) revealed bilateral hydroureteronephrosis with thick-walled bladder. CECT scan (Fig. 10.11.3) revealed thick-walled bladder with SPC tract.
Salvage cystoprostatectomy with ileal conduit diversion was done. Figures 10.11.4A to C show cystoprostatectomy specimen. Histopathology revealed features of cystitis. No evidence of malignancy was seen in multiple sections in the bladder. Bilateral seminal vesicles, vas and ureter were free of tumor. There was no evidence of tumor in the prostate.
Postoperative recovery was uneventful. Ureteric stents were removed after 2 weeks. Ileal conduit stoma was working well. Complete blood count (CBC) and renal function test (RFT) were normal. Patient returned to his normal work.

**FOLLOW-UP**

Patient was doing well with no complaints at 1 year. His PSA level was 0.05 ng/mL.

**COMMENTS**

Surgical management of post radiation cystitis offers good functional outcomes.

**BIBLIOGRAPHY**

11.1 CONGENITAL ANTERIOR URETHRAL OBSTRUCTION

INTRODUCTION

Congenital obstruction of the distal urethra is much less common than obstruction of the proximal urethra. It can be due to anterior urethral valves (congenital diverticulum of urethra), valvular obstruction of the fossa navicularis, and cystic dilatation of the ducts of Cowper’s gland (syringocele). Of these, the most common cause is anterior urethral valves. The diagnosis may be confirmed by endoscopy when a sharp valvular ridge should be easily seen.

CLINICAL PRESENTATION

A 30-year-old male presented with acute urinary retention necessitating suprapubic cystostomy (SPC) 4 months ago. Past history of obstructive voiding with straining since childhood, history of circumcision 8 years ago, repeated dilatation 6 monthly since last 6 years.

EXAMINATION

Circumcised, no evidence of balanitis xerotica obliterans (BXO), urethra soft to feel. Oral hygiene and mucosa were healthy.

INVESTIGATIONS

Retrograde urethrogram (RGU) revealed dilatation of the proximal urethra with narrow anterior urethra (Fig. 11.1.1).
TREATMENT

Urethroscopy revealed valve in the anterior urethra for which laser incision was done.

COMMENTS

Anterior urethral obstruction is rare and can be diagnosed by retrograde urethrogram or micturating cystourethrogram (MCU) and can be managed endoscopically.

BIBLIOGRAPHY

INTRODUCTION

There are several options for the management of failed urethroplasty like optical internal urethrotomy, core through internal urethrotomy, redo-urethroplasty, etc. In some cases, all these procedures may not be successful and patient may become incontinent. In such a situation, continent urinary diversion is the option.

CLINICAL PRESENTATION

A 12-year-old male suffered road traffic accident in August 1987. He had fracture femur and pelvic bones. He underwent suprapubic cystostomy (SPC) and internal fixation of a fractured femur. Transpubic urethroplasty was done in March 1988. He had retention 1 month after catheter removal. SPC was done again.

He had multiple urethral calibrations or dilatations. He developed completely choked urethra by December 1988. Core through (cold knife) internal urethrotomy was done. He could not pass urine after catheter removal.

Scrotal tube urethroplasty was done in March 1989. He required multiple calibrations or dilatations postoperatively. He developed false passage in the urethra. Optical internal urethrotomy (OIU) was done in 1990.

Subsequently, he had continuous urine leak per urethra and was on SPC.

EXAMINATION

Perineal urethrostomy opening was seen. Stone felt in blind ending tube. There was wide gaping of rectus muscle and pubic diastases. Small penile length (Figs 11.2.1 and 11.2.2).

Fig. 11.2.1: Clinical picture—Lower abdomen  Fig. 11.2.2: Clinical picture—Perineum
INVESTIGATIONS

Micturating cystourethrograph showed small capacity bladder with wide opening bladder neck (Fig. 11.2.3). Endoscopic evaluation revealed small capacity, smooth-walled bladder, wide open bladder neck, complete block distal to veru, hair ball and calculus seen inside the blind end of scrotal tube and stone extracted, bony hard callus around the scrotal tube.

![Micturating cystourethrogram](image)

**Fig. 11.2.3: Micturating cystourethrogram**

TREATMENT

In view of multiple failed urethroplasties and repeated failed OIU, ileocecal augmentation cystoplasty and continent urinary diversion using appendix (Penn’s pouch) and closure of bladder neck was done in 2008 (Figs 11.2.4 to 11.2.11).
Fig. 11.2.4: Operative pictures—Bladder neck closure

Fig. 11.2.5: Ileocecal segment isolation
Figs 11.2.6A to C: Detubularization and refashioning of ileocecal segment

Figs 11.2.7A to D: Reconstruction of continence mechanism
Figs 11.2.8A and B: Closure of augmented bladder

Figs 11.2.9A and B: Creation of Mitrofanoff stoma

Fig. 11.2.10: Postoperative micturating cystourethrogram
**FOLLOW-UP**

From the last 5 years, patients is doing regular intermittent self-catheterization, is asymptomatic and continent with normal renal and urinary parameters.

He developed a small stone in the continent pouch in 2011, which was removed endoscopically through the appendiceal opening.

**COMMENTS**

Continent urinary diversion is the last salvage procedure in management of cases of stricture urethra with multiple failed urethroplasty with or without urinary incontinence.

**BIBLIOGRAPHY**

INTRODUCTION

Foreign bodies in lower urinary tract are rare. The common causes of such cases are curiosity, auto erotic stimulation, psychotic diseases and sexual behavior in inebriated state and medical procedures. The foreign bodies vary from wire, screw, ball point pen and even rare cases of animals such as a snake have been reported in literature. The common symptoms are severe pain, hematuria, frequency and urinary tract infection. Procedures to remove such foreign bodies should be as simple as possible and should result in minimal damage to the bladder and urethra.

CASE 1

Clinical Presentation

A 25-year-old male presented in emergency with difficulty in passing urine for 3 days. He gave history of playing with his genitalia with a safety pin.

Examination

External genitalia were normal. On palpation, a firm object was palpable in the area of perineum.

Investigations

Plain X-ray of the pelvis revealed a safety pin in the perineal region. RGU confirmed safety pin in the region of bulbomembranous urethra (Fig. 11.3.1).

Fig. 11.3.1: Case 1—Safety pin in urethra
**Treatment**

Cystourethroscopy revealed safety pin which was caught in a forceps and gently removed. Bladder was found normal. Urethral catheter was passed and kept for 3 days.

**CASE 2**

A 16-year-old boy presented with history of dysuria persisting for last 6 months. He had history of catheterization in the past.

**Examination**

External genitalia were normal.

**Treatment**

Cystoscopy revealed a piece of catheter in the urethra which was removed endoscopically (Fig. 11.3.2).

![Fig. 11.3.2: Case 2—Broken catheter in urethra](image)

**COMMENTS**

Foreign body in urethra is rare. In case of unusual history and symptoms, patient should be enquired for insertion of a foreign body. If found, can be managed accordingly.
BIBLIOGRAPHY

11.4 GUNSHOT INJURY WITH STRICTURE URETHRA AND RECTO-URETHRAL FISTULA

**INTRODUCTION**

Recto-urethral fistula may have a congenital or acquired origin. Acquired cases include inflammatory, neoplastic, or traumatic etiologies. Traumatic etiological factors are injury due to surgical procedures, road traffic, accidents and rarely by gunshot injury. Magnetic resonance imaging (MRI) helps in identification of extent of injury and helps in planning of management. The aim of the surgical approach is the closure of all types of fistulas. Diversion of urine by suprapubic cystostomy is mandatory and temporary fecal diversion by colostomy is required. There are different approaches for management like perineal, posterior sagittal, anterior, combined, endoscopic, etc. The approach is to be individualized according to etiology, and location of the fistula. Surgery of recto-urethral fistulas is technically demanding and requires collaboration with rectal surgeons.

**CLINICAL PRESENTATION**

A 25-year-old male had gunshot injury during a local fight. Patient was admitted in a local hospital and SPC was done. Patient presented to us with recurrent fever.

**EXAMINATION**

Local examination revealed no significant findings.

**INVESTIGATIONS**

Intravenous pyelogram (IVP) revealed normal upper tracts with pellets in the pelvic region (Fig. 11.4.1). Retrograde urethrogram (RGU) and Micturating Cystourethrogram (MCU) revealed stricture bulbomembranous junction with communication with rectum (Figs 11.4.2 and 11.4.3).
Fig. 11.4.1: Intravenous pyelogram showing normal upper tracts with pallets

Fig. 11.4.2: Retrograde urethrogram and micturating cystourethrogram showing stricture bulbomembranous urethra
**TREATMENT**

After control of infection, combined perineal and transpubic repair of rectourethral fistula with end to end anastomosis of urethra with omental wrap was done. Postoperative recovery was uneventful. Follow-up MCU revealed normal urethra (Fig. 11.4.4).

![Fig. 11.4.3: Micturating cystourethrogram showing communication with colon](image)

![Fig. 11.4.4: Postoperative micturating cystourethrogram](image)
**FOLLOW-UP**

At follow-up of 12 years patient was voiding well. He had urinary incontinence and erectile dysfunction for which artificial urinary sphincter and penile prosthesis surgery was done.

**COMMENTS**

Managements of recto-urethral fistula is challenging. Patients need detailed evaluation, imaging and multi disciplinary surgery.

**BIBLIOGRAPHY**

11.5 LATERAL PERINEOSCROTAL FLAP URETHROPLASTY

INTRODUCTION

For urethral stricture more than 2 cm in length, substitution urethroplasty is recommended, which can be done in one or two stages. Genital skin is preferred but in some cases perineoscopic flap urethroplasty may be required. Inverted U-shaped flap is done but in cases of previously operated by perineal incision, blood supply will be compromised and same inverted U-shaped flap cannot be used. In such a situation, lateral perineoscopic flap urethroplasty is recommended.

CASE 1

Clinical Presentation

A 25-year-old male presented with a history of traumatic urethral stricture 12 years ago. He had history of fall and injury to perineum.

Examination

He had suprapubic cystostomy (SPC) in situ. Multiple midline scars noted in the perineum.

Past History

He underwent SPC and multiple stage urethroplasty and urethral dilatation 12 years ago which failed. He underwent fistulectomy and evacuation of abscess post-surgery. He also developed persistent vesicocutaneous fistula for which SPC was done.

Investigations

X-ray pelvis was normal (Fig. 11.5.1). RGU revealed complete stricture at the distal bulbar urethra (Fig. 11.5.2). MCU showed vesicoureteral reflux on the left side (Fig. 11.5.3A). MCU lateral film showed nonopening of the bladder neck (Fig. 11.5.3B). Noncontrast MRI lower abdomen revealed features of prostatitis. SPC with Foley’s bulb was seen in urinary bladder. Dilated posterior urethra was seen. Status of rest of the urethra should not be commented as it was collapsed (Fig. 11.5.4).

Urethroscopy and suprapubic cystoscopy revealed anterior urethra to be normal with complete block at distal bulbar urethra, bladder neck and prostatic urethra were normal, veru was normal, totally obstructed urethra beyond veru.
Fig. 11.5.1: Case 1—X-ray pelvis

Fig. 11.5.2: Case 1—Retrograde urethrogram
Right lateral perineoscrotal flap was created and first stage urethroplasty was done (Fig. 11.5.5). Completely fibrosed urethra adhered to bulbospongiosus muscle. Stricturous urethra identified and opened. The proximal urethra identified by Hay groove dilator antegrade and opened. The gap was approximately 8 cm, per rectal finger was kept throughout dissection to prevent injury to rectum, urethral opening anastomosed to dartos-based lateral perineoscrotal flap. Two catheters were kept in the proximal and distal urethra and in the SPC tract.
Urethral catheter was removed after 2 weeks. SPC catheter was clamped. Patient started voiding well. SPC catheter was removed after 4 weeks.

Fig. 11.5.5: Case 1—Postoperative clinical picture

Fig. 11.5.6: Case 2—Clinical picture
CASE 2

Clinical Presentation

A 10-year-old male child presented with history of urethral injury following road traffic accident. He underwent end to end urethroplasty (perineal approach) 1 year ago. There was gradual slowing of urinary stream. He developed perineal abscess followed by urinary leak.

Examination

External genitalia were normal. Urethral fistula noted in the perineum (Fig. 11.5.6). Rectal examination was normal.

Investigations

Retrograde urethrogram revealed stricture at bulbomembranous junction with large cavity with discharging sinus (Figs 11.5.7A and B). MCU revealed vesicoureteral reflux on the right side (Fig. 11.5.8).

Figs 11.5.7A and B: Case 2—Retrograde urethrogram

Fig. 11.5.8: Case 2—Micturating cystourethrogram
Treatment

Right lateral perineoscrotal flap first stage urethroplasty with excision of cavity and sinus was done. Right lateral perineoscrotal flap incision (Fig. 11.5.9). Proximal urethra was identified by Hay groove dilator antegradely (Fig. 11.5.10A). Excision of the fistulous tract and cavity was done. Anastomosis of the lateral flap with the proximal urethra was done (Figs 11.5.10B and C). Figure 11.5.11 showing picture after complete anastomosis. Two catheters were kept in the proximal and distal urethra and in SPC tract.

Urethral catheter was removed after 2 weeks. SPC catheter was clamped. Patient started voiding well. SPC catheter was removed after 4 weeks. Second stage closure was done after 6 months.

**Fig. 11.5.9:** Case 2—Right lateral perineoscrotal flap incision

**Figs 11.5.10A to C:** Case 2—Intraoperative pictures
COMMENTS

Lateral perineoscrotal flap staged urethroplasty is indicated in cases of previously failed urethroplasty done by use of perineal flap. Blood supply of the flap is maintained by blood vessels coming from lateral side.

BIBLIOGRAPHY

11.6 NEOPHALLUS WITH URETHRAL FISTULA

**INTRODUCTION**

Phalloplasty is the construction or reconstruction of a penis, or the artificial modification of the penis by surgery, often for cosmetic purposes after amputation of the penis or for sexual reassignment. An operation using the forearm as a donor site is used in large number of cases. The chances of complications of the extended urethra are higher, averaging 55%. The most common complications reported are urinary fistula (hole) requiring perineal urethrostomy, urinary retention (from stenosis or narrowing of the new urethra), etc.

**CLINICAL PRESENTATION**

A 35-year-old male presented with history of dysuria and urinary leak from the perineum for 1 year duration.

**PAST HISTORY**

He had traumatic amputation of penis at the age of 22 years. He had undergone neophallus reconstruction using forearm graft with ulnar bone.

**EXAMINATION**

On examination of external genitalia, it was noted that he had well-made phallus but no erection. He had a fistulous tract at the junction of scrotum and penis.

**INVESTIGATIONS**

Retrograde urethrogram (Fig. 11.6.1) revealed neophallus with urethral fistula.
He was counseled for the staged repair of urethral fistula, but he did not come back.

**COMMENTS**
Reconstructed neophallus can have several complications including urinary fistula. They need a regular follow-up.

**BIBLIOGRAPHY**
11.7 POST-HYPOSPADIAS STRICTURE URETHRA

**INTRODUCTION**

Stricture urethra is a significant complication of hypospadias reconstruction. Treatment should initially be dilation, direct vision internal urethrotomy. Open surgical repair will usually be successful, but should be reserved for difficult strictures, late strictures or failures of endoscopic procedures.

**CLINICAL PRESENTATION**

A 25-year-old male presented with complaints of difficulty in passing urine since last 2 years.

**PAST HISTORY**

He had hypospadias repair during childhood. The detail of the repair was not available.

**EXAMINATION**

Meatus was normal. Penis was circumcised. Loose skin was present over ventral side. Penis was short. Both testes were normal. He was having normal erections.

**INVESTIGATIONS**

Retrograde urethrogram revealed dilatation of repaired urethra with proximal urethral stricture (Fig. 11.7.1).

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**Fig. 11.7.1:** Retrograde urethrogram
**TREATMENT**

Cystourethroscopy revealed presence of skin at the repaired site with stricture at the proximal end. Optical interval urethrotomy (OIU) was done.

**COMMENTS**

Following hypospadias repair, stricture urethra is common at the proximal or distal anastomosis. They can be managed endoscopically otherwise complete reconstruction in one or two stages may be required.

**BIBLIOGRAPHY**

11.8 STRICTURE URETHRA—SYRINGOCELE

INTRODUCTION
The dilated Cowper’s gland duct is referred to as a syringocele (in Greek, syringe means “tube” and cele means “swelling”). Syringoceles are cystic dilations of Cowper’s gland duct within the bulbous urethra. They are usually small, inconsequential lesions. But rarely, they can be of sufficient size to cause varying degrees of outlet obstruction.

The urethrographic and endoscopic characteristics of dilated Cowper’s gland ducts are classified in four groups:

1. *Simple syringocele:* A minimally dilated duct.
2. *Perforate syringocele:* A bulbous duct that drains into the urethra via a patent ostium and appears as a diverticulum
3. *Imperforate syringocele:* A bulbous duct that resembles a submucosal cyst and appears as a radiolucent mass
4. *Ruptured syringocele:* The fragile membrane that remains in the urethra after a dilated duct rupture.

Tubular or cystic dilatation of Cowper’s gland duct has been called syringocele while congenital urethral narrowing is known as Cobb’s collar.

CLINICAL PRESENTATION
A 23-year-old male presented with complaints of slow stream, straining and post-void dribbling since last 8 years. There was no history of fever, hematuria, dysuria, trauma, surgery or instrumentation in the past, no other comorbidities.

EXAMINATION
External genitalia—glans and meatus were normal, no visible swelling.

INVESTIGATIONS
Urine R/M: RBC 2+, WBC: 3–4/hpf, Urine culture: Sterile
Blood urea: 23mg/dL, Serum creatinine: 0.9 mg/dL. Uroflowmetry: Plateau-shaped curve, Qmax: 5.6 mL/s, voided volume: 224 mL
Retrograde urethrogram revealed a stricture at the proximal bulbar urethra with a diverticulum-like structure ventrally (Fig. 11.8.1).
**TREATMENT**

Patient underwent retrograde urethroscopy on March 12, 2008. Per operative finding was stricture at proximal bulbar urethra (2.5 cm). An opening proximally on ventral surface of bulbous urethra. Excision of stricture along with the dilated sac followed by end to end urethroplasty was done. Postoperative period was uneventful. Patient discharged on 5th postoperative day. Patient was on catheter and voiding normally after catheter removal.

**HISTOPATHOLOGY**

Focal squamous metaplasia with subepithelial fibrosis compatible with stricture disease.

**COMMENTS**

It is important to identify the etiology of stricture urethra. Syringocele is a rare finding and can be diagnosed on imaging and treated according to presentation.

**BIBLIOGRAPHY**


Fig. 11.8.1: Retrograde urethrogram
11.9 SUPRAPROSTATIC STRICTURE URETHRA

INTRODUCTION

The preprostatic urethra or supraprostatic urethra is one of the four parts of the male urethra. The preprostatic urethra is also known as the intramural urethra, as it is the portion of the urethra which passes almost vertically through the wall of the urinary bladder, before it enters the prostate gland. Causes of supraprostatic strictures are: (1) Anastomotic strictures following radical prostatectomy; (2) Bladder neck strictures after transurethral resection of the prostate (TURP) and open prostatectomy; (3) Post-traumatic. The treatment modalities are endoscopic, open repair or continent cutaneous diversion. The endoscopic are cold knife incision, balloon dilation, filiform or follower, laser core through urethrotomy and endoprosthesis. They require repeated courses and failure rates are as high as 27–75%. If first attempt fails, repeated attempts have poorer success rates. The open procedures of urethral reconstruction are by vascularized tissue transfer techniques using bladder. Several techniques are described.

CLINICAL PRESENTATION

A 32-year-old male presented with suprapubic catheter with retention of urine.

PAST HISTORY

He had road traffic accident in October, 2005. He was in shock. He had pelvic fracture and blunt abdominal trauma. He underwent exploratory laparotomy, repair of intraperitoneal and extraperitoneal bladder rupture and railroading was done. Patient was discharged on SPC and per urethral catheter (PUC). PUC removed after 8 weeks. He developed obstructive urinary flow after catheter removal. His urine flow gradually worsened and went into acute retention of urine. RGU and MCU were done which revealed stricture of the supraprostatic region (Fig. 11.9.1). Laser core through urethrotomy was done in another hospital twice which failed. Patient was then referred to us.

INVESTIGATIONS

Retrograde urethrogram revealed normal anterior and posterior urethra with high lying bladder neck with obstruction (Figs 11.9.2A and B).

Retrograde urethroscopy showed normal anterior urethra and posterior urethra up to supramontanal region. On antegrade cystoscopy bladder neck could not be seen, except for scarring in the region of bladder neck.
Fig. 11.9.1: Retrograde urethrogram

Figs 11.9.2A and B: Retrograde urethrogram and micturating cystourethrogram

**TREATMENT**

Bladder neck reconstruction was done by transpubic approach. Rectangular flap was made from anterior wall of bladder and swung downwards. Bladder wall flap was fashioned into a tube around the catheter using 3–0 vicryl. Prostatic urethra was opened over the bougie. Reconstructed tube was anastomosed to the prostatic urethra (Fig.11.9.3). SPC was placed. He developed fecal fistula which was managed conservatively; transverse loop colostomy was done with daily dressing. Patient was discharged on SPC and PUC.
FOLLOW-UP

Three months later, pericatheter RGU was done revealing no extravasation of contrast (Fig. 11.9.4). PUC was removed. MCU showed wide patent urethral tube (Fig. 11.9.5). He was continent. Uroflowmetry revealed maximum flow rate of 31 mL/sec (Fig. 11.9.6).

Three months later, he was voiding well. Colostomy was closed and SPC was removed.
Management of supraprostatic stricture is challenging and treatment is to be individualized. If endoscopic treatment fails, open surgery by vascularized flap is recommended.

**BIBLIOGRAPHY**

INTRODUCTION

Transitional cell carcinoma (TCC) is the most common type of bladder cancer and cancer of the ureter, urethra and urachus. Synchronous and asynchronous urethral transitional cell carcinoma, in relation to bladder cancer in male and female can occur. Four to eighteen percent of patients will develop recurrent urethral TCC in the remnant urethra after cystectomy.

CLINICAL PRESENTATION

A 72-year-old diabetic male with bilateral cataract presented with complaints of poor flow of urine, scrotal swelling and pus discharge from the left hemiscrotum.

PAST HISTORY

In July 2012, the patient presented outside with bladder outlet obstruction and posted for TURP. During cystourethroscopy, bulbar urethra showed growth, posterior urethra and bladder were normal. Transurethral resection of the urethral tumor was done, patient was sent home with catheter. Catheter slipped out at home and patient presented 2 days later with high fever and large scrotal swelling, incision and drainage of the scrotal abscess was done. Cystoscopy done at the same time showed a fistulous opening in bulbar urethra leading to left hemiscrotum. Histopathology of resected growth showed transitional papilloma (grade 1 TCC). Subsequently, fistula healed and cystoscopy revealed no growth.

In March 2013, the patient presented with poor general condition, bladder outlet obstruction and pus discharge from scrotal opening. Cystourethroscopy revealed large recurrence of urethral growth with fistulous tract in bulbar urethra. Transurethral resection (TUR) of urethral tumor was done and catheter placed. Histopathology revealed grade 1 TCC.

EXAMINATION

Abdomen—soft, nontender and no organomegaly.
Local examination revealed a large scrotal swelling on left side with erythema, induration and a pus discharging opening in the left hemiscrotum (Figs 11.10.1A and B). Urethra—a hard growth was palpable in bulbar urethra. Digital rectal examination (DRE) revealed grade 1 prostate, firm, smooth and nontender.
Urethral Diseases

INVESTIGATIONS

Hemoglobin (Hb): 12.8 gm/dL, Total leukocyte count (TLC): $15.16 \times 10^3/\mu\text{L}$; Serum creatinine: 1.50 mg/dL; Urine R/M: pus cells: numerous, albumin: positive; Urine C/S: *Escherichia coli* sensitive to amikacin, gentamycin, levofloxacin and nitrofurantoin, Urine for malignant cytology: negative. Ultrasonography (USG) kidney, ureter and bladder (KUB): Bilateral kidneys normal in shape, size and echotexture, Bilateral hydronephrosis (HDUN); Bladder: normal; Prostate: 18.6 cc. RGU revealed extravasation of contrast and fistula to perineum (Figs 11.10.2A and B). MRI revealed a large tumor involving left hemiscrotum and base of the penis. There was thickening at the base of the bladder but no tumor (Figs 11.10.3A to C).

Figs 11.10.1A and B: Clinical picture

Figs 11.10.2A and B: Retrograde urethrogram
TREATMENT

Urethrocystoscopy revealed an ulceroproliferative growth in the anterior urethra (Figs 11.10.4A and B). Bladder and prostate were not involved.
Total scrotectomy with penectomy and anterior urethrectomy with perineal urethrostomy was done under spinal on June 18, 2013. Figures 11.10.5A and B show the operative specimen. Figure 11.10.6 is shows postoperative picture.

Figs 11.10.5A and B: Operative specimen

Fig. 11.10.6: Postoperative picture
**FOLLOW-UP**

The patient came for follow-up at 1 week and 2 weeks, postoperatively. Catheter was removed after 2 weeks. Patient started voiding well. Wound healed well (Fig. 11.10.7). He was advised adjuvant chemotherapy.

**HISTOPATHOLOGY**

High-grade urothelial carcinoma of the urethra, stage: pT3 pNx pMx

Both testes were free of tumor, resected margins were free, tumor was invading corpus spongiosum and corpus cavernosum.

**COMMENTS**

Transitional cell carcinoma of urethra is rare and can occur after cystectomy or in isolation. Radical surgery can salvage the patient.

**BIBLIOGRAPHY**

11.11 COMPLEX URETHRAL STRICTURE IN CHILDREN

**INTRODUCTION**

Urethral strictures in children are rare but difficult urological problem. There are similarities as well as marked differences between children and adults in the nature of the lesions. The etiology of stricture urethra can be congenital, infectious and traumatic which can be iatrogenic or noniatrogenic.

In post-traumatic stricture urethra in children, the level of the traumatic posterior urethral distraction defect is less predictable. There are three distinct types of urethral injury: (1) supraprostatic; (2) transprostatic; and (3) prostatomembranous. The divided urethral ends are more markedly separated and bladder neck is involved commonly due to rudimentary prostate and delicate puboprostatic ligaments.

The management is temporary diversion, urethral dilatation, direct visual internal urethrotomy (DVIU) which can be electrosurgical or neodymium-doped yttrium aluminum garnet (Nd-YAG), holmium laser and urethroplasty. When there has been failure of dilation or internal urethrotomy, or when above procedures are not desirable, urethroplasty is the preferred therapy. In complex posterior urethral strictures, transpubic urethroplasty is recommended.

**CLINICAL PRESENTATION**

A 9-year-old male child presented with history of road traffic accident resulting in fracture pelvis and rupture urethra. In emergency, he had suprapubic cystostomy. Later on, he had perineal urethroplasty which failed. He underwent laser OIU which also failed. He came to us with a suprapubic cystostomy.

**EXAMINATION**

Suprapubic and perineal scars were present. Penis was normal (Fig. 11.11.1).

**INVESTIGATIONS**

Retrograde urethrogram revealed normal anterior urethra and complete block at membranous urethra. X-ray also showed healed fracture of pubic bones (Fig. 11.11.2). MCU revealed normal bladder, no vesicoureteral reflux, and bladder neck was competent (Fig. 11.11.3).
Fig. 11.11.1: Clinical picture

Fig. 11.11.2: Retrograde urethrogram

Fig. 11.11.3: Micturating cystourethrogram
TREATMENT

Transpubic urethroplasty with combined transpubic and perineal approach was done. In lithotomy position, lambdoid perineal incision was made (Fig. 11.11.4). Distal urethra was mobilized (Fig. 11.11.5). Suprapubic incision was made. Bladder was mobilized and proximal end of urethra was identified (Fig. 11.11.6). Mucosa was everted at both ends of the urethra (Fig. 11.11.7). End to end urethral anastomosis was done (Fig. 11.11.8). Omental wrap was done around the anastomosis (Fig. 11.11.9). Postoperative recovery was uneventful.
Fig. 11.11.6: Proximal end of urethra

Fig. 11.11.7: Everted urethral mucosa

Fig. 11.11.8: End to end urethral anastomosis

Fig. 11.11.9: Omental wrap
FOLLOW-UP

Urethral catheter was removed after 3 weeks. Patient started voiding well. MCU revealed open bladder neck and urethra (Fig. 11.11.10).

![Postoperative micturating cystourethrogram](image)

**Fig. 11.11.10:** Postoperative micturating cystourethrogram

COMMENTS

Transpubic urethroplasty is preferred treatment in children for the management of complex urethral stricture.

BIBLIOGRAPHY

11.12 CONGENITAL URETHRAL DIVERTICULUM

**INTRODUCTION**

A congenital urethral diverticulum is a transitional cell epithelium lined pouch that is the result of either distention of a segment of the urethra or the attachment of a structure to the urethra by a narrow neck (i.e., a Müllerian remnant). In males, a congenital anterior urethral diverticulum may result from incomplete development of the urethra with a defect in only the ventral wall and subsequent distention of this segment by the hydraulic force of the voiding stream. Another possible etiology can be injury of the urethra that may cause an intraspongiosal hematoma; this could create a paraurethral space and subsequent diverticulum. It has been suggested that congenital diverticula may represent giant cystic dilation of Cowper’s ducts.

Congenital anterior urethral diverticulum (CAUD) is an uncommon condition in children usually presenting as a fluctuant ventral penile swelling. Primary excision and repair is the preferred mode of treatment for CAUD.

**CLINICAL PRESENTATION**

A 14-year-old male presented with history of swelling on the ventral aspect of penis and dribbling of urine after micturition.

**EXAMINATION**

There was a soft swelling on the ventral aspect of penis which increased at the time of voiding. Penis was normal.

**INVESTIGATIONS**

Retrograde urethrogram revealed a round diverticulum on the ventral side of anterior urethra (Fig. 11.12.1). MCU revealed normal bladder and urethra with a diverticulum in anterior urethra (Fig. 11.12.2).

**TREATMENT**

Surgical excision with repair of urethra was done. Patient started voiding well.
COMMENTS

Congenital urethral diverticulum is a rare anomaly and can be corrected surgically.

BIBLIOGRAPHY

11.13 POST-URETHROPLASTY URETHRAL DIVERTICULUM WITH CALCULI

INTRODUCTION

Urethral diverticulum of male urethra is uncommon. The etiology can be congenital, infection, obstruction, trauma, or post-surgery. Majority of them are acquired. Urethral diverticulum can be associated with stone due to stasis and hair growth following skin flap urethroplasty.

CLINICAL PRESENTATION

A 56-year-old Nigerian male presented with difficulty in passing urine for last 3 years.

PAST HISTORY

Forty two years back, he had history of terminal hematuria for a week which was diagnosed as bilharziasis and was treated in Nigeria. He then started having poor urinary stream for which multiple urethral dilatations were done in 1970s and early 1980s. In 1986, he was diagnosed as bladder neck stricture and bladder neck incision was done. In 1989, he had significant decrease in urine flow and urethroplasty was done in Nigeria. He was symptom free from 1989 to 2010.

Since 2010, he had noticed progressive reduction in urine flow. Digital rectal examination was normal. Retrograde urethrogram revealed a large urethral diverticulum in bulbar urethra with multiple calculi (Figs 11.13.1 to 11.14.3).

Fig. 11.13.1: Plain X-ray showing multiple calculi

Fig. 11.13.2: Right oblique urethrogram showing urethral diverticulum
Urethral Diseases

**Fig. 11.13.3:** Left oblique urethrogram showing urethral diverticulum

**TREATMENT**

Urethral diverticulectomy with removal of five calculi and urethral reconstruction was done in one stage. (Figs 11.13.4 to 11.13.9).

**Fig. 11.13.4:** Dissection of urethral diverticulum
Fig. 11.13.5: Urethral diverticulum with calculus

Fig. 11.13.6: Calculi with hairs in the diverticulum
Fig. 11.13.7: After excision of diverticulum

Fig. 11.13.8: Urethral reconstruction
COMMENTS

Post-urethroplasty dilatation and diverticulum formation is common due to stricture at the distal anastomosis. Chronic urinary retention precipitates stone formation. Treatment is removal of stones with excision and repair of the diverticulum.

BIBLIOGRAPHY

12.1 NON-HODGKIN’S LYMPHOMA OF TESTIS

**INTRODUCTION**

Testicular lymphoma was first reported and defined by Malassez and Curling in 1866. Incidence rate is around 0.26 per 100,000 people per year. Testicular lymphoma is the most common testicular malignancy in patients older than 60 years of age. Testicular lymphoma accounts for only 4% of all extranodal non-Hodgkin’s lymphomas (NHLs) and 5% of all testicular malignancies. Median survival is approximately 12–24 months.

Testicular lymphoma may be primary or secondary. Secondary involvement of the testis is far more common. A stage IV testicular lymphoma is virtually indistinguishable from an advanced stage nodal lymphoma with testicular involvement. Rate of testicular involvement in advanced stage diffuse large B cell is 10–18%.

**DIFFERENTIAL DIAGNOSIS**

In a young man, the differential diagnosis includes: infections, testicular torsion (painful), neoplastic lesions (germ cell tumor) and metastasis to testis (usually from prostate, lung, melanoma, kidney).

**DIAGNOSIS AND STAGING**

Usual investigations include: Testicular ultrasound which will show normal homogeneous echogenic testis being replaced focally or diffusely with hypoechoic vascular lymphomatous tissue; Complete blood count; Serum lactate dehydrogenase; Bone marrow aspiration or biopsy; Positron emission tomography-computed tomography (PET-CT) scan for nodal evaluation and staging.
**PROGNOSTIC MARKERS**

Prognosis is favorable in case of younger age, localized disease and presence of sclerosis at pathologic analysis. Smaller tumor size, lower histological tumor grade, lack of epididymal or spermatic cord involvement also has got favorable prognosis.

**HISTOPATHOLOGY**

Diffuse large B-cell most common and is seen in about 70% of the cases. Follicular lymphoma is in pediatric age is common. CD10 or Bcl-6 expression favors better overall survival group, CD10 or Bcl 6 negative are poor prognostic indicates.

**TREATMENT**

Radical orchidectomy provides initial diagnosis and treatment, once diagnosis is confirmed referral to medical oncologist is advisable for staging and further manegement. After only locoregional treatment (orchidectomy and radiation therapy), relapse is high, ranging approximately from 50 to 80%, mostly in central nervous system (CNS) or contralateral testis.

Connors et al. have shown that a doxorubicin-based chemotherapy regimen program of 6–9-week duration [CHOP (cyclophosphamide, hydroxydaunorubicin, oncovin, prednisolone) or doxorubicin, cyclophosphamide, vincristine and bleomycin] and prophylactic radiotherapy to the uninvolved contralateral testis, leads to a 4-year actuarial survival of 93%.

Cyclophosphamide, hydroxydaunorubicin, oncovin, prednisolone with prophylactic intrathecal therapy and adjuvant scrotal radiotherapy seems promising. Forty nine patients treated with 6–8 cycles of CHOP with rituximab and intrathecal chemotherapy ± locoregional radiotherapy showed 3-year overall survival and progression free survival (PFS) of 87% and 84%, respectively, with no contralateral testis relapses and 2.5% CNS relapse at 3 years.

Management of contralateral testis is important because it has recurrence rate ranging from 0–30%. In all elderly patients irradiation of contralateral testis is recommended.

**CASE 1**

**Clinical Presentation**

A 63-year-old male presented with complaints of a nodule and swelling at base of penis and right testis for 1 month duration. The swelling was painless, gradually increasing in size. He also had obstructed voiding symptoms for 1 month duration.

There was no history of hematuria, trauma or fever. He was also hypertensive and asthmatic for which he was on medications.
Examination

Examination revealed diffuse swelling of penis and scrotum (Figs 12.1.1A and B). Meatus was normal, a 3 × 3 cm hard swelling in right hemiscrotum inseparable from right testis and epididymis was noted. There was a nodule in left hemiscrotum separable from left testis. Digital rectal examination (DRE) revealed grade 1 prostate, no growth felt.

![Figs 12.1.1A and B: Case 1: Clinical pictures](image)

Investigations

Hemoglobin (Hb): 14.7 g/dL, Total leukocyte count (TLC): 8,100/cumm, Platelets: 299,000/cumm; Urea/Creatinine: 41/1.1 mg/dL, liver function test (LFT): within normal limits. Chest X-ray: normal, Serum prostate-specific antigen (PSA): 2.4 ng/mL, Uroflowmetry (Qmax): 7mL/sec. Ultrasound: 3.2 × 3.1 × 1.8 cm, hyper- or hypoechoic mass in right scrotal sac inseparable from testis. Both corpora engorged and swollen, increased flow in corpora. A 0.9 × 0.6 × 0.7 cm hypoechoic lesion in distal left cavernosa. Magnetic resonance imaging (MRI) lower abdomen: T1 images (Figs 12.1.2A and B) and T2 images (Figs 12.1.3A and B) of penis. MRI scrotum: T1 images (Fig. 12.1.4) and T2 images (Fig. 12.1.5). PET-CT scan (Figs 12.1.6A to C): metabolically active disease in penis and both hemiscrotum, bilateral inguinal lymph nodes, left external iliac nodes and aortocaval lymph nodes.
Figs 12.1.2A and B: Case 1—magnetic resonance imaging T1 images of penis

Figs 12.1.3A and B: Case 1—Magnetic resonance imaging T2 images of penis

Fig. 12.1.4: Case 1—Magnetic resonance imaging T1 images of scrotum
Fig. 12.1.5: Case 1—Magnetic resonance imaging T2 images of scrotum

Figs 12.1.6A to C: Case 1—Positron emission tomography-computed tomography
**Treatment**

Right sided high inguinal orchiectomy and trocar suprapubic cystostomy (SPC) was done. Findings were—hard mass in right hemiscrotum inseparable from testis, thickened corpora at base (Figs. 12.1.7A and B). 8 Fr infant feeding tube could be placed in the bladder.

![Image of operative and cut open specimen pictures](image)

**Figs 12.1.7A and B:** Case 1—Operative and cut open specimen pictures

**Postoperative Course**

Swelling started decreasing 2 days after chemotherapy, and patient started passing urine per urethra after which the SPC was clamped. SPC catheter was removed after 7 days.

**Histopathology**

Non-Hodgkin’s lymphoma B cell (Fig. 12.1.8A) large beta cell (Fig. 12.1.8B) CD20 positive. (Fig. 12.1.8C) and CD3 negative.
**Adjuvant Treatment**

Patient was started on chemotherapy on postoperative day 4 (dose according to body surface area, BSA) on R–CEOP, [rituximab (700 mg), cyclophosphamide (1,400 mg), epirubicin (140 mg), vincristine (2 mg), prednisolone (60 mg)] regimen for 5 days. Six cycles of above treatment was given. This was followed by prophylactic intrathecal chemotherapy once every 3 months for 1.5 years and radiation to the contralateral testis. Figure 12.1.9 shows clinical picture after 2 years of treatment.
Follow-Up

At 3 years, patient was asymptomatic without any recurrence of tumor. He complained of erectile dysfunction for which phosphodiesterase inhibitors were given.

CASE 2

Clinical Presentation

A 28-year-old male presented with right testicular swelling of 3-month duration. No history of pain.

Examination

Right testicular enlargement, hard and nontender.

Treatment

Right high inguinal orchidectomy was done. Figure 12.1.10 showing radical orchidectomy specimen.

Histopathology

Non-Hodgkin’s lymphoma (NHL).
Follow-Up
Adjuvant chemotherapy was given.

COMMENTS
Non-Hodgkin’s lymphoma of the testis is a rare disease. Histopathology confirms the diagnosis. Radical orchidectomy with chemotherapy can provide good prognosis. In spite of chemotherapy, high incidence of relapse in the central nervous system (CNS) was observed. Prophylactic treatment against such recurrence may be necessary to improve the treatment outcome of patients with testicular NHL.

BIBLIOGRAPHY
12.2 POST-ORCHIDOPEXY TESTICULAR TUMOR

INTRODUCTION
Patients with cryptorchidism have a fourfold to eightfold increased risk of developing germ cell tumors when compared to their normal counterparts. Orchidopexy, even at an early age, appears to reduce the incidence of germ cell tumor only slightly. For an undescended testis, the most common malignant histology is seminoma. For those who undergo early orchidopexy, the most common malignancy is non-seminoma.

CLINICAL PRESENTATION
A 35-year-old male presented with history of swelling in the right side of scrotum for 6-months duration.

PAST HISTORY
There was history of right orchidopexy at the age of 3 years and its details were not available.

EXAMINATION
Right testis was palpable in the scrotum, firm in consistency and nontender. Cord structures were normal.

INVESTIGATIONS
Alpha-Fetoprotein (AFP): 5 IU/mL, Human Chorionic Gonadotropin (HCG): 20 IU/mL. Ultrasound (Fig. 12.2.1): right testis enlarged with altered echogenicity. Contrast-enhanced computed tomography (CECT) abdomen: no evidence of any lymphadenopathy. X-ray chest was normal.

TREATMENT
Right high inguinal orchidectomy was done.

HISTOPATHOLOGY
Seminoma testis, no lymphovascular invasion and cord was not involved.

ADJUVANT TREATMENT
2,500 rad radiotherapy was given to abdomen and pelvis.
**FOLLOW-UP**

Patient was followed up for 15 years and there was no recurrence of tumor. Tumor markers were normal.

**COMMENTS**

A close observation for the size of testes after orchidopexy is important. If the size increases, immediate attention is required.

**BIBLIOGRAPHY**

12.3 NON SEMINOMATOUS GERM CELL TUMOR

**INTRODUCTION**

Testicular cancer is a relatively rare cancer with an incidence rate of 7.0 per 100,000 population. It affects males between the age of 20–40 years which is the productive age group for men and is a cause of great concern. Testicular germ cell tumors (GCTs) can be subdivided into seminoma and non seminomatous germ cell tumors (NSGCTs). Teratomas are subdivided into teratoma differentiated, malignant teratoma intermediate, malignant teratoma undifferentiated and malignant teratoma trophoblastic. Management of NSGCT is chemotherapy followed by surgery for residual disease. The prognosis for malignant teratoma is poor.

Testicular cancer spreads to retroperitoneal lymph nodes in the abdomen. For complete treatment, management involves retroperitoneal lymph node dissection (RPLND). Retroperitoneal lymph node spread is usually the first and only site of metastatic disease. About 15–40% of patients are clinically understaged, particularly in the retroperitoneum. Untreated, retroperitoneal lymph node metastasis are usually fatal. The most common site of late recurrence of both teratoma and viable GCT is the retroperitoneum.

Retroperitoneal lymph node dissection is done as primary treatment for stage I and II NSGCT. It is also done post chemotherapy in the following settings:

- **Standard RPLND:** It is done in patients after induction chemotherapy who have disseminated testicular cancer and present with residual radiographic disease in the retroperitoneum with normal tumor markers
- **Salvage RPLND:** Done in cases that are post second line salvage chemotherapy with normal tumor markers
- **Desperation RPLND:** Done in cases that are post second line salvage chemotherapy with elevated tumor markers
- **Redo RPLND:** It is done in patients who had previous RPLND with in-field recurrence.

Retroperitoneal lymph node dissection is the most challenging operation in urology as lymph nodes surrounds the great vessels—aorta and vena cava. In the past, this operation was done by open surgery and had significant operation related complications. Currently, this operation is being done laparoscopically with or without robotic assistance.

Three cases of NSGCTs are presented.
CASE 1: METASTATIC NONSEMINOMATOUS GERM CELL TUMOR

Clinical Presentation

A 31-year-old male presented with complaints of right scrotal swelling associated with occasional pain for last 6 months. He was seen by local practitioner and was treated for epididymo-orchitis. He did not improve and there was progressive increase in the size of the swelling.

Examination

No pallor, cyanosis, clubbing, icterus, lymphadenopathy or raised jugular venous pressure noted. Patient was afebrile. Pulse Rate was 86/min; Respiratory Rate (RR) was 20/min; Blood Pressure (BP) was 130/80 mm Hg. Chest was clear bilaterally clear.

Abdomen was soft, bowel sounds present, no organomegaly and no tenderness. Local examination revealed grossly enlarged, hard, smooth and nontender right testis (Fig. 12.3.1). Left testis was normal.

Investigations

Ultrasound revealed right testicular mass. CECT abdomen revealed lymph node mass in the retroperitoneum (Fig. 12.3.2). CECT chest revealed multiple metastases in the lungs (Figs 12.3.3A and B).
Treatment

Right high inguinal orchidectomy was done on July 30, 2013. There was a large testis with thickened tunica vaginalis. Figures 12.3.4A and B showing large testis with cord is specimen with variegated appearance on cut specimen.

Histopathology

Mixed GCT (seminoma—22%, choriocarcinoma—3%, teratoma—50%, yolk sac tumor—25%) limited to testes. Lymphovascular invasion was not seen. Pathological stage pT1 Nx, spermatic cord resected margin was free.
Adjuvant Treatment

He was started on chemotherapy (bleomycin, etoposide, cisplatin; BEP).

**COMMENTS**

It is unfortunate that patient present late due to ignorance and as a result disease advances and the outcome is poor.

**CASE 2: GIANT MALIGNANT TERATOMA IN AN UNDESCENDED TESTIS**

**Clinical Presentation**

A 30-year-old male presented with left-sided abdominal swelling of 6-months duration. Swelling was gradually increasing in size without pain. Left testis was not noted in the scrotum.

**Examination**

Per abdominal examination revealed a left hard tumor involving the left side of the abdomen. The mass was fixed and did not move with respiration. Investigations revealed raised AFP and HCG levels. CECT abdomen revealed large tumor with variegated appearance involving left half of abdomen and crossing the midline (Figs 12.3.5A to C).

Fine needle aspiration cytology (FNAC) revealed poorly differentiated malignant teratoma testis.

He was advised chemotherapy but was lost in follow-up.
Tumors in undescended testis are usually diagnosed at late stages and they are more aggressive with poor prognosis.

**CASE 3: NONSEMINOMATOUS GERM CELL TUMOR—POSTCHEMOTHERAPY RELAPSE**

**Clinical Presentation**

A 31-year-old male presented with a left testicular swelling in December 2007.

**Investigations**

Alpha fetoprotein: 2,386 ng/mL (normal upto 15 ng/mL), Lactate dehydrogenase (LDH): 279 U/L (110–216 U/L), HCG: 527 mIU/mL. Computed tomography (CT) abdomen (in December 2007): 2.6 × 2.5 cm necrotic left paraaortic adenopathy.

He underwent left radical orchidectomy on December 07, 2007.
**Histopathology**

Histopathology revealed mixed germ cell tumor with embryonal carcinoma, yolk sac, and teratoma components. Angioinvasion was present. Tumor was confined to testis with free surgical margins.

Staging: pT2 N2 M0

**Treatment**

Patient received 4 cycles of chemotherapy (BEP) from January to March 2008. Postchemotherapy tumor markers were normal. Postchemotherapy CT scan showed presence paraaortic lymph nodes an (no uptake on PET scan).

Repeat CECT in May 2011 revealed increase in size of the lymph node to 33 × 32 mm with tumor surrounding inferior branch of renal artery (Figs 12.3.6A to E). PET-CT scan (on May 11, 2011) revealed mildly fluorodeoxyglucose (FDG) avid hypodense left paraaortic lymph node at L2–L3 vertebral level, measuring 4.3 × 3.6 × 5.1 cm, AFP (on May 08, 2011): 1.39 IU/mL (<5.8 IU/mL), serum HCG (on May 07, 2011) was less than 2.0 mIU/mL.
Treatment for Residual Disease

Robotic retroperitoneal lymph node dissection was done. Figure 12.3.7 shows the specimen of lymph node mass with spermatic vessels. Histopathology metastatic carcinoma of immature teratoma, other paraaortic lymph nodes were free of tumor.

Fig. 12.3.7: Specimen of lymph node mass excised with spermatic vessels

Follow-Up

At one-year of follow-up, PET scan was normal with normal tumor markers.

COMMENTS

Usually postchemotherapy residual cancer has poor prognosis and disease may not be eradicated. It has been shown that in testicular cancer complete eradication of the disease can be achieved by RPLND. Postchemotherapy RPLND has good prognosis and survival rate is good.

BIBLIOGRAPHY

13.1 CORPORAL ABSCESS

INTRODUCTION
Spontaneous cavernositis is a distinctly uncommon entity. Corporeal infection and abscess formation have been described in association with priapism, cavernosography, intracavernous injection therapy, trauma and penile prostheses. It can also be caused by hematogenous spread or through the urethra.

CLINICAL PRESENTATION
A 48-year-old male presented with complaints of pain and swelling in the perineum and scrotum for 2 months along with poor urinary stream. He had fever since last 3 days. He also gave the history of trauma in perineum as well. The symptoms of poor stream were present before the trauma as well. Patient was diabetic and hypertensive for past 1½ years. He had percutaneous transluminal coronary angioplasty (PTCA) done in May 2010.

EXAMINATION
Examination revealed diffuse oedema present over scrotum and perineum. Digital rectal examination revealed prostate of normal size, firm, smooth and rectal mucosa free.

INVESTIGATIONS
Retrograde urethrogram (RGU) and micturating cystourethrogram (MCU) revealed normal penile urethra (Figs 13.1.1A to C). There was seen a short segment stricture of the bulbar urethra. Smooth passage of contrast was seen into the bladder. There was no secondary tract. The bladder filled normally and was smooth in contour. There was no vesicoureteral reflux bilaterally. A normal posterior urethra was identified during voiding. No periurethral leak of contrast was noted.
Ultrasonography (USG) scrotum revealed right testicle measured 4.1 x 2.0 x 3.1 cm (volume 13.4 cc) was normal in size, configuration and echogenicity and of homogeneous echotexture. Left testicle measured 4.4 x 2.3 x 3.3 cm (volume 18 cc). It is enlarged and globular in shape. Normal color flow Doppler signal was seen within the right testis. There is increase in vascularity on the left side. Right epididymis measured 9.6 x 7.0 mm and was normal in size. Left epididymis measured 14.1 x 11.8 mm and was enlarged in size and showed increased vascularity. The spermatic cord was enlarged and thickened on left side and also showed increased vascularity. There was increased peritesticular fluid bilaterally. The overlying scrotal skin was thickened and edematous (left more than right). The above findings were suggestive of left epididymo-orchitis with funiculitis. There was no evidence of testicular torsion or epididymitis. USG kidney, ureter and bladder (KUB) (on November 07, 2011) revealed kidneys to be normal and prostatic volume was 18.3 cc.

Magnetic resonance imaging (MRI) pelvis (contrast) (Figs 13.1.2A to E): the left corpus cavernosum of the mid shaft of the penis appeared hypointense on T1 and hyperintense on T2 imaging and fluid attenuated inversion recovery (FLAIR) sequences and showed peripheral enhancement. There was disruption of the tunica albuginea of the left corpus cavernosum in the mid
penile shaft with surrounding hypointense soft tissue, which showed multiple areas of necrosis with peripheral enhancement on postcontrast study.

The right corpus cavernosum and the tunica albuginea were intact. The corpus spongiosum along with the penile urethra were intact. The membranous urethra was compressed. The prostatic urethra was normal. There was subcutaneous soft tissue edema noted in the scrotum. Bilateral testis showed normal signal intensity with minimal fluid in the scrotal sac and diffuse enhancement of its covering, i.e. tunica albuginea and epididymis. The

Figs 13.1.2A to E: Magnetic resonance imaging (MRI) pelvis with contrast
spermatic vessels were prominent. The urinary bladder was well distended and does not show any abnormal wall thickening. The prostate and seminal vesicles were unremarkable. There were prominent nodes seen in bilateral inguinal regions and external and internal iliac grooves. The largest node in the left external iliac region measured $1.5 \times 2.2$ cm in size. The pelvic side walls were free and bony pelvis was unremarkable.

## TREATMENT

Cystourethroscopy revealed narrow urethra which did not permit 10 or 6 Fr feeding tube. The bladder was filled with saline using a syringe. Ultrasound-guided percutaneous suprapubic cystostomy was done after filling the bladder with Gentamycin and saline under general anesthesia on November 08, 2011. An incision was made in the perineum. Corporal abscess was drained. Appropriate antibiotics were given for 6 weeks and wound healed well. Patient started passing urine normally. Suprapubic cystostomy (SPC) catheter was removed.

## COMMENTS

Corporal abscess is rare but can occur in immunocompromised men with diabetes mellitus (DM). It can present with urethral obstructive symptoms. Proper imaging by MRI can diagnose the condition accurately. Surgical intervention is required in these case.

## BIBLIOGRAPHY

13.2 FRACTURE PENIS

**INTRODUCTION**

Penile fracture is a well-recognized clinical entity. It is relatively uncommon and is considered an urological emergency. The most common cause is trauma caused during vigorous sexual activity by hitting of erect penis to hard object (perineum). Conservative management has been practiced but surgical repair of the torn corpus cavernosum minimizes complications. Late penile deformity, suboptimum painful erections, difficulty in coitus, pulsatile diverticulum, and prolonged hospital stay are associated with conservative management.

**CLINICAL PRESENTATION**

Figures 13.2.1A and B are two cases who presented in the emergency with history of trauma during sex.

Physical examination shows swelling of penis. In the first case (Fig. 13.2.1A) there was classical deformity after injury to corpora cavernosa. Color Doppler ultrasound was done to confirm the injury to corpora cavernosa. Both cases were explored by subcoronal incision. Degloving of the penis and surgical repair of corpora cavernosa was done.

Figs 13.2.1A and B: Clinical picture following penile trauma
**COMMENTS**

Penile fracture is a clinical diagnosis. The ideal management has evolved and remains largely surgical. Preoperative imaging should not delay surgical repair.

**BIBLIOGRAPHY**

INTRODUCTION

Penile strangulation or entrapment is an unusual condition that requires urgent treatment due to its potential complications. Non-metallic, thin objects are easy to remove but can cause severe injury. Metallic objects are difficult to remove but the injuries are usually less severe. Penile ulceration and edema in children may well indicate the presence of a strangulatory object.

CASE 1

Clinical Presentation

A 26-year-old male factory worker was playing with a spring coil for sexual pleasure and after some time he was unable to remove it due to distal swelling. He came to emergency after 2 days with history of swelling of penis. On examination, there was a spring coil strangulating at the base of penis with edema, swelling and discoloration of the penile skin (Figs 13.3.1A to C).
Treatment

In emergency, he was taken up for surgery. Due to strength and thickness of spring coil, it was not possible to remove it over the penis. With the help of engineering department, special tools were used to open the spring coil and was removed (Fig. 13.3.2). There was discoloration of distal penile skin which was necrosed and sloughed out (Fig. 13.3.3). Partial thickness skin grafting was done (Fig. 13.3.4). His penis could be salvaged.
CASE 2

Clinical Presentation

A 24-year-old male used a bottle opener for sexual pleasure which was unable to remove. He presented in emergency within 6 hours. Examination revealed a bottle opener strangulating at the base of the penis with distal swelling (Fig. 13.3.5). In emergency, he was taken up for surgery under anesthesia. The bottle opener was removed easily (Fig. 13.3.6A and B). Penile swelling subsided.

**Figs 13.3.6A and B:** Case 2—Clinical picture during and after removal of bottle opener

COMMENTS

Penile strangulating injuries can present in emergency and should be managed as early as possible to minimize damage to the penis otherwise such injuries can be fatal.

**BIBLIOGRAPHY**

13.4 PEYRONE’S DISEASE

**INTRODUCTION**

Chronic inflammation of the tunica albuginea is a connective tissue disorder involving the growth of fibrous plaques and is known as Peyronie’s disease. The thick sheath of scar tissue surrounding the corpora cavernosa cause pain, abnormal curvature, erectile dysfunction, indentation, loss of girth and shortening of the penis. An ultrasound can provide conclusive evidence of Peyronie’s disease, ruling out congenital curvature or other disorders. Etiology can be trauma during intercourse or it is an autoimmune disease and can be associated with fibrosis in other parts of the body. As exact etiology is not known, there are variety of treatment like medications, intracavernosal injections, and finally surgery for patients having severe deformity and sexual dysfunction.

**CLINICAL PRESENTATION**

A 52-year-old man presented with complaints of ventral chordee at the time of erection, unable to perform intercourse and painful ejaculation since last 1 year. He was married for 28 years and had 3 children. Previously, he had normal erections and sexual activity. Patient was diabetic (type II) for past 1 year. He had coronary artery bypass graft (CABG) done in September 2010.

**PHYSICAL EXAMINATION**

Penis size was normal. A fibrous plaque was palpable on the ventral side of penis. There was left lateral deviation of penis. Digital rectal examination (DRE) revealed prostate grade I, firm, smooth and mucosa free.

![Clinical picture](image-url)
INVESTIGATIONS

Urine and blood parameters were normal. Artificial erection with injection papaverine revealed lateral deviation of penis with poor erection (Figs 13.4.1A and B). X-ray penis revealed no calcification (Fig. 13.4.2). Ultrasound of the penis revealed a fibrous plaque of 1 cm size on the dorsum of the penis (Fig. 13.4.3). Color Doppler ultrasound injection papaverine revealed good arterial flow with high diastolic pressure suggestive of venous leak (Fig. 13.4.4).

Fig. 13.4.2: X-ray penis

Fig. 13.4.3: Color Doppler ultrasound of the penis showing fibrous plaque
TREATMENT

Under general anesthesia, artificial erection was done which revealed left deviation of the penis. X-ray penis (Fig. 13.4.5). Malleable penile prosthesis (spectra) insertion with remodeling of the penis was done. Figures 13.4.6 to 13.4.11 shows various steps of the procedure. Patient tolerated the procedure well. His postoperative period was uneventful and Foley’s catheter was removed next day.
Fig. 13.4.6: Intraoperative picture showing dilatation of corpora cavernosa

Fig. 13.4.7: Intraoperative picture showing measurement of length of the corpora cavernosa

Fig. 13.4.8: Intraoperative picture after insertion of prosthesis
Fig. 13.4.9: Intraoperative picture showing insertion of prosthesis proximally

Fig. 13.4.10: Intraoperative picture showing insertion of prosthesis distally

Fig. 13.4.11: Intraoperative picture showing closure of wound
FOLLOW-UP

He had straight erect penis and was able to perform intercourse satisfactorily (Fig. 13.4.12).

Figs 13.4.12: Clinical picture after surgery

COMMENTS

Peyronie’s disease is a rare condition and patients present with painful erection and sexual dysfunction. Most of the patients can be managed conservatively, but some may require surgery.

BIBLIOGRAPHY

13.5 PENILE CARCINOMA IN SITU

**INTRODUCTION**

Cancer cells that are only in the skin of the penis and have not spread to any deeper tissues beyond the basement membrane are called carcinoma in situ (CIS) or penile intraepithelial neoplasia (PIN). They are also referred to as Bowen’s disease or erythroplasia of Queyrat and may lead to invasive squamous cell carcinoma. The condition can stay at this stage for several years. Multiple treatments for squamous cell carcinoma in situ (SCCIS) of the penis have been used with variable success and morbidity. The treatment are Imiquimod 5% cream, laser treatment and surgical excision. They have high incidence of recurrence indicating the need for careful follow-up and patient self-examination.

**CLINICAL PRESENTATION**

A 63-year-old male presented with ulcerative lesion over the glans penis for 1 month duration. There was no history of sexual contact.

**Examination**

Red colored ulcerative lesion around 1 cm in size over the meatus was seen on the left side (Fig. 13.5.1). No induration noted in the surrounding structures. A 5 mm ulcerative lesion was present over prepuceal skin and there was reddish discolouration of the glans on the right side close to meatus. The other side of glans and corpora cavernosa were normal. There was no inguinal lymphadenopathy.

![Fig. 13.5.1: Clinical picture showing the lesion](image_url)
**Treatment**

Penile (Glans) excision biopsy with prepuzial skin biopsy was done under general anesthesia on 06.08.2013. He tolerated the procedure well. The post-operative period was uneventful. The Foley’s catheter was removed next day. He voided well and the wound healed well.

**Histopathology**

It revealed stratified squamous epithelium with full thickness dysplasia. Focal koiocytic change was also seen. The epithelium was dysplastic showing moderately pleomorphic cells with nuclear membrane irregularities. The dermo-epidermal junction and the underlying dermis showed dense lymphoplasmacytic infiltrate. Margins and deep tissues were free. Prepucial biopsy revealed stratified squamous epithelium with papillomatosis and broad and thickened rete ridges and mild dysplasia. Basement membrane was intact.

**REFERENCES**

13.6 PREPUTIAL TUMOR

[INTRODUCTION]
Penile cancer is a rare cancer and can involve prepuce, glans and can extend and involve corpora cavernosa. Isolated involvement of preputial skin is rare. There are several treatment options for penile cancer, depending on staging. They include surgery like wide local excision, microsurgery, laser surgery, circumcision, penectomy, radiation therapy, chemotherapy and biological therapy.

[CLINICAL PRESENTATION]
A 71-year-old nondiabetic and normotensive male presented with complaints of swelling over prepuce for 2 months. There was no history of bleeding. He had past history of coronary artery disease (CAD) for 10 years, hypothyroidism. CABG was done 10 years ago and thyroid surgery done 15 years ago along with hernia repair.

[EXAMINATION]
There was an ulcerated growth 1.5 cm in size on prepuce (Figs 13.6.1A and B). The glans penis was normal. The corpora cavernosa were normal.

Figs 13.6.1A and B: Clinical picture
TREATMENT

Circular incision was made 1 cm away from ulcerated growth and circumcision was done. Figure 13.6.2 shows the picture of the specimen. Hemostasis was achieved. Postoperative recovery was uneventful.

Fig. 13.6.2: Surgical specimen

HISTOPATHOLOGY

Histopathology revealed well-differentiated squamous cell carcinoma. Resected margins were free of tumor.

FOLLOW-UP

Wound healed well. There was no recurrence of tumor in 1 year of follow up.

COMMENTS

Surgery is the mainstay of treatment of penile cancer and according to grade and stage further adjuvant treatment can be planned. Careful follow-up is important.

BIBLIOGRAPHY

13.7 PENILE MELANOMA

■ INTRODUCTION
Melanomas of the penis are rare tumors with poor prognosis. The identified risk factors for the development of penile melanoma are melanosis and a pre-existing nevus. Delayed diagnosis explains the usually bad prognosis. Classical surgical treatment used to be radical; but recently, conservative surgery has been proposed. For an early diagnosis, genital melanosis requires surgical excision when technically feasible, and any atypical lesion of the penis should be submitted to a biopsy.

■ CLINICAL PRESENTATION
A 65-year-old male presented with history of abnormal swelling over glans penis for 6-month duration.

■ EXAMINATION
Examination revealed cherry colored swelling over glans penis (Fig. 13.7.1) which was bleeding on touch. There were also reddish patches over the glans penis. Inguinal lymph nodes were not palpable.

Fig. 13.7.1: Clinical picture
**TREATMENT**

Partial amputation of the penis was done. Histopathology confirmed melanoma of the penis. Patient was lost on follow-up for further management.

**COMMENTS**

Melanoma of the penis is a rare disease and needs aggressive treatment.

**BIBLIOGRAPHY**

13.8 LATERAL CURVATURE OF THE PENIS

**INTRODUCTION**

Lateral curvature of the penis is caused by overgrowth or hypoplasia of one corporal body. It is usually congenital and remains unnoticed in a flaccid penis. During erection this disparity is noticed and there can be a problem with penetration during sexual intercourse. Surgical correction of this deformity can be done by Nesbit plication. During correction, one should be careful to avoid injury to neurovascular bundles.

**CLINICAL CASES**

**Case 1**

A 20-year-old male presented with complaints of left lateral curvature of penis during erection (Fig. 13.8.1) and unable to penetrate during intercourse. On examination, left lateral curvature was more than 15 degree. He had no other problem. He was counselled for the surgical correction. Nesbit plication was done at two sites of maximum deformity (Fig. 13.8.2). Figure 13.8.3 shows clinical picture after artificial erection during surgery. Postoperative recovery was uneventful 3 months after surgery, examination showed straight penis following erection (Fig. 13.8.4).

*Follow up:* Patient was able to perform intercourse with satisfaction to both partners.

*Fig. 13.8.1:* Case 1—Clinical picture before surgery
**Fig. 13.8.2**: Case 1—Intraoperative picture showing Nesbit’s procedure

**Fig. 13.8.3**: Case 1—Intraoperative picture showing straight penis following artificial erection

**Fig. 13.8.4**: Case 1—Postoperative clinical picture after erection
Case 2

A 25-year-old unmarried male, presented with abnormal curvature of penis. On examination, there was left curvature of penis on erection (Fig. 13.8.5). He was counseled for the surgical correction. As he was unmarried, he decided for surgery before marriage.

![Case 2—Clinical picture](image)

Fig. 13.8.5: Case 2—Clinical picture

**COMMENTS**

Lateral curvature of penis can be corrected surgically if there is problem during intercourse.

**BIBLIOGRAPHY**

14.1 BLADDER STONE OVER TENSION-FREE VAGINAL TAPE

**INTRODUCTION**

Tension-free vaginal tape (TVT), a less-invasive variation of the suburethral sling, has been rapidly gaining popularity worldwide in the treatment of female stress urinary incontinence. Tape erosion is a known complication following the procedure and stone can form over the eroded tape. The presenting symptoms can be dysuria, recurrent urinary tract infection (UTI), etc.

**CLINICAL PRESENTATION**

A 43-year-old female presented with recurrent UTI following TVT procedure for stress urinary incontinence 5 years back. She had past history of bilateral round ligament plication, right ovarian cystectomy, left tubal cornual reimplantation, cystocele repair, Kelly’s suture for right chocolate cyst of ovary and left tube blockage at isthmus in 1995. She also had wedge resection of bladder neck in 1997 for retention of urine. She had stress urinary incontinence for which TVT repair was done in September 2005 (cystoscopy intraoperative shows no tape in bladder). No history of another comorbid illness.

**EXAMINATION**

No positive finding. She was fully continent.

**INVESTIGATIONS**

Ultrasound examination revealed normal upper tract, an echogenic focus 2–4 cm adjacent to left anterior wall of urinary bladder. The shadow was not changing position. Postvoid residual (PVR) urine: 57 mL. X-ray pelvis showed radiopaque shadow in region of urinary bladder (Fig. 14.1.1).
Cystoscopy revealed a stone fixed over left anterior wall of urinary bladder over the eroded tape. Holmium laser cystolithotripsy was done and eroded tape was removed endoscopically. Clinical picture showing fragmented stone (Fig. 14.1.2). Postoperative X-ray showed no radiopaque shadow in the bladder (Fig. 14.1.3). She stopped having recurrent urinary infections and was continent.
**COMMENTS**

Following TVT/transobturator tape (TOT), tape erosion is a rare complication and possibilities to be considered if patient has persistent dysuria or urinary tract infection. Endoscopic management is possible.

**BIBLIOGRAPHY**

14.2 CHORIOCARCINOMA URETHRA

INTRODUCTION

Primary extrauterine choriocarcinoma is a rare entity with high malignant potential and is mostly found in genital tract in patients with coexisting or antecedent pregnancy. Most of the extrauterine cases reported are gestational choriocarcinomas located in uterine cervix. Saito et al. described diagnostic criteria for this entity as: (1) absence of disease in uterine cavity; (2) pathologic confirmation of the disease and (3) exclusion of molar pregnancy and coexistence of normal intrauterine pregnancy. Choriocarcinomas which originate from genital regions outside uterus are reported in ovary, tube and vulva. Extragenital sites include gastrointestinal tract, brain, urinary bladder and heart. Gestational trophoblastic diseases have a varying potential for local invasion and metastases, and they generally respond to chemotherapy.

CLINICAL PRESENTATION

A 40-year-old female presented with the history of bleeding per vagina for 3-month duration. On examination, she has a large tumor dark-colored protruding from the urethra (Fig. 14.2.1). Other examination was normal. A wedge biopsy was done which confirmed the diagnosis of choriocarcinoma. She was advised for pelvic exenteration but she did not agree and lost to follow-up.

Fig. 14.2.1: Clinical picture showing tumor
**COMMENTS**

Choriocarcinoma is a very rare tumor with poor prognosis.

**BIBLIOGRAPHY**

14.3 FEMALE EPISPADIAS IN ADULTS

INTRODUCTION

Isolated female epispadias without bladder extrophy is an extremely rare congenital anomaly. It is characterized by a bifid clitoris, flattening of the mons and separation of the labia. Most patients reported in literature are young children, in whom the condition comes to light when they do not attain continence by 2–3 years of age. In developing countries such as India, where illiteracy and poverty are common and women have a low status in society, patients tend to hide their problem and suffer in silence, hence their late presentation.

CASE 1

Clinical Presentation

A 17-year-old female presented with a history of total urinary incontinence since birth.

Examination

External genitalia revealed flat mons pubis, bifid clitoris, patulous urethral meatus with deficient roof and intermittent effortless trickle of urine (Figs 14.3.1 and 14.3.2). The labia minora were swollen and sodden. Vaginal

Fig. 14.3.1: Case 1—Clinical picture
examination was normal. Bonney’s test was negative for the hypermobility of urethra.

**Investigations**

Urine, hemogram and renal function tests were normal. Plain X-ray kidney, ureter and bladder (KUB) region revealed spina bifida from L5 vertebra and below. Intravenous urogram revealed normal upper tracts. Voiding cystourethrogram (VCUG) revealed normal capacity bladder with short urethra and no vesicoureteral reflux. Cystometrogram (CMG) revealed normal bladder function. Leak point pressure was 20 cm. Cystopanendoscopy revealed a wide patulous external urethral meatus, short urethra. Bladder and ureteric orifices were normal.

**Treatment**

Urethral lengthening and Young Dee’s bladder neck reconstruction was done.

The postoperative course was uncomplicated. The urethral catheter was removed after 2 weeks and a VCUG performed. There was no extravasation of contrast. Subsequently, the suprapubic cystostomy catheter was removed.

**Follow-up**

The patient was continent (dry interval >4 hours) and the residual urine was insignificant.
CASE 2

Clinical Presentation

A 24-year-old woman presented with a history of total urinary incontinence since birth. She had been married for 7 years but was separated from her husband because of her urinary incontinence. She had undergone a vaginal operation (Kelly’s stitch) 2 years back, which did not relieve her problem.

Examination

The physical examination was normal, except for the external genital abnormalities which were similar to those described in the first case. Bonney’s test for urethral hypermobility was negative.

Investigations

X-ray KUB did not reveal any abnormality; specifically there was no spina bifida. Intravenous pyelogram (IVP) showed normal upper tracts but the bladder was not completely distended. VCUG showed a short urethra with a wide funnel-shaped bladder neck. There was grade I vesicoureteric reflux on the left side. CMG was normal. Cystoscopy showed a patulous urethra 1 cm long, a wide bladder neck, normal bladder mucosa, a wide “golf hole”-shaped right ureteric orifice and a “horseshoe”-shaped left ureteric orifice.

Treatment

The patient underwent urethral and bladder reconstruction and bilateral ureteric reimplantation by Paquin’s technique.

COMMENTS

Isolated female epispadias without bladder exstrophy is extremely rare; however, total reconstruction can give rise to complete continence and cosmetic appearance.

BIBLIOGRAPHY

**14.4 FAILED TENSION-FREE VAGINAL TAPE**

**INTRODUCTION**

Tension-free vaginal tape (TVT), a less-invasive variation of the suburethral sling, has been rapidly gaining popularity worldwide in the treatment of female stress urinary incontinence.

Majority of studies reported high success rates of 80–92% at 1-year postoperatively, and low complication rates such as urinary retention (1.5%), voiding difficulty (5.4%), or vaginal erosion (6.2%).

**CLINICAL PRESENTATION**

A 40-year-old female presented with stress urinary incontinence for which she underwent TVT 1 year back. She started having difficulty in passing urine and developed retention of urine.

**EXAMINATION**

On examination, there was no erosion of the mesh. Cystoscopy revealed tightening of urethra and bladder was normal.

**TREATMENT**

Per vaginal incision was made over the mesh and partial excision of the mesh was done (Figs 14.4.1 and 14.4.2). Postoperatively, catheter was removed after 5 days and she startedvoiding well.

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Figs 14.4.1A and B: Intraoperative pictures
**COMMENTS**

Tension-free vaginal tape can be associated with complications like erosion or obstruction. Surgical correction can be done.

**BIBLIOGRAPHY**


**Fig. 14.4.2:** Excised mesh
INTRODUCTION

Bladder exstrophy is a rare congenital malformation of the bladder and can be associated with complete urethral epispadias. Usually, these defects are noticed immediately after birth and managed accordingly. But in India, due to unawareness, poverty and lack of medical care, the patients present late in life. Due to prolonged exposure of bladder mucosa, malignant changes can occur and bladder loses its pliability, and patient may require cystectomy.

CLINICAL PRESENTATION

A 26-year-old female, resident from remote village, presented with abnormal swelling in lower abdomen and continuous leakage of urine since birth. She was covering the lesion with towels and changing them frequently.

EXAMINATION

She had exposed bladder in lower abdomen with normal ureteric orifices. The covering mucosa was not soft and supple (Fig. 14.5.1).
TREATMENT

In view of the age of the patient and bladder mucosa changes, her cystectomy and continent urinary diversion by rectosigmoid pouch and abdominal wall reconstruction was done (Fig. 14.5.2). On follow-up, she was asymptomatic and got married with a man with exstrophy bladder and had normal sexual life.

COMMENTS

Adult female exstrophy with complete epispadias is a very rare congenital deformity. In India, patients present late and can be managed by cystectomy with continent diversion.

BIBLIOGRAPHY

14.6 MALIGNANT MELANOMA FEMALE URETHRA

**INTRODUCTION**

Malignant melanoma is one of the rarest tumors of the female urethra. It is associated with a rapid and high local recurrence rate. Total urethrectomy with bilateral inguinal lymph node dissection should be done as the initial form of the treatment in patients with this disease who have no evidence of distant metastasis. The postoperative adjuvant therapy, consisting of alpha-interferon and chemotherapy, should be administered immediately; therefore, vesicostomy should be done along with surgery. The prognosis of urethral melanoma is poor.

**CLINICAL PRESENTATION**

A 35-year-old female presented with bleeding per vagina.

**EXAMINATION**

On examination (Fig. 14.6.1), there was a protruding tumor from the urethra, highly vascular and was bleeding on touch.

![Fig. 14.6.1: Clinical picture](image-url)
**TREATMENT**

On per vaginal examination, the uterus and cervix were normal. Biopsy confirmed the diagnosis of melanoma urethra. Patient was counseled for the treatment but lost to follow up.

**COMMENTS**

Malignant melanoma of female urethra is a very rare tumor of high malignant potential with poor prognosis.

**BIBLIOGRAPHY**

14.7 GARTNER’S CYST

INTRODUCTION

A Gartner’s duct cyst (sometimes incorrectly referred to as vaginal inclusion cyst) is a benign vaginal cystic lesion that arises from the vestigial remnant of a mesonephric duct or Gartner’s duct. They are typically small asymptomatic cysts that occur along the lateral walls of the vagina, following the course of the duct. They can, however, enlarge to substantial proportions and be mistaken for urethral diverticulum or other structures. These lesions are an uncommon and extremely rare.

CLINICAL PRESENTATION

A 38-year-old female presented with a swelling in the vagina for 6-month duration. The swelling was gradually increasing in size. There was no bleeding per vagina.

EXAMINATION

On examination (Fig. 14.7.1), there was a cystic lesion protruding from the vagina.

Fig. 14.7.1: Clinical examination
**TREATMENT**

On cystoscopic examination (Fig. 14.7.2), a cystic lesion was seen arising from the lateral side of the urethral meatus with normal bladder and urethra. Local excision of the cyst was done. Histopathology confirmed Gartner’s cyst.

**COMMENTS**

Gartner’s cyst is a rare benign condition and should be considered in the diagnosis of cystic lesions in vagina.

**BIBLIOGRAPHY**

14.8 PARAURETHRAL CYST

■ INTRODUCTION

A paraurethral cyst results from retained secretions in skene’s glands, secondary to ductal obstruction. The cyst displaces the meatus in an eccentric manner. Most periurethral cysts can be diagnosed by physical examination. The diagnosis may be confirmed with transvaginal sonography. Cystourethroscopy should be performed to rule out other pathology, but may be done in the same setting as surgical excision. Complete surgical excision is effective and is associated with minimal risk of recurrence.

■ CLINICAL PRESENTATION

A 39-year-old female presented with abnormal swelling in the vagina. She had no urinary complains. No history of bleeding per vagina.

■ EXAMINATION

On local examination (Fig. 14.8.1), there was a nontender cystic swelling just below and right to the urethral meatus.

Fig. 14.8.1: Clinical picture
**TREATMENT**

Cystoscopy revealed eccentric swelling from the urethral meatus on the right side. Remaining urethra and bladder was normal. Urethral catheter was passed (Fig. 14.8.2) which showed eccentric swelling from the urethral meatus. Local excision of the cyst was done. Patient recovered and no complaints after that. Histological examination showed a fibrocollagenous cyst wall lined by transitional epithelium.

![Fig. 14.8.2: Clinical picture with catheter](image)

**COMMENTS**

Paraurethral cysts are rare and can be diagnosed by eccentric position and can be managed surgically.

**BIBLIOGRAPHY**

14.9 VAULT RECURRENCE FOLLOWING CARCINOMA CERVIX

**INTRODUCTION**

Ten to twenty percent recurrence is reported following surgery or radiotherapy in women with stage IB/IIA cervical tumor with no evidence of lymph node involvement. Majority of recurrence occur within 2 years of diagnosis, and prognosis is poor. Treatment decision is based on performance status of the patient; site of recurrence and/or metastasis; extent of metastatic disease and prior treatment. The treatment options are radical radiation, pelvis exenteration, concurrent chemotherapy and radiotherapy. If disease is localized to pelvis, then pelvis exenteration (en bloc removal of bladder, genital organ and rectum) with urinary diversion (wet colostomy) is recommended.

**CLINICAL PRESENTATION**

A 37-year-old female presented with history of nausea and vomiting on March 10, 2010. Investigations revealed serum creatinine—7 mg/dL. Ultrasound revealed left gross hydronephrosis and mild right hydroureteronephrosis (HDUN). In emergency, ultrasound-guided right percutaneous nephrostomy (PCN) was done. Serum creatinine came down to 2.2 mg/dL next morning. She was married in 2001 but had no children.

Past history of right hemicolectomy with left oophorectomy in 2002. Histopathology revealed adenocarcinoma colon and left ovary. Postoperatively, she was given 6 cycles of chemotherapy. She had recurrence of tumor in right ovary for which right oophorectomy was done in 2004.

She again presented with bleeding per vagina in 2007. She was diagnosed as carcinoma cervix with left HDUN with poor functioning left kidney. Neoadjuvant chemotherapy followed by Wertheim’s hysterectomy with lymphadenectomy was done. Histopathology revealed adenocarcinoma cervix involving uterus, parametrium, both tubes, posterior vagina and involvement of three lymph nodes. Postoperatively, she was given radiotherapy—28 sessions.

**INVESTIGATIONS**

Magnetic resonance imaging (MRI) (Figs 14.9.1A to D) revealed recurrent tumor 2.8 cms infiltrative ill-defined mass encasing both ureters and infiltrating into the anterior wall of the rectum and adjacent small intestinal loops. Clinical stage T4aNOMx. X-ray chest—normal. Positron emission tomography-computed tomography (PET-CT): active recurrent disease involving soft tissue density mass between bladder and rectum and involving both ureters.
TREATMENT

Anterior exenteration with anterior resection of rectum with ileal conduit diversion with diverting ileostomy was done (Figs 14.9.2 and 14.9.3). The bowel and omentum were adherent, adhesiolysis done. Histopathology revealed adenocarcinoma with mucin component less than 50% involving the wall of the rectum and urinary bladder in a known case of carcinoma colon and carcinoma cervix. All surgical margins were free.

Postoperatively, she recovered well. Serum creatinine was 1.6 mg/dL. The ileostomy was functioning well.
Fig. 14.9.2: Operative specimen

Fig. 14.9.3: Operative specimen
Cervical cancer, despite being potentially preventable, remains an important cause of morbidity and mortality. Vault recurrence with involvement of bladder, ureter and rectum can occur in spite of surgery, radiation and chemotherapy. Pelvic exenteration can prolong survival following recurrence if disease is localized in the pelvis.

14.10 URETEROVAGINAL FISTULA

INTRODUCTION

Ureteric injury is an uncommon yet serious complication of gynecologic surgery for inflammatory pelvic masses, ovarian and uterine neoplasm, and endometriosis. The reported incidence of ureteric injuries following abdominal hysterectomy is 0.5–2.0%.

These injuries can manifest by urinary leak/intraperitoneal leak following surgery, by flank pain, fever, etc. Recognition of injuries can be confirmed by imaging, intravenous urography (IVU) or contrast-enhanced computed tomography (CECT) scan and confirmed by cystoscopy and retrograde ureterogram. Management can be by primary repair or percutaneous nephrostomy and delayed ureteric reconstruction.

CLINICAL CASES

Case 1

A 38-year-old female underwent abdominal hysterectomy for fibroid uterus. Five days after surgery, there was leak of urine per vagina. Abdominal examination was normal. Intravenous urogram (Fig. 14.10.1) revealed left mild hydroureteronephrosis. Cystoscopy revealed normal bladder, and left retrograde pyelogram (RGP) (Fig. 14.10.2) confirmed narrowing of lower ureter. Left ureteric reimplantation with psoas hitch was done. Her urinary leak stopped.

Fig. 14.10.1: Case 1—Intravenous urogram

Fig. 14.10.2: Case 1—Left RGP
Case 2

A 42-year-old female underwent abdominal hysterectomy. Seven days after surgery, she had urinary leak per vagina. Intravenous urogram (Fig. 14.10.3) revealed right hydroureteronephrosis. Right RGP (Figs 14.10.4 and 14.10.5) confirmed ureteric injury at the level of bifurcation of iliac vessels with urinary leak. Right ureteric reimplantation with Boari’s flap was done. Her urinary leak stopped.
Case 3

A 36-year-old female had lower segment cesarean section (LSCS) for obstructed labor in August 2009. She also had past history of LSCS in 2003. Ten days after surgery, patient developed urinary incontinence. She had normal voiding in between.

Her general examination was normal. Per abdomen examination revealed Pfannenstiel scar + local examination—no abnormality detected. Ultrasound examination (Fig. 14.10.6) revealed left hydronephrosis. Intravenous urogram (Fig. 14.10.7) revealed left hydronephrosis till lower end. Cystoscopy revealed

Fig. 14.10.6: Case 3—Ultrasound

Fig. 14.10.7: Case 3—Intravenous urogram
normal bladder, and left RGP confirmed narrowing of lower ureter. Left ureteric reimplantation was done. Her urinary leak stopped.

**COMMENTS**

Ureteric injury can occur following gynecologic procedure. Ureteric injury should be suspected in case of urinary leak or any abdominal signs. Imaging suggests upper tract changes which can be confirmed by RGP. Endoscopic management is possible in some cases otherwise ureteric reimplantation with ureteral substitution is required according to the level of injury.

**BIBLIOGRAPHY**

# 14.11 FEMALE URETHRAL CARCINOMA

## INTRODUCTION

Female urethral carcinoma is a rare tumor with predominantly poor survival. Both the disease and its treatment can affect sexual and urinary function. The natural history of urethral carcinoma varies, therefore, the appropriate application of surgery, radiation, and chemotherapy remains unknown. Management of this disease remains driven by individual clinician’s experience and data derived from small case series.

Circumferential local excision of the distal urethra and adjacent portions of the anterior vaginal wall can be accomplished for small, exophytic, well-differentiated lesions of the external meatus or distal third of the urethra. Spatulation of the urethra and approximation to the subjacent vagina and labia preserve urinary continence while helping to prevent urethral meatal stenosis.

For proximal invasive lesions or lesions involving the entire urethra, an extensive resection is required, including cystourethrectomy (anterior exenteration) with a wide margin of vagina, or in some cases, the entire vagina. The perineal incision is similar but wider than the incision for standard transvaginal anterior pelvic exenteration. Adjuvant radiation and chemotherapy are recommended in these cases.

## CASE 1

**Clinical Presentation**

A 65-year-old female presented with the complaints of a small swelling in the urethra. She had no history of vaginal bleeding. She was not sexually active.

**Examination**

Examination (Fig. 14.11.1) revealed a small 0.5 cms size red-colored lesion close to ureteral meatus. Cold cup biopsy revealed well-differentiated squamous cell carcinoma.

**Treatment**

Distal uretherectomy was done. Postoperatively, she was given radiotherapy. If 2-years follow-up, she was asymptomatic with no recurrence of the tumor.
CASE 2

Clinical Presentation

A 59-year-old female presented with a foul-smelling swelling from the vagina for 6-months duration. The swelling bled on touch.

Examination

She had a fungating mass lesion protruding from the vagina which was infiltrating the vagina (Fig. 14.11.2). She had palpable inguinal lymph nodes. Biopsy confirmed poorly differentiated squamous cell carcinoma. She was counseled for the treatment but refused any treatment.

COMMENTS

Female urethral tumors are rare clinical problem. Distal tumors are easy to manage with good prognosis. Proximal/panurethral tumors are more aggressive and needs multimodality treatment, and the prognosis is poor.

BIBLIOGRAPHY

INTRODUCTION

Urethral caruncle is a common problem in females, more frequently after menopause. These lesions presents as exophytic, often ulcerated or polypoidal mass, at or near the urethral meatus. It is usually eccentrically placed and it is to be differentiated from the prolapsed urethral mucosa. Urethral caruncle is a benign polypoid mass but conclusive association with malignancy, urologic disorder, or systemic disease has not been established. Often the lesion carries a challenging clinical differential diagnosis that includes malignancy. The treatment of urethral caruncle is surgical excision.

CLINICAL CASES

Case 1

A 60-year-old female presented with an abnormal swelling from the urethral meatus. On examination (Fig. 14.12.1), there was a small cherry-colored lesion arising from the right side of the urethral orifice. Local excision was done. Histopathology confirmed urethral caruncle.

Fig. 14.12.1: Case 1—Clinical picture
Case 2

A 37-year-old female presented with an abnormal swelling from the urethra for 6-months duration. On examination (Fig. 14.12.2), a red-colored swelling was protruding from right side of urethra anterolaterally. Cystoscopy revealed normal urethra and bladder. Local excision of the swelling was done. Histopathology confirmed urethral caruncle.

Case 3

A 45-year-old female presented with an abnormal swelling from the urethral opening which was bleeding on touch. Examination (Fig. 14.12.3) revealed red-colored large swelling from the urethral orifice from left side. Cystoscopy revealed normal urethra and bladder. Local excision of the swelling was done. Histopathology confirmed urethral caruncle.

COMMENTS

Urethral caruncle can be small or big tumor usually eccentric at the urethral opening. Excision and biopsy are necessary for confirmation of diagnosis.

BIBLIOGRAPHY

### INTRODUCTION

Uterovesical fistula or “Youssef’s syndrome” is characterized by cyclical hematuria, the absence of vaginal bleeding and complete urinary continence. It is a rare complication of cesarean section when bladder injury occurs and a fistula develops. Most of the cases require operative treatment; however, some cases can be managed conservatively.

### CLINICAL PRESENTATION

A 32-year-old female presented with history of blood mixed with urine at the time of menstruation following second cesarean section. She had first child birth also by cesarean section. She had no urinary incontinence.

### EXAMINATION

She had induration at the vault of the vagina.

### INVESTIGATIONS

Ultrasound revealed normal kidneys, ureter and bladder. Hysterosalpingogram (Fig. 14.13.1) revealed leakage of contrast in the vagina giving typical “cup and saucer” appearance. Cystogram revealed no leakage of contrast in the vagina. Cystopanendoscopy revealed induration at the dome of the bladder.

![Fig. 14.13.1: Hysterosalpingogram](image-url)
**TREATMENT**

On exploration, communication was detected between uterus and bladder. Bladder and uterus were mobilized and repair of uterovesical fistula was done in double layers. Patient recovered well and Foley’s catheter was removed after 14 days.

**FOLLOW-UP**

At 12 years of follow-up, she was asymptomatic.

**COMMENTS**

Uterovesical fistulas are to be expected to increase due to an increasing rate of repeated cesarean sections. They can be suggested by their typical symptoms, easily diagnosed by imaging examinations and successfully treated by surgical closure.

**BIBLIOGRAPHY**

14.14 VESICOVAGINAL FISTULA WITH BLADDER CALCULI

**INTRODUCTION**

The presence of vesical calculus with vesicovaginal fistula is rare. The etiology for stone formation can be due to urinary contamination, a high or supratrigonal fistula location, residual urine in the bladder and a long history of disease. These cases can be managed by staged repair by removal of calculi endoscopically, followed by fistula repair or in one stage by transabdominal repair or by vaginal approach.

**CLINICAL PRESENTATION**

A 42-year-old female presented with continuous leakage of urine per vagina following abdominal hysterectomy for fibroid uterus for a duration of 6 years. Urine leak started after 5 days of surgery. She did not consult because of poverty. Once she also started having dysuria, she came for consultation.

**EXAMINATION**

She had lower abdominal incision scar and had continuous leakage of urine per vagina. Per vaginal (PV) examination revealed a 2-cm fistulous opening in the anterior wall of vagina.

**INVESTIGATIONS**

Urine R/M revealed pus cells and RBCs. Urine C/S revealed proteous infection. Plain X-ray KUB region revealed a large bladder calculus. Intravenous urogram

**Fig. 14.14.1: Intravenous urogram**
urogram revealed normal upper tracts with small capacity bladder with vesical calculus and leakage of contrast in the vagina (Fig.14.14.1). Cystopanendoscopy revealed a 2 cm in size supratrigonal fistula with normal ureteric orifices.

**TREATMENT**

Treatment was done by surgery—Open transabdominal O’Conors repair of vesicovaginal fistula was done and bladder stone was removed. She had no incontinence after surgery.

**COMMENTS**

VVF along with bladder can be managed simultaneously in the same sitting or separately if associated with inflammation/infection.

**BIBLIOGRAPHY**

14.15 POST-HYSTERECTOMY BILATERAL URETERIC AND BLADDER INJURY

**INTRODUCTION**

The incidence of ureteral and bladder lesions after hysterectomy is 0.3–4.3%. The incidence of ureteral and bladder lesions after laparoscopic hysterectomy is the most encountered urinary complication in gynecological surgery. Late diagnosis of ureteral lesions, with or without bladder injury, requires further intervention.

**CLINICAL PRESENTATION**

A 52-year-old female was diagnosed as endometrial carcinoma and she underwent abdominal hysterectomy on March 23, 2013. After surgery, the patient developed pain in abdomen and leakage of urine per vagina postoperatively. She also had hypertension and type II diabetes mellitus.

**INVESTIGATIONS**

She was investigated and found to have bilateral ureteric and bladder injury. Contrast enhanced computed tomography (CECT) abdomen revealed mild bilateral hydronephrosis (HDN). There was complete disruption of right ureter with extravasation of contrast, along with disruption of posterior wall of bladder with rent of size 1.6 cm. There was also partial disruption of lower end of left ureter. There was moderate amount of loculated fluid collection posterior to the urinary bladder (Fig. 14.15.1).

Cystoscopy revealed large defect in the bladder. Right retrograde pyelography (RGP) and DJ stenting was done. Left ureteric catheter could not be passed and left percutaneous nephrostomy (PCN) was done on April 18, 2013.

CECT abdomen after 3 months revealed normal functioning of both kidneys with right DJ stent in situ with left ureteric obstruction at the level of iliac vessel with extravasation of contrast in the vagina (Figs 14.15.2A and B).

Per vaginal examination revealed a large defect palpable in anterior vaginal wall. There was no palpable tumor. Cystoscopy revealed normal urethra. Right DJ stent in situ with normal left ureteric orifice. There was an 8 × 6 cm posterior bladder wall defect with visible posterior wall of vagina. Right RGP revealed normal ureter and DJ stent changed. Left ureter was completely blocked 1 cm above the ureteric orifice. There was no visible tumor in the bladder defect.

Multiple bladder biopsies were taken from fistulous site. Histopathology of all the margins of vesicovaginal fistula (VVF) showed hyperplastic stratified epithelium. Underlying stroma was infiltrated by lymphocytes. There was no evidence of dysplasia.
TREATMENT

O’Conors repair of VVF with augmentation cystoplasty and left lower ureteric substitution by ileal replacement of ureter was done. The patient recovered well.
**FOLLOW-UP**

Nephrostomy catheter was removed. Postoperative cystogram (Fig. 14.15.3) revealed normal bladder with no leak of contrast and well-filled ileal segment. She started voiding well.
Large VVF with ureteral defect can be managed by repair of fistula along with augmentation cystoplasty with ileal replacement of ureter.

**BIBLIOGRAPHY**


**Fig. 14.15.3:** Postoperative cystogram
14.16 URETEROVAGINAL FISTULA WITH URETERAL DUPLICATION

**INTRODUCTION**

Complete duplication of ureter with ureterovaginal fistula is a very rare complication. With history of normal voiding with urinary leak, ureterovaginal fistula should be suspected. Proper evaluation, cystoscopy, 3 swab test and retrograde pyelogram (RGP) can clinch the diagnosis.

**CLINICAL PRESENTATION**

A 35-year-old married female, having three children, presented with complaints of continuous urinary leakage for a duration of 1 year following abdominal hysterectomy, appendectomy, pelvic floor repair for dysfunctional uterine bleeding. She had normal postoperative recovery. Urinary leak started 6 weeks postoperatively. She had no response to bladder catheterization. She was then further evaluated. Cystoscopy revealed two ureteric orifices on the left side. No vesicovaginal fistula (VVF). Both orifices were catheterized. Retrograde urethrogram (RGU) was not done as there was no C-arm facility available. She was treated for stress urinary incontinence.

She was leaking urine per vaginum and also voided normally per-urethra. There was no history of stress incontinence, urgency and dysuria. Per vaginal examination revealed indurated area at vault. Three swab test revealed that upper swab was soaked with clear urine.

**INVESTIGATIONS**

Ultrasound revealed left lowerpole hydronephrosis. Intravenous urogram (Fig. 14.16.1) revealed left duplication of ureter with hydroureronephrosis of lower moiety. Micturating cystourethrogram (MCU) revealed no VVF (Fig. 14.16.2). Cystoscopy revealed two ureteric orifices on left side (Fig. 14.16.3). Bladder was normal. Retrograde pyelogram revealed normal upper moiety ureter (lower orifice) and extravasation of contrast from lower moiety ureter 2 cm above the ureteric orifice.

![Fig. 14.16.1: Intravenous urogram](image-url)
TREATMENT

Modified Lich-Gregoir stented left lower moiety ureteric reimplantation was done. She had total continence on stent removal.

COMMENTS

In ureteral duplication with ureterovaginal fistula, injury can only be in one moiety and diagnosis can be missed. Proper history, examination and evaluation can clinch the diagnosis. Common sheath reimplantation can be done if injury is close to the ureteric orifice, otherwise single ureter can be reimplanted.

BIBLIOGRAPHY

INTRODUCTION

The rupture or fistulization of lymph vessels into the urinary system, known as chyluria (milky urine), is caused mainly by bancroftian filariasis. On rare occasions, chyluria may also be caused by neoplasia, lymphatic malformation, abdominal trauma, as well as other infectious diseases such as tuberculosis. The patients suffering from milky urine in bancroftian filariasis can be from endemic and nonendemic areas. A detailed history of illness, careful physical examination and examination of urine for chyle confirms the diagnosis. Imaging studies like intravenous urogram, magnetic resonance imaging (MRI), lymphangiogram and retrograde ureterogram can demonstrate abnormal communications. The management is based on patient education and adjustment to a low lipid, high protein diet in addition to increased fluid intake. If the patient does not respond, he can be managed by instillations of radiographic agents, diluted silver nitrate on the site of leak. Intractable cases with severe chyluria are managed surgically by stripping of lymphatic communications previously by open approach and now laparoscopically robotically.

CLINICAL PRESENTATION

A 45-year-old male presented with passing of whitish color urine for 2 years duration (Fig. 15.1.1). He had been treated previously but did not respond to treatment. Intravenous urogram revealed normal functioning kidneys. Retrograde pyelogram revealed communication of pelvicalyceal system on the right side. Right retroperitoneososcopic stripping of lymphatics was done. Patient improved of symptoms.
COMMENTS

Chyluria is a common clinical condition in endemic areas in India. Patients can be managed effectively by diet control, endoscopic injections and surgery.

BIBLIOGRAPHY


Fig. 15.1.1: Clinical picture of chyle in urinary bag
15.2 Filarial Scrotum

**INTRODUCTION**

Genital bancroftian filariasis may manifest in several ways including hydrocele, lymph varix, lymph scrotum, filarial penis or elephantiasis of the genitalia and chyluria. Hydrocele accounts for 90% of the morbidity due to genital filariasis. Acute presentation of genital filariasis is rarely seen and includes acute funiculitis, orchitis and epididymitis.

Diagnosis of genital filariasis can be confirmed by direct demonstration of microfilaria in blood or aspirated fluid. Various studies have established the role of high-frequency, high-resolution scrotal ultrasonography for diagnosing scrotal filarial infection in symptomatic patients.

**TREATMENT**

After medical treatment, surgical excision can be done if the patient has physical discomfort.

**CLINICAL CASES**

**Case 1**

A 55-year-old man presented with huge scrotal swelling hanging up to knee joint with difficulty in walking (Fig.15.2.1). He was obese, diabetic and hypertensive. On advise, he agreed for bariatric surgery and surgery of scrotum. Both surgeries were done together. Complete excision of the

![Fig. 15.2.1: Case 1—Clinical picture](image)
filarial scrotum was done with preservation of testis. He tolerated the surgery well. After 3 months follow-up, his weight reduced by 50% and he had no discomfort in walking. Scrotum returned to normal shape and size.

**Case 2**

A 60-year-old man presented with swelling over penis and scrotum for many years (Fig. 15.2.2). He was counseled for surgery but decided for observation only.

![Fig. 15.2.2: Case 2—Clinical picture](image)

**COMMENTS**

Filariasis of genital tract is common in endemic areas. In case of functional deformity, surgery is advisable.

**BIBLIOGRAPHY**

15.3 INTERSEX—TRUE HERMAPHRODITE

**INTRODUCTION**

True hermaphroditism is an intersex condition in which an individual is born with ovarian and testicular tissue. There may be an ovary underneath one testicle or the other, but more commonly one or both gonads are an ovotestis containing both type of tissues. The karyotypes can be 47,XXY; 46,XX/46,XY; or 46,XX/47,XXY, and various degrees of mosaicism [with one interesting case of an XY predominant (96%) mosaic giving birth].

On examination, external genitalia are often ambiguous, the degree depending mainly on the amount of testosterone produced by the testicular tissue between 8 and 16 weeks of gestation.

The management is individualized according to external genitalia and sex of rearing. A detailed discussion and counseling is necessary.

**CLINICAL PRESENTATION**

A 21 years young gentleman is a known case of hermaphroditism. The patient was born with indeterminate genitalia and brought up as a male child. Several reconstructive surgeries were done on the penis but the details were not available.

Magnetic resonance imaging was done in the year 2000 which showed no female organs but in the year 2007, it showed normal uterus, ovaries and vagina.

![Fig. 15.3.1: Clinical picture of the patient](image-url)
EXAMINATION

Examination revealed poor development of secondary sexual characters (Fig. 15.3.1), normal size penis with scars of previous surgery and no testis was not present in the scrotum (Fig. 15.3.2). Breasts were poorly developed (Fig. 15.3.3). Cystoscopy revealed normal urethra and bladder.

Fig. 15.3.2: External genitalia

Fig. 15.3.3: Clinical picture of the chest
**TREATMENT**

The patient was counseled for his condition various and treatment options were discussed. Parents insisted to rear him as a male to avoid any harassment of their son.

Robotic hysterectomy with bilateral oophorectomy was done under general anesthesia on December 01, 2010. Figure 15.3.4 shows specimen of uterus and both the ovaries. The patient recovered well.

![Operative specimen](image)

**Fig. 15.3.4:** Operative specimen

**FOLLOW-UP**

He was given testosterone supplementation.

**COMMENTS**

Clinical management of intersex conditions is controversial because the available evidence is limited and conflicting; with no long-term population-based studies comparing matched controls.

**BIBLIOGRAPHY**

15.4 PELVIC LIPOMATOSIS

INTRODUCTION

Pelvic lipomatosis is a rare condition of overgrowth of normal fat in the perivesical and perirectal spaces. Although it is usually an incidental finding from a gastrointestinal or genitourinary evaluation, various symptoms, usually urologic, have been ascribed to its presence. Diagnosis of pelvic lipomatosis can be made confidently by cystographic features and computed tomography (CT) scan. In 75% of pelvic lipomatosis, cystitis cystica glandularis may be associated. Pelvic lipomatosis frequently progresses to obstructive uropathy and renal failure, and cystitis cystica glandularis into adenocarcinoma.

Management for obstructive uropathy is by surgery and excision of fat.

CASE 1

Clinical Presentation

A 32-year-old male, presented with the history of single episode of gross hematuria with passage of clots. He also had history of dysuria. Examination revealed a diffuse mass in lower abdomen.

Investigations

Intravenous urogram (Figs 15.4.1A and B) revealed normal upper tract with a normal capacity elongated and pear-shaped bladder with multiple filling defect at the base of the bladder. The bladder was displaced superiorly and anteriorly. CT scan (Fig. 15.4.2) and MRI showed compression of the bladder by fat with intravesical lesion.

Cystoscopy revealed four polypoidal masses ranging from 2 x 3 cm in diameter in the region of bladder base. Trans urethral resection was done and histopathology confirmed cystitis cystica glandularis.

Treatment

The patient was advised weight reduction and a regular follow-up. During follow he developed bilateral hydroureteronephrosis and bilateral ureteric stenting could not be done. Exploration with excision of fat at the dome of the bladder and bilateral ureteric reimplantation was done.
CASE 2

Clinical Presentation

A 42-year-old male, presented with left flank pain for 1 year duration along with urinary frequency and dysuria. Five years ago he had received shock wave lithotripsy for left renal calculus with complete clearance of the calculus. Six moths back he was diagnosed with left ureteric stone for which ureteroscopy was attempted in another hospital but failed due to inability to visualize the left ureteric orifice.

Investigations

Investigations revealed normal urine R/M. Urine culture was sterile. Renal function tests were normal. Plain X-ray KUB region revealed radiopaque shadow on the left side of the pelvis (Fig. 15.4.3). Intravenous urogram
Fig. 15.4.3: Case 2—Plain X-ray KUB region

Fig. 15.4.4: Case 2—Intravenous urogram

Fig. 15.4.5: Case 2—Computed tomography (CT) scan
revealed normal functioning kidney and medial deviation of ureters with calculus in left lower ureter (Fig. 15.4.4). CT scan revealed fat around the bladder with lower ureteric calculus (Fig. 15.4.5).

**Treatment**

Cystoscopy revealed inflammatory changes in lower part of the bladder and ureteric orifices could not be located. Exploration, excision of fat at the dome of the bladder and bilateral ureteric reimplantation with removal of left ureteric stone was done.

**COMMENTS**

Pelvic lipomatosis is a rare disease characterized by an abnormal accumulation of adipose tissues around the pelvic organs. The symptoms are vague and nonspecific. The diagnosis is facilitated by typical radiological features. CT scanning is extremely useful and typical. The course of the disease is usually benign in older men, but may be dangerous in the young and it may require urinary diversion.

**BIBLIOGRAPHY**

15.5 POSTNEPHRECTOMY RESIDUAL KIDNEY

**INTRODUCTION**

During difficult nephrectomy due to perirenal adhesions and bleeding, surgeon can leave partial functioning kidney which can present as renal fistula.

**CLINICAL PRESENTATION**

A 26-year-old male presented with history of recurrent urinary tract infection (UTI) in 1996. On evaluation, he was detected to have left solitary kidney (congenitally absent right kidney) with grade 4 vesicoureteral reflux (VUR) (BU: 30 mg/dL, serum creatinine: 1.1 mg/dL). Left ureteric reimplantation was done in 1996. He again presented in 2007, with complaints of recurrent nausea, vomiting and decreased urine output.

**INVESTIGATIONS**

Blood urea was 200 mg/dL and serum creatinine was 8 mg/dL. Ultrasound revealed absent right kidney (Fig. 15.5.1), hydronephrotic left kidney (Fig. 15.5.2) and dilated lower left ureter (Fig. 15.5.3).

![Fig. 15.5.1: Ultrasound (absent right kidney)](image-url)
**Fig. 15.5.2:** Ultrasound (hydronephrotic left kidney)

**Fig. 15.5.3:** Ultrasound (dilated lower left ureter)
**TREATMENT**

Patient was diagnosed with end-stage renal disease (ESRD). The patient was started on hemodialysis. Left PCN was done in December 2007. He was planned for renal transplant, and referred for pre-transplant nephrectomy. He underwent left open simple nephrectomy somewhere else on January 30, 2008. Intraoperative findings: Kidney was badly adherent to surrounding tissues (peritoneum, psoas). Ureter was thickened and dilated. Left subcapsular nephrectomy with excision of the ureter up to pelvic brim was done.

**HISTOPATHOLOGY**

7.8 × 6 × 4 cm kidney, with chronic pyelonephritis; dilated, thickened ureter showing ureteritis.

**FOLLOW-UP**

Drain was removed on 2nd postoperative day. The patient developed watery discharge from the main wound on 5th postoperative day. Regular dressings done—watery discharge stopped for 2 days. The patient was put on maintenance hemodialysis.

The patient again developed watery discharge from main wound after 7 days. Ultrasonography KUB region revealed 7 × 6 cm loculated collection in left renal fossa (Fig. 15.5.4). Left pigtail was placed which drained 150 mL clear fluid. Antegrade pyelogram (Fig. 15.5.5) revealed hydronephrotic residual kidney. CT scan revealed left residual kidney (Figs 15.5.6A and B). Combined cystogram and nephrostogram revealed residual ureter (Fig. 15.5.7).

Drain fluid creatinine: 8 mg/dL. PCN was draining at 100–150 mL/day. Patient was planned for cystopanendoscopy (CPE) and exploration on April 07, 2008. CPE revealed left hemitrigone, right ureteric orifice was not seen and left native ureteric orifice—could not be cannulated. New ureteric orifice was wide open on the posterolateral wall.
Fig. 15.5.4: Repeat ultrasound

Fig. 15.5.5: Antegrade pyelogram showing dilatation of residual kidney
Figs 15.5.6A and B: Contrast enhanced computed tomography scan

Fig. 15.5.7: Combined cystogram and nephrostogram
Exploration done via 11th rib flank approach. Intraoperative findings: residual portion of left kidney, badly adherent to surrounding tissues, renal vessels ligated. Nephrectomy of residual kidney was done (Figs 15.5.8A and B).

Histopathology revealed $8.5 \times 6.5 \times 2.5$ cm kidney showing chronic pyelonephritis, with hydronephrosis and loss of CMD. Ureter was not identified.

The patient had uneventful postoperative course. Drain was removed on 2nd postoperative day. Patient was put on maintenance hemodialysis, and he underwent renal transplant later.

**Figs 15.5.8A and B:** Specimen of nephrectomy of residual kidney

**COMMENTS**

Precaution should be taken for complete removal of kidney during nephrectomy.
15.6 RENAL ARTERY ANEURYSM

**INTRODUCTION**

Renal artery aneurysms, previously considered to be rare, have been diagnosed more frequently in recent years mainly due to the extensive use of angiography. They can be asymptomatic; otherwise, common presentation is hypertension. Most cases were manifestations of medial hyperplasia or atherosclerosis of the renal arteries. Symptomatology is not pathognomonic. Expanding aneurysm, intractable hypertension, hematuria and renal infarction represent the most common indications for surgical repair. Reconstruction and repair of these aneurysms, with preservation of the kidney, is the preferred treatment.

**CLINICAL PRESENTATION**

A 38-year-old female, who is married and has three children, during executive health check-up detected to have left renal artery aneurysm on ultrasound examination. She has normal blood pressure and was asymptomatic. She had no history of hematuria. On abdominal examination a bruit could be heard in the area of left renal artery.

Digital subtraction angiography (Figs 15.6.1A to D) confirmed the presence of left renal artery aneurysm involving distal part of the renal artery and its branches.

**TREATMENT**

She was counseled for the conservative treatment vs surgery but she could not be followed up.
Figs 15.6.1A to D: Digital subtraction angiography

**COMMENTS**
Symptomatic renal artery aneurysm merits early surgical treatment to prevent any emergency catastrophe.

**BIBLIOGRAPHY**
15.7  RENAL LYMPHANGIOMA

INTRODUCTION

Lymphangiomas are developmental malformations due to failure of developing lymphatic tissue to establish normal communication with remainder of lymphatic system. Renal lymphangiomatosis is a rare disorder characterized by unilocular or multilocular cystic masses in the peripelvic or perirenal areas. Imaging plays a key role in differentiating lymphangiomas from other renal cystic diseases. It is usually a benign tumor.

Renal peripelvic lymphatic cysts present with palpable abdominal mass, elevated blood pressure, normal renal function, although obstructive uropathy reported. They are usually bilateral; no sex/age predilection. They may exacerbate during pregnancy, possibly rupture producing perinephric fluid and ascites. Location of cysts can be peripelvic (most common) perinephric (capsular cysts separated by thin septations) and rarely intraparenchymal which are indistinguishable from renal cysts. Subcapsular collection are the rarest.

Histologically they are thin-walled with a single layer endothelium containing lymph/proteinaceous-gelatinous material.

Management: Asymptomatic cases do not warrant any intervention. Percutaneous drainage is effective in many cases. Surgical intervention consists of cyst marsupialization and the last therapeutic measure is open/ laparoscopic or robotic ablation.

CASE 1

Clinical Presentation

A 25-year-old male presented with ill-defined right flank fullness not associated with urinary or bowel complaints.

Clinical Examination

The clinical examination was unremarkable and he was normotensive.

Investigations

Screening USG abdomen revealed a multiloculated right perirenal fluid collection. Serum, urine biochemical analysis and hematological investigations were normal. CECT (Fig. 15.7.1) revealed a hypodense (fluid density) mass lesion in right perinephric region well-contained within Gerota’s fascia and well-circumscribed hypodense lesions within renal parenchyma. Both kidneys demonstrated good function. Spleen, liver and pancreas were devoid of cystic lesions. T2-weighted MR image (Fig. 15.7.2) revealed hyperintense perinephric “capsular” cysts.
In the absence of troublesome symptoms and lack of functional consequences, the patient was kept under follow-up. At one year of follow-up, the patient was symptom free and there was no further increase in the size of lymphangioma. He continues to be managed conservatively.
CASE 2

Clinical Presentation

A 13-year-old girl presented with insidious onset abdominal distention for a duration of 1 year. No history of fever, anorexia, jaundice, oliguria, respiratory distress. Family history was unremarkable.

Clinical Examination

Clinical examination revealed normal blood pressure, mild pallor no lymphadenopathy/pedal edema, abdomen was protuberant with bimanually palpable, bosselated, cystic flank masses. Respiratory/cardiovascular systems were unremarkable.

Investigations

Investigations revealed normal renal and liver function parameters. Urinalysis was normal. Chest X-ray was also normal. USG abdomen revealed bilateral renal subcapsular fluid collection without hydronephrosis. The liver, spleen, pancreas and gall bladder were normal. CECT abdomen

Fig. 15.7.3: Case 2—Contrast enhanced computed tomography (CECT)
revealed bilateral renal subcapsular hypodense lesions, displacing normally functioning renal parenchyma (Fig. 15.7.3). Intraparenchymal or peripelvic cysts were absent. MRI revealed hyperintense bilateral subcapsular renal lesions, severely distorting the parenchyma akin to subcapsular hematomas (Figs 15.7.4A to C). A band of renal tissue was seen stretched between capsule and surface of kidney on left side.

![Figs 15.7.4A to C: Case 2—Magnetic resonance imaging (MRI)](image)

**Treatment**

Therapeutic aspiration revealed clear fluid, high globulin content and lymphocytes confirming the diagnosis of lymphangiomatosis. Due to re-accumulation of fluid—definitive transperitoneal laparoscopic ablation was performed. Bilateral wide excision of lymphangioma cyst wall, equivalent to decorticating kidneys, was done. 5.6 liters of fluid was evacuated from lymphangioma. The band of renal tissue, stretched between the renal capsule and parenchymal surface, was excised with a wedge of renal cortex and the defect was sutured intracorporeally. Postoperative period was uneventful. She had abdominal lymphorrhea for 3 days. Follow-up at six months showed
no recurrence. At 1 year the patient was asymptomatic, but he had developed mild ascites and diagnostic aspiration was consistent with lymph.

**COMMENTS**

Perirenal lymphangioma is a rare condition, can be detected and diagnosed by imaging. It can be managed effectively by surgery.

**BIBLIOGRAPHY**

INTRODUCTION

Retroperitoneal fibrosis is a rare disease characterized by a fibrosclerotic tissue in the retroperitoneum, often leading to encasement of the ureters. About two-thirds of the cases of retroperitoneal fibrosis are idiopathic, while the remaining cases are secondary to a variety of different causes, including drugs, tumors and infections. Most patients present with abdominal and/or low back pain or detected during investigations for chronic kidney disease. Serum markers of inflammation are usually, but not invariably, elevated. The diagnosis can be confirmed by computed tomography or magnetic resonance imaging, which typically show an enhancing retroperitoneal mass medially dislodging the ureters. Positron emission tomography can be useful to document the extent and metabolic activity of the inflammatory process. Basically, it is a medical disease and treatment rests on glucocorticoids with a tapering scheme variably combined with immunosuppressive agents. In cases of ureteric obstruction not responding to medical treatment, relief of obstruction by stenting or ureterolysis is required. Ureterolysis can be performed by open surgery, laparoscopically and recently robotically as well. Ureterolysis is combined with retroperitonization or by omental wrap to prevent further ureteric obstruction.

CLINICAL PRESENTATION

A 52-year-old female presented with the history of loss of appetite and weakness for 3 months duration. There was no history of calculus disease and no urinary symptoms. He had a history of hypertension for past 3 years on medication. Abdominal examination was normal. Per vaginal (PV) examination was also normal.

INVESTIGATIONS

Serum creatinine was 5.6 mg/dL. Ultrasound revealed mild hydronephrosis with dilated upper ureter. Cystoscopy revealed normal bladder. Bilateral retrograde pyelography (RGP) was done which revealed bilateral hydroureteronephrosis with medial deviation of middle part of ureters (Fig. 15.8.1).

TREATMENT

Bilateral ultrasound-guided percutaneous nephrostomy (PCN) was done. Serum creatinine returned to normal level in 3 weeks time. Abdominal exploration, bilateral ureterolysis (Fig. 15.8.2) with bilateral omental wrap (Figs 15.8.3A and B) and bilateral DJ stenting was done. She recovered well after surgery. Plain X-ray (Fig. 15.8.4) shows bilateral DJ stent in position with lateralization of ureters. DJ stents were removed after 6 weeks. Her renal functions remained normal on follow up.
Fig. 15.8.1: Bilateral retrograde pyelogram (RGP)

Fig. 15.8.2: Intraoperative picture showing ureterolysis

Figs 15.8.3A and B: Intraoperative pictures showing omental flaps and omental wrapping of ureter
Idiopathic retroperitoneal fibrosis is a rare condition and a high index of suspicion is required in the diagnosis in cases of bilateral midureteric obstruction. In early stages, it can be managed by medical therapy, otherwise surgery is required.

**BIBLIOGRAPHY**

15.9 RETROVESICAL HYDATID CYST

INTRODUCTION

Hydatid disease is caused by the parasite, *Echinococcus granulosus* and is endemic in many parts of the world. Hydatid cysts may develop in almost any part of the body and hydatid cysts located at some unusual sites may create diagnostic confusion. Hydatid cysts located in the peritoneal cavity or pelvic hydatid cysts are usually secondary to spontaneous rupture from a primary liver focus or surgical inoculation. However, isolated retrovesical hydatid cysts are extremely rare with only a few cases reported in literature.

PRESENTATION

Retrovesical hydatid cysts may have a varied and nonspecific presentation. Patients may present with palpable mass, flank pain, frequency, urinary retention and pain on micturition. Patients may also present with constipation, weight loss and renal insufficiency.

ETIOPATHOGENESIS

According to the classical theory of Deve, fissuring or rupture of a primary hepatic, splenic or mesenteric cyst would seed its contents in the abdominal cavity. This primary cyst might then heal and even disappear, leaving behind a scar that could be overlooked by any diagnostic modality. The pouch of Douglas would then be the preferred site for development of a secondary cyst in the pelvis, initially intraperitoneal and later subperitoneal. When a primary visceral lesion is absent and there is no evidence of peritoneal seeding, hematogenous dissemination could explain the pathogenesis of a solitary retrovesical lesion.

DIFFERENTIAL DIAGNOSIS

In view of its rarity, a hydatid cyst may not be the first differential diagnosis in a patient presenting with an isolated pelvic cyst. On imaging, a retrovesical hydatid cyst may mimic the following conditions: rectal duplication cyst, rectosigmoid neoplasm, posterior bladder diverticulum, cyst of the seminal vesicle, hydronephrosis in a pelvic kidney and large ectopic ureterocele. In female patients, however, a retrovesical hydatid cyst may mimic any one of the following gynecological conditions: ovarian neoplasm, Müllerian remnant, hydrosalpinx, pseudomyxoma peritonei and tubal pregnancy.

TREATMENT

Medical treatment followed by open/laparoscopic/robot-assisted laparoscopic surgical excision.
CLINICAL CASE PRESENTATION

A 64-year-old male, Hindi professor, from Nepal presented with history of urgency for 3 years. He had increased frequency of urination. There were no obstructive voiding symptoms, dysuria, hematuria, lithuria, pyuria, hydatiduria, fever, etc. He had occasional dull aching suprapubic pain.

He had a pet dog. He was diagnosed as having a cyst in retrovesical region on ultrasound in 2007. Initially took homeopathic medicine, then albendazole for 7 months. No history of tuberculosis. History of appendicectomy 30 years ago.

EXAMINATION

On examination, no mass palpable in abdomen, appendectomy scar noted. Digital rectal examination revealed grade 1 benign prostate enlargement and cystic boggy swelling anterior to rectum.

INVESTIGATIONS

Enzyme-linked immunosorbent assay (ELISA) for *Echinococcal* antibody was 1.80 IU/mL (positive > 1.1 IU/mL). USG pelvis showed multicystic lesion posterior to urinary bladder. CECT (Figs 15.9.1A and B) showed the multicystic mass located in the rectovesical pouch pushing the posterior bladder wall anteriorly.

Figs 15.9.1A and B: Contrast enhanced computed tomography (CECT)

TREATMENT

Robot-assisted laparoscopic excision of the hydatid cysts was done. The procedure was uneventful and the patient was relieved of his symptoms. Cut open section of the specimen with multiple daughter cysts is shown in Figure 15.9.2.
COMMENTS

Retrovesical hydatid cyst is a rare condition and can be diagnosed by imaging showing daughter cysts. Surgical removal is advisable in symptomatic patients.

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