Retrocaval ureter: Report of two cases and literature review

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Retrocaval ureter is a rare congenital anomaly in which the ureter passes behind, and is compressed by, the inferior vena cava. Its etiology is assumed to be abnormal embryologic development of the inferior vena cava as a result of atrophy failure of the right subcardinal vein in the lumbar portion. We report two cases of retrocaval ureter and review the relevant literature. One patient was a 7-year-old boy who presented with right flank pain. The other was a 40-year-old male who was found to have right hydronephrosis accidentally on abdominal sonography during a health examination. Both underwent retrograde pyelography which showed the typical S-shape of a retrocaval ureter. Abdominal computerized tomography and magnetic resonance imaging confirmed the diagnosis of retrocaval ureter. Ureteroureterostomies were performed. One patient showed focal squamous metaplasia of the ureter and the other had chronic inflammation and fibrosis. Follow-up studies showed improvement in hydronephrosis and renal function in both patients. We conclude that retrocaval ureter is a rare disorder and surgical correction is usually effective.

**Key Words:** retrocaval ureter, computerized tomography, MRI


Retrocaval ureter is a rare embryologic developmental anomaly. The ureters develop from the mesonephros, which travels with the kidney through the lumbar venous ring. Six venous channels in the lumbosacral region are noted in the embryo. In man, atrophy of the posterior cardinal vein, which is one of the six veins, should occur in early fetal development. If the posterior cardinal vein persists, retrocaval ureter results. Here, we report two cases of retrocaval ureter and review the relevant literature.

**CASE PRESENTATIONS**

**Case 1**

A 7-year-old boy presenting with right flank pain was admitted after right hydronephrosis was detected using abdominal sonography. Laboratory examination was unremarkable. Retrograde pyelography (RP) revealed an S-shaped ureter (Figure 1). Magnetic resonance imaging (MRI) showed right hydronephrosis and hydrourereter; the right ureter deviated medially and was dorsal to the inferior vena cava (IVC) (Figure 2). 131I renogram showed delayed excretion of radioisotope in the right kidney. Under the impression of retrocaval ureter, ureteroureterostomy was performed and the pathologic report showed focal squamous metaplasia of the resected redundant ureter. Postoperative convalescence was smooth and right hydronephrosis improved during follow-up (Figure 3).
Case 2
A 40-year-old male was noted to have right hydronephrosis incidentally on abdominal sonography during a routine physical examination. Intravenous urography showed a normal left kidney and moderate right hydronephrosis with a reversed J-shaped ureter (Figure 4). RP revealed the same picture. A 26 cm double-J was inserted smoothly to give a typical S-aspect in the retrocaval ureter. Abdominal computerized tomography (CT) clearly showed that the ureter passed behind the IVC (Figure 5). Renal scan also showed obstruction. We performed ureterolysis and ureteroureterostomy during which a stenotic segment and dilated proximal ureter were found (Figure 6). Histopathologic examination of the resected stenotic ureter revealed chronic inflammation and fibrosis. The patient recovered well during follow-up.

**Figure 1.** Retrograde pyelography shows the typical S-shaped course of the ureter. Marked hydronephrosis and hydroureter are also noted.

**Figure 2.** Magnetic resonance imaging reveals hydronephrosis and hydroureter in which the ureter deviates medially to the inferior vena cava (arrows).

**Figure 3.** Abdominal sonography reveals that the right hydronephrosis has improved postoperatively.

**Figure 4.** Intravenous urography shows right hydronephrosis with a reversed J-shaped ureter.
Retrocaval ureter

DISCUSSION

The first recorded case of retrocaval ureter was seen on autopsy and was described by Hochstetter in 1893 [1]. Retrocaval ureter, also termed circumcaval ureter or preureteral vena cava, is an anomaly of embryologic development of the IVC in which the ventral infrarenal subcardinal vein persists, forcing the more dorsally placed ureter to pass behind it [2]. In a postmortem series, Nielsen reported retrocaval ureters in 0.9 of 1,000 cases [3].

The incidence of retrocaval ureter is higher in men than in women (2.8:1), and most patients do not present with symptoms until the third or fourth decade of life [4]. Symptoms depend on the degree of ureteral obstruction or the presence of complications. Intermittent flank pain is often noted as the first complaint. Occasionally, recurrent urinary tract infection, hematuria, pyelonephritis, or stone formation is noted [5]. In our first case, focal squamous metaplasia of the ureter was discovered. However, no malignant change associated with retrocaval ureter has been reported.

Different diagnostic modalities, including intravenous pyelography, RP, inferior vena cavagraphy, abdominal sonography, abdominal CT, and MRI, have been utilized. With regard to cost effectiveness and invasiveness, abdominal CT may be the diagnostic procedure of choice [4]. However, MRI can be used if the patient has poor renal function.

In 1982, Bergman classified retrocaval ureter into two clinical types [6]. Type I (low loop) is the most common, with the dilated proximal ureter assuming the shape of a reverse “J”. Usually, this type of ureter is obstructed. Type II (high loop) is seen less frequently. The ureter passes behind the IVC at the level of, or just above, the pelvic-ureteral junction. This type of ureter is frequently not obstructed. Both of our cases belonged to type I and showed obstructive uropathy; surgical intervention was indicated.

Retrocaval ureter may co-exist with a solitary kidney or other congenital anomalies, including contralateral renal hypoplasia or ectopia [7], horseshoe kidney [8, 9], aberrant renal artery [2], Turner’s syndrome [10], Goldenhar syndrome [11], retroperitoneal fibrosis [12], and polycystic disease of the kidneys [13]. In our two cases, no concomitant anomalies were found.

Surgical correction most commonly involves ureteral division, stenotic and redundant ureter resection with relocation, and ureteroureteral or ureteropelvic reanastomosis. It can be accomplished by a conventional intra-abdominal or retroperitoneal approach, or the newly developed laparoscopic approach [14]. Both of our patients underwent conventional retroperitoneal ureteroureterostomy.

\(^{131}\)I renogram has been used clinically in preoperative evaluation of obstructive uropathy and

Figure 5. Computerized tomography reveals right hydronephrosis and hydroureter with medial deviation. Two segments of the ureter are also noted on each side of the vena cava (arrows).

Figure 6. Laparotomic view of the right ureter passing posterior to the vena cava (white arrow) shows mobilized ureter, stenotic segment (curved arrow), and dilated proximal ureter (black arrow).
the degree of postoperative recovery through measuring individual glomerular filtration rate. Surgical correction is indicated only when obstruction or complications are present.

CONCLUSION

Although retrocaval ureter is a relatively rare disorder, it should be suspected when imaging studies reveal the S-shape of the right ureteral orientation. Abdominal CT is the diagnostic procedure of choice and surgical correction is indicated when obstructive uropathy is evident.

REFERENCES