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Retrocaval Ureter- A Rare Cause Of Hydronephrosis In Children

Abstract

Retrocaval ureter is a rare congenital anomaly, with the ureter passing posterior to the inferior vena cava. Its rarity and non-specific presentation poses a challenge to surgeons & radiologists in making the correct diagnosis. Though congenital,it commonly manifests in the third to fourth decade of life and very rarely in children. Symptomatic patients need surgical intervention. We present a child who presented with flank pain and diagnosed to have right hydronephroureterosis on radio imaging. During surgery, retrocaval anomaly was noticed. It was successfully treated by ureteral dissection, excision of the retrocaval segment and uretero-ureteral anastomosis.

Keywords: Retrocaval ureter; Hydroureteronephrosis

Case Report

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Introduction

Retrocaval ureter is a rare condition that results due to anomalous development of the inferior vena cava [1]. The incidence is reported to be approximately 1:1000, with male predominance [2]. The anomalous vessel compresses the ureter, causing varying degrees of hydronephrosis. Due to gradual development of hydronephrosis the patients usually present in third or fourth decade of life.

Surgical intervention is mandatory in cases with significant functional obstruction. Open or laproscopic pyelopyelostomy and ureteroureterostomy is the treatment of choice [3].

Case Report

5 year old boy came to our outpatient department with intermittent flank pain since one week. There were no other systemic complaints. Patient was previously investigated & operated for right non palpable testis a year ago. An incidental finding of right mild hydronephrosis (AP Diameter of 12mm) were noted during sonological evaluation. General and systemic examination were normal. Haematological workup was normal. Ultrasonography showed hydronephrosis with antero-posterior diameter of 3.5cm associated with dilatation of upper ureter (dia-9.2 mm). Micturating cystourethrogram was done to rule out vesico ureteric reflux. DTPA renal scan was suggestive of preserved parenchymal function & GFR with significant subrenal obstruction at right peviureteric junction along with upper hydroureter. CT-IVP demonstrated right hydronephrosis with dilatation of the upper ureter due to kinking at junction of upper & middle ureter suggestive of obstruction (Figures 1&2). Left kidney was normal.

We decided to operate this child in view of the increased hydronephrosis. Child underwent open surgical intervention via right upper abdominal transverse incision. On exploration the ureter was found to dip behind the inferior vena cava after circumscribing the vessel it pursued its normal course, below the bifurcation of the iliac vessels (Figures 3&4).

The entire upper ureter was mobilized and dissected & brought anterior to in the inferior vena cava and placed in its normal position. End to end uretero-ureteral anastomosis over 4 fr Dj stent was done & wound closed in layers keeping drain in situ. Drain was removed after 6 days. Postoperative course was uneventful. Cystoscopic stent removal done 6 weeks later. 3 months postoperative USG revealed a residual hydronephrotic changes in right kidney of hydronephrosis. Child doing well on further 3 month follow-up.

Discussion-The first recorded case of retrocaval ureter was seen on autopsy and was described by Hochstetter in 1893 [4,5] Retrocaval ureter is a rare congenital disease. The prevalence of the disease is reported to be 1 in 1100 live births. It mostly manifests itself in the third or fourth decade, and occurs three times more frequently in men than in women [6]. Its etiology is presumed to be the abnormal persistence of the subcardinal vein in embryologic development of the IVC resulting from the major portion of the infrarenal IVC, which formed from the subcardinal vein that lies ventral to the ureter [7]. Patients present with to right flank pain, urinary tract infections, and hematuria or urolithiasis, which are not always associated with symptomatic obstruction. Retrocaval ureter has been previously diagnosed by IVU and RGP, but nowadays, CT scan is the best modality for diagnosis [5].

In 1982, Bergman classified retrocaval ureter into two clinical types [8]. Type I (low loop) is the most common, with the dilated proximal ureter assuming the shape of a reverse "J". Usually, this type of ureter is obstructed. Type II (high loop) is seen less frequently. The ureter passes behind the IVC at the level of, or just above, the pelvic-ureteral junction. This type of ureter is frequently not obstructed.

Treatment is ureteroureterostomy with anterior transposition of ureter, which has been performed by using open, laparoscopic [9] and retroperitoneoscopic [10] approaches. If there is severe

hydronephrosis, Anderson Hynes pyeloplasty has been advocated. Occasionally, nephrectomy may be required in the presence of a thinned out cortex, poor function or severe infection. Open surgery was the first line successful treatment of this condition [11]. However, laparoscopic surgery is now used has been successfully described in small number of cases [9]. Recently LESS is emerging in an effort to raise the standard of laparoscopic procedures through a scar-free approach [12].

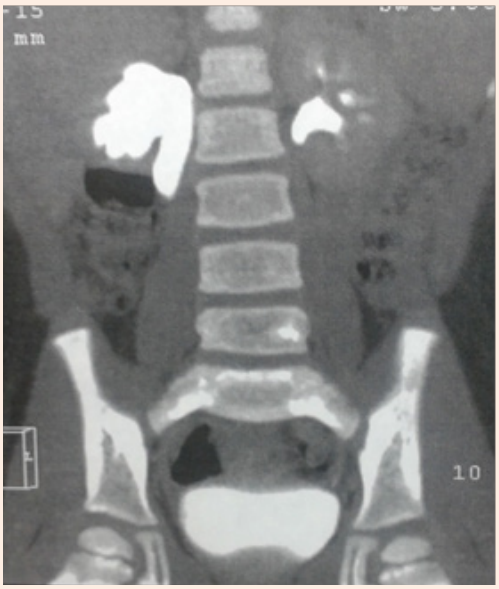

Figure 1: CT IVP image.

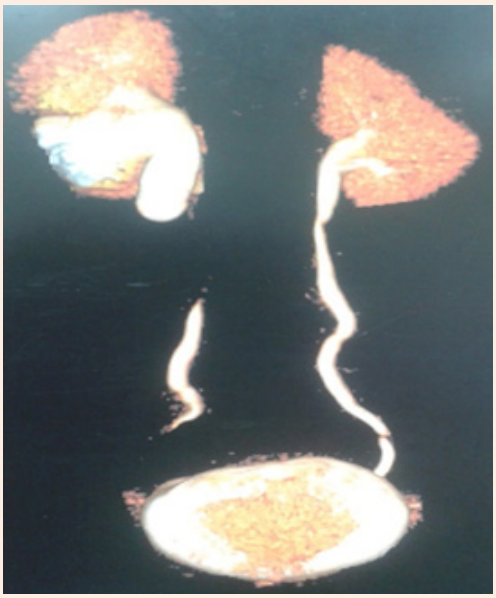

Figure 2: 3D reconstructed image.

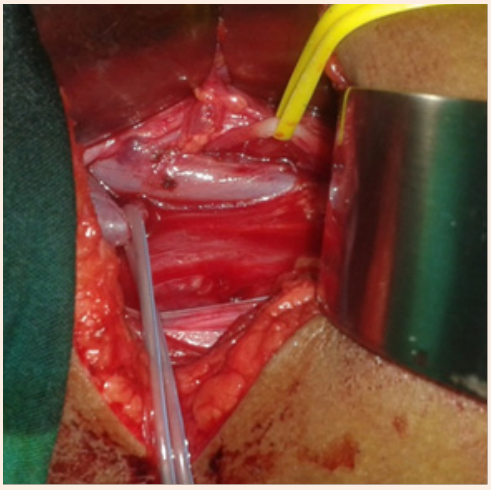

Figure 3: Intraop pic –IVC & Retro due to retrocaval ureter.

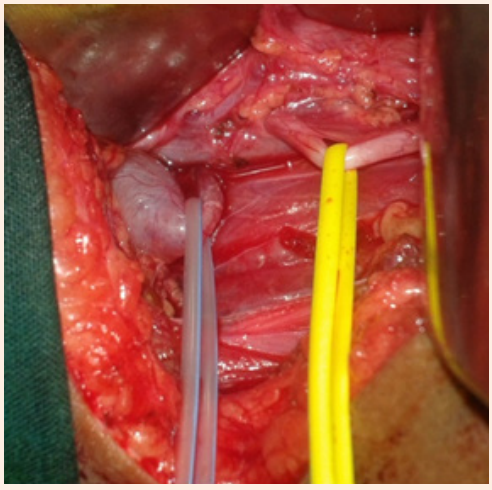

Figure 4: Rt hydroureteronephrosis cavalureter.

Conclusions

Retrocaval ureter is a congenital anomaly presenting clinically late, in the third and fourth decades of life, It is rarely noticed in paediatric age groups. Imaging studies are sufficient for making an accurate diagnosis of a retrocaval ureter. Treatment is surgical, which allows for correction of the anomaly, with resolution of symptoms. Although Minimally invasive surgery emerging new the major trend in current management,time tasted open surgery still remains gold standard treatment for retrocaval ureter.

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