

# RETROCAVAL RIGHT URETER: A VERY RARE CONGENITAL ANOMALY SUCCESSFULLY REPOSITIONED ANTECAVALLY



✍ Col. Dr. B.K. Hamal, MBBS, MS, FICS  
 ✍ Dr. Ganesh Thapa, MS Resident  
 ✍ Dr. B.G. Karmacharya, MS Resident

## Abstract

Retrocaval ureter is one of the rarest congenital anomalies. In this condition, the right ureter passes behind the (inferior venacava) IVC and is then grossly obstructed leading to hydroureter and hydronephrosis. A 21 years man who presented with this rare condition was corrected successfully.

**Key words:** retrocaval ureter, IVU, ureteroureterostomy, pyeloplasty

## Introduction

Retrocaval ureter is one of the rarest congenital anomalies, the etiology being abnormal embryologic development of the IVC. Male to female ratio is 3 or 4:1. Most patients present with right lumbar pain. They may have recurrent urinary tract infections or episodes of acute pyelonephritis. We report such a case of retrocaval right ureter in a 21 years male patient. The patient was investigated and was found to have a gross proximal ureteric dilatation and gross hydronephrosis but the cause for the obstruction could not be identified preoperatively. The patient underwent exploration of

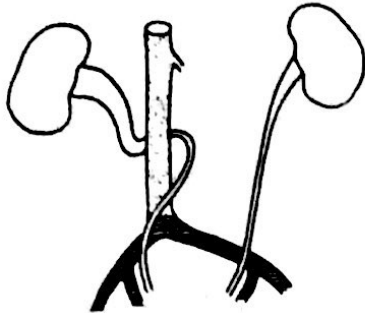
right kidney and ureter and was found to have one of the rarest forms of congenital anomaly i.e. the retrocaval ureter. The excision of the retrocaval part of ureter was done followed by successful anteriorisation and end to end anastomosis.

## Case Report

A 21 years male serving personnel presented with pain in the right loin of three weeks duration associated with fever. He also had burning micturition and increased frequency of urination for one week. On examination, the positive findings were raised temperature, tenderness in right lumbar area and right renal angle. Routine hematological and biochemical renal parameters were normal except leucocytosis. Urine routine and microscopic examination revealed plenty of pus cells and culture was negative. KUB x-ray was unremarkable. USG of abdomen and pelvis showed right sided ureterohydronephrosis but no calculi were seen. Intravenous urogram (IVU) revealed normal left sided kidney and right kidney was poorly visualized. Retrograde pyelogram revealed narrowing at the L3 level and dilated proximal ureter and grossly dilated kidney (Fig 1). Based on the above findings we assumed that obstruction could be due to radiolucent stone, band or fibrosis. He underwent exploration of right kidney and



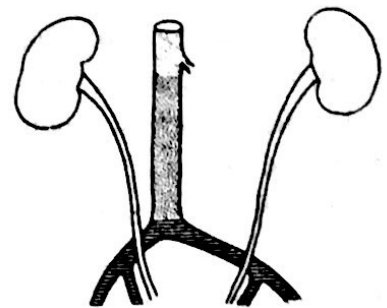
ureter through right subcostal incision. The operative findings were severely compressed retrocaval right ureter at L3 level with gross dilatation and tortuosity of proximal ureter and extra renal pelvis.



The ureter was mobilized and compressed retrocaval segment was excised



The mobilized ureter was spatulated and anastomosed over 28x6F double J stent antecavally by using 3-0 Vicryl®



The patient had an uneventful recovery and symptomatic relief. He was reviewed after four week and DJ stent was removed. He is due for an IVP study.



## Discussion

Retrocaval ureter is a rare congenital abnormality, the incidence is one in 1500 cadavers<sup>1</sup>. It is a condition in which the ureter deviates medially and passes behind the IVC<sup>2</sup>. Though the abnormality is congenital, it does not present until the 3<sup>rd</sup> or 4<sup>th</sup> decades of life<sup>3</sup>. However, a case has been reported even at the age of two years<sup>4</sup>. The IVC normally develops from the posterior cardinal, sub cardinal and supra cardinal veins, which undergo sequential development, anastomosis and regression to become the IVC and azygos venous system. Normally the right sub cardinal veins form the pre renal IVC, the sub cardinal, supra cardinal anastomosis form the renal segment and the right supra cardinal vein forms the post renal IVC. The left supra cardinal and lumbar portions of the right posterior cardinal veins atrophy. If the sub cardinal vein in the lumbar portion fails to atrophy and become primary right side vein, the ureter is trapped dorsal to it<sup>1</sup>. Variation of this includes duplication of venacava, with ureter lying behind or beside the vascular limbs.

The common presentations of retrocaval ureter are right lumbar pain-dull aching or intermittent, recurrent urinary tract infections and microscopic or gross hematuria. There is a high incidence of calculi due to stasis.

Diagnosis is confirmed by USG and IVU. IVU typically shows S shaped or fish hook deformity in the dilated proximal ureteric segment with moderate hydronephrosis. Spiral CT and MRI help to delineate the anatomy clearly and non-invasively. So far CT has been the procedure of choice to confirm the diagnosis of retrocaval ureter, however, MRI is likely to replace CT<sup>5</sup>. The retrocaval ureter is classified into two types based on its radiographic appearance and site of narrowing of ureter<sup>4</sup>.

Type I- commoner, ureter crosses the IVC at the level of L3 and has a fishhook or S shaped deformity of ureter proximal to obstruction, marked hydronephrosis is seen.

Type II- cross over occurs at the level of renal pelvis, lesser degree of hydronephrosis or none at all, renal pelvis and upper ureter lie nearly horizontal before encircling the IVC in a smooth curve (sickle shaped curve).

## Our case falls in the type I variety

Various anomalies associated with retrocaval ureter are : horseshoe kidney, double IVC<sup>6</sup> and left retrocaval ureter with Goldenhar syndrome<sup>7</sup>.

Treatment mainly consists of excising the retrocaval segment, anteriorisation and ureteroureterostomy. This can be performed retroperitoneoscopically and eight cases of such procedures have been reported so far<sup>8</sup>. If there is severe hydronephrosis, pyeloplasty with precaval transposition has been advocated. Occasionally nephrectomy may be required in presence of thin renal cortex, poor function and severe infection.

## References

1. Richard N, Schlüssel, Alan B Relik. Anomalies of upper urinary tract-anomalies of ureteric position: Walsh PC, Retik AB, Vaughan ED Jr. ed: Campbell's Urology, 7<sup>th</sup> ed. Philadelphia, WB Saunders, 1998;2: 1850-57
2. Resnick MI, Kurush ED. Extrinsic obstruction of the ureter, in Walsh P, Retik AB, Stamey TA, Vaughan ED Jr. ed: Campbell's Urology, 6<sup>th</sup> ed, Philadelphia, WB Saunders, 1992; 1:533-69
3. Bateson EM, Atkinson D. Circumcaval ureter, A new classification. Clinical Radiol 1969; 20: 173
4. Dalal A; Barsiwala N; Vaidya A; Dalaa S Retrocaval ureter: a case report and review of literature Journal of Indian Association of Paediatric Surgeons. 1999 Jul-Sep;4(3):147-50
5. MC Uthppa, D Anthony and C Allen Retrocaval ureter: MR appearance. Bri J Rad 75(2002), 1779
6. Kokubo T, Okada Y, Yashiro N, et al. CT diagnosis of retrocaval ureter associated with double inferior venacava: report of a case. Radiat-Med 1990;8(3):96-8
7. Ishitoya S, Arai Y, Okudo K, et al. Left retrocaval ureter associated with Goldenhar syndrome (brachial arch syndrome). J Urol 1997; 158(2): 572-3
8. Miyazato M et al. Retroperineoscopic ureteroureterostomy for retrocaval ureter. Hinyokika Kyo 2002 Jan;48(1):25-8

