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A Cause of Right-Sided Flank Pain: Retrocaval Ureter

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ABSTRACT

Background: The retrocaval ureter represents a rare congenital anomaly that occurs in 0.1% of the population and causes progressive ureterohydronephrosis, most of which are asymptomatic.

Case Report: In this case report, the retrocaval ureter detected in an adult male complaining of right-sided colicky flank pain is discussed.

Conclusion: This condition, whose main cause is an anomaly in the development of the vena cava, is easily diagnosed with imaging methods, and its symptoms can be completely corrected with surgical treatment.

Keywords: Retrocaval ureter, congenital abnormalities, hydronephrosis, chronic pain, diagnosis.

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INTRODUCTION

Retrocaval ureter (RCU) is an unusual congenital anomaly characterized by the passage of the right ureter behind the inferior vena cava (IVC). It is also called circumcaval ureter because it descends by going around the inferior vena cava from behind. This congenital anomaly was first described in a cadaver by Hochstetter in 1893.[1] Harrill made the initial clinical diagnosis in 1940.[2] Since it has been shared as case reports since its first description and comprehensive incidence studies are few, its prevalence cannot be stated with certainty, but it is estimated at 0.1%. There are studies worldwide reporting rates between 0.06% and 0.17%.[3] It is three times more common in men than in women.[4]

This anomaly arises during the 4th to 8th weeks of intrauterine development, stemming from an aberrant formation of the infrarenal inferior vena cava. Specifically, the abnormality involves the anteriorly positioned subcardinal vein giving rise to the IVC instead of the usual posteriorly located supracardinal vein. In a typical scenario, the infrarenal inferior vena cava derives from the dorsally positioned supracardinal vein. However, in cases where it develops from the ventrally located subcardinal vein, the ureter becomes entrapped posteriorly, resulting in a pre-ureteral vena cava. It is noteworthy that some authors prefer using the term "preureteral vena cava" as the root cause of this variant lies in a developmental abnormality of the vena cava rather than the ureter itself.

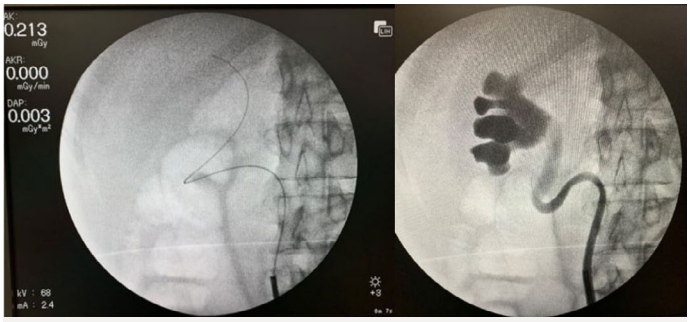

Figure 1. The path followed by the hydrophilic guidewire sent through the ureterorenoscope and "Fish hook" view of the ureter in retrograde pyelography.

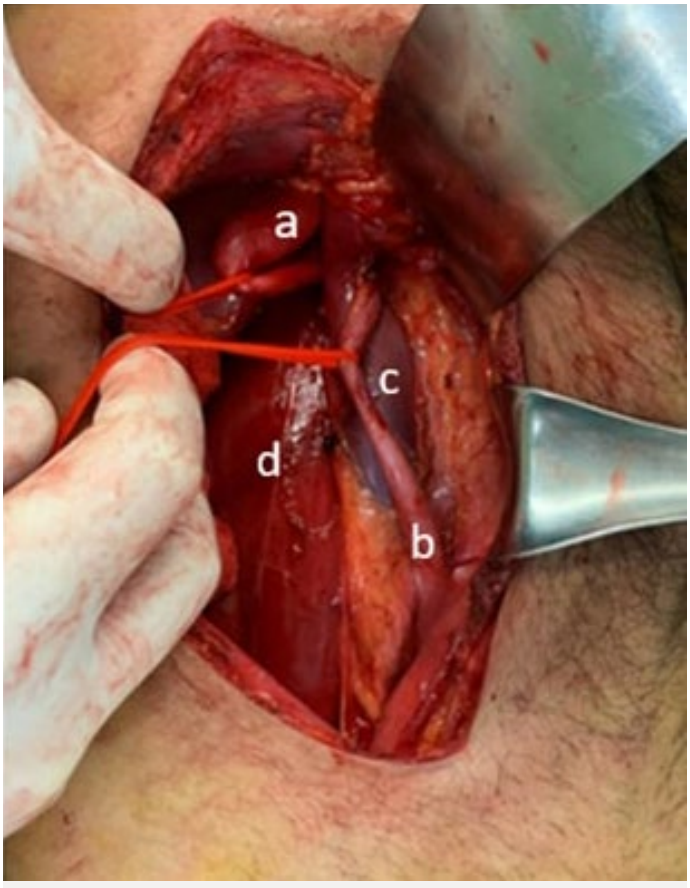

Figure 2. Image of the crossed ureter before ureteral incision: **(a)** Proximal part of the ureter. **(b)** Distal part of the ureter. **(c)** Inferior vena cava. **(d)** Psoas muscle.

Although most cases are clinically silent, it is associated with progressive ureterohydronephrosis and may present with various symptoms such as right-sided flank pain, renal colic, frequently recurring urinary infections, and hematuria.

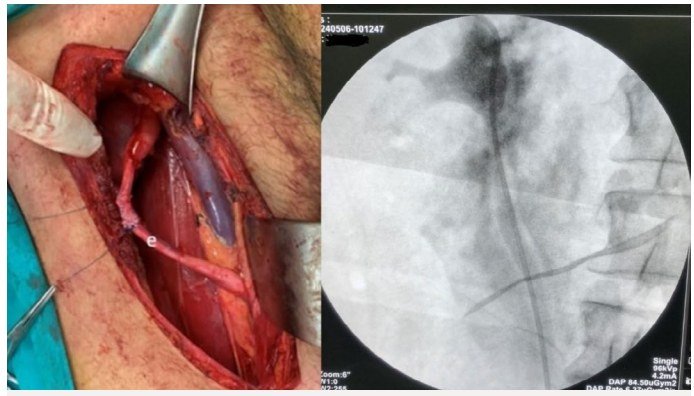

Figure 3. Image of the final state of the ureter: **(e)** Uncrossed anastomotic ureter and postoperative imaging of the right collecting system.

CASE REPORT

A 31-year-old male patient with no history of urological surgery presented with the complaint of right-sided flank pain for the last two years. Abdominal examination was normal. Laboratory analysis, including urinalysis and assessment of blood parameters such as full blood count, urea, creatinine, and electrolytes, demonstrated results that fell within the expected normal ranges. Abdominal ultrasonography revealed right-sided hydronephrosis. In computed tomography imaging, the right renal pelvis was seen in a grade 2 dilated impression, and the anterior-posterior diameter of the renal pelvis was 18 mm. The proximal one-third of the ureter was dilated, and hydronephrosis did not continue beyond this level. It was decided to perform Diagnostic Ureterorenoscopy and Retrograde Pyelography. The imaging showed that the ureter narrowed after the middle segment of the right ureter. Opaque material was administered at this level, and right hydroureteronephrosis and a "fish hook" deformity were observed in the proximal ureter (Fig. 1).

The condition of RCU was diagnosed. Although opaque material and a hydrophilic guidewire passed through the narrow segment, a stenosis was observed that did not allow the passage of the rigid thin Ureterorenoscope (URS), and a decision for surgery was made. Right subcostal lumbotomy was performed, the RCU was reached by exploration, and the ureter was dissected (Fig. 2). The excessive retrocaval segment was maneuvered and surgically removed, the ureteral narrow segment was resected, and the uretero-ureteral anastomosis was performed using Vicryl 3/0 around a 4.8 Fr 24 cm double J stent (Fig. 3). A closure procedure was then executed in three planes, with the placement of a drain in the right renal compartment. Postoperative imaging is shown in Figure 3. Postoperatively, the patient's follow-up was uneventful, culminating in the removal of the ureteral catheter on the

30th day after the surgery. Notably, the patient's symptoms ameliorated during the follow-up period. Hydronephrosis was not observed in the postoperative checks.

DISCUSSION

As mentioned before, the main cause of this rare congenital anomaly is not a developmental anomaly of the ureter but the abnormal formation of the IVC. It occurs because the IVC originates from the anteriorly located subcardinal vein instead of the posteriorly located supracardinal vein. Hence, it is also recognized as preureteral vena cava. It usually progresses silently and is detected incidentally with the help of various imaging studies. It most commonly occurs in the 3rd and 4th decades with symptoms associated with upper urinary tract obstruction: flank pain, abdominal pain, hematuria, urinary infection, and stone formation. It most commonly occurs with urinary infection in children.[5]

The diagnosis of RCU is commonly established through imaging methods like intravenous urogram (IVU), retrograde pyeloureterogram (RGP), or computed tomography (CT) scans. Magnetic Resonance Imaging (MRI) may be considered a preferred alternative to CT scans, as it provides a radiation-free depiction of the ureter and inferior vena cava and can delineate their courses more effectively.[6] Additionally, a nuclear renal scan proves valuable in assessing the degree of obstruction and evaluating renal function.

In 1976, Kenawi and Williamsen proposed an anatomical classification system for RCU, categorizing it into two types based on the height of the retrocaval segment of the ureter. This classification relies on radiographic observations and identifies distinct locations of ureteral narrowing.[7] Also called Type 1 Low Loop, the majority of cases fall into this group. The ureter dilates until it crosses the ureteral vena cava from below. The specific appearance of this group is called an "S" or "fish hook" deformity. Type 2 crosses the vena cava at a higher level, and the renal pelvis and ureter appear horizontal. It is defined as High Loop because it crosses high. It has a sickle-shaped smooth curve. It constitutes 10% of cases.[8] In Type 2, hydronephrosis and symptoms are less severe, and therefore, the need for surgery is less frequent than in the other group. Our case was in the Type 1 group and required surgery due to urinary obstruction.

Conservative surgery plays a predominant role in treating symptomatic cases. The commonly employed technique involves ureteral uncrossing to restore the continuity of the excretory pathway. In Type 2 cases, this is typically accomplished through direct plasty and end-to-end uretero-ureteral anastomosis, while in Type 1 cases, as in ours, resection-anastomosis of the retrocaval segment is performed. Various authors have outlined laparoscopic and robotic reconstructive techniques, advocating these as preferred methods in surgically addressing RCU. This approach presents multiple advantages compared to traditional open surgery, including its minimally invasive nature, early mobilization, short hospitalization, avoidance of complications such as postoperative wall pain or gastrointestinal ileus, and resulting in a shorter overall recovery period.[9]

CONCLUSION

RCU has increasing reporting rates with the rise in imaging methods. This anomaly, which is mostly asymptomatic, typically appears as low back pain in symptomatic patients. It is easily diagnosed with imaging methods, and its symptoms can be corrected with surgical treatment, leading to full recovery.

Informed Consent: Written informed consent was obtained from patients who participated in this study.

Author Contributions: Concept – İEE, Design – AÖ; Supervision – HS; Resource – AFV; Materials – AÖ; Data Collection and/or Processing – İEE; Analysis and/or Interpretation – İEE; Literature Search – AÖ; Writing – AFV; Critical Reviews – HS.

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