

Case Report

Congenital distal ureteric stricture presenting as obstructive megaureter in an infant: a case report

Sreedhar Dayapule, Abhiram Kucherlapati, Sandeep Donthiri*, Ifrah Ahmad Qazi

Department of Urology, Narayana Medical College, Andhra Pradesh, India

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*Correspondence:

Dr. Sandeep Donthiri,

E-mail: sandyreddy19@gmail.com

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ABSTRACT

Congenital distal ureteric stricture is a rare but most important cause of hydro ureteronephrosis in infants presenting as obstructive megaureter. A 8 months old child suffering from recurrent episodes of fever and poor growth had giant hydronephrosis due to distal ureteric stricture. Often misdiagnosed as primary megaureter or pelviureteric junction obstruction, congenital ureteral strictures and valves are the main causes of congenital ureteric obstruction. He was treated with excision of narrow ureteric segment with tapering ureteroplasty and a ureteral reimplantation was performed. This case is presented and discussed with reference to etiology of this rather rare anomaly.

Keywords: Megaureter, Di-mercapto succinic acid, Di-mercapto pentetic acid, UTI

INTRODUCTION

Congenital obstructive megaureter has been observed during fetal age or any stage at the time of childhood.¹ Upper renal tract dilatation is readily diagnosed in utero, and congenital ureteric obstruction may present with prenatal HN.² The aim of this report was to present a male child with obstructive megaureter due to distal ureteric stricture. He was surgically treated with excision of narrowed distal ureter in addition to tapering ureteroplasty with ureteroneocystostomy.

CASE REPORT

A 8 -months-old infant with antenatal history of giant left hydronephrosis (11 mm) initially admitted under paediatrics department with history of recurrent episodes of fever for evaluation since 6 months, later he was referred to our department of urology as USG was showing gross left hydronephrosis. Laboratory tests were otherwise normal except signs of UTI

including 25 to 30 pus cells /hpf. The microbiological urinalysis was positive for UTI revealing pseudomonas 105 CF µ/ml. Urinary ultrasonography revealed a gross enlargement of the left kidney, pelvis, parenchymal thickness of 4 mm and a dilated ureter. MCUG was showing no evidence of reflux. Scintigraphy using di-mercapto succinic acid (DMSA) and di-mercapto pentetic acid (DTPA) showed a dilated left kidney with reduced scintigraphy uptake with scars in upper pole and lateral aspect of cortex with chronic pyelonephritis changes revealing a differential renal function of 31% for the left and 69% for the right kidney with signs and findings showing UVJO. Contrast enhanced CT urography showed gross enlargement of the left pcs and huge dilatation of left ureter with contrast filling entire pcs and ureter, however distal part of ureter couldn't be opacified due to stricture (Figure 1 and 2). Due to progressive worsening of the upper urinary tract function and repeated UTIs, surgical intervention became a necessity. Surgical intervention revealed a severely narrowing of the left distal ureter for 2 cm in accordance with UVJO

resembling bird's beak and the diameter of the ureter proximal to UVJO was measured nearly 4 cm (Figure 3). Excision of the distal ureter and a segment of megaureter with a tapering ureteroplasty of the proximally dilated megaureter for a length of 8 cm around a 8 Fr feeding tube and reimplantation using modified lich Gregoire technique was performed (Figure 4). The urinary bladder was drained via 6 Fr silicone foley catheter and 10 fr foleys was placed as spc. Postoperatively he did well with no episode of pain, nausea or vomiting. On the second postoperative day, the patient started oral feedings and was discharged home in good condition. Post operative follow up was done till 6 months, baby had no episodes of fever. On follow-up ultrasound showed left mild hydroureteronephrosis. Dpta and Dmsa scan at 6 months showed split function was stabilized and no evidence of obstruction was noted.

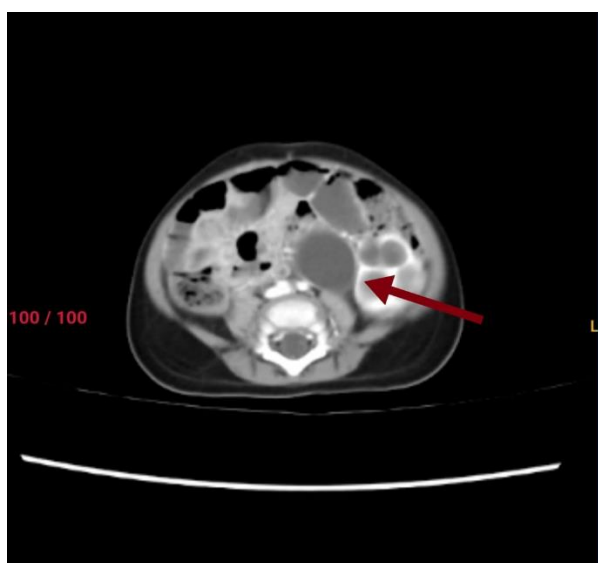


Figure 1: CT urography showing the gross enlargement of the left kidney with reduction of cortical thickness and giant pelvis and ureter.

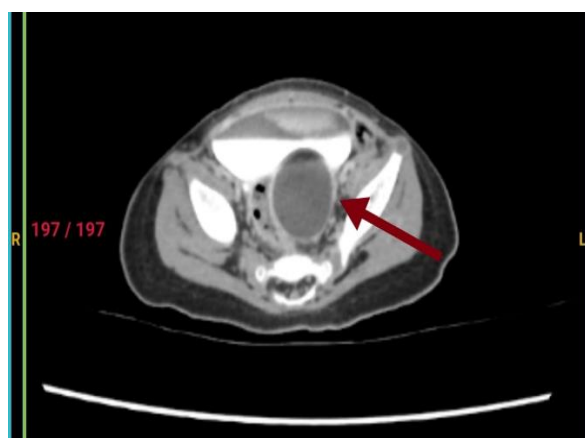


Figure 2: CT urography showing the gross dilatation of left ureter with contrast draining in to bladder from right kidney.

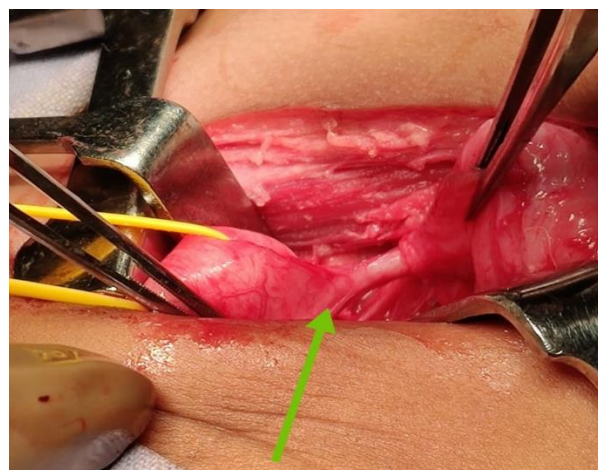


Figure 3: Operative view showing marked narrowing of the Left distal ureter with dilation of the proximal ureter.

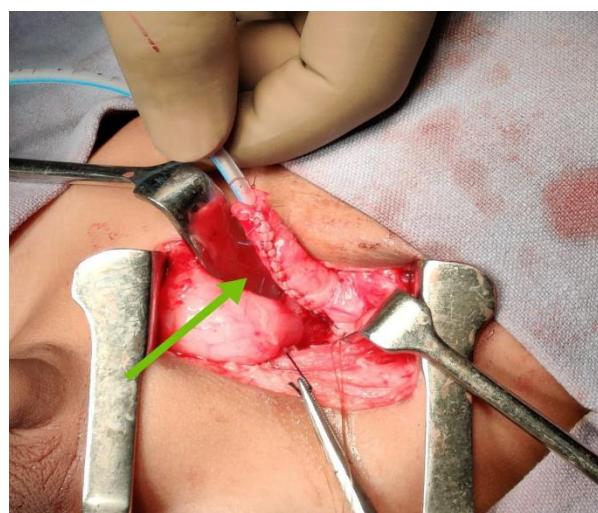


Figure 4: Operative view showing distal part of dilated left ureter after tapering ureteroplasty around a 8 Fr catheter before reimplantation.

DISCUSSION

Muscularization of the ureteral tube begins at the 12th week of gestation, continues throughout pregnancy. Vascular compression can interfere with this process, resulting in incompletely formed segment in the upper ureter, the mid ureter at the pelvic brim, and the lower ureter proximal to the bladder wall, and tends to be related to contiguous vessels. These vessels, however, it is postulated that they disappear as part of the embryological developmental process, leaving behind the stricture.³ Cussen analysed 124 obstructed ureters. Stricture was responsible in 81 specimens and ureteral valves was seen in the rest.⁴ Strictures will have a 60% decrease in luminal diameter with diminished population of smooth muscle cells. Furthermore, in severe strictures the smooth muscle layer is replaced by fibrous tissue, which we found in our case.⁴ Tartari et al study showed

ureteral valves can be associated with strictures, here in our case valves were absent.⁵ Normal ureteral development undergoes an initial solid phase followed by a phase of re-canalisation. Congenital ureteric strictures have been attributed to incomplete recanalisation of the ureter.⁶ Most common cause of giant hydronephrosis in newborns is megaureter which accounts for approximately 20% of cases and occurs in bilateral in 15-25 of patients.⁷ Ureteral dilatation has been classified into four categories, they are namely non-refluxing non-obstructed ureter, refluxing non-obstructive ureter, refluxing obstructive ureter and non-refluxing obstructive ureter.⁸ Diagnostic work-up performed in our case did not reveal vesico-ureteral reflux and so our patient can be classified as non-refluxing obstructive ureter due to long length distal ureteric stricture. It had been reported that of the 27% of pediatric patients with kidney failure due to obstructive uropathy, obstructive megaureter accounts for 3.5% of cases.⁹ Normal findings in the left kidney and ureter were noted so no evidence of kidney failure was observed in our case. Among different etiologic factors in HUN, congenital distal ureteric stricture is rare, associated with an dilated segment of the entire ureter with abrupt narrowing at distal ureter near vuj.¹⁰ Most of these cases can be detected during antenatal ultrasound screening of urinary tract.^{11,12} Prenatal ultrasonography also revealed giant HUN in our case during routine follow-up. It is most likely these patients with obstructive mega ureter show symptoms during childhood. Although the age of our patient was found to be within the limits of infancy period, surgical treatment was performed in our case since there was reduced scintigraphic function with repeated UTIs positive for pseudomonas and severe hydronephrosis (>30 mm). Histological analysis in our patient revealed fibrosis of tissues removed at surgery.

CONCLUSION

Congenital obstructive megaureter due to distal ureteric stricture is a rare but important cause of HUN in children and may occur at any stage during infancy. It is commonly associated with UTIs and deterioration of upper urinary tract. Timely and accurate management of this entity is important for the preservation of upper urinary tract function. The health providers dealing with such kinds of patients should keep this anomaly in mind and a prompt surgical consultation is recommended and the patient should be treated accordingly.

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