Pediatr Urol Case Rep 2020; 7(4):88-93 DOI: 10.14534/j-pucr.2020461252

PEDIATRIC UROLOGY CASE REPORTS
ISSN 2148-2969
http://www.pediatricurologycasereports.com

Incomplete duplex renal system with severe obstruction at the uretero-ureteral junction and minor obstruction at the common stem-vesical junction: Challenging diagnosis and conclusive laparoscopic treatment

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ABSTRACT

Twin obstructive megaureters associated with duplex renal system is a rare malformation, particularly when a giant ureter dilation is present. We report an original case, initially misinterpreted, of a right incomplete duplex renal system with huge disproportion between the upper pole giant megaureter and the lower pole moderate megaureter, with severe obstruction at the uretero-ureteral junction and minor obstruction at the uretero-vesical junction. Uro-magnetic resonance imaging allowed conclusive diagnosis and the laparoscopic approach was effective for definitive, successful treatment.
Key Words: Incomplete duplex renal system, uretero-ureteral junction obstruction, giant megaureter, laparoscopic heminephrectomy.

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E mail: i.marianna@hotmail.it
Received: 2020-04-03 / Revisions 2020-04-30
Accepted: 2020-05-01 / Publication Date: 2020-07-01

Introduction

Twin obstructive megaureters associated with duplex system is a rare malformation, particularly when a giant ureteral dilation is present, with few cases described in the literature [1-14].

We report the case of female infant with an initially misinterpreted right incomplete duplex renal system, associated with upper pole (UP) giant megaureter and lower pole (LP) moderate megaureter, due to severe obstruction at the uretero-ureteral junction and minor obstruction at the uretero-vesical junction.

Case report

A 3-month-old girl had a prenatal diagnosis of severe right hydroureteronephrosis, confirmed at birth (Fig. 1).

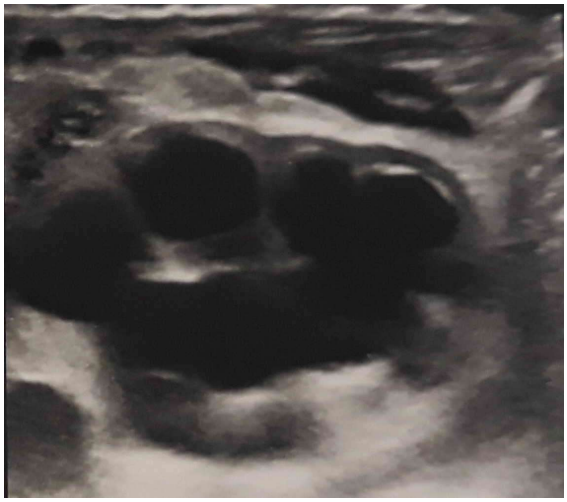

Fig. 1. The appearance of the right hydroureteronephrosis on ultrasonography.

A normal voiding cystourethrography and a dynamic renal scan [Tc-99m MAG3 (mercaptoacetyltriglycine)], performed at 4 months of age, showed an apparently single renal system with primary obstructive megaureter (POM), measuring a right renal functional contribution of 40%. At that stage, the presence of a non-functional upper pole (UP) and its giant megaureter was unrecognized and the disproportion between the huge dilation of the right urinary tract shown by ultrasound (US) and the relatively moderate POM shown at MAG3 did not lead to suspect the presence of a more complex pathology. Because of the unchanged dilation, at 5 months of age, a double J stent (3 Fr, 14 cm) was inserted in a single right ureteral orifice and the patient started antibiotic prophylaxis. The procedure did not produce any change at the US controls 2 and 4 weeks after. Persevering in the misunderstanding, the persistence of hydroureteronephrosis was interpreted as the result of insufficient drainage of the double J stent. Meanwhile, the baby's clinical condition worsened for breathing difficulties due to abdominal distention and diaphragm elevation, nevertheless, she never developed urinary tract infection (UTI). So, we decided to perform a right ureterostomy. At the surgical exploration of right peri-vesical side, a moderately dilated POM was identified and disconnected from the bladder, without open the bladder. At that moment, there was the unexpected finding of an enormously dilated thin-walled structure connected to the distal ureter with a short non-canalized segment. It was interpreted as possible giant ureteric diverticula. The structure was emptied of the clear liquid contained inside and removed through the small peri-vesical approach. A few minutes of observation did not detect the presence of other liquid in the retro-peritoneum and we assumed that we had completely removed the cystic structure. The ureterostomy showed a good function with some modest improvements of urinary dilation at US, which were judged as promising. Clinical conditions were good as well. To mark only a febrile episode 3 months later and a retracting scar at the ureterostomy site which needed revision. At 11 months of age, we did not find any reason to postpone the planned undiversion. The ureteral right reimplantation in the bladder was performed with a classical intravesical trans-trigonal transverse anti-reflux technique. Subsequently, no change in the permanent US finding of upper urinary tract dilation was revealed, now associated with a palpable mass on the upper right abdominal quadrant, in a totally asymptomatic patient. An ascending pyelography was performed, according to De Castro's percutaneous procedure [1], showing a good outcome of the re-implanted POM and no communication with the mass (Fig. 2).

An extra-renal cystic mass, like a cystic lymphangioma, was suspected and even suggested by several international consultations. Supplementary investigations were essential. Uro-magnetic resonance

imaging (uro-MRI) showed a giant megaureter coming from a small renal non-functioning UP, finally bringing to definitive diagnosis of incomplete duplex renal system with twin megaureters (Fig. 3).

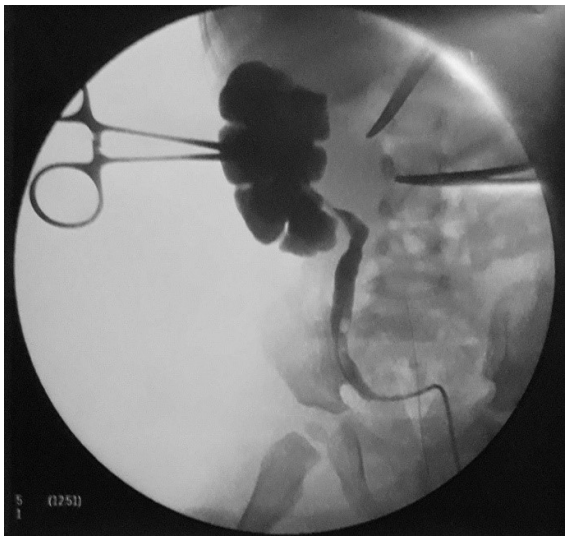

Fig. 2. An ascending pyelography view of the re-implanted primary obstructive megaureter. (POM).

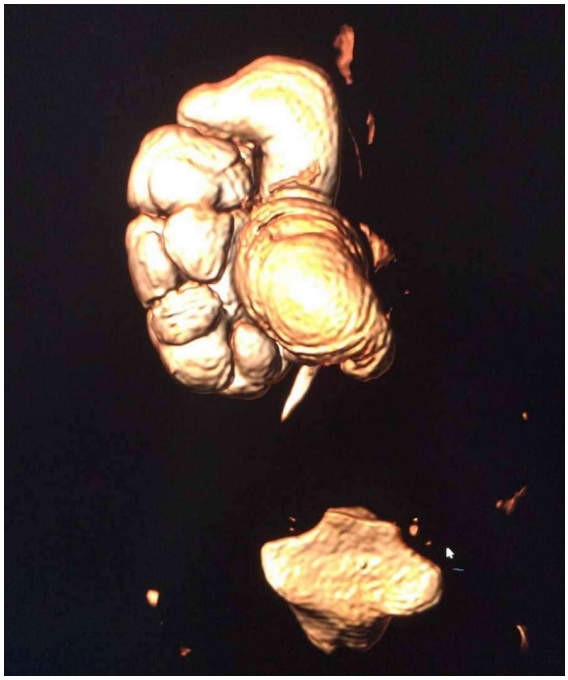

Fig. 3. Uro-magnetic resonance imaging (uro-MRI) showed a giant megaureter coming from a small renal non-functioning UP.

A laparoscopic UP hemi-nephroureterectomy was planned when patient was 17-month-old and 9, 7 kg of weight. A retrograde endoscopic LP ureteral catheterization, according to De Castro's percutaneous procedure [1], was performed (Fig. 4), followed by transperitoneal laparoscopic approach (4 trocars).

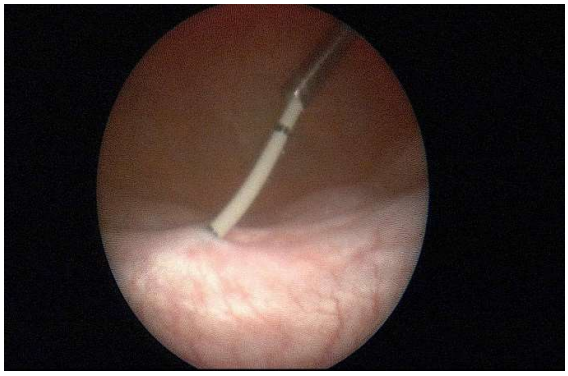

Fig. 4. A retrograde endoscopic LP ureteral catheterization.

The giant megaureter coming from an hypoplastic UP, compressing the LP and extremely adherent to the LP ureter and to the adjacent structures was located and removed together with the small UP (Fig. 5).

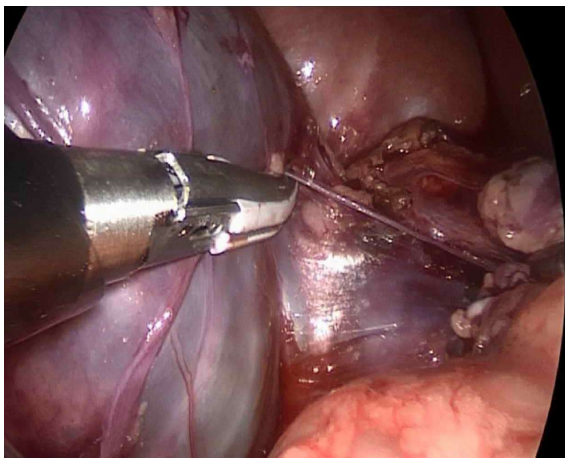

Fig. 5. Laparoscopic removal of the giant megaureter with small UP.

Length of surgery was 160 minutes. No need of conversion neither intraoperative bleeding

or urine leakage were observed. Post-operative recovery was uneventful. The histological exam confirmed a (n) hypoplastic UP and a giant blindly ending megaureter. At 1 year follow up we reported a good outcome and no loss of renal function on the residual kidney moiety.

Discussion

Megaureter may be correlated with unilateral renal agenesis, complete or incomplete double renal system, ectopic kidney, contralateral cystic and dysplastic kidney, and horseshoe kidney [2-7]. Differential diagnoses of megaureter include mesenteric and adnexal masses in females. However, these masses are usually spherical or oval with complete septa inside in contrast to tubular and tortuous shape of megaureter. Dilated bowel loops must be differentiated as well but they may demonstrate peristalsis and have materials (mixture of solid, liquid, and gas) in their lumen. Rastogi R. [8] in 2008 described a case of a child with massive megaureter misinterpreted as a cystic lymphangioma.

Treatment is indicated when megaureter is secondary to severe reflux or obstruction. Nephroureterectomy may be indicated, when, function of the interested kidney is absent or severely impaired.

Megaureter associated with double system is rare. Weinstein et al. [9] described a primary ureterovesical junction obstruction in the common stem of an incomplete double system managed successfully with a common sub-mucosa tunnel reimplantation of both tapered ureters. POM of the LP moiety of a double system has been described for the first time in 1991, by Lee et al. [10]. In 2012, Annigeri et al. [11] described the first case of congenital giant megaureter with double system presenting as abdominal lump in a neonate.

There are only a few cases reported in the literature of double system in which the ureter has assumed massive proportions, overshadowing the renal pelvis and parenchyma. Uson et al. [12] and Lelli-Chiesa et al. [13] reported a case of a giant ureter presenting as an abdominal mass in a newborn. Whitmore et al. [14] reported a case of giant hydronephrosis of a duplex kidney associated with an ectopic ureter.

In our case, despite the erroneous diagnostic itinerary and misinterpretations, correct diagnosis was finally reached: an incomplete duplex renal system with an UP giant megaureter joining the LP ureter with an atretic segment and a short common stem with obstruction at the junction with the bladder. Several US and MAG3 renal scan gave no suspicion of double system and the presence of single ureteral meatus in the bladder at cystoscopy did not help to reach the correct conclusion. Retrospectively reviewing our management, the duplex renal system could have been suspected in advance, primarily because of the dissonant findings between US and MAG3. Uro-RMN should have been performed earlier and before any surgical procedure. Furthermore, the unexpected finding at the first surgery of a cystic structure connected to the LP ureter should have been deepened and explained; probably we have resected the lower part of the UP giant megaureter, misinterpreted as ureteral diverticula. Uro-MRI was certainly decisive to reach the right and complete diagnosis and the laparoscopic approach was particularly effective for definitive treatment. Minimally invasive procedure on the upper urinary tract in small infants weighting less than 10 kg is technically challenging and require expertise [15-16]. It was feasible and effective in our hands.

Conclusion

We have reported an original case of complex congenital anomalies of kidney and urinary tract (CAKUT). The main message is that any unexpected and unclear findings during diagnostic itinerary or surgery need correct detailed interpretation. Uro-MRI finally allowed conclusive diagnosis and the laparoscopic approach was effective for definitive, successful treatment.

Compliance with ethical statements

Conflicts of Interest: None.

Financial disclosure: None.

Consent: Informed and written consent were taken from patient and her parents to publish this case report.

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