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Case Series

Spectrum of presentation and management of retrocaval ureter: a 10-year single centre experience

Sanjula Jethwani*, Harikrishna R., A. V. Venugopalan

Department of Urology and Renal Transplant, Government Medical College, Kozhikode, Kerala, India

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***Correspondence:**
Dr. Sanjula Jethwani,
E-mail: sanjulaurology@gmail.com

ABSTRACT

Retrocaval ureter is a rare congenital abnormality with an incidence of 0.06-0.17%. With the frequent use of radiological imaging in modern practice, there has been an increase in the identification as well as evolution in the management options for these cases. We present a series of 18 cases of retrocaval ureter managed at our institute from August 2015 to May 2025. The initial evaluation included routine blood investigations and ultrasound KUB followed by CT scan, IVU and RGP for confirmation. This case series aims to highlight the spectrum of presentation of retrocaval ureter that we encountered and the management option chosen. In our study, there were 17 cases of Type 1 retrocaval ureter and one case of Type 2 retrocaval ureter. 14 cases presented with flank pain, 3 cases with urinary tract infection and 1 case were an incidental finding. One patient had an associated renal pelvic calculus and 1 patient had a poorly functioning right kidney. One patient presented with a broken forgotten DJ stent in situ placed initially in view of recurrent urinary infections. In our study, 1 patient had left renal agenesis and 1 patient was a known case of ventricular septal defect. Retrocaval ureter, being a rare entity, has been reported sporadically over decades. The existing case series report a few numbers of cases or dwell on the traditional management done with the increasing trend towards minimal invasive approach. In our study, we aim to highlight the varied presentations of retrocaval ureter that we encountered in our institute, along with the management tailored accordingly. Retrocaval ureter is a rare clinical entity. With modern diagnostic tools, there is an increasing chance of early diagnosis, thus enabling us to safeguard the renal function with prompt intervention.

Keywords: Retrocaval ureter, Flank pain, Hydroureteronephrosis

INTRODUCTION

Retrocaval ureter, also known as circumcaval ureter, is a rare congenital abnormality with an incidence of 0.06-0.17%.[1] It was first described by Hochstetter in 1893.[2] It is in fact a congenital vascular maldevelopment, presenting in the 3rd to 4th decade of life, being three times more common in males. It can be an incidental finding or present with abdominal pain and recurrent urinary tract infections.[3] Complications including urolithiasis and varying degrees of renal dysfunction can also be seen as a result of ureteric obstruction due to compression by inferior vena cava (IVC).[4] Initially, most of these cases were incidentally detected during autopsy, but with the frequent use of radiological imaging in modern practice, there has been an increase in the identification as well as evolution in the management options for these cases.

CASE SERIES

We present a series of 18 cases of retrocaval ureter managed at our institute from August 2015 to May 2025. There were 11 males and 7 females with age ranging

from as young as 19 years to as late as 69 years. The most common presentation was right-sided flank pain in 14 cases, followed by a history of recurrent urinary tract infections in 3 cases, followed by incidental detection in 1 case. There was no history of hematuria or other lower urinary tract symptoms.

The initial evaluation included routine blood investigations and ultrasound (USG) KUB followed by CT (Computed Tomography) scan, IVU (Intravenous Urogram) and RGP (Retrograde Pyelography) for confirmation of the diagnosis. The characteristic finding on the USG was that of right side hydroureteronephrosis with a non-dilated distal ureter. Contrast enhanced CT scan demonstrated the retrocaval ureteric segment, which could also be visualised as the medial deviation of the right ureter on IVU (Intravenous Urogram) (Figure 1) and RGP (Retrograde Pyelogram). Out of the 18 cases, 17 had Type 1 retrocaval ureter and 1 case had Type 2 retrocaval ureter.

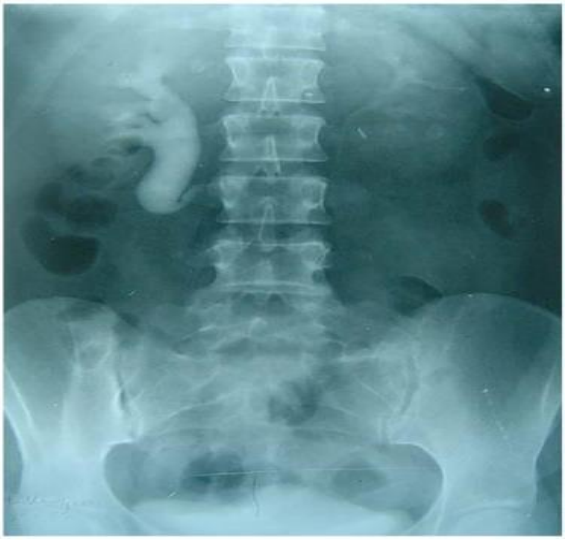

Figure 1: IVP showing the characteristic medial deviation of the retrocaval ureter.

This case series aims to highlight the spectrum of presentation of retrocaval ureter that we encountered and the management option chosen.

Case 1

A 46-year-old male, with chronic myeloid leukaemia, who presented with right sided flank pain in the past 1 month and was diagnosed to have retrocaval ureter. He was managed with right DJ stenting, following which he symptomatically improved. Post stent removal, he was asymptomatic and kept on regular follow up.

Case 2

A 69-year-old male, who presented with right sided flank pain of 6 months duration. He was diagnosed to have Right renal pelvic calculus measuring 1.2 x 1.1 cm (1400 HU) along with retrocaval ureter. He was managed with PCNL (Percutaneous Nephrolithotomy) for the pelvic calculus followed by uretero-ureterostomy for the retrocaval ureter.

Case 3

A 27-year-old female with hypertension, who presented with right sided flank pain in the past 1 year. On evaluation, she was found to have retrocaval ureter with a poorly functioning right kidney with a 12% Split renal function on DTPA. In view of persistent symptoms, she underwent a Right Simple Nephrectomy.

Case 4

A 19-year-old male who presented with right sided flank pain and fever with chills, He was found to have retrocaval ureter with right pyelonephritis along with a contralateral renal agenesis (Figure 2). He was managed with IV antibiotics for the pyelonephritis and later underwent surgical management in the form of uretero-ureterostomy with excision of atretic segment in view of management of the solitary functioning kidney.[5]

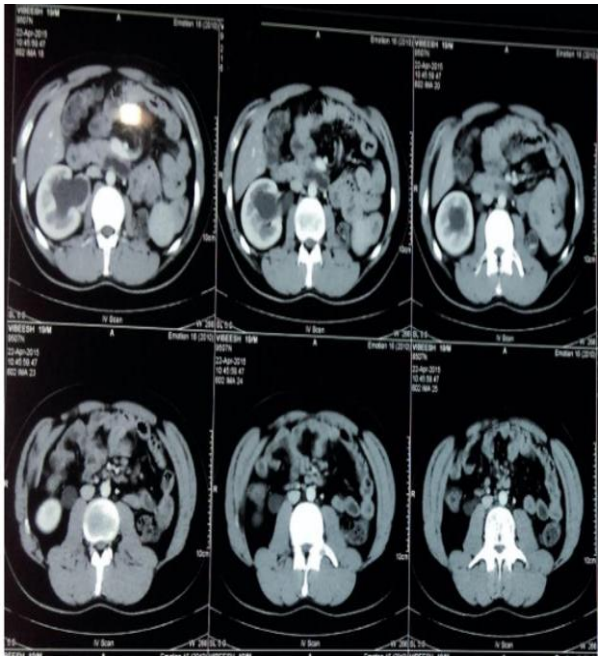

Figure 2: CT scan image showing right hydronephrosis and hydroureter upto level L3 where the ureter crosses behind the IVC, along with left renal agenesis.

Case 5

A 27-year-old female, known case of ventricular septal defect (VSD), presented with right flank pain of 3 years duration. She was found to have retrocaval ureter and underwent laparoscopic uretero-ureterostomy (Figure 3) in view of persistent symptoms.

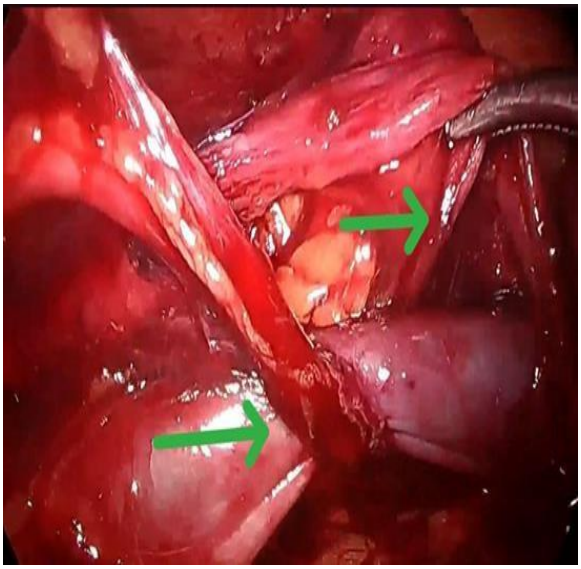

Figure 3: Intra-operative image showing the right ureter passing behind the IVC during laparoscopic uretero-ureterostomy.

Case 6

A 62-year-old female with a history of Right recurrent pyelonephritis for which she was managed outside by Right DJ stenting 2 years back. The patient was lost to follow up and presented to us with complaints of right sided flank pain and a history of passage of stent fragment. On evaluation, she was diagnosed to have retrocaval ureter along with retained proximal fragment of the stent (Figure 4). She was managed by uretero-ureterostomy along with ureteroscopic removal of the retained stent fragment.

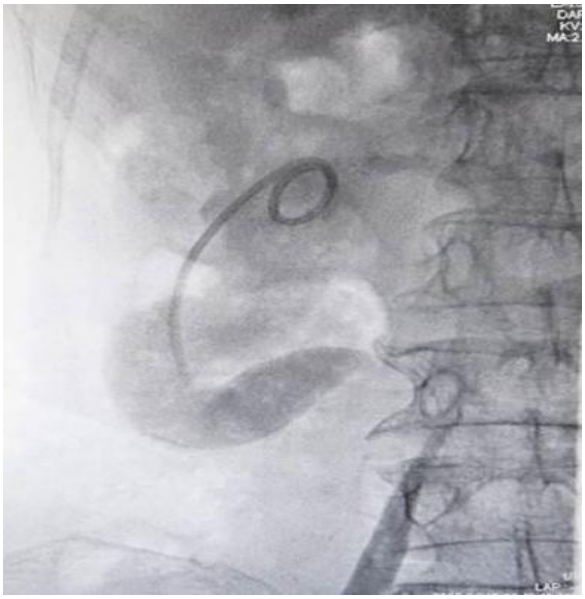

Figure 4: RGP demonstrating the retrocaval ureter along with the retained stent fragment.

Case 7

A 51-year-old male who presented with right sided flank pain with a duration of 4 months. He was diagnosed to have retrocaval ureter and was managed with uretero-ureterostomy. Intraoperatively the ureter was found to continue downward, medial to the IVC, after passing behind the IVC rather than returning to the normal lateral course (Figure 5).

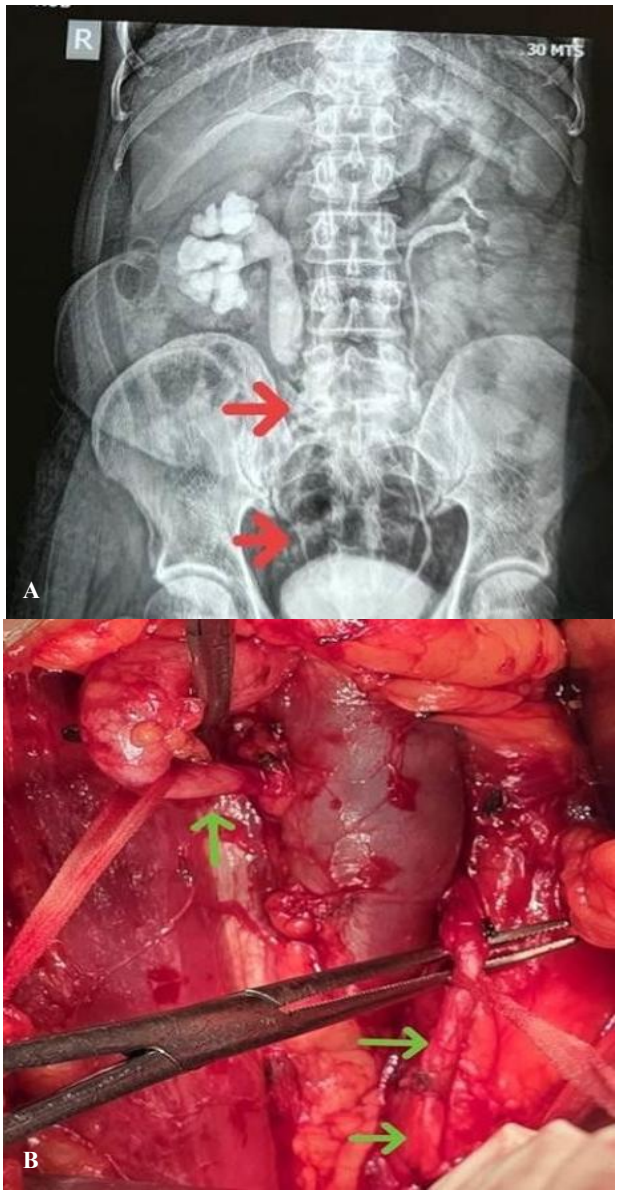

Figure 5: (A) IVP and (B) intra-operative photo demonstrating the continued medial course of the right ureter after crossing behind the IVC.

Cases 8-14

The case 8-14 were also managed with uretero-ureterostomy (Figure 6), out of which 1 case was done by laparoscopic approach.

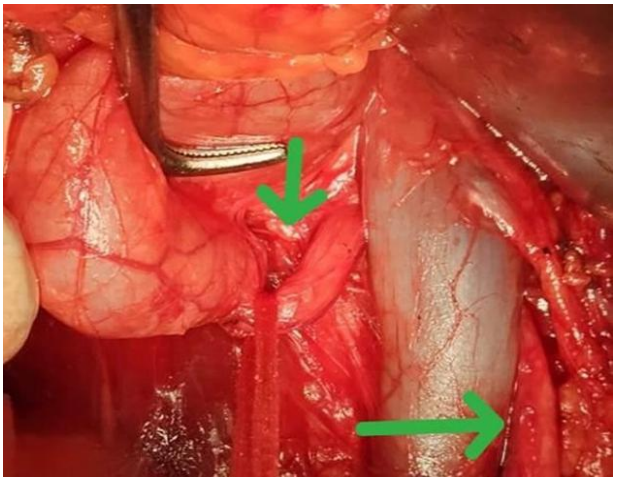

Figure 6: Intra-operative image showing the retrocaval segment during open uretero-ureterostomy.

Table 1: Summary of all cases.

Variables	N (%) / Mean±SD
Age (years)	40.7±17.1
Range	19-69
Sex	
Male	11 (61.11)
Female	7 (38.89)
Clinical presentation	
Flank pain	14 (77.78)
Recurrent urinary infection / pyelonephritis	3 (16.67)
Incidental	1 (5.55)
Associated conditions / previous urological history	
Renal pelvic calculus	1 (5.6)
Hypertension	1 (5.6)
Left renal agenesis	1 (5.6)
Ventricular septal defect	1 (5.6)
Chronic myeloid leukemia	1 (5.6)
Broken DJ stent	1 (5.6)
Type of retrocaval ureter	
Type 1	17 (94.4)
Type 2	1 (5.6)
Management	
Uretero-ureterostomy	12 (66.7)
Pyelo-ureterostomy	3 (16.7)
Nephrectomy	1 (5.6)
DJ stenting	1 (5.6)
Conservative management	1 (5.6)
Surgical approach	
Open	12 (66.7)
Laparoscopic	4 (22.22)

Cases 15-17

The case 15-17 were managed by pyelo-ureterostomy out of which 2 were done by laparoscopic approach.

Case 18

A 40-year-old female, who was incidentally detected to have type 2 retrocaval ureter. In view of absence of symptoms, normal renal function and minimal hydroureteronephrosis, she was conservatively managed.

The post-operative period of all the cases was uneventful. There were no significant intraoperative or post-operative complications. On follow up, the patients had both symptomatic along with clinical improvement. The histopathology of the atretic excised ureteric segment showed areas of fibrosis along with chronic inflammation.

DISCUSSION

Retrocaval ureter is a rare congenital vascular anomaly in which a segment of the proximal ureter crosses behind the IVC. It occurs due to an abnormal development of the venous system rather than the urinary tract.[6] It is usually found on the right side with the exception of conditions like situs inversus and double IVC, wherein it occurs on the left side.[7] This occurs between the 4th-8th week of development. The subcardinal veins, which drain the mesonephros, connect to the posterior cardinal veins via multiple anastomoses, with both lying ventral to the developing ureter. During the 6th week, the supracardinal veins develop between the two systems, lying dorsal to the developing ureter. The renal portion of IVC is formed by the right subcardinal vein and the infrarenal portion by the distal right supracardinal vein. When the renal segment of IVC develops from the subcardinal vein along with the abnormal persistence of right posterior cardinal vein (which fails to atrophy), the ureter gets trapped posteriorly, leading to the formation of retrocaval ureter (Figure 7).[8]

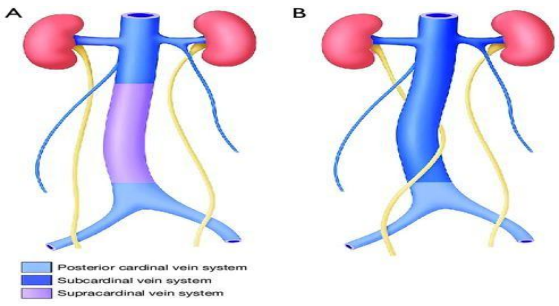

Figure 7: The normal development of the IVC and the abnormality leading to the development of retrocaval ureter.[9]

Bateson et al described 2 types of retrocaval ureter, Type 1 being more common (90% cases). Type 1 (low loop type) occurs due to an intrinsic abnormality of the ureteric segment which passes behind the IVC at the level of L3 vertebra, thus visualised as a S shaped or fish hook deformity (Figure 8). It causes significant hydronephrosis and thus usually needs surgical resection. Type 2 (10%

cases) also known as high loop type, occurs due to extrinsic compression of the normal ureter by IVC when it passes at the level of the renal pelvis, which is visualised as a sickle shaped deformity. It rarely causes obstruction to the kidney and thus is usually conservatively managed.[10]

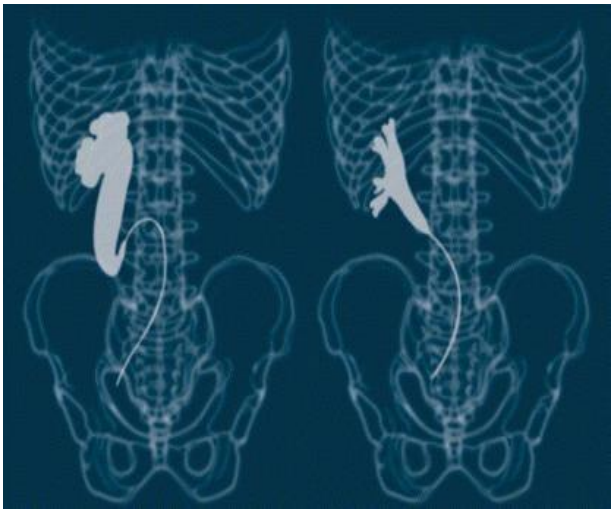

Figure 8: Type 1 and Type 2 retrocaval ureter as per Bateson and Atkinson classification.[11]

In our study, there were 17 cases of type 1 retrocaval ureter (94.44%) and one case of type 2 retrocaval ureter (5.56%). Although congenital, the usual presentation is in the 3rd-4th decade, with male: female ratio being 2.8:1.[8] In our study, the age of presentation varied from 19 years to 69 years with the mean age being 40.67 years. There were 11 males (61.11%) and 7 females (38.89%) with a ratio of 1.57:1. The spectrum of clinical presentation varies from asymptomatic and incidental detection to the most common symptoms being right sided flank pain, recurrent urinary tract infections and hematuria. Complications include calculi and renal failure which may be associated with renovascular hypertension.

In our study, there were many varied presentations. 14 cases (77.78%) presented with flank pain, 3 cases (16.67%) with urinary tract infection and 1 case (5.55%) were an incidental finding. One patient had an associated renal pelvic calculus and 1 patient had a poorly functioning right kidney.

One patient presented with a broken forgotten DJ stent in situ placed initially in view of recurrent urinary infections. Retrocaval ureter may have associated congenital anomalies (21% cases) mainly in the urogenital and cardiovascular systems. These include horseshoe kidney, ureteropelvic junction obstruction (UPJO), contralateral renal anomalies- agenesis, malrotation or ectopic position, ureteral duplication, hypospadias, congenital lack of vas deferens, uterine and vaginal agenesis in the genitourinary system. Cardiovascular anomalies include variations of IVC, situs inversus. Others like brachial arch syndrome, Turner syndrome, myelomeningocele, yolk sac tumour, oesophageal atresia and imperforate anus are also found.[12] In our study, 1 patient had left renal agenesis and 1 patient was a known case of ventricular septal defect.[5]

These abnormalities are important to identify as they may affect treatment choice. Solitary functioning kidney needs prompt intervention to prevent deterioration of renal function. Cardiovascular abnormalities may increase the morbidity of the procedure, thus impacting the outcome.

The initial investigation usually done is ultrasound of the abdomen, which demonstrates the hydronephrosis, occurring mainly due to the compression of the retrocaval ureteric segment by the vena cava and psoas muscle, ultimately leading to inflammation and fibrosis. This can be seen in the histopathology of the excised segment.

The best modality to confirm the diagnosis is CT scan, as it will identify the entire course of the ureter and also identify any other associated anomalies. MRI can be used in doubtful cases and when CT is contraindicated. IVU can also help to demonstrate the characteristic loop, but may need to be combined with RGP when it fails to demonstrate the entire ureteric course, which can happen due to the obstruction. In cases of doubt about renal function, we can use isotope renal scans to help decide the therapeutic option to be chosen.

The management options depend on the clinical presentation, degree of hydronephrosis, type of retrocaval ureter, impairment of renal function and the management of associated complications. Conservative management with regular follow up can be considered for patients with mild hydronephrosis, asymptomatic with incidental detection and in type 2 cases. Ureteral stenting may offer temporary relief in acute setting and facilitate drainage. Surgical management was first demonstrated by Kimbrough in 1935.[13] He described the division of the ureter with or without excision of the narrowed or aperistaltic segment, anteriorisation of the ureter, followed by uretero-ureterostomy or dismembered pyeloplasty over a stent. Another technique described by Harril was the ureteropelvic anastomosis, wherein the division was made at the level of the renal pelvis just above the pelviureteric junction[14] This technique has the advantage of the good blood supply of the pelvis and upper ureter, thus decreasing the chances of post-operative stricture at the anastomotic site.

Complications like renal and ureteric calculi can be dealt with in a single step or a staged procedure. Occasionally, nephrectomy may be needed in the case of severe impairment of renal function or refractory hypertension.

Currently, in the era of minimal invasive surgery, Laparoscopic or retroperitoneoscopic approach for the reconstruction of retrocaval ureter has become popular with all the advantages of reduced post-operative pain and shorter convalescence time. However, they can increase the operative time and can be technically demanding.

In our study, we have the complete spectrum of management options described above. Case 1 was managed by DJ stenting and followed up post stent removal, during which the patient remained asymptomatic and there was no impairment of renal function. Cases 4-14 were managed by uretero-ureterostomy along with excision of the retrocaval segment, out of which 2 were completed laparoscopically. Case 4 had a contralateral renal agenesis associated with the retrocaval ureter, in addition to recurrent pyelonephritis of the solitary functioning kidney. Case 6 had a broken fragment of DJ stent which was removed intra-operatively by ureteroscopy through the cut proximal end of the ureter. Case 7 had a variation detected intra-operatively, wherein the ureter after passing behind the IVC continued the medial course downward rather than returning to the lateral position. Cases 15-17 were managed by pyelo-ureterostomy, out of which 2 were done laparoscopically.

Case 2 presented with a renal pelvic calculus along with the retrocaval ureter, which was managed by PCNL in the first stage followed by uretero-ureterostomy in the second stage. Case 3 presented with a poorly functioning right kidney along with hypertension and recurrent urinary tract infections and thus was managed by Right Simple Nephrectomy.

Case 18 had a Type 2 retrocaval ureter and was conservatively managed in view of absence of symptoms and hydronephrosis.

The histopathology of the atretic excised ureteric segment showed areas of fibrosis along with chronic inflammation. In the post-operative period, patients showed complete symptomatic recovery along with regression of the hydronephrosis on repeat follow-up imaging. Thus, early diagnosis and prompt treatment can preserve renal function and prevent future complications.

Retrocaval ureter, being a rare entity, has been reported sporadically over decades. The existing case series report a few numbers of cases or dwell on the traditional management done with the increasing trend towards minimal invasive approach. In our study, we aim to highlight the varied presentations of retrocaval ureter that we encountered in our institute, along with the management tailored accordingly.

CONCLUSION

Retrocaval ureter is a rare clinical entity. With modern diagnostic tools, there is an increasing chance of early diagnosis, thus enabling us to safeguard the renal function with prompt intervention. This case series aims to highlight the spectrum of presentation and complications that one can encounter and the management options available to deal with them, with minimal invasive approach becoming the trend.

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