Case Report

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Retrocaval ureter: a rare cause of urinary tract obstruction

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ABSTRACT

Retrocaval ureter is an uncommon cause of ureteric obstruction due to a rare congenital anomaly of inferior vena cava. We report a case of 23 year old male who presented with right flank pain and was diagnosed with ultrasound and intravenous urography. He underwent open retroperitoneal ureteroureterostomy.

Keywords: Retrocaval ureter, Flank pain, Intravenous urography, Ureteroureterostomy

INTRODUCTION

Retrocaval ureter also referred to as circumcaval ureter is a rare congenital anomaly wherein ureter passes posterior to vena cava causing obstruction of upper urinary tract leading to hydronephrosis.¹ Despite its congenital etiology, symptoms appear in 3rd or 4th decade of life.² Diagnosis requires high index of suspicion and if symptomatic, may require surgical intervention.³

CASE REPORT

A 23 year old male presented with right flank pain since three months, which was gradual in onset and dull aching in nature. Patient had no other complaints and no history of operative intervention in the past. There were no significant findings on general and per-abdominal examination. Complete blood counts, urine analysis, blood urea and creatinine levels were normal.

Abdominal ultrasound reveals a right renal gross hydronephrosis with dilated right upper ureter having constriction 3 cm distal to PUJ (pelvic urteric junction). Intravenous urography showed right sided hydronephrosis with characteristic "fish hook" shape of ureter along with medial deviation. The findings were consistent with right retrocaval ureter (Figure 1).

Patient operated with right subcostal incision and abdominal wall opened in layers and retroperitoneum approached, operative findings revealed right hydronephrosis with dilated right upper ureter with stenosed retrocaval part of ureter and normal lower ureter. Proximal and distal part of ureter was cut and spatulated, ureteroureterostomy was performed to restore ureteral continuity over Double-J stent with vicryl 4-0. Retroperitoneal drain was placed and wound was closed in layers (Figure 2).

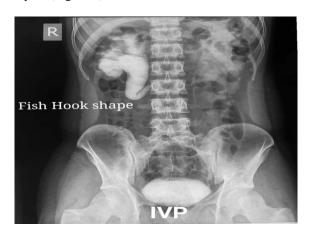


Figure 1: Intravenous pyelography showing fish hook shape of ureter.

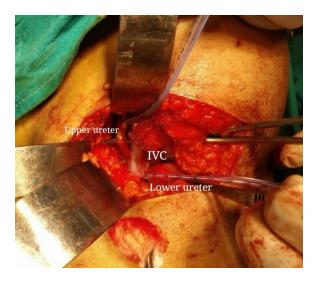


Figure 2: Intraoperative picture of ureter showing presence of ureter behind inferior vena cava.



Figure 3: Post-operative X-ray KUB showing right ureteric DJ stent after correction.

Post-operative period was uneventful. The drain was removed on 3rd post-operative day and the patient was discharged on 5th post-operative day. Removal of double – J stent was carried out after 4 weeks. In follow up patient was asymptomatic with normal kidney and ureteric function with sterile urine (Figure 3).

DISCUSSION

Retrocaval ureter is a rare congenital anomaly that arises from dysgenesis of inferior vena cava and should be more appropriately called as pre-ureteral vena cava.⁴ Retrocaval ureter was 1st described by Hochstetter in 1893 but the first case of retrocaval ureter was published by Anderson and Hynes in 1949.⁶ The incidence is approximately 1 in 1500 people with a 3:1 male preponderance but a few clinical cases have been reported worldwide.⁷

During fetal development the pre-renal, renal and postrenal segments of inferior vena cava develop from the right vitelline vein, right subcardinal and right sacrocardinal veins respectively. During the growth of fetus, instead of right subcardinal vein as renal segment of inferior vena cava; right posterior cardinal vein persists, leading to retrocaval ureter. Retrocaval ureter is also called as circumcaval ureter as right posterior cardinal vein lies ventral to ureter; the ureter effectively comes to lie posterior to vena cava.⁴

The anomaly mostly involves the right ureter, as was reported in this case. If it involves the left ureter then it is usually associated with either partial or completes situs inversus or duplication of inferior vena cava.⁸

The proposed probable causes for abnormal development of inferior vena cava are maternal exposure to diethylene glycol and monomethyl ether which are used as industrial solvent.

Based on classical imaging findings, two types of retrocaval ureter have been described which are: type 1 or low loop, the more prevalent variety where in the level of obstruction to the ureter is usually a little farther from the lateral margin of aberrantly developed inferior vena cava at level of 3rd lumbar vertebrae. The ureter shows a sharp medial swing from this point curving up to the pedicle of vertebral body thus resembling a "fish hook" or S-shaped appearance on intravenous urography. Depending on amount of compression due to the aberrant vessel a varying degree of hydronephrosis and hydroureter can be present.¹⁰

Type 2 or high loop variety comprises only 10% of the cases and intravenous urography reveals a smooth "sickle shaped" curve of right ureter with the level of obstruction at lateral margin of 3rd lumbar vertebrae.¹⁰

Most of the patients remain asymptomatic but those with symptoms commonly present with flank or abdominal pain that can be intermittent, dull aching and is commonly due to ureteric obstruction and associated hydronephrosis. Some patients may also present with recurrent urinary tract infection and haematuria. Renal calculi and pyonephrosis may complicate the condition.

Ultrasound of abdomen is non-invasive and an initial investigation, used mostly to demonstrate hydronephrosis and ureteric obstruction. Intravenous urography shows dilatation of renal pelvis, calyces and upper ureter showing "reverse J" or typical "fish hook" appearance but cannot demonstrate middle and lower ureter. Hence, it can be combined with antegrade and ascending urography to confirm the diagnosis but is an invasive procedure. ¹¹⁻¹³

Abdominopelvic CT scan is helpful in excluding these conditions. Spiral CT scan may define ureter and inferior vena cava anomalies and is considered as an investigation

of choice. MRI can demonstrate course of preureteral vena cava and may be more detailed and less invasive imaging modality without exposure to radiation. ¹⁴ In this case, the diagnosis was made using intravenous urography.

Treatment depends primarily on clinical symptoms, severity of hydronephrosis and impairment of renal function. Patients who are asymptomatic, without hydronephrosis, infection or urolithiasis and with no renal impairment can be followed up conservatively with periodic examinations. ¹⁵

Surgical management includes both open and laparoscopic approach which includes division of dilated renal pelvis with transposition and reanastomosis, ureteroureterostomy over double-J stent with or without resection of stenotic retrocaval segment and ligation or transection of inferior vena cava with or without reanastomosis. Some patients may require nephrectomy if kidney is non-functioning. ¹⁵ In this case ureteroureteral renastomosis anterior to vena cava was done.

Transperitoneal or retroperitoneal laparoscopic ureterolysis and reconstruction of retrocaval ureter are a time consuming and technically demanding procedure. It is associated with a satisfactory success rate, less intraoperative bleeding, early return to normal activity, minimal pain and a cosmetically better scar. ^{2,15,16} Robotic approach to retrocaval ureter was 1st published for paediatric patient by Gundeti et al in 2006. ¹⁷

CONCLUSION

Retrocaval ureter is a rare congenital anomaly which occurs due to abnormal development of inferior vena cava which is diagnosed incidentally and can be confirmed with CT scan and needs to be corrected surgically unless patient is asymptomatic. Surgical repair is associated with excellent prognosis.

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