

Preureteral Venacava: An Unusual Cause of Ureteral Obstruction

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ABSTRACT

Preureteral venacava is a rare congenital anomaly in which ureter passes behind the inferior venacava. It is due to abnormal persistence of right subcardinal vein such that the ureter is trapped posterior to it. We present a case of 35 years old male who presented with right flank pain. Diagnosis was made by Computed Tomography (CT) urography which showed proximal ureteral dilatation with “J” shaped deformity. Open surgical repair resolved the symptoms.

KEY WORDS: Preureteral venacava; Retrocaval ureter; Ureteral obstruction, Venous anomaly.

INTRODUCTION

Preureteral venacava is a rare congenital abnormality in which ureter passes behind the inferior vena cava causing ureteral obstruction. Earlier this condition was known as retrocaval or postcaval ureter (as initially it was thought of as an anomaly of ureteral development); however these terms are anatomically descriptive but developmentally misleading. It is a venous anomaly and the basic pathology is the abnormal persistence of right subcardinal vein (instead of the right supracardinal vein) forming the main portion of the inferior venacava, which traps the ureter dorsal to it.¹ The first observed case of Preureteral venacava was reported by Hochstetter in 1893,² while first case of surgical repair was described by Anderson and Hynes in 1949.³

Although it is a congenital anomaly, patients usually present with symptoms at 3rd and 4th decades of life from a resulting hydronephrosis.⁴ Intravenous urography, retrograde pyelography, Computed Tomography (CT), and Magnetic Resonance Imaging

(MRI) are main diagnostic investigations. Surgical intervention is required in most symptomatic of cases. Exploration of ureter, its transaction and end to end anastomosis anterior to venacava can be performed by open method, but more recently laparoscopic and robotic techniques are becoming more popular.

CASE REPORT

A 35 years male presented with right flank pain for three years. Pain was mild to moderate, dull aching, intermittent and associated with occasional burning micturition. No abnormality was found on general and abdominal physical examination. Routine investigations were normal. Ultrasound of abdomen showed right hydronephrosis and proximal hydroureter. CT urography revealed moderate proximal hydroureteronephrosis with ureter lying posterior to inferior venacava at the level of forth lumbar vertebra, showing “S” or “J” shaped deformity (Figure 1). Patient operated under general anesthesia by open method. Retrocaval ureter was transected, redundant part excised, ureter transposed anteriorly, both ends spatulated, and end to end anastomosis of ureter and pelvis (Uretero-pyelostomy) was performed keeping double J stent (Figure 2 and 3). His symptoms were relieved and ultrasound study after 12 weeks demonstrated regression of hydronephrosis.

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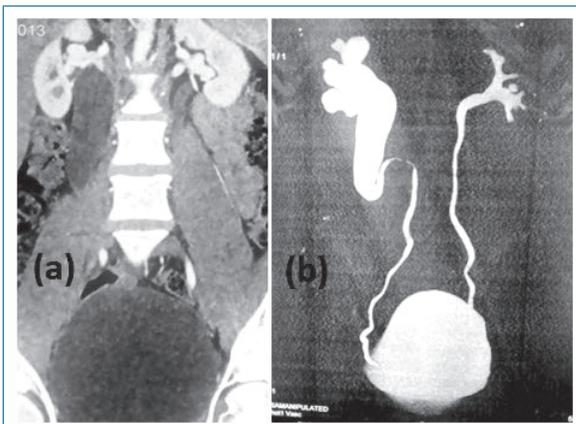


Figure 1: The CT urography findings suggestive of retrocaval ureter: proximal dilated ureter (a) and (b), and “fishhook sign” (b).

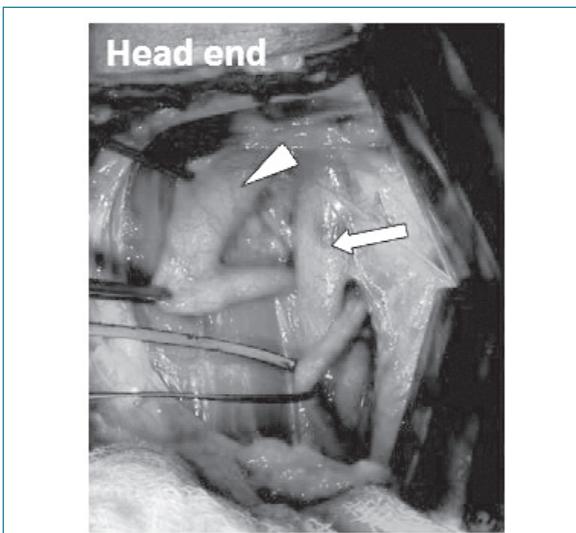


Figure 2: Intra-operative picture showing dilated proximal ureter (arrowhead) and inferior venacava anterior to segment of ureter (arrow).

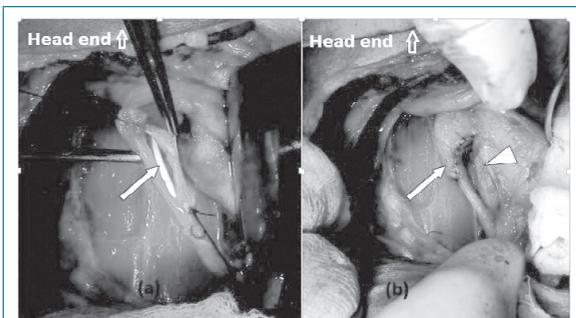


Figure 3: After transaction and excision of retrocaval segment, ureter was transposed anteriorly and anastomosed. (a), ureteral stent (arrow); (b), funnel shaped anastomosis (arrow) adjacent to inferior venacava (arrowhead).

DISCUSSION

The prevalence of the preureteral venacava is reported to be 1 in 1000 live births.⁴ It is three to four times more common in males than females. This abnormality is mostly observed in the right side, but in some cases such as situs inversus or duplication of inferior venacava, it may be in left side.^{4,5} Here, the ureter deviates medially behind the inferior venacava, winding about it and crossing in front of it to resume a normal course distally to the bladder. Based on radiological appearance and site of narrowing, it can be classified into two clinical types.⁶ Type I is more common (90%) and typically shows obstructed pattern, where the Ureter crosses behind the inferior vena cava at the level of the third lumbar vertebra and has fish hook shaped (S shaped) deformity at the point of obstruction. Significant hydronephrosis is seen in 50% of the patients. In type II (10%), the upper ureter is not kinked and crosses the venacava at higher level such that renal pelvis and retrocaval segment lie almost horizontal and exhibits sickle shaped deformity. There is lesser degree of hydronephrosis or none at all and thus are less symptomatic.

Preureteral venacava is associated with other congenital anomalies in up to 21% of cases. These are mainly related to the cardiovascular and urogenital systems such as horseshoe kidney, ureteropelvic junction obstruction, congenital lack of the vas deferens, hypospadias, extra vertebra, diverticulum, kidney agenesis, syndactyly, intestinal malrotation, and Goldenhar syndrome etc.⁷

The symptoms are because of ureteral obstruction and include flank pain, infection, stone formation, hematuria etc. Imaging studies are sufficient for its diagnosis and include urtrasonography, excretory urography, retrograde urography, CT urography and MRI. Three dimension CT urography is regarded as the procedure of choice to confirm the diagnosis. Some has suggested MRI as less invasive modality than CT and retrograde pyelography with more detailed information.⁸

Asymptomatic cases with minimal hydronephrosis require no treatment, but should be followed up regularly. Surgical correction involves ureteral division, anterior transposition, excision of redundant pelvis and retrocaval segment (which can be aperistaltic), and ureteroureteral or ureteropelvic reanastomosis. Open

surgical repair is the standard procedure and the most commonly performed one. However, minimal invasive techniques are emerging as the treatment of choice with encouraging result. Laparoscopic and robotic reconstruction of ureter via both transperitoneal and retroperitoneal approaches has been described.^{9,10} A double-J stent is placed into ipsilateral ureter cystoscopically beforehand and kept for four to six weeks postoperatively. Minimal invasive repairs have advantages as compared to open surgery in terms of minimal postoperative morbidity, early convalescence and short hospital stay. Occasionally nephrectomy may be required when there is presence of thinned out cortex, poor function or severe infection.

Our patient had symptomatic type I Preureteral venacava which was diagnosed with CT urography. Open surgical repair improved patient symptomatically and radiologically.

CONCLUSION

Preureteral venacava is a rare congenital venous anomaly due to persistence of right subcardinal vein. Though congenital, patients become symptomatic in their third or fourth decades with flank pain. CT urography is the diagnostic modality of choice, which shows typical fishhook or "S" shaped deformity and also the retrocaval segment. Although open surgical repair is the standard of care in symptomatic cases, minimally invasive method may be the preferred approach in future.

REFERENCES

1. Lerman I, Lerman S, Lerman F. Retrocaval ureter: report of a case. *J Med Soc N J* 1956;53(2):74–7.
2. Hochstetter F. Beitrage zur entwicklungsgeschichte des venen-systems der amnioten: III. Sauger Morph Jahrb. 1893;20:542.
3. Anderson JC, Hynes W. Retrocaval ureter: A case diagnosed pre-operatively and treated successfully by a plastic operation. *Br J Urol* 1949;209-14.
4. Rubinstein I, Cavalcanti AG, Canalini AF, Freitas MA, Accioly PM. Left retrocaval ureter associated with inferior venacaval duplication. *J Urol* 1999;162:1373-4.
5. Watanabe M, Kawamura S, Nakada T, Ishii N, Hirano K, Numasawa K et al. Left preureteral vena cava (retrocaval or circumcaval ureter) associated with partial situs inversus. *J Urol* 1991;145:1047–8.
6. Bateson EM, Atkinson D. Circumcaval ureter: a new classification. *Clin Radiol* 1969;20(2):173–7.
7. Perimenis P, Gyftopoulos K, Athanasopoulos A, Pastromas V, Barbalias G. Retrocaval ureter and associated abnormalities. *Int Urol Nephrol* 2002;33(1):19-22.
8. Uthappa MC, Anthony D, Allen C. Case report: retrocaval ureter: MR appearances. *Br J Radiol* 2002;75(890):177–9.
9. Miyazato M, Kimura T, Ohyama C, Hatano T, Miyazato T, Ogawa Y. Retroperitoneoscopic ureteroureterostomy for retrocaval ureter. *Hinyokika Kyo* 2002;48(1):25–8.
10. Hemal AK, Rao R, Sharma S, Clement RGE. Pure robotic retrocaval ureter repair. *Int Braz J Urol* 2008;34:734-8.