An unusual case of hydronephrosis: Retrocaval ureter

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ABSTRACT

Retrocaval ureter, circumcaval ureter, or most accurately pre-ureteral vena cava is a rare vascular anomaly involving the ureter. It is believed to be due to the persistence of the right subcardinal vein. As a result, the right ureter deviates medially and goes dorsal to the inferior vena cava, crossing over it and resumes its ordinary course distally, causing varying degrees of hydroureteronephrosis. We describe a case report of a middle-aged female who was a known diabetic, presented with the right flank pain and fever. On abdominal examination, she had tenderness in the right renal angle. Her laboratory investigations were normal except for elevated glucose levels. Urine analysis showed *Escherichia coli* infection, which was treated with antibiotics. On imaging, revealed right retrocaval ureter with Grade IV hydronephrosis. She underwent the right laparoscopic ureteroureterostomy through the transperitoneal approach. Retrocaval ureter is one of the rare causes of hydronephrosis. The treatment is a straightforward procedure, has a good outcome.

Key words: *Circumcaval ureter, Hydroureteronephrosis, Retrocaval ureter*

Retrocaval ureter or circumcaval ureter is a rare congenital anomaly. It is more accurately termed pre-ureteral vena cava due to the aberration in the development of inferior vena cava (IVC), wherein there is the persistence of the right posterior cardinal vein [1]. As a result, the ureter courses medially and passes behind the IVC winding around and crossing the front of it from medial to lateral side to reach the bladder. Patients may be asymptomatic or present with proximal hydronephrosis mimicking congenital pelviureteric obstruction. It is of two types with type 1 presenting with marked hydronephrosis [2]. Hydronephrosis in retrocaval ureter is described as a consequence of ureteral compression by psoas muscle, spinal column, and IVC. Occasionally, calculi may form above the obstruction.

We, thus, describe a middle-aged female patient presenting with the right hydronephrosis with the rare cause being retrocaval ureter. This case report highlights the possibility of retrocaval ureter being one of the differential diagnoses in the evaluation of the right-sided hydronephrosis.

CASE REPORT

A 47-year-old female from Kerala, known case of diabetes mellitus, presented with the right flank pain for 2 months, and low-grade fever for 1 week, not associated with chills and rigors. The pain was a dull-aching type, non-radiating, not referred, no aggravating factor, and relieved on taking over the counter analgesics. She did not complain of hematuria, burning micturition, or lower urinary tract symptoms.

On general physical examination, she was moderately built, well-nourished. The vitals were stable. On abdominal examination, minimal tenderness in the right renal angle was noted. A provisional diagnosis of the right pyelonephritis was made.

She was evaluated with complete blood count, the total leukocyte count was normal (5400 cells/cubic mm). Blood urea (16.7 mg/dl) and serum creatinine (0.69 mg/dl) were normal. Random blood glucose level was elevated (259 mg/dl). Urine analysis was normal, except for the presence of glucose. Urine culture showed the presence of *Escherichia coli*, for which culture-specific intravenous antibiotic was given.

Ultrasound of abdomen and pelvis showed the right kidney pelviureteric junction calculus 2.9×1.9 cm with severe hydronephrosis; the left kidney was normal. X-ray of the kidney ureter bladder was essentially normal. Computed tomography (CT) scan of the abdomen and pelvis with IV contrast revealed the right proximal ureter to be dilated and seen up to the level of the lower border of the third lumbar vertebra beyond which it was coursing in the retrocaval region with no evidence of dilation, with Grade IV hydronephrosis. Two calculi measuring 19×13 mm and 13×7 mm were noted in the dependent portion of lower pole calyces. With the CT scan of the abdomen suggesting the possibility of the retrocaval ureter, a retrograde pyelography was planned.

Retrograde pyelography showed the classic "fishhook" sign confirming the diagnosis (Fig. 1). The diethylenetriaminepentaacetic acid scan was not done as the excretory function of the kidney was maintained. The endocrinologist evaluated her for the diabetes mellitus. After obtaining informed consent, she underwent the right laparoscopic ureteroureterostomy through the transperitoneal approach. Intraoperative findings being dilated tortuous right upper ureter traversing behind the IVC with distal ureter being normal in caliber (Fig. 2). The two calculi were noted at the time of ureterostomy and were extracted. Ureteroureterostomy was done over a double-J ureteric stent. The post-operative period was uneventful, and the patient was discharged on the 5th post-operative day. After 1 month, cystoscopy was done, and double-J ureteric stent was removed.

DISCUSSION

The retrocaval ureter is a rare, congenital urologic anomaly. Hochstetter first described it in 1893. The incidence has been reported as 1 in 1500 cadavers [1], with a prevalence of 0.13% [3]. The male:female ratio is 2.8:1 [1]. The right side is commonly involved, with the left side occurring in situs inversus or double IVC. Although it is a congenital anomaly, it presents at the third to fourth decade of life as was in this case. Few cases have been reported in children [4]. A vast majority of patients are asymptomatic or present with flank pain, recurrent urinary tract infections, hematuria, and features of obstruction. Renal calculi and pyonephrosis may complicate the condition. In 1969, Bateson and Atkinson classified retrocaval ureter into two types: Type 1 wherein the ureter crosses the level of the third lumbar vertebra, and there is marked hydronephrosis and type 2 is less common and characterized by the ureter crossing at the level of the renal pelvis. In our case, it was a type 1 retrocaval ureter.

The differential diagnosis of the retrocaval ureter may include conditions displacing the ureter from its ordinary course such as retroperitoneal mass or retroperitoneal fibrosis and obstructive lesion mimicking congenital pelvic ureteric junction obstruction. The diagnosis of the retrocaval ureter can be confirmed by

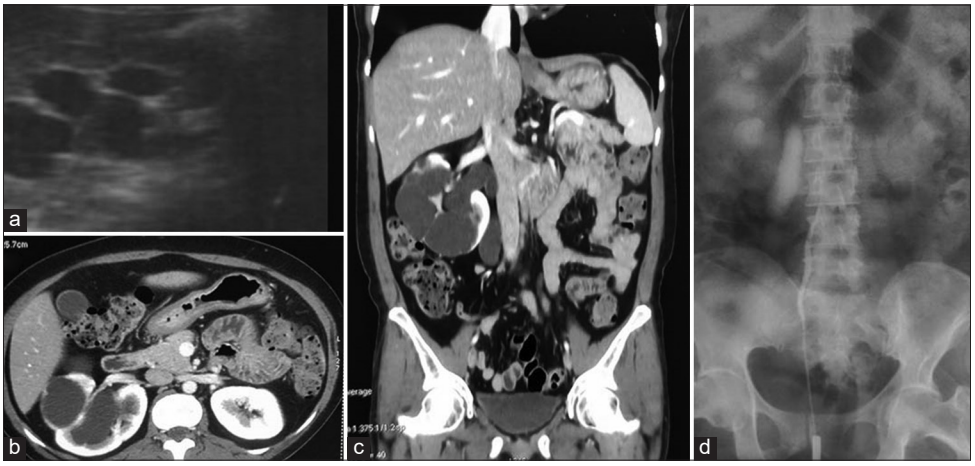

Figure 1: (a) Ultrasound showing the right gross hydronephrosis. (b and c) Computed tomography showing inferior vena cava and right ureter coursing behind it. The right side Grade IV hydronephrosis. (d) Retrograde pyelography showing "fishhook sign"

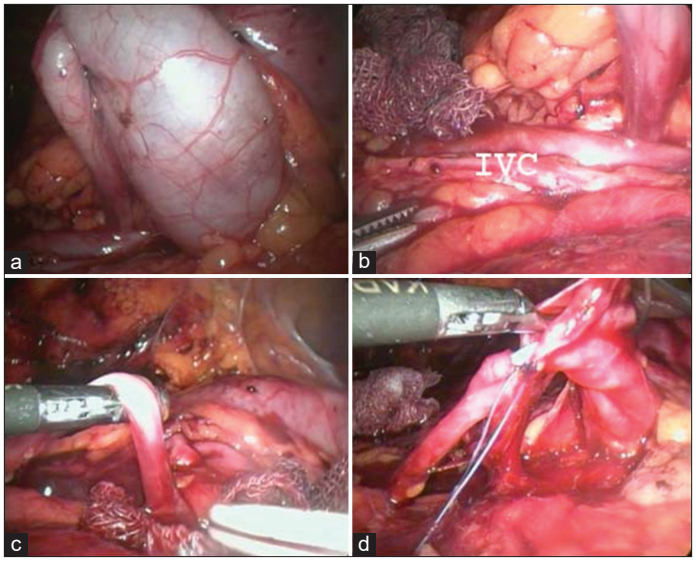

Figure 2: Intraoperative images: (a) The dilated proximal part of the right ureter. (b) Distal ureter traced going behind the inferior vena cava. (c) Anteriorization of the right ureter. (d) Ureteroureterostomy over the double-J stent

imaging. Retrograde pyelography describes the "classic fishhook sign" [5]. Ultrasound of the abdomen and pelvis would reveal hydronephrosis, but it poorly delineates the ureter. CT scan is the investigation of choice as both the ureter and IVC are better visualized. Magnetic resonance imaging may be preferred in comparison to other imaging modalities as it is less invasive and is without any radiation exposure.

Treatment is by conservative management in asymptomatic patients. Intervention is indicated in patients with functionally significant obstruction causing pain or renal function deterioration. In this case, the patient had Grade IV hydronephrosis and pain, which demanded a surgical intervention. Surgical treatment involves an open or laparoscopic approach, the steps of which include resection, relocation, and reanastomosis of the ureter [6]. Conventionally, open repairs were a gold standard for many years. In 1949, Anderson and Hynes first reported the successful dismembered pyeloplasty technique for a case of retrocaval ureter [7]. In 1994, Baba *et al*. reported the first successful laparoscopic pyeloplasty for retrocaval ureter [8]. With the advances in technology and growing experience, the laparoscopic approach through transperitoneal or retroperitoneal route is considered the best as it reduces post-operative pain, shortens the length of hospital stay, aids early recovery, and provides cosmetically better scar [9]. Patients who have undergone treatment generally have an uneventful recovery and excellent prognosis.

A high index of suspicion is essential to diagnose this condition preoperatively. With the increasing awareness among clinicians, these cases are being reported, and with the advances in technology, the approaches have been minimally invasive over the years [10].

CONCLUSION

The retrocaval ureter is a rare cause of hydronephrosis, which may mimic as congenital pelvic ureteric junction obstruction. While considering the differential diagnosis for causes of hydroureteronephrosis, retrocaval ureter must be considered.

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