

CASE REPORT

RETROCAVAL /CIRCUMCAVAL URETER: RARE CONGENITAL ANOMALY OF URETER OR INFERIOR VENA CAVA

Samir M Shah¹, Chirag K Patel², Smit M. Mehta², Vikram B Gohil³

Author's Affiliations: ¹Professor & Head; ²Resident doctor; ³Associate Professor, Department of General Surgery, Govt. Medical College, Bhavnagar

Correspondence: Dr Chirag K Patel Email: chiragkpatel89@gmail.com

ABSTRACT

Retrocaval ureter also referred to as pre-ureteral vena cava is a rare congenital anomaly with the ureter passing posterior to the inferior vena cava and coming medial to it. Though it is a congenital anomaly, patients do not normally present with symptoms until the 2nd and 3rd decades of life from various presenting complain resulting due to hydronephrosis. We present a case reported in Bhavnagar, Gujarat; a 19-year-old male presented with right flank pains of 2 yr and associated right moderate hydronephrosis. Diagnoses were confirmed with intravenous pyelography (IVP) and computed tomography of abdomen with IVP. And patient was treated with open surgery, including resection of stenosed retrocaval ureter and spatulated end to end uretero-ureterostomy in front of IVC.

Key words: Retrocaval ureter, Circumcaval ureter, Flank pain, Hydronephrosis, uretero-ureterostomy

INTRODUCTION

Retrocaval ureter also referred to as circumcaval ureter or preureteral vena cava is a rare congenital anomaly with the ureter passing posterior to the inferior vena cava. The ureter classically course medially behind the inferior vena cava winding around it and then passes laterally in front of it to then course distally to the bladder. Though it is a congenital anomaly, patients do not normally present with symptoms until the 2nd and 3rd decades of life, with various complain resulting due to back pressure changes leads to hydronephrosis. The hydronephrosis may be due to kinking of the ureter, a ureteric segment that is adynamic or compression of inferior vena cava. It was initially considered as aberration in ureteric development; however current studies in embryology have led to it being considered as an aberration in the development of the inferior vena cava.^{1,2} Hence it is being suggested that the anomaly be referred to as a pre-ureteral vena cava.³

CASE HISTORY

A 19 year old male patient, presented with history of right flank pain since 2 year, and taking analgesics from general practitioner for right flank pain, gradually pain was increase. Patient had no any other complains and no operative intervention in the past. There were no significant findings on general and per

abdominal examination. Full blood count, urinalysis and blood urea and creatinine were normal. Abdominal ultrasonography revealed a right moderate hydronephrosis and right upper hydroureter. An intravenous pyelography showed right moderate hydronephrosis and hydroureter of the proximal ureter with non-visualization of the rest of the right ureter with normal left kidney and ureter (figure 1). As we were suspecting a benign cause of ureteric stricture or external compression of ureter, patient was subjected to computed tomography with intravenous pyelography which was suggestive of right circumcaval ureter with right hydronephrosis and right upper hydroureter (figure 2). Patient was undergone laprotomy and excision of retrocaval stenosed segment of right ureter and spatulated end to end uretero-ureterostomy with double J stent kept insitu. Post operative course was normal. Patient was discharge from hospital on 4th post operative day. Stitch removal on 8th post operative day. Double J stent removed on 21th post operative day. Follow up ultrasonography after 2 month showed normal, no hydronephrosis and hydroureter. With normal renal function test.

DISCUSSION

Retrocaval ureter is a rare congenital anomaly occurring with incidence of about 1 in 1500 people with a three to four times male predominance in autopsy

studies.⁴ Though few clinical cases have been reported worldwide. The first observed case of retrocaval ureter was described by Hochstetter in 1893⁵. Though initially thought of as an anomaly of ureteric development studies in embryology has revealed an anomaly related to the development of the inferior vena cava. The appropriate term giving the correct description of the anomaly is preureteral vena cava. The anomaly predominantly involves the right ureter, as was observed in this reported case. If it involves the left ureter then it is usually associated with either partial or complete situs inversus or duplication of the inferior vena cava (IVC).⁶ The ureter typically deviates medially behind the inferior vena cava, winding about and crossing in front of it from a medial to a lateral direction, to resume a normal course, distally, to the bladder. The renal pelvis and upper ureter typically appear elongated and dilated in a "J" or fishhook shape before passing behind the vena cava. Although it is a congenital anomaly it normally presents in the second and third decade of life as typified by the ages of the presented cases. Majority of patients presenting with symptoms, 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. Renal calculi and pyonephrosis may complicate the condition. Some cases are found incidentally during radiographic imaging for other conditions.

Retrocaval ureter classify into two clinical types. Type 1 is commonest and has moderate to severe hydronephrosis in about 50% of cases with extreme medial deviation of middle ureteric segment and the ureter assuming an S or 'fish hook' deformity. Type 2

has less medial deviation of the ureter with mild or no associated hydronephrosis and forms about 10% of cases⁷. Surgical management is reserved for the type 1 cases that are usually symptomatic. Retrocaval ureter has hence been defined as a rare congenital anomaly that requires surgical correction in the symptomatic patient.

Abdominal ultrasound demonstrates hydronephrosis. IVU usually does not demonstrate the middle and distal ureter may require a retrograde ureteropyelogram to demonstrate the ureter. Spiral CT scan may define the ureter and inferior vena cava anomalies obviating the need for a retrograde ureteropyelogram and is considered an investigation of choice. Important differential diagnosis includes retroperitoneal fibrosis and retro peritoneal masses displacing the ureter from its normal course. Abdomino pelvic CT scan is helpful in excluding these conditions. MRI can nicely demonstrate the course of a preureteral vena cava and may be a more detailed and less invasive imaging modality, without exposure to radiation, when compared with CT and retrograde ureteropyelography. Treatment is surgical and involves division of the ureter and repositioning it anterior to the inferior vena cava. This may be achieved through an anastomosis between the renal pelvis and the ureter or a uretero-ureteric anastomosis over a double-J stent. The segment behind the inferior vena cava which may be aperistaltic is either excised or left in situ. In this reported case, the segment was excised. Surgical intervention is for symptomatic cases and changes of hydronephrosis and altered renal function. Patients with minimal caliceal dilatation and no significant symptoms do not need surgery but need to be followed up.

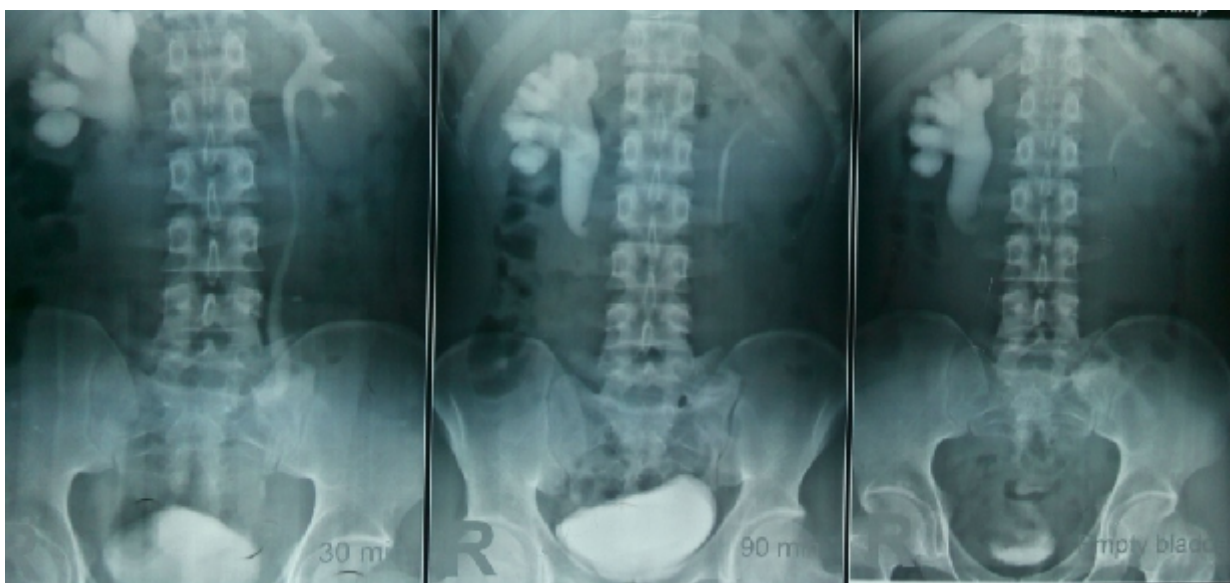


Figure 1: Intravenous pyelography of patient, showing right side moderate hydronephrosis and upper hydroureter with kinking of upper ureter

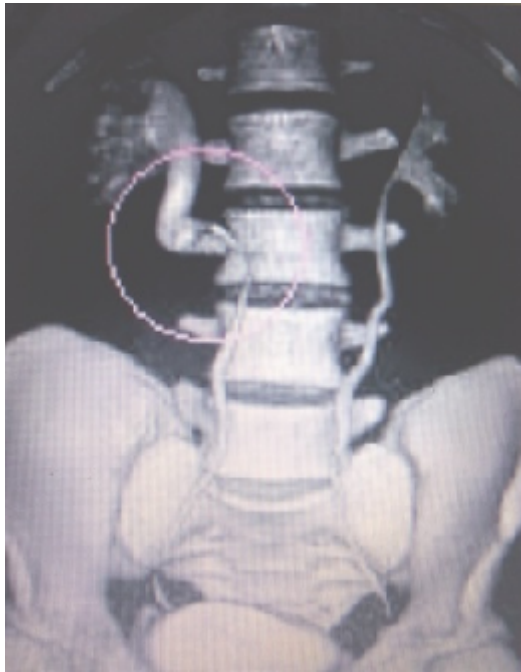


Figure 2: CT Scan of abdomen with IVP showing abnormal course of right ureter, coming posterior and medial to inferior vena cava

CONCLUSION

Retrocaval ureter is a rare congenital anomaly that presents clinically late in the second and third decades of life. Very few clinically symptomatic cases

have been reported worldwide. Treatment is surgical allowing for correction of the anomaly with resolution of symptoms. There is the need to research whether it is developmental anomaly of ureter of inferior vena cava.

REFERENCES

1. Chuang VP, Mena CE, Hoskins PA. Congenital anomalies of the inferior vena cava. Review of embryogenesis and presentation of a simplified classification. *Br J Radiol.* 1974; 47:206–213.
2. Schlüssel RN, Retik AB. Preureteral Vena Cava. In: Kavoussi LR, Novick AC, Partin AW, Peters CA, editors. *Campbell-Walsh Urology.* 9th ed. Elsevier Saunders; 2007. pp. 3418–3420. .
3. Dreyfuss W. Anomaly simulating a retrocaval ureter. *J Urol.* 1959; 82:630.
4. Heslin JE, Mamonas C. Retrocaval ureter: Report of four cases and review of literature. *J Urol.* 1951; 65:212–222.
5. Olson RO, Austen G., Jr *N Engl J Med.* 1950; 242:963–968.
6. Watanabe M, Kawamura S, Nakada T, et al. Left preureteral vena cava (retrocaval or circumcaval ureter) associated with partial situs inversus. *J Urol.* 1991; 145:1047–1048.
7. Bateson E, Atkinson D. Circumcaval ureter: a new classification. *Clin Radiol.* 1969; 20:173–177.