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Case Report

Retrocaval ureter: a rare congenital anomaly presenting as renal colic with hematuria

Mayuri A. Kamble, Anand P. Thawait*, Ashok T. Kamble

Department of Surgery, Mahatma Gandhi Institute of Medical Sciences, Sewagram-442102, Wardha, Maharashtra, India

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***Correspondence:**
Dr. Anand P. Thawait,
E-mail: anandthawait@yahoo.co.in

ABSTRACT

Retrocaval ureter, a congenital venous anomaly of inferior venacava development was first reported by Hochstetter in 1893. Though rare, it causes wide variety of symptoms ranging from asymptomatic individuals to end stage renal disease. Preoperative diagnosis is usually confirmed by imaging. In symptomatic cases, surgical intervention should be performed and renal function improves after operation. This article presents here, a case of retrocaval ureter presenting as renal colic with hematuria because of more degree of obstruction by inferior venacava with symptoms and signs, operative and perioperative management, as well as literature review of this rare clinically important entity.

Keywords: Retrocaval ureter, Congenital, Renal colic

INTRODUCTION

Retrocaval ureter or circumcaval ureter or postcaval ureter[1] is a rare venous congenital anomaly related to abnormal embryological development of inferior venacava. The passage of ureter posterior to inferior venacava, from dorsal lateral position above to ventral medial position below, causes upper urinary tract obstruction. Other congenital anomalies causing ureteric obstruction are accessory or aberrant renal vessels.[1] The anomaly is usually observed on right side with male preponderance. The prevalence of disease is reported to be 1 in 1000 live births.[1]

The symptoms appear in 3rd or 4th decade of life, despite its congenital cause.[2]

The diagnosis requires high index of suspicion. In symptomatic cases, surgical intervention is often required.[3]

CASE REPORT

A 30 years old male presented with right lumbar pain, burning micturition and hematuria since six months. The pain was sudden onset, rapidly progressive, colicky, off and on, severe, which used to relieve by medications and was accompanied by vomiting. Patient also had mild fever off and on, associated with dysuria and hematuria. Clinical examination revealed pallor in thin built male with right renal angle tenderness. Kidneys were not palpable.

Investigations revealed hemoglobin 8.9 gm%, urea 18 mg%, creatinine 1.2 mg%, sodium 144 mEq/L and potassium 4.0 mEq/L. Urine analysis showed pus cells with RBCs. Urine culture was sterile. Ultrasonography demonstrated right hydronephrosis with proximal hydroureter (Figure 1). Plain abdominal radiograph was normal. Intravenous pyelography detected right sided hydronephrosis with characteristic fish-hook (reversed "J") shape of ureter along with medial deviation. No renal

tract calcification was noticed. The findings were consistent with retrocaval ureter (Figure 2). Colour doppler study confirmed right retrocaval ureter.

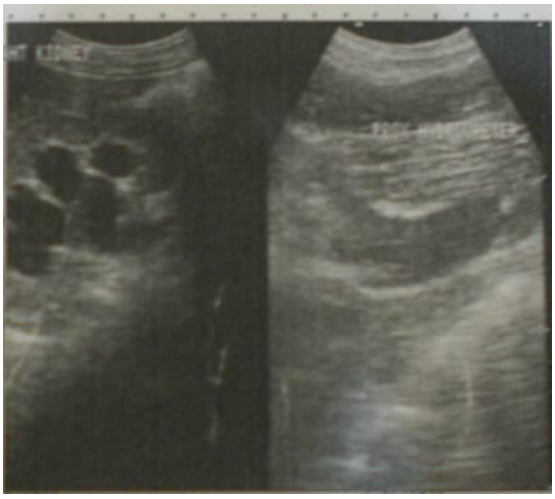

Figure 1: Ultrasonography demonstrating right hydronephrosis with proximal hydroureter.

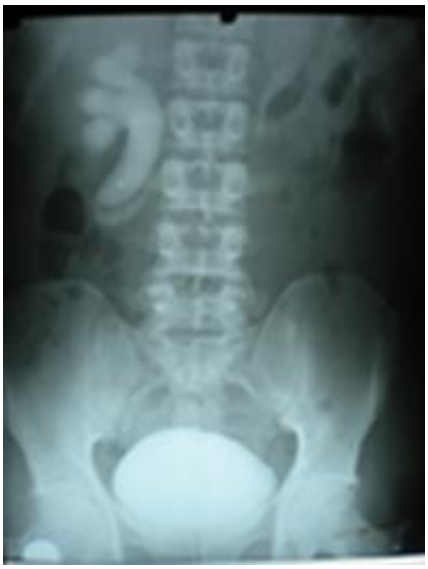

Figure 2: Intravenous pyelography showing right hydronephrosis with hydroureter and reversed J shaped ureter suggestive of retrocaval ureter.

The patient was explored by right lumbar incision. Intraoperatively, dilated right upper ureter was seen compressed, while crossing behind inferior vena cava. The lower ureter was normal in caliber. After meticulous dissection and mobilization, the compressed and a dynamic ureteral segment, behind inferior vena cava was excised. The ureteral continuity was restored by ureteroureterostomy after proximal and distal spatulation and DJ stenting. The incision was closed in layers after keeping a tube drain. Postoperative period was uneventful. DJ stent was removed after 4 weeks. In follow-up, the patient was asymptomatic with normal upper tracts and sterile urine.

DISCUSSION

Retrocaval ureter was first described by Hochstetter in 1893,[4] but first case of retrocaval ureter repair was published by Anderson and Hynes in 1949.[1] Approximately 200 cases have been reported worldwide. Degironcoli reviewed 138 cases in 1961 and further review of 74 cases made a total of 212 cases, till 1966.

Retrocaval ureter is embryological anomaly of inferior venacava and not ureter. A definite right sided inferior venacava forms after subsequent development, anastomosis and regression of (1) right subcardinal vein to form prerenal inferior venacava, (2) subcardinal supracardinal anastomosis to form renal segment and (3) right supracardinal vein to form post renal inferior venacava. If subcardinal vein in lumbar region fails to atrophy and becomes primary right sided vein, the ureter is trapped dorsally to it and forms retrocaval ureter.[5] Other anomalies associated with retrocaval ureter is seen in 21% cases and include double inferior venacava, brachial arch syndrome, horse shoe kidney, myelomeningocele, hypospadias, syndactyly in both feet, intestinal malrotation, turner syndrome, oesophageal atresia, abnormal left kidney (agenetic, ectopic, malrotated), anterior urethral calculus, pelviureteric junction obstruction, congenital lack of vas deferens, extra vertebrae, diverticulum and cardiovascular anomalies such as situs inversus with left retrocaval ureter and Goldenhar syndrome.[6] None of these anomalies were seen in our patient.

Maternal exposure to diethylene glycol and monomethyl ether (as industrial solvent) during fetal period is proposed probable cause for abnormal development of inferior venacava.[7]

Only sporadic cases are reported in literature. Heslin et al. showed incidence of retrocaval ureter at autopsy to be 1 in 1500 (0.06 - 0.17% of autopsy materials), while the prevalence is reported to be 1 in 1000 livebirths.[8] Although congenital, it does not present until 3rd to 4th decades of life due to gradual development of hydronephrosis.[9] This anomaly is more common in children than in adults. The incidence is greater in males than females with ratio of 2.8:1.[5] The anomaly is usually observed on right side. Left sided anomaly is seen in some cases like situs inversus and duplication of inferior venacava.[3] Bilateral retrocaval ureter has been reported once in acardiac fetus.

The main clinical features include dull aching flank pain in 75% of cases or recurrent attacks of right loin pain (intermittent renal colic) due to consequent proximal ureterohydronephrosis with hematuria, microscopic or gross, in 25% cases. Other modes of presentation are recurrent urinary tract infections, asymptomatic hydronephrosis, recurrent pyelonephritis, concomitant stone formation (usually ureteric calculus) due to stasis above obstruction. Clinical symptoms and

hydronephrosis improve by 6 months postoperatively. Other disorders associated with retrocaval ureter are retroperitoneal fibrosis, carcinoma of ureter and renovascular hypertension.

Ultrasonography is non-invasive method of demonstrating anatomy of retrocaval ureter. Intravenous pyelography is initial investigation of choice and shows dilatation of renal pelvis, calyces and upper ureter above obstruction. Antegrade and retrograde pyelography reveals characteristics S shape midline deviation of ureter and on oblique view, ureter hugging lumbar spine. Retrograde pyelography combined with inferior venocavography clearly confirms diagnosis but is invasive.[10] CT is the best modality for diagnosis today, being most efficacious, least invasive with inherent ability to define three dimensional relationship of retroperitoneal structures. Enhanced CT scan with ureteral catheterisation can demonstrate opacified catheter posterior to inferior venacava.[11] New imaging studies e.g. spiral CT scan and MRI clearly delineate anatomy noninvasively with MRI being equally efficacious, having no radiation risk and no use of iodinated contrast material. An isotope renal scan reveals degree of obstruction and differential renal function to decide therapeutic modalities. In our case, the diagnosis was made by intravenous pyelography.

Bateson and Atkinson classified retrocaval ureter into two types, according to radiological appearance and site of ureteral narrowing. In more common type I (low loop), ureter crosses behind inferior venacava, at level of third lumbar vertebrae, usually medial to pedicle and has fish hook (S shaped) deformity at point of obstruction. Marked hydronephrosis is seen in 50% of patients. In type II (high loop), renal pelvis and upper ureter lie horizontally and retrocaval segment of ureter is at same level, as the renal pelvis (segment becomes retrocaval at renal pelvis). There is less medial deviation of the ureteral segment. The inferior venacava is encircled in a smooth curve (sickle shaped curve) as seen in retrograde pyelogram with mild hydronephrosis. Type II is less common, around 10% of all cases.[9] The main causes of hydronephrosis are adhesion of retrocaval segment, lumen stenosis due to compression by psoas muscle and spinal column posteriorly and venacava anteriorly, leading to inflammation, fibrosis and torsion.

Treatment primarily is based on clinical presentations, grade of hydronephrosis and derangement of renal function. In cases of mild hydronephrosis, without obvious symptoms, no demonstrable progression of hydronephrosis, no worsening of renal function, no stone formation or infection, conservative treatment with periodic examination is followed. Symptomatic patients generally need surgical treatment. Various operative methods proposed for retrocaval ureter include ureteroureteral reanastomosis, with or without resection of retrocaval segment, ligation or transection of inferior venacava and nephrectomy. In 1935, Kimbrough performed the first successful surgical correction. Presently, ureteroureteral reanastomosis, anterior to venacava, with resection of retrocaval segment is most favoured surgical treatment. When kidney is severely damaged, (thinned out cortex and poor function) or when marked infection is present, nephrectomy is treatment of choice, provided contralateral kidney is normal.

Anderson Hynes ureteropelvic pyeloplasty (1945) and precaval transposition of ureter has advantages over ureteroureterostomy or uretroneocystostomy, as both ends of anastomosis are dilated and have a good blood supply. If retrocaval portion of ureter is stenosed, ureteroureteral anastomosis is performed with DJ stent in situ for 4-8 weeks, to prevent stricture. However, Chung and Gill performed pyeloplasty, in case of retrocaval ureter having atretic retrocaval portion and demonstrated normal urine flow at 6 months of followup.[12] Extensive dissection of ureter strips off its blood supply and causes infection and necrosis. Cathro performed section of inferior venacava and transposited retrocaval ureter anteriorly, but post-operative complication included residual oedema of lower extremities for 6 months. Goodwin, Burke and Muller demonstrated good results with section and reanastomosis of inferior venacava in solitary kidney.

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. However, it is still time consuming and technically demanding with intracorporeal suturing as the main time consuming step.[13] Laparoscopic pyelopyelostomy is technically more easier to perform than ureteroureterostomy which requires extra fourth port insertion to facilitate dissection and has more stricture rate.[14] The robotic approach to retrocaval ureter was first published for pediatric patient by Gundeti et al. in 2006.[15]

There is decrease in grade of hydronephrosis and symptoms 6-12 months post operatively. Complications include urine leakage and urosepsis seen more commonly in ureteroureterostomy.

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