

Retrocaval ureter: a rare cause of intermittent right flank pain in a 37 years old lady

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Abstract

A 37 years old female presented with 6 months history of intermittent right flank pain. She was otherwise fit and had no other complaints. Her lab investigations were normal. Abdominal ultrasonography revealed moderate hydronephrosis and proximal hydroureter but no calculus. Retrograde ureteropyelography findings were consistent with the diagnosis of retrocaval ureter, a rare clinical condition. Surgical exploration confirmed the presence of right retrocaval ureter. The ureter was transected and transposed anterior to inferior vena cava and ureteropelvic anastomosis was performed. Postoperative recovery was uneventful.

Keywords: Retrocaval ureter, hydronephrosis, ureter, ureteropelvic junction obstruction.

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Introduction

The retrocaval ureter is a rare congenital anomaly, in which the ureter passes posterior to the Inferior vena cava (IVC). The condition occurs when either subcardinal vein or posterior cardinal vein form infrarenal vena cava instead of the supracardinal vein. As the subcardinal vein is anterior to the developing ureter during embryogenesis, the fully developed ureter hooks around the IVC as it courses down. Retrocaval ureter is sometimes termed as "preureteral vena cava," because, the condition is actually developmental anomaly of IVC and not ureter.¹ It is the right ureter which is predominantly involved in this condition. A retrocaval ureter on the left is seen when there is persistence of left cardinal vein in patients with complete situs inversus or duplication of IVC.² The reported incidence of retrocaval ureter is 1 in 1000, with male predominance.³ In nearly 20% cases, the condition may be associated with concomitant anomalies, mainly from cardiovascular system and genitourinary tract.⁴

Although, it is a congenital anomaly, but patients usually present in 3rd and 4th decades of life with intermittent flank or abdominal pain or dull ache due to ureteric

obstruction and associated hydronephrosis. Because of the rarity of condition, it is not suspected during routine workup of patients with flank pain. A report of a 37 years old lady who was treated as a case of ureteric stone but later investigations confirmed the presence of retrocaval ureter as the cause of her right flank pain is presented.

Case Report

A 37 years old female presented with intermittent pain of 6 months duration. Pain was mostly dull with intermittent exacerbations. She had no symptoms related to urination. She was otherwise fit and healthy with no co-morbidities and was not taking any medications. She was suspected of having stone in upper ureter. On general physical and abdominal examination, no abnormality was detected. Lab evaluation including urinalysis, complete blood counts, renal function tests, were within normal limits. Abdominal ultrasonography revealed moderate hydronephrosis and proximal hydroureter but no calculus was identified in ureter. The renal cortical thickness of affected kidney was variable but overall it was fair. The contralateral kidney was normal. No other pathology was observed on sonography. Plain X-ray KUB (kidney, ureter and bladder) was requested, it did not show any radio-opaque calculus in the line of ureter. The CT scan abdomen and pelvis was requested but as the Institution's CT scan machine was out of order, the retrograde pyelogram (RGP) was performed. The RGP depicted "J" shaped or "fish hook" right ureteropelvic junction with an associated hydronephrosis and medially deviated middle and distal ureter (Figure-1). On the basis of above findings, a diagnosis of right retrocaval ureter was made and surgery was planned. The right kidney and upper ureter was approached by right subcostal lumbar incision. There was moderate hydronephrosis and proximal hydroureter. The dilated right ureter was seen to be curving medially beneath the inferior vena cava (IVC). The ureter finally curved anteromedial to IVC and took a downward course. The distal ureter appeared normal. Careful mobilization of retrocaval ureter was achieved which exposed atretic and scarred portion of ureter posterior to IVC. The ureter

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Figure-1: Retrograde ureteropyelogram showing typical "fish hook ureter".



Figure-2: The ureter is seen crossing beneath the inferior vena cava from medial to lateral side.

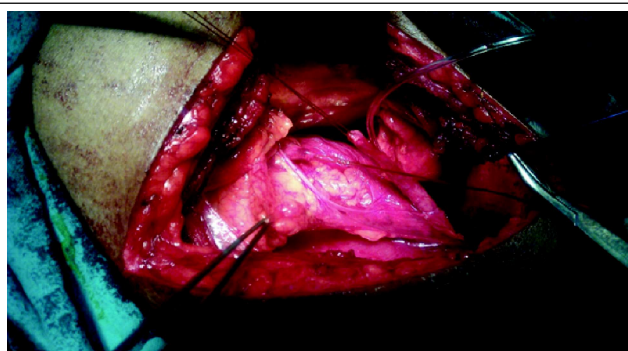


Figure-3: Ureter transected and transposed anterior to IVC.

was transected near pelvis and ureter was delivered on the medial aspect of IVC. The atretic and redundant portion

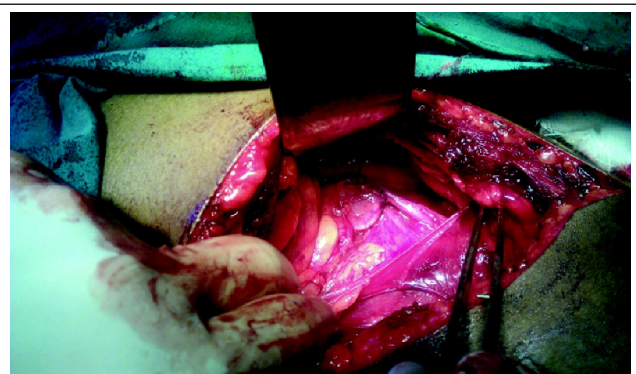


Figure-4: Ureteropelvic anastomosis completed over double "J" ureteral stent.

of the upper ureter was excised and an oblique ureteropelvic anastomosis was performed over double "J" ureteral stent (Figure-2-4). A tube drain was placed and wound was closed in layers. The postoperative course was uneventful. The drain was removed on 3rd postoperative day and patient was discharged the following day. The double "J" ureteral stent was removed at 4 weeks under local anaesthesia. The patient was followed up in clinic at 3 months. She was asymptomatic. Her urinalysis was normal and an abdominal ultrasonography revealed mild hydronephrosis. She was discharged from regular follow up.

Discussion

In the literature, retrocaval ureter has been classified into two clinical types, in accordance with the common radiological appearance. The more common Type 1 (also named "low loop"), is characterized by the so-called "typical S" or "fishhook" deformity of the ureter to the level of the obstruction. The point of obstruction is placed some distance from the lateral margin of the IVC at the level of the third lumbar vertebra. In the Type 2 variant (also called "high loop"), the ureter has a "sickle-shaped" curve. The point of obstruction is at the lateral margin of the IVC. Type 2 variant is rare, and represents around 10% of the known cases of retrocaval ureter.⁵

The imaging studies are important for an accurate preoperative diagnosis and successful surgical management. Abdominal ultrasound demonstrates hydronephrosis and proximal hydroureter without calculus. The intravenous urography (IVU) shows typical "fish hook" or "J" or "S" shaped deformity in the proximal dilated ureter with moderate hydronephrosis. However, an IVU may not demonstrate the middle or distal ureter

requiring retrograde ureteropyelogram (RGP) to visualize the ureter and confirm diagnosis. Nowadays, Computed Tomography Urography (CTU) is preferred over IVU or RPG as it may depict the anomaly in three dimensions giving the most "wholesome" solution to its diagnosis.⁶ However, other authors are of the opinion that MRI can nicely demonstrate the course of a retrocaval ureter better than CT.² MRI has high cost and is available in limited hospitals.

In terms of differential diagnosis, circumcaval ureter must be differentiated from retroperitoneal mass, retroperitoneal fibrosis, non-radio-opaque calculus or prior surgery. In retroperitoneal mass, the IVU often reveals lateral deviation of the ureter from the compression of the mass and in retroperitoneal fibrosis, the ureter is involved on both sides and the whole segment of the ureter is stiff in the IVU.

In patients diagnosed to have a retrocaval ureter, conservative surgery and periodical examination is recommended when there is no significant hydronephrosis, infection or stone formation. Surgery is required in symptomatic patients or in those with worsening kidney function. The standard repair of retrocaval ureter is open surgical pyelopyelostomy. The procedure involves identification and dissection of the ureter, dilated pelvis and IVC. The dilated renal pelvis is then transected. The ureter is transposed anterior to IVC and pyeloplasty is performed in a tension free, watertight manner over internal ureteral stent.⁷ The disadvantages of open surgery for retrocaval ureter are a large skin incision, significant postoperative pain and longer hospital stay.

With advancement in minimally invasive surgery, and experienced gained in laparoscopic approach, various authors have reported successful repair of retrocaval ureter. The procedure can be performed transperitoneally¹ or retroperitoneally.⁸

Compared with open surgery, the advantages of laparoscopic repair are minimal invasion and shorter hospital stay. Hemal et al⁹ published the first case of pure robotic repair of retrocaval ureter. The authors reported that robotic repair was associated with ease of dissection and intracorporeal suturing and apart from the ergonomic

and technical benefits, there was no other advantage over laparoscopic repair. Occasionally nephrectomy may be required in the presence of a nonfunctional kidney.¹⁰

Conclusion

This case highlights the importance of retrocaval ureter in the differential diagnosis of patients presenting with right flank pain. Multidetector CT scan, where available, remains the imaging of choice in diagnosing this rare anomaly. Surgical correction remains the standard of care to correct this anomaly. Although open surgery is usually employed but good outcomes have been reported with new minimally invasive laparoscopic and robotic techniques.

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