



A duplex renal anomaly in an infant: challenges in its diagnosis and management

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ABSTRACT

Duplex renal systems are common congenital anomalies having a female preponderance with an estimated prevalence ranging between 0.3% and 6% of the pediatric population. Here, a rare case of duplex anomaly in an infant is presented with its diagnostic challenges and management.

Key Words: Infant, kidney abnormalities, diagnostic imaging, megaureter, duplex. hydroureter.

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Introduction

Duplex renal system anomalies can be diagnosed antenatally, but most commonly postnatally due to symptoms. Although females are more affected than males, the left and right sides are equally affected. Congenital giant megaureter (CGM), a very rare unilateral urinary anomaly; It is defined as "a ureter whose lumen is focal and segmentally dilated to more than 10 times the normal diameter" [1-5]. Here, a rare case of duplex anomaly in an infant is presented with its diagnostic challenges and management.

Case report

A two-month-old male baby born with a full-time vaginal delivery with a birth weight of 3 kg was referred to an out center with left hydronephrosis detected antenatally at 24 weeks and postpartum sonology and Tc-99m MAG3 (mercaptoacetyltriglycine Tc MAG3)

tests. Dynamic urogram scanning showed that he had reduced tracer clearance as well as left hydronephrosis with 15.63% differential function (Fig. 1A). The baby had a vague palpable non tender left flank mass with normal renal function tests. Diagnostic cystoscopy and retrograde ureterography via orthotopic left ureteric orifice showed the left duplex kidney with dilated upper pole ureter as well as its pelvis with its unrecognizable lower opening; therefore left upper loop ureterostomy was performed. The baby was taken up for definitive surgery by laparoscopically; the left upper pole moiety was not appreciated, but two ureters were identified, so the previous upper pole ureterostomy was dismantled and the end to end anastomosis as well as side to side anastomosis to the other normal non-dilated lower pole ureter was made, and a double J (JJ) stent was placed across the anastomosis. After 6 weeks, JJ stent was removed from the previously unidentified stenotic orifice located above the bladder neck with the help of fluoroscopy, and therefore, this time, the diagnosis of left duplex kidney with upper pole orifice opening just above bladder neck was

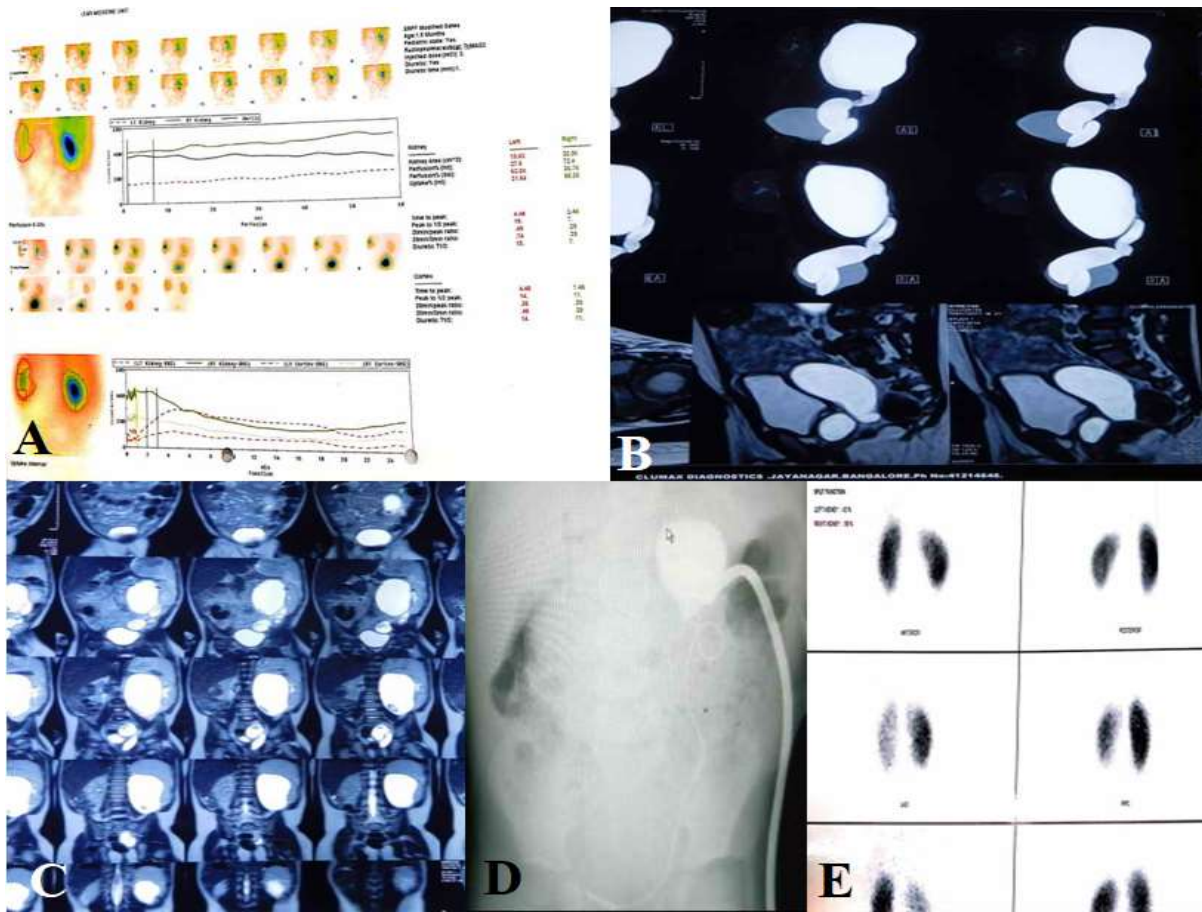


Fig. 1. Imaging studies. **A)** Pre-operative MAG3 Scan showing left hydronephrosis with a differential function of 15%. **B, C)** MRU (Magnetic resonance urography) images - showing severe hydronephrotic pelvis with giant tortuous dilated, tortuous, sacculated partially communicating upper pole ureter with thinned out upper pole moiety renal parenchyma, having its lower end opening abruptly in the posterior urethra with its stenotic opening. **D)** Dye study through the pigtail catheter showing sacculated non communicating pelvis. **E)** Post-operative DMSA Scan showing markedly improved function without any scars/pyelonephritic focus with a differential function of 42%.

made as per Wagers Meyer's law. Because of such a diagnostic dilemma, a magnetic resonance urography (MRU) was taken, showing the giant sacculated, tortuous, partially communicating or non-communicating mega-hydroureter with a maximum diameter of about 5 cm and severe hydronephrotic pelvis with a maximum anteroposterior diameter of about 12 cm covering the entire left abdomen, and confirming the left duplex kidney (Fig. 1 B, C, D, E). To explore the left duplex moiety, a left

lateral incision was made, where only the lower moiety kidney was appreciated, again with the giant mega-hydroureter and hydronephrotic pelvis. The previous uretero-ureteric anastomosis between both moieties of the ureters was then dismantled and the continuity of the lower ureter was reestablished. The giant hydroureter and the mega pelvis, all about 20 cm long, were almost completely excised as its upper pole moiety was not appreciated and therefore could not be traced. (Fig. 2. A, B, C, D). Post-operatively

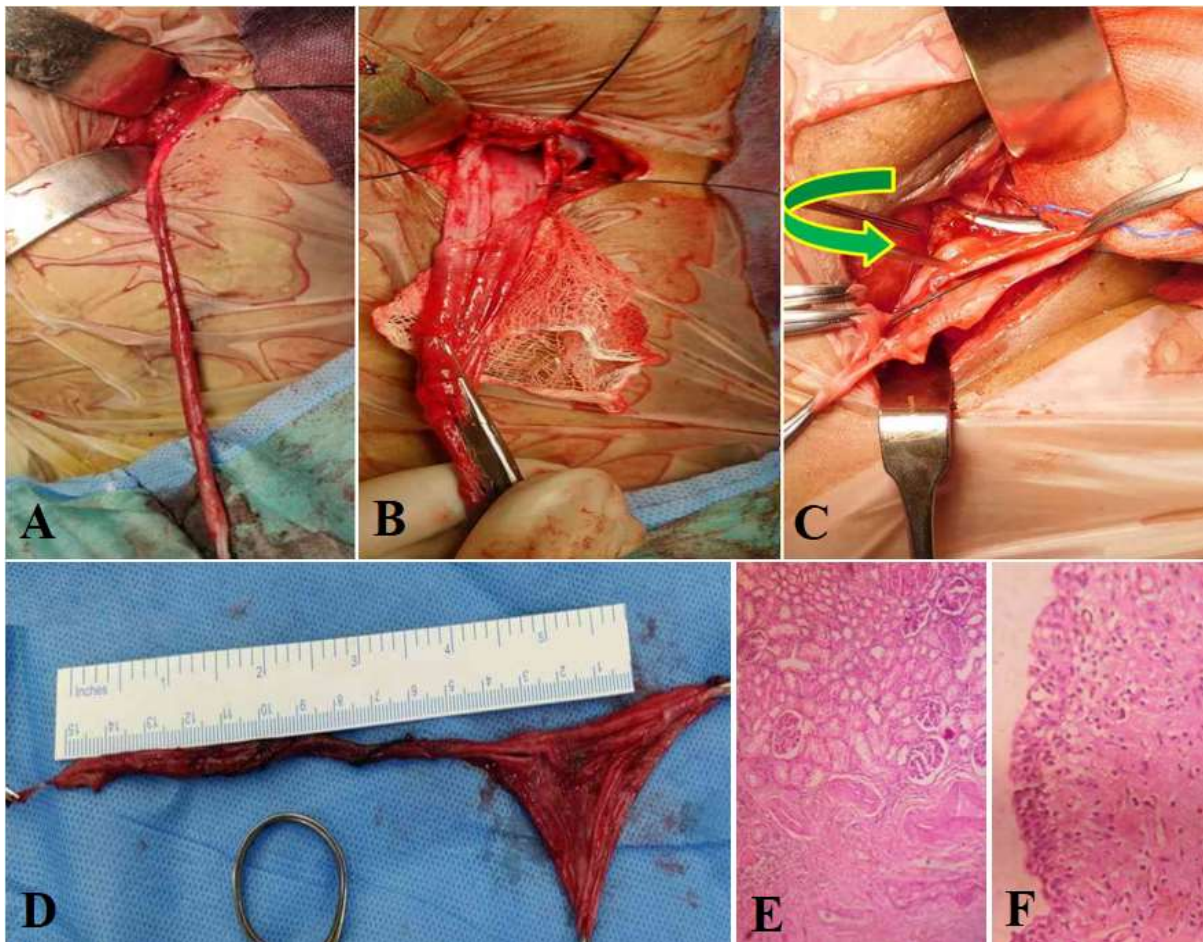


Fig. 2. Intra operative pictures. **A)** A dilated pelvis with part of its excised ureter. **B)** Laid opened giant hydronephrotic pelvis. **C)** Left hydronephrotic pelvis with a green arrow pointing at its upper moiety parenchyma-completely excised. **D)** 50 % of the excised pelvis with its ureter around 20cm in length. **E,** **F)** Histopathological pictures. **E)** 40X (High power fields) showing upper pole renal parenchyma with chronic inflammatory infiltrates. **F)** Ureter showing atrophic epithelium.

baby persisted to have giant mega hydroureter as well as pelvis in spite of dry out put through the abdominal drain, hence a pigtail catheter was inserted in massively hydronephrotic upper moiety pelvis to decompress the system; dye study through this catheter surprisingly revealed, non-communicating dilated pelvis without having urinary ascites without any symptoms (Fig. 1 D). Baby underwent left dorsal lumbotomy approach to trace the upper moiety which was easily got with its capsule intact without disturbing lower pole moiety; excised and sent for histopathological examination which showed chronic

inflammatory infiltrate in the renal parenchyma with atrophic to ulcerated urothelium of the pelvis as well as of the giant hydroureter (Fig. 2 D, E, F). Post operatively dimercaptosuccinic acid (DMSA) nuclear scan done showed drastically improved overall left kidney function with 42% function without any scars or pyelonephritic focus (Fig. 1E). Baby is doing well without any symptoms.

Discussion

The nephric duct from intermediate mesoderm, at its cranial end branching gives rise to the ureter, renal pelvis and calyces, whereas the

distal part of the nephric duct (ureteric bud) incorporates into the urogenital sinus giving rise to the trigone of the bladder. Premature branching of the ureteric bud results in an incomplete duplex with ureters. If more than one bud develops and migrates to the metanephros a duplex kidney with two separate complete ureters are formed [1,2], wherein the upper pole ureter usually drains caudal and medial to the lower pole ureter as per Weigert Meyer rule (law) having associated poorly formed, dysplastic upper moiety. The ureter from the lower pole can either have a normal insertion or associated with vesico ureteral reflux (VUR) [1-7].

Common complications of duplex system include vesicoureteric reflux, ureterocele, pelviureteric junction obstruction, and recurrent urinary tract infection. Few cases of hydroureter associated with duplex system have been reported [1-7].

Pediatric magnetic resonance (MR) urography can be used to thoroughly evaluate renal and urinary tract abnormalities that are difficult to identify or fully characterize with other imaging techniques [6,7].

Author had a 2 month male baby with antenatally detected left hydro nephrosis, post-natal evaluation by imaging as well as cystoscopy revealed varied clinical diagnosis. Finally MR Urography confirmed the diagnosis of left duplex moiety having hydronephrotic giant tortuous, sacculated adynamic, aperistaltic ureter opening with its stenotic opening in the posterior urethra just above the bladder neck. In view of its rarity in its presentation, the confounding unusual imaging as well as intra operative findings due to altered embryological mechanism and hence challenges in its successful management.

Ureteral recanalization at 28 days of gestation follows transient luminal obstruction at 37 to

40 days of gestation, before re canalizing; begins in the mid ureteric region and extends cranially and caudally due to cellular apoptosis. The Chwalle membrane, between bladder and ureter, if abnormal results in ureterocele and incomplete ureteral luminal cellular apoptosis [6,7].

In this case, the defective or incomplete cellular apoptosis of ureteric bud as well as defective Chwalle membrane resorption may be the possible embryological explanation for giant, sacculated, tortuous non draining megaureter.

Conclusion

Alertness as well as completely focused attitude towards the knowledge on the possible mechanisms of embryopathology on the part of pediatric urologist as well as radiologist in assessing these intriguing pediatric urological anomalies can be successful in avoiding morbidity in neonates as well as in children for their better outcome.

Compliance with ethical statements

Conflicts of Interest: None.

Financial disclosure: None.

Consent: Patient confidentiality has been maintained and written consent has been obtained from the patient's parents for the publication of patient information and clinical pictures and can be provided as required.

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