Retrocaval Ureter Causing Obstructive Hydronephrosis in a Pediatric Patient Managed with Minimal Invasive Surgery: A Case Report

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Abstract

Retrocaval ureter is a rare congenital anomaly leading to varying degrees of ureteral obstruction and potential renal deterioration, often necessitating surgical management. We report a case of an 11-year-old boy who presented with a 6-month history of intermittent right flank pain, computerized tomography urogram (CTU) confirmed the diagnosis of retrocaval ureter, demonstrating right-sided hydronephrosis and medial displacement of the right proximal ureter as it coursed posterior to the inferior vena cava (IVC) at the level of L4 vertebra. The patient underwent successful laparoscopic transperitoneal repair. The procedure involved incision of the renal pelvis, anterior transposition of the ureter relative to the IVC, and subsequent pyeloplasty over a double-J (DJ) stent. The postoperative course was uneventful, and follow-up demonstrated complete resolution of both symptoms and hydronephrosis. This case highlights the importance of early diagnosis and the efficacy of minimal invasive surgery in the management of retrocaval ureter, with favorable outcomes in preserving renal function.

Keywords: Retrocaval ureter, Hydronephrosis, Minimally invasive surgery (MIS), Pyeloplasty, Case report.

Introduction

Retrocaval ureter is a rare congenital vascular anomaly resulting from aberrant development of the inferior vena cava (IVC) or its embryonic precursors. As a result, the ureter passes posterior to the IVC, becoming compressed between the IVC anteriorly and the lumbar vertebral body posteriorly, causing varying degrees of functional ureteral obstruction and progressive deterioration of renal function if left untreated.[1] The anomaly was first described by Hochstetter in 1893 in a cadaver,[2] while the first clinically diagnosed case was reported by Harrill in 1940.[3] To date, more than 200 cases have been documented worldwide, with an estimated incidence of approximately 0.13%. The condition demonstrates a marked male predominance, with a reported male-to-female ratio of approximately 3:1.[4,5] The exact number of reported cases in Middle Eastern countries remains uncertain, as the available literature is limited to isolated case reports and small case series, most of which involve adult patients.[6,7] A review of the available literature revealed no previously reported cases of retrocaval ureter in the Sultanate of Oman. This likely reflects both the rarity of the condition and the possibility of underreporting.

The precise etiology of retrocaval ureter remains incompletely understood. However, some authors have proposed a potential association with maternal exposure to teratogenic substances such as monomethyl ether.[8] Retrocaval ureter may also coexist with a spectrum of congenital anomalies, including horseshoe kidney, ureteropelvic junction obstruction, duplication of the IVC, congenital absence of the vas deferens, hypospadias, supernumerary vertebrae, diverticula, anterior urethral calculi, renal agenesis, bilateral foot syndactyly, intestinal malrotation, and Goldenhar syndrome.[9]

We present a case of a symptomatic retrocaval ureter in an 11-year-old boy who was diagnosed and successfully managed with minimally invasive surgical approach at Sultan Qaboos University Hospital (SQUH) with excellent clinical outcomes. The study outlines the clinical presentation, imaging findings, and evaluates an individualized management strategy, emphasizing the role of minimally invasive approach. Informed consent for publication, including accompanying images, was obtained from the patient's parents. This manuscript was prepared following the CARE guidelines (https://www.care-statement.org).

Case Report

An 11-year-old boy with no significant past medical or surgical history presented with a 6-month history of intermittent right flank pain. There were no associated symptoms, including urinary complaints or constipation. There was no family history of renal anomalies, or urological disorders. Abdominal examination was unremarkable. Initial laboratory investigations were within normal limits. Complete blood count, blood urea and creatinine levels were normal. Urinalysis revealed microscopic urinary tract infection (UTI) . The patient was initially managed as a case of urinary tract infection (UTI); however, urine culture was negative.

Abdominal ultrasonography (US) demonstrated a mildly increased echogenicity of the right kidney, with grade IV hydronephrosis involving the right pelvis and calyces, without significant cortical thinning. The proximal ureter was markedly dilated, measuring 18 mm in diameter [Figure 1]. The left kidney exhibited normal echogenicity and preserved corticomedullary differentiation, with mild fullness of the renal pelvis, and no evidence of calculi. The urinary bladder was adequately distended, with no wall thickening or intraluminal calculi, and a post-void residual volume of approximately 10 mL.

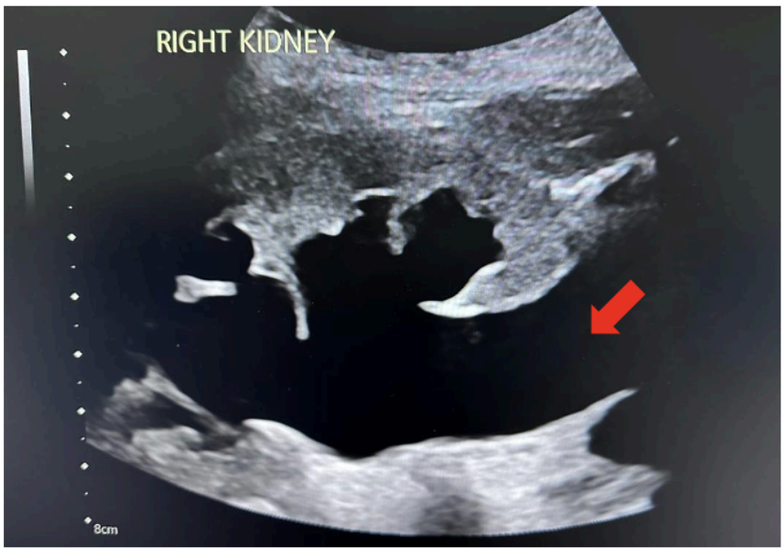

Figure 1: Renal ultrasound demonstrating grade IV hydronephrosis affecting the right renal pelvis and calyces, with significant dilatation of the proximal ureter (red arrow).

A renal MAG3 diuretic renogram revealed preserved right renal function, contributing 45% of total renal function, but demonstrated delayed tracer clearance with dilatation of the pelvicalyceal system and proximal ureter. The left kidney showed normal function and drainage, contributing 55% of total renal function [Figure 2]. These findings were suggestive of a proximal ureteric obstruction, with differential considerations including intrinsic or extrinsic ureteric pathology such as ureteric calculi, crossing vessels, or retrocaval ureter.

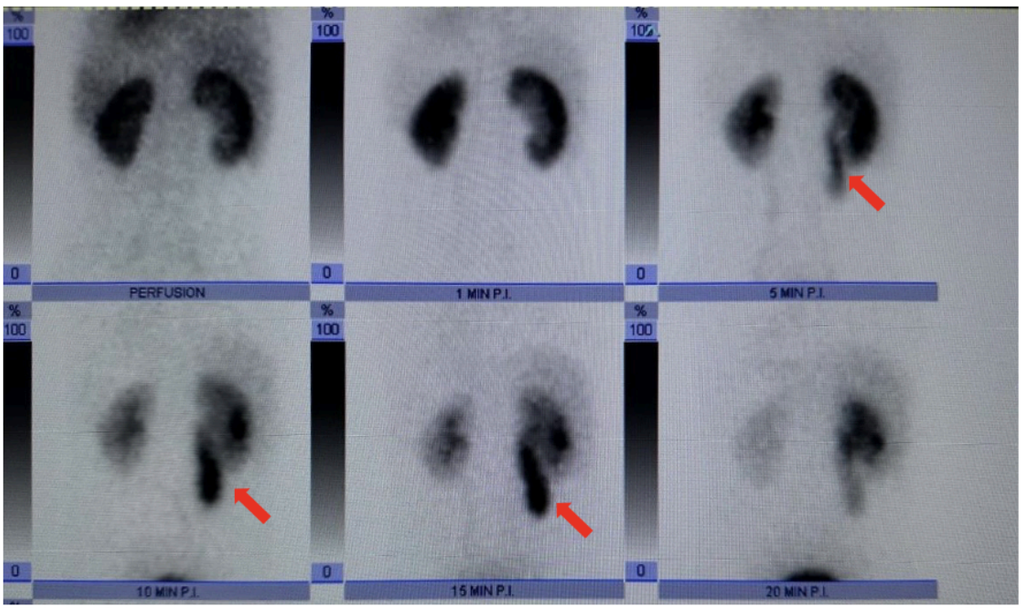

Figure 2: Renal MAG3 diuretic renogram demonstrating delayed tracer clearance, with associated dilatation of the pelvicalyceal system and proximal ureter (red arrow).

Subsequent CT urography confirmed right-sided hydronephrosis and demonstrated medial displacement of the right proximal ureter as it coursed posterior to the IVC at the level of the superior endplate of L4 vertebra. The distal ureter beyond this segment was of normal diameter, and both ureters inserted normally into the urinary bladder. These findings were consistent with a diagnosis retrocaval ureter [Figure 3].

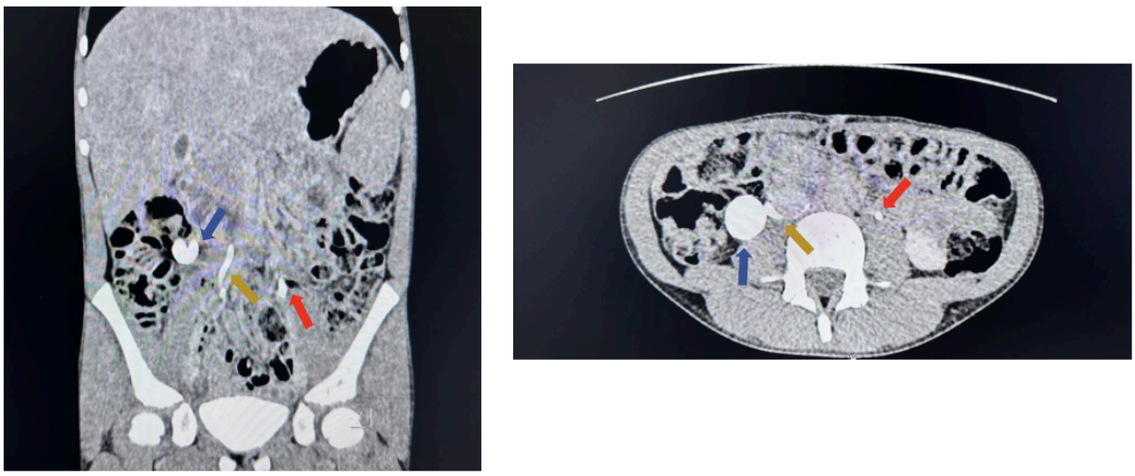

Figure 3: CT urography demonstrating right-sided hydronephrosis (blue arrow) and medial deviation of the proximal right ureter as it courses posterior to the IVC with a normal-caliber distal ureter (yellow arrow). The left ureter appears normal in both course and diameter (red arrow).

A laparoscopic transperitoneal pyeloplasty was performed under general anesthesia. Bladder catheter was inserted under aseptic technique. The patient was positioned in the right lateral decubitus position, with appropriate padding and placement of an axillary roll. Prophylactic Cefazolin was administered at induction. A 5 mm laparoscopic optic port was inserted transumbilical using Hasson open technique, two 3 mm working trocars were inserted under direct vision at epigastric and left lower quadrant. A transmesocolic window was created and the dilated right renal pelvis was identified [Figure 4a]. Careful dissection allowed identification and mobilization of the proximal ureter and IVC. The pelvic most dependent point was identified, and the pelvis was anchored at its highest point to the abdominal wall using 5-0 Prolene as transcoropral traction suture for exposure. The renal pelvis incision was made at the most dependent, and the ureter was repositioned anterior to the IVC. The ureter was incised at the normal-caliber segment, and cut ends were spatulated. A running suture of the posterior wall was taken to approximate the pelvis prominent point to the angle of spatulated ureter using 5-0 Vicryl RB-II. A 4 Fr x 14 cm double J (DJ) stent was placed antegrade prior to completing the anterior wall anastomosis [Figure 4b]. Standard closure of the port sites was done at the end of the procedure. Postoperative period was uneventful, urinary catheter was removed on postoperative day two, and the patient was subsequently discharged home in stable condition.

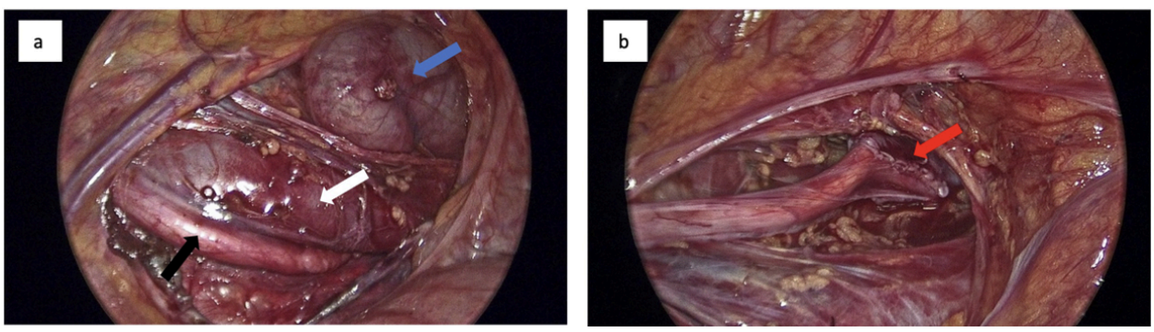

Figure 4: Intraoperative views. (a) The ureter coursing posterior to the IVC, with a dilated proximal segment (blue arrow), the IVC (white arrow), and a normal-caliber distal ureter (black arrow). (b) The ureter following transection, anterior transposition, and end-to-end anastomosis (red arrow).

The patient was followed for a total duration of 6-months postoperatively. At the 6-week follow up, the DJ stent was removed by cystoscopy. At 2-months post-procedure, the patient demonstrated significant clinical and radiological improvement, with resolution of flank pain and marked reduction in hydronephrosis. US showed a decrease in right ureteric diameter to 8 mm. After 3-months, the patient remained asymptomatic, and repeated US demonstrated further reduction in right ureteric diameter to 4 mm. Given the sustained clinical and imaging improvement, the patient was subsequently discharged from follow-up.

Discussion

Retrocaval ureter is a rare congenital vascular anomaly caused by aberrant development of IVC. Embryologically, the IVC develops through a coordinated process of development, anastomosis, and regression of the right vitelline, right subcardinal, and right sacrocardinal veins, which give rise to the hepatic (pre-renal), renal, and post-renal segments, respectively. Failure of regression of the right posterior cardinal vein that leis anterior to the right ureter in the lumbar region causes the ureter pass in a circumcaval course.[10] Left-sided retrocaval ureter is extremely rare and usually associated with other congenital anomalies such as persistent left cardinal vein, complete situs inversus, or IVC duplication.[1]

Although retrocaval ureter is a congenital condition, symptomatic cases are most commonly diagnosed in adulthood due to the progressive nature of hydronephrosis. The most frequently reported symptom is flank pain, however, patients may also present with recurrent UTI, hematuria, or urolithiasis. With advances in imaging modalities, incidental detection of retrocaval ureter has become increasingly common.[11]

Imaging is essential in the diagnosis of retrocaval ureter. Abdominal US is often the initial modality, typically demonstrating right-sided hydronephrosis and proximal hydroureter.[7] However, its ability to delineate the ureter's course relative to the IVC is limited, and it therefore serves primarily as a screening tool prompting further evaluation.[12] While intravenous pyelography (IVP) and retrograde ureteropyelography were traditionally used for diagnosis, CT urography is now the preferred modality due to its superior anatomical resolution and utility in preoperative planning. Magnetic resonance imaging (MRI) provides comparable diagnostic accuracy without ionizing radiation, though its use may be limited by availability, cost, and longer acquisition times. A characteristic "fish-hook" or "S-shaped" deformity of the ureter is characteristic on imaging.[13] Renal scintigraphy and diuretic renography are used to assess renal function and the degree of ureteral obstruction.[11]

Bateson and Atkinson classified retrocaval ureter into two types based on the level at which the ureter crosses the IVC on imaging. Type I (low loop) is more common variant, in which the ureter crosses the IVC at the level of L3-L4, and is characterized by marked ureteral dilation, typically demonstrating a reverse "J" or "S" shaped configuration. In contrast, type II (high loop) is less common, with the ureter crossing the IVC at a higher level (L1–2), usually associated with lesser ureteral dilatation and minimal hydronephrosis.[11]

The primary objective in the management of retrocaval ureter is to relieve ureteral obstruction, restore normal urine flow, and preserve renal function. Asymptomatic patients may be managed conservatively with regular imaging and monitoring of renal function. Ureteral reconstruction is indicated in symptomatic individuals, or in those with evidence of obstruction and declining renal function. The standard approach involves division of the ureter, anterior transposition relative to the IVC, and reanastomosis. This debate centers on the potential risk of injury to adjacent vascular structures, particularly the lumbar veins, which may necessitate conversion to open surgery. Preservation of the retrocaval segment is generally recommended when it appears macroscopically normal, exhibiting appropriate color, peristalsis, and caliber. Conversely, excision is favored when the segment is dysplastic, stenotic, kinked, or demonstrates abnormal peristalsis, as this may improve long-term outcomes. Nephrectomy may be indicated in symptomatic patients with a non-functioning kidney (≤10% function on renal scintigraphy) to prevent complications, particularly recurrent infections.[11]

Historically, open surgery—via lumbotomy or transabdominal approaches—was considered the standard treatment. However, the morbidity associated with large incisions has led to the widespread adoption of minimally invasive techniques, which offer advantages such as reduced postoperative pain, shorter hospital stays, and faster recovery. The transperitoneal laparoscopic approach provides excellent anatomical exposure and facilitates intracorporeal suturing, whereas the retroperitoneal approach allows direct access to the ureter and IVC without mobilization of intraperitoneal organs, thereby potentially reducing intra-abdominal complications. Nevertheless, the limited working space in the retroperitoneal approach may increase technical difficulty, particularly during suturing. Robotic-assisted surgery has also been described in several case series.[11] In the present case, a transperitoneal approach was chosen due to the aforementioned advantages, particularly as this represented the first procedure of its kind performed in the department. However, we believe that a retroperitoneal approach may be a feasible alternative in future cases. Despite the demonstrated efficacy of minimally invasive approaches, potential postoperative complications must be considered, including ureteral stricture, recurrent hydronephrosis, and urinary leakage. Meticulous surgical technique is essential to ensure precise ureteral reconstruction, avoid iatrogenic injury, and confirm that the ureter is adequately transposed anterior to the IVC prior to completion of the anastomosis.[11]

A retrospective study conducted between 2013 and 2023 in two Middle Eastern countries (Egypt and Saudi Arabia) reported three cases of adult right-sided retrocaval ureter. All patients underwent standard open ureteroureterostomy via a flank incision. In each case, the ureter was dissected proximal to the site of entrapment, followed by transposition anterior to the IVC and end-to-end anastomosis over a double-J stent. The duration of surgery was not specified. Follow-up at 6 weeks to 4 months demonstrated complete resolution of symptoms, with regression of both hydroureter and hydronephrosis in all patients.[7]

In another retrospective study conducted in China between January 2015 and September 2021, five pediatric cases of right-sided retrocaval ureter were managed using laparoscopic ureteral reconstruction via an extraperitoneal approach. The stenotic ureteral segment was resected, and ureteroureterostomy was performed. The reconstructed ureter was transposed anterior to the IVC and sutured over a a double-J stent. The operative time ranged from 130 to 243 minutes, with a mean duration of 180.2 minutes. Follow-up at 6 months and 2 years revealed complete symptom resolution and significant improvement in hydronephrosis.[12]

Conclusion

Retrocaval ureter is a rare congenital anomaly in which imaging plays a fundamental role in establishing the diagnosis, with CT urography being the modality of choice. Early diagnosis and timely surgical intervention are essential to relieve the obstruction and prevent irreversible renal damage. Surgical correction, most commonly through minimally invasive techniques such as laparoscopic or robotic ureteroureterostomy, represents the treatment of choice in symptomatic patients. These approaches provide superior visualization and operative precision, facilitating careful dissection and ureteral mobilization while offering the advantages of smaller incisions, reduced postoperative pain, shorter hospital stay, and faster recovery. In the present case, follow-up demonstrated favorable outcomes, including complete resolution of symptoms and significant improvement of hydroureter on repeat imaging. Long-term follow-up remains crucial for monitoring renal function and identifying potential complications.

Disclosure

Informed consent for the publication of this case has been obtained from the patient's parents.

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