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Congenital Mid Ureteric Stricture: Case Report and Review of the Literature

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Abstract **Case Report**

Introduction*:** Most congenital ureteral strictures occur at the ureteropelvic or ureterovesical junction in children. Rare causes of hydronephrosis are congenital midureteral strictures. Definitive preoperative diagnosis is clinically challenging, and most patients are misdiagnosed preoperatively. ***Case presentation: We report 9 years old female child hydronephrosis. Renal scintigraphy with DTPA; revealed a normally functioning left kidney with split renal function of 47% and an obstructive pattern. Furthermore; Congenital Mid ureteric stricture is typically not diagnosed preoperatively, and definite diagnoses have been reached via retrograde assessment of the ureter; and surgical resection-anastomoses of the obstructed right ureteral tract allowed establishing normal ability of urine outflow. ***Conclusion***: Pre-operative detection of Congenital Midureteric stricture is clinically challenging and requires a high level of attention. Whenever we observe a dilation of only the proximal segment of the ureter, we should keep this diagnosis in mind and discuss the images in detail with an experienced radiologist. The diagnosis should not be delayed in order to apply the appropriate approach surgery.

Keywords: Midureteric, stricture, diagnostic.

INTRODUCTION

There are many etiologies involved in obstructive uropathies. Rare causes of hydronephrosis are congenital midureteral strictures. The ureteral strictures are narrowing of the lumen of the ureter [9]. Strictures are the decrease of ureteral lumen diameter by 60%. These obstructions are usually due to valves [1].

Definitive preoperative diagnosis is clinically challenging, and most patients are misdiagnosed preoperatively. Intraoperative identification is therefore very important [2].

CASE PRESENTATION

A 9-year-old female child presented with recurrent abdominal pain in the right lower part of abdomen. A renal ultrasound revealed right hydronephrosis. The left kidney was normal; A voiding cystourethrogram showed a normal bladder and urethra and no evidence of vesicoureteric reflux.

Renal scintigraphy with DTPA; revealed a normally functioning left kidney with split renal function of 47% and an obstructive pattern.

Based on a preoperative results investigations, we performed posterior lombotomy for right pyeloplasty.however; we finded a dilation of the proximal ureter; the other segments are not explored.

Congenital Midureteric stricture is not diagnosed preoperatively, and definite diagnoses have been reached via retrograde assessment of the ureter; CT U scan showed dilation of the proximal ureter and midureteral stricture .figure 1and 2 a surgical revision via the right parailiac retroperitoneal approach was performed and the ureteric segment was excised and an end-to-end ureteroureterostomy was fashioned following spatulation of the two ends over a DJ stent.figure 3 and 4 Postoperatively, the stent was removed at 6 weeks. The patient had uncomplicated postoperative course. Long-term follow-up revealed the regression of hydronephrosis.

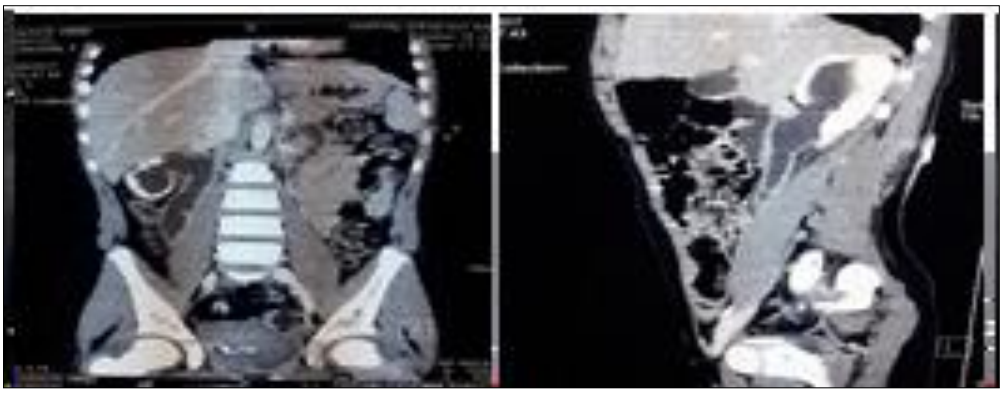

Fig 1 **Fig 2**

Figure 1-2: CTU images on coronal and sagittal show dilation of the proximal ureter and midureteral stricture (Narrowed zone at junction of dilated upper two thirds and narrowed lower one third of ureter)

Fig 3 **Fig 4** **Fig 5**

Figure 3: Photograph showing dilated ureter proximal to the stricture and distal normal caliber ureter
Figure 4: Ureterectomy of the stenosed segment
Figure 5: Anastomosis of ureter

DISCUSSION

Midureteric stricture is a rare cause of hydronephrosis in neonates and is often misdiagnosed as pelviureteric junction obstruction in the first instance. Accurate preoperative diagnosis with IVP and radionuclide scans may not be possible in all cases [3].

Congenital mid-ureter stricture is represented by a narrowing that occurs between the pelviureteric junction and vesicoureteric junction. Campbell reported only 4% had mid-ureter stricture in an autopsy of a sequence of 72 children who had ureteral stricture.1 Most recently, Meng *et al*., reported that middle ureteral stricture accounts for only 1.6% of all ureteral strictures [4].

In pathology, ureteral stricture is defined as a mechanical obstruction due to structural abnormalities in the wall. Two pathogenesis of ureteral stricture exist: ureteral valve and true ureteral stricture. The ureteral valve is a transverse fold of ureteral mucosa with anatomically proven [4-5].

Te pathogenesis of this condition is unclear, and many theories attribute it to abnormal embryonic development, including abnormal fetal vessel compression, intrauterine infammation, incomplete ureteral recanalization, ischemia due to abnormal branches of blood vessels and localized developmental arrest [5].

Furthermore, the ureteral valves are associated with other anomalies of the urinary system in 41% of cases: the anomalies encountered are multiple such as renal duplicity, vesico-renal reflux, renal agenesis, kidney in Horseshoe. The frequency of these associations is not explained by the two etiological theories proposed and suggests an anomaly of ureteral embryogenesis [6].

Congenital Midureteric junction is mostly diagnosed as a unilateral disorder; however, cases with bilateral anomalies have been reported in the literature [7].

Diagnosis of ‘ureteric valves’ in a patient presenting with hydrouretronephrosis can be met if following there is proven presence of transverse folds of the ureteric mucosa containing bundles of smooth muscle fiber on histological examination, signs of obstructive disease above the valve with a normal ureter below it, and no other evidence of mechanical or functional obstruction [8].

According to Rabinowitz, [3] ureteric valves can be classifi ed as Type I or Type II, with Type I having smooth muscle present within the leaflet and Type II having smooth muscle at the base only [9].

The most common presentation of this condition includes flank pain, gross hematuria and urinary infection or can be detected incidentally while evaluating suspected cases of megaureters, ureteropelvic or ureterovesical junction obstruction [8].

Gery and in children with unclear diagnoses. With the development of ultrasound techniques for ureteral examination and improvements in the diagnostic rate, ultrasound can serve as a noninvasive and repeatable efective method to diagnose ureteral stenosis or stricture. Ultrasound can be used to determine the location of a ureteral obstruction according to the degree of ureteral dilation and morphological changes, as well as to determine the etiology and extent of stenosis according to diferences in the acoustic images of the stenosis site. At our center, the diagnosis of ureteral stenosis mainly relies on ultrasound, IVU and CTU, while MRU and retrograde pyelography are performed irregularly. Te accuracy of the ultrasound examination is highly dependent on the personal experience of the examiner [5].

The treatment of choice for congenital mid ureter stenosis consists of excision of the affected ureteric segment and anastomosis; using a retroperitoneal approach adapted to the location and can be managed by either an endoscopic approach or surgical reconstruction, including open or minimally invasive (laparoscopic/robotic) techniques [10-2].

Laparoscopic and robotic technology has been successfully used to treat ureteral stenosis. Compared with open surgery, minimally invasive surgery has the advantages of less postoperative pain, a shorter hospitality stay, less scarring, and less parental anxiety. In the future, more children will undergo treatment with laparoscopic and robotic surgery [5].

CONCLUSION

Although congenital narrowing of the ureter at its proximal or distal end is common, congenital mid ureteral obstruction is rare [12]. It should be included in the differential diagnosis of ureteral obstruction in children [13].

Pre-operative detection of this condition is clinically challenging and requires a high level of attention. Whenever we observe a dilation of only the proximal segment of the ureter, we should keep this diagnosis in mind and discuss the images in detail with an experienced radiologist [14]. Tension-free primary ureteroureterostomy is the most effective treatment option [2].

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Authors' Contributions:

MR design the study, performed the treatment, wrote the manuscript and approve the final manuscript. HK perform literature search, acquisition of the data, manuscript review and revising the manuscript. All authors read and approved the final manuscript.

Consent for Publication:

Written informed consent for publication of clinical details and clinical images was obtained from the parents of both patients.

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