CASE REPORT

Retrocaval ureter presenting at 6 years of age in a girl child — An extreme rarity

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Abstract
Retrocaval ureter is one of the very rare congenital anomalies. We report a case of retrocaval ureter in a 6-year-old girl who presented with right flank pain and hydronephrosis. The diagnosis was made on intravenous urography which showed typical “J” shape deformity in the proximal dilated ureter with moderate hydronephrosis. CT scan delineated the course of ureter. The patient was operated and findings were confirmed. The ureter was transected near the pelvis and a pyeloureteric anastomosis with pre-caval transposition of the ureter was performed and the patient was discharged in fair health.

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1. Introduction

Retrocaval ureter, also referred to as pre-ureteral vena cava is a rare congenital anomaly in which the ureter deviates medially and passes behind the inferior vena cava (IVC), winding around and crossing in front of it from medial to lateral side. Though it is a congenital anomaly, patients usually present with symptoms like right lumbar pain in 3rd and 4th decades of life from a resulting hydronephrosis, but our patient developed symptoms at a very young age of 6 years and was managed successfully. In view of the rarity of this anomaly, that too in a girl child, presenting at a very young age, the case is being presented here with review of literature.

2. Case report

A 6-year-old female child came to our out-patient department with complaint of pain in the right flank that was of sudden onset and initially colicky but had become constant now and it was moderate in intensity. There was no

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haematuria. The blood pressure at presentation was 106/74 mmHg. There were no significant findings on physical examination. Haematological reports read: haemoglobin 133 g/L, blood urea nitrogen 3.93 mmol/L and serum creatinine 36.24 mmol/L. Routine examination of urine was normal. On microscopic examination, there were 0–2 white blood cells and 0–1 epithelial cells per high power field but no red blood cell was detectable. Abdominal ultrasonography revealed a right hydronephrosis and proximal hydroureter but no calculus. So a presumptive diagnosis of right pelviureteric junction obstruction was made. Intra-venous urography showed delayed excretion of the contrast and subsequently a typical “J” shaped or “fish hook” deformity in the proximal dilated ureter. It also showed moderate hydronephrosis with non-visualization of the rest of the right ureter (Fig. 1). On diethylene triamine penta acetate renal scan, the right kidney had a differential renal function of 32.14% and glomerular filtration rate of 2.48 mL/min while the left kidney had differential renal function of 67.86% and glomerular filtration rate of 68.58 mL/min. A CT scan of the abdomen revealed right sided hydronephrosis with non-visualization of middle and distal ureter on ipsilateral side on urographic phase. It excluded any extrinsic lesion as a possible cause of the above findings. A diagnosis of a symptomatic right retro-caval ureter was made and the patient was taken up for an open surgery through a right loin oblique incision. The intraoperative findings confirmed the diagnosis of right retrocaval ureter and associated hydronephrosis (Fig. 2). The right ureter was divided at the lateral border of the IVC and distal part dissected free from the IVC. The redundant distal segment of right renal pelvis and the ureter coursing behind the IVC were excised. Pyeloureteric anastomosis was done over a double-J ureteric stent with vicryl 4-0 interrupted sutures anterior to the IVC (Fig. 3). The postoperative period was uneventful and the patient was discharged on 5th postoperative day. The double-J ureteric stent was removed after 3 weeks. The patient is on regular follow-up and asymptomatic as per the serial ultrasound abdomen reports and there is no evidence of any hydronephrosis on the ipsilateral side since then.

Figure 1  Intravenous pyelogram showing a typical “J” shaped or “fish hook” deformity in the proximal dilated ureter with moderate hydronephrosis with non-visualization of the rest of the right ureter.

Figure 2  Feeding tubes around the right ureter with the ureter coursing behind the inferior vena cava.

Figure 3  Pyeloureteric anastomosis over a double-J ureteric stent anterior and lateral to the inferior vena cava.
3. Discussion

Retrocaval ureter is a rare embryological anomaly involving the venous system and resulting in the ureter coursing behind and being obstructed by the IVC. It was first reported by Hochstetler in 1893 [1]. The incidence of retrocaval ureter is 1 in 1500 cadavers and male to female ratio is 3 or 4 to 1 [2].

The anomaly predominantly involves the right ureter, as observed in this reported cases. A retrocaval ureter on left is seen only with persistence of left cardinal vein, with complete situs inversus or duplication of the IVC [3,4].

Majority of the symptomatic patients present with flank or abdominal pain that can be intermittent, dull and aching and is commonly due to ureteric obstruction and associated hydronephrosis. Some patients may present with recurrent urinary tract infection and haematuria which can be microscopic or even gross haematuria. Renal calculi and pyonephrosis may complicate the condition. Some cases are found incidentally during radiographic imaging for other conditions. Abdominal ultrasound demonstrates hydronephrosis. IVU usually does not demonstrate the middle and distal ureter requiring a retrograde ureteropyelogram to demonstrate the ureter and hence confirm the diagnosis. Spiral CT scan may define the ureter and IVC anomalies obviating the need for a retrograde ureteropyelogram and is considered an investigation of choice.

Retrocaval ureter is classified into two types based on its radiographic appearance and the site of narrowing of ureter [5,6]. Type I is more common. In this, the ureter crosses behind the IVC at the level of the third lumbar vertebra and has a “fish hook” shaped (“S” shaped or reversed “J” shaped) deformity of the ureter at the point of obstruction. Marked hydronephrosis is seen in 50% of the patients. In Type II, cross-over occurs higher at the level of the renal pelvis and there is lesser degree of hydronephrosis or none at all. In this, renal pelvis and upper ureter lie nearly horizontal before encircling the vena cava in a smooth curve (sickle shaped curve).

The hydronephrosis may be due kinking of the ureter, adynamic ureteric segment or compression against the psoas muscle. The various anomalies associated with retrocaval ureter are horse shoe kidney, double IVC and left retrocaval ureter with Goldenhar syndrome [7,8].

The clinical diagnosis of retrocaval ureter cannot be excluded sheeerly on the basis of age and it can present even in a very young child as is evident in our case. Once diagnosed, prompt surgical intervention is the answer as renal function deteriorates very fast in children. Treatment is surgical transection of ureter at pelvis, dissection of the ureter anteriorly from the IVC, anteriorisation and uretero-ureteral anastomosis. There may be severe hydronephrosis. Anderson Hynes pyeloplasty with precastal transposition of the ureter has been advocated and the same was done in our case. Occasionally nephrectomy may be required in the presence of thinned out cortex, poor function or severe infection [9]. The other modalities of treatment used are percutaneous nephrostomy, ureteric stenting and retroperitonioscopic dissection of ureter, transection and ureteric reanastomosis using automatic suture device [10].

4. Conclusion

Retrocaval ureter is a rare congenital anomaly that usually presents late in the 3rd and 4th decades of life but sometimes it can present as early as 6 years of age as in our case. Therefore, in every child presenting with right lumbar pain, the treating clinician should have a high degree of clinical suspicion and retrocaval ureter should always be considered one of the differential diagnoses and patient should be investigated and managed accordingly. Any delay in surgical intervention is likely to jeopardize the renal function in symptomatic children. Therefore, early and prompt surgery should be the rule in symptomatic children who have been diagnosed with retrocaval ureter.

Conflicts of interest

The authors declare no conflicts of interest.

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