

Retrocaval Ureter: A Rare Cause of Hydronephrosis

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Abstract

Retrocaval ureter or circumcaval ureter or postcaval ureter is a rare venous congenital anomaly where the vena cava compresses the ureter, causing varying degrees of hydronephrosis. We report one case of retrocaval ureter in 34-year-old women who presented with right lumbar pain. The diagnosis was made on radiographic imaging, which showed right hydronephroureterosis. During surgery, retrocaval anomaly was noticed. It was successfully treated by ureteral dissection, excision of the retrocaval segment and uretero-ureteral anastomosis.

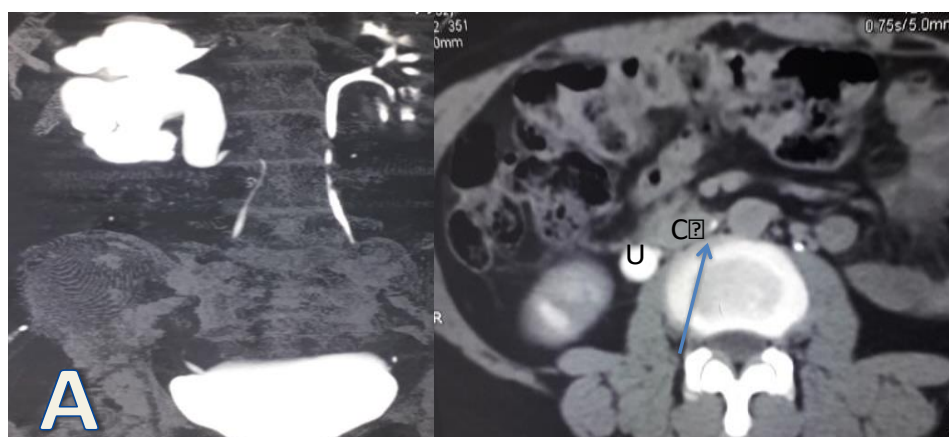
Keywords: Retrocaval ureter; Hydroureteronephrosis, surgery

Introduction

Retrocaval ureter is a rare congenital venous anomaly of inferior vena cava [1]. It is characterized by the passage of ureter posterior to inferior vena cava (IVC), from dorsal lateral position above to ventral medial position below; it causes upper urinary tract obstruction. Hochstetter reported the first case in 1893 [2]. More than 200 cases have been reported in the literature [3,4]. We present a woman who presented with flank pain and was diagnosed to have right hydronephroureterosis on radiographic imaging. Open surgery is the treatment of choice.

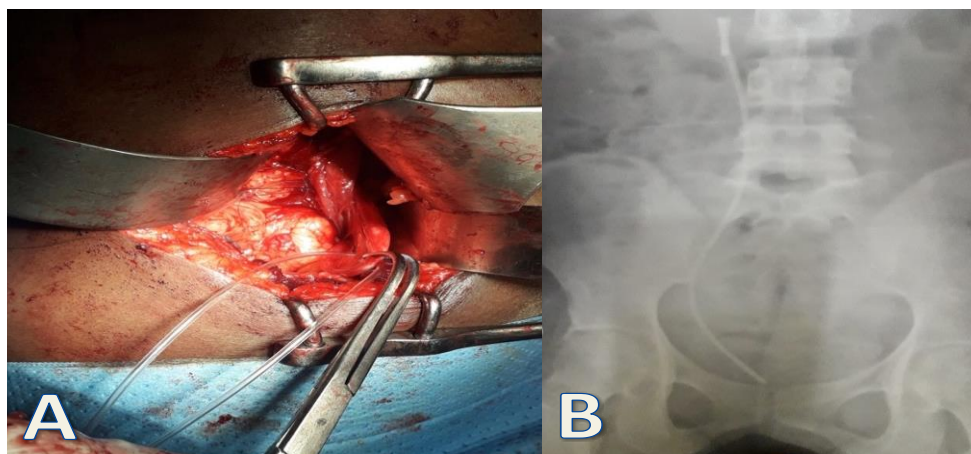
Clinical presentation

A 34-year-old woman came to our outpatient department with intermittent lumbar pain she has experienced for a week. There were no other systemic complain. Physical examination revealed tenderness at the flank. Haematological workup was normal. His serum creatinine was 5 mg/dl and urine analysis were negative. Ultrasonography showed right hydronephrosis. Computed tomography (CT) scan after intravenous pyelography study revealed right hydronephrosis with dilatation of the upper ureter above the crossing of IVC and a narrowing of the urethra distal to the crossing of IVC (Figure 1).



CT scan : (A) right ureter with a dilated upper portion with a 'fish-hook' deformity at this level. (B) and the ureter passed behind the inferior vena cava and exited between IVC and aorta.

The ureteral segment passing behind the IVC and exiting between IVC and aorta was also shown (Figure 2).



(A) The surgical management
(B) RX ASP showed the normal course of the ureter containing catheter JJ.

Left kidney was normal. The patient underwent open surgical intervention via right lombotomy incision. On exploration the ureter was found deep behind the inferior vena cava. The entire upper ureter was mobilized and dissected; the ureter was divided and placed anterior to the IVC prior to and end-to-end anastomosis over a 4 fr double J stent. Trans urethral Foley catheter was removed at postoperative day 6. Postoperative course was uneventful. The ureteral stent was removed 1 month later. He was clinically well after the operation.

Discussion

Retrocaval ureter is a rare congenital anomaly [1,2]. The prevalence of disease is reported to be 1 in 1000 live births [5]. Its incidence is greater in males than females with a sex ratio of 2.8:1[6].

Postcaval ureter is a rare congenital venous anomaly due to the ureter passing posterior to the inferior vena cava. Its etiology is presumed to be the abnormal persistence of the subcardinal vein in embryologic development of the inferior vena cava (IVC). The IVC results from the evolution of three paired and symmetrical systems: posterior, subcardinal and supracardinal cardinal systems. During embryological development, metanephros migrates proximally from the renal pelvis and normally passes between the posterior supracardinal and cardinal veins. The lower segment of the lower caval vein consists of supracardinal veins while the cardinal posterior veins undergo atrophy. The persistence of the posterior cardinal vein is the key element of this malformation. A retrocaval ureter on left is seen only with persistence of left cardinal vein or with complete situs inversus [1,6].

Several cases of retrocaval ureter are reported in autopsy (500- 1500) [1]. Patients seldom report symptoms until the third or fourth decades of life. Common presentations

include flank pain, recurrent urinary tract infections, stone formation and microscopic or macroscopic haematuria [8,9,10].

Ultrasonography, antegrade and retrograde pyelography with inferior venocavography, intravenous urogram (IVU) were commonly used to diagnose circumcaval ureter. Currently, CT scan and magnetic resonance imaging (MRI) have been considered the best efficacious methods of confirming the diagnosis, least invasive with inherent ability to describe the lesion in a three-dimension mode [10-13].

In the literature retrocaval ureter has been classified into two types in accordance with the radiographic appearance and the site of ureter narrowing [5].

The **Type I** is more common. The ureter is in normal position until the height and usually crosses behind the IVC at the level of the third or fourth lumbar vertebra and IV pyelography study reveals a typical “fish-hook” shaped deformity of the ureter, displaying a figure of inverted J or S over the place where is the obstruction [5,7]. The ureter passes then behind the lower vena cava, bypasses her and appears on its median edge. The obstruction causes the dilation of the upper urinary tract proximal to the level of lateral side of IVC.

The **Type II** is less common. The Post caval segment of the ureter crosses higher at the level of the renal pelvis [5]. Various techniques for the management of retrocaval ureter have been reported [11-13]. Treatment primarily is based on clinical presentations, grade of hydronephrosis and existence of impairment of renal function. In patients with no subjective symptoms and no hydronephrosis, surgical correction is not mandatory, therapeutic

abstention is justified. Periodical examination has been required.

When either obstructive symptoms or kidney function caused by circumcaval ureter worsen, surgical correction is indicated to preserve renal function and to provide long-term symptomatic relief [8-13].

Various operative methods proposed for retrocaval ureter include ureteroureteral reanastomosis, with or without resection of retrocaval segment, ligation or transection of inferior vena cava and nephrectomy. Open surgery is the standard treatment and is usually successful.

It consists in realising a surgical transection of ureter at pelvis, dissection of the ureter anteriorly from the IVC, with transposition and uretero-ureteral anastomosis. Most of the authors that suggested an ureteropelvic anastomosis also advocated for Harril's method [3] by which a section is made at the level of the pelvis just above the ureteropelvic junction.

Laparoscopic or retroperitoneoscopic ureterolysis and reconstruction has become popular in recent years with satisfactory success rate, less intraoperative bleeding, early return to routine activities, minimal pain, cosmetically acceptable surgical scar and shorter convalescence time while preserving therapeutic efficacy [11,12].

Conclusion

IVC abnormalities and circumcaval ureter are rare. CT scanner and MRI are currently effective options to determine the anatomic relationship of the IVC and ureter. Surgical intervention is often necessary. Laparoscopic surgery offers many advantages, including a less invasive approach.

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