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Segmental Cystic Dilatation of Ureter in Children

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Segmental cystic dilatation of ureter is extremely rare in children. In this paper, we present and discuss an infant with a segmental cystic dilatation of ureter. While the proximal and distal ureteral segments were normal calibration and peristaltic activity, a cystic dilatation in the middle of ureter was found. Excision of the dilated segment of ureter and end-to-end ureteric anastomosis was performed. This rare anomaly should be kept in mind in the differential diagnosis of the abdominal masses in infancy.
Key Words: Ureter, Cystic Dilatation of Ureter, Segmental Megaureter, Megaureter, Children

Çocuklarda Üreterin Segmental Kistik Dilatasyonu

"Segmental megaüreter" veya "segmental üreteral kistik dilatasyon" oldukça nadir görülen bir patoloji olup literatürde sadece birkaç olgu sunumu bildirilmiştir. Bu çalışmada sol taraf segmental megaüreter ve karşı taraf inkomplet üreteral duplikasyonu olan bir olgu sunulmuştur. Skrotal şişlik şikâyeti ile başvuran 2 aylık erkek olguda sağ hidrosel ve sol inmemiş testis saptandı. Olgunun antenatal ultrasonografisinde sağ renal pelviste dilatasyon saptanması üzerine postnatal değerlendirmesinde sol üreter orta kısmında kistik dilatasyon ile beraber sol renal pelvis ile distal üreterde hafif dilatasyon gözlendi. Laparotomide sol üreter orta kısımda belirgin kistik dilatayon ile birlikte sol renal pelvisin ve üreter proksimal ve distalinin normal olduğu görüldü. Dilate segment eksize edildi ve üretere uç uca anastomoz yapıldı.
Anahtar Kelimeler: Segmental Megaüreter, Üreteral Kistik Dilatasyon, Megaüreter, Üreter, Çocuk

Introduction

Congenital anomalies of the kidney and urinary tract account for more than 50% of abdominal masses found in neonates and involve some 0.5% of all pregnancies.[1,2] Megaureter is a considerable condition in these anomalies. Megaureter is a generic term indicating the presence of an enlarged ureter with or without concomitant dilatation of the upper collecting system. The normal diameter of ureter is usually 5 mm in children and rarely exceeds this size.[3,4] In practice, a ureter with a diameter of 7 mm or more should be considered as a megaureter. Megaureter may be categorized as primary or secondary, refluxing or nonrefluxing, obstructed or non-obstructed, and nonrefluxing unobstructed.In all of these conditions ureteric dilatation is found completely. However, segmental megaureter or segmental cystic dilatation of ureter is a very rare entity and only a few case reports were reportedin the literature.[5,6]

In this paper, we present a 2-mo-old infant with a segmental cystic dilatation of left mid-ureter and discuss in regard to differential diagnosis.

A 2-mo-old infant presented with scrotal swelling. Physical examination revealed a right congenital hydrocele and left undescended testis. His postnatal history presented that his ultrasound on postnatal 7th day demonstrated a normal right collecting system and a minimally dilated left renal pelvis with distal ureteric cystic dilatation suggesting an ureterocele. The voiding cystourethrogram showed no vesicoureteral reflux and no filling defect in the bladder. Intravenous urography (IVU) showed incomplete duplication of the right upper urinary tract. Left ureter was not visualized on IVU. Renal scintigraphy demonstrated a decreased function on the left side (right 64%, left 36%). Computed tomography scan showed a cystic mass with 5x5x4 cm in size (Fig 1). The blood and urine analysis were normal.

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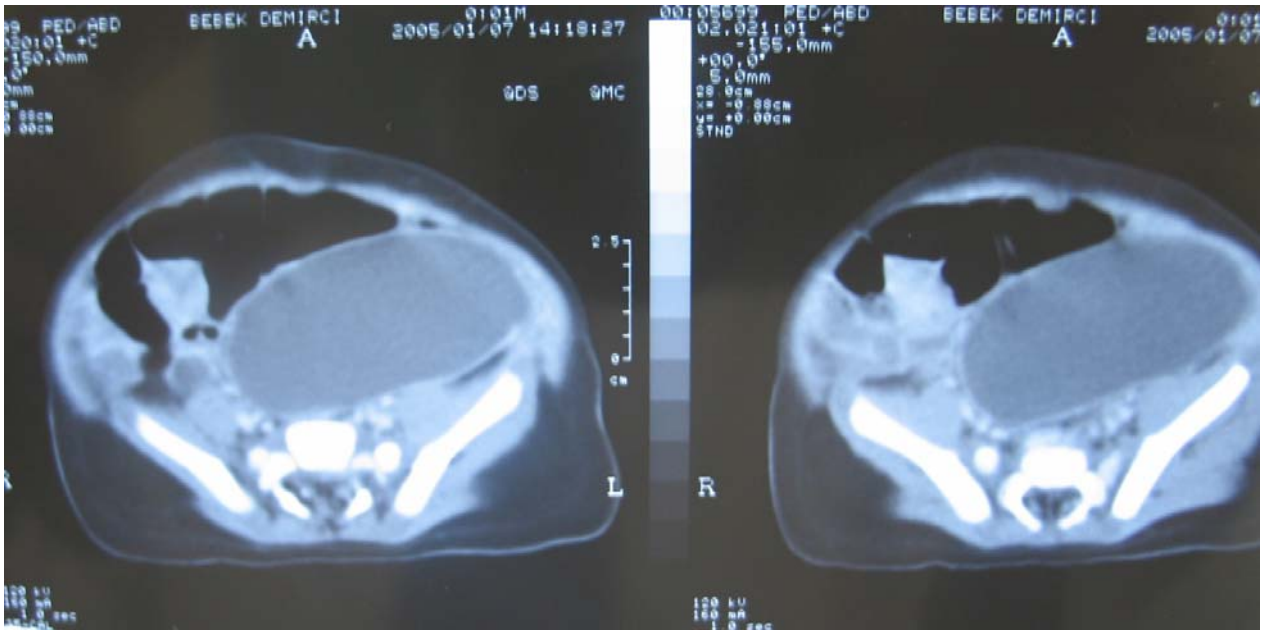

Fig 1. Computed tomography scan shows an extremely cystic mass in the abdomen.

During laparotomy, cystic dilatation of the left mid-ureter and incomplete duplication of the right upper urinary tract was found. Approximately 60 ml urine was aspirated in from dilated segment (Fig 2). The left renal pelvis and proximal and distal ureteral segments showed normal calibration and peristaltic activity (Fig 3A). Excision of the dilated segment of ureter and end-to-end ureteric anastomosis over a transanastomotic stent was performed (Fig 3B). The postoperative period was uneventful.

Histological examination of the dilated segment of

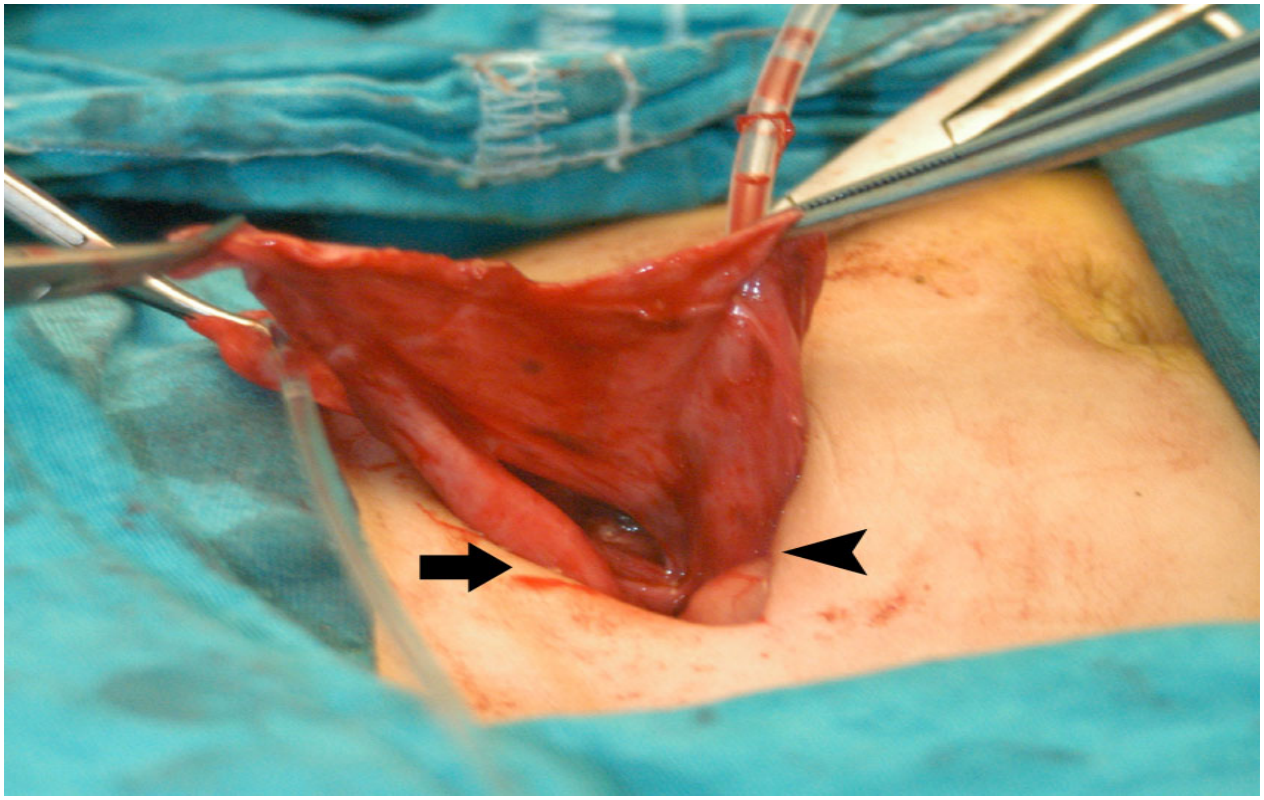

Fig 2. Cystic dilatation of the left mid-ureter with normal proximal (arrow head) and distal ureter (arrow).

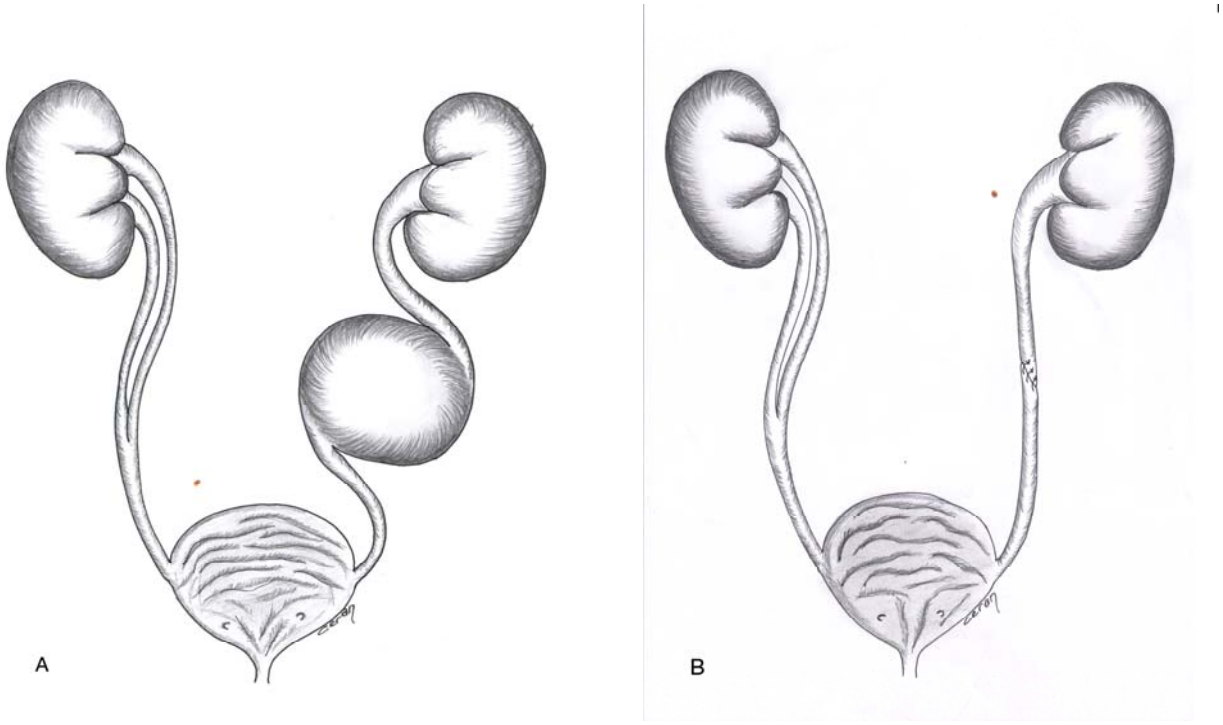

Fig 3. This illustration shows an incomplete duplication of the ureter and a bifidrenal pelvis on the right side and midureteric segmental dilatation on the left side **(A),** after midureteric excision, the remaining elongated ureteral segments were enough to perform an end-to-end anastomosis **(B).**

ureter revealed loss of normal urothelium.

Discussion

Only a few case reports have been found about segmental megaureter or segmental cystic dilatation of ureter in the literature. In 1986, four cases of congenital megacalycosis associated with ipsilateral segmental megaureter have been presented by Mandell et al.[5] In all of their cases, distal ureteric dilatation had been found and those are similar to primary megaureter secondary to a narrowed and aperistaltic distal ureteral segment more than segmental megaureter. In 1995, a case of congenital segmental megaureter with sparing of the proximal and distal ureter, as in our case, has been reported by Ramaswamy at al.[6] A similar case but with bilateral segmental megaureter was presented Pinter et al in 1997.[7] In 2004, from Spain, Soler reported a case of a case of unilateral multicystic dysplastic kidney and contralateral megacalycosis associated with ipsilateral distal segmental megaureter.[8] Recently, Prieto et al reported a case of a congenital severe midureteral dilatation associated with mild proximal ureteral dilatation and a distal ureter with a normal diameter.[9] Distinctly, the present case that had segmental megaureter associated with contralateral incomplete ureteric duplication is the first case in literature.

Several theories have been speculated regarding the pathophysiology of segmental megaureter. Ramaswamy suggested that segmental megaureter was a variant of nonrefluxing megaureter and attenuated nexuses and thin myofilaments might be responsible from this entity, as in their case.[6,10] Although, in another study, it was suggested that aganglionosis might be liable the absence of ganglion cells has not subsequently been confirmed.[5] Pinter speculated that recanalization of the solid ureteral duct, if abnormal, might produce segmental ureteral dilatation.[7] However, there is no information about innervation patterns in dilated ureteric segment.

The treatment of segmental cystic dilatation of ureter should be planned according to the ipsilateral kidney function and length of the normal ureteric segment. If the remaining parts of distal and proximal ureter have enough length, end-to-end ureteroureterostomy after excision of the segmental megaureter should be performed as in our case. If excision is not possible, tailoring or trimming of the dilated segment should be considered. If the ipsilateral ureterorenal unit is poorly functioning, hypoplastic or dysplastic, a nephroureterectomy should be planed.

Segmental cystic dilatation of ureter must be distinguished from ureterocele and other conditions

such as primary megaureter or ureteral diverticulum. At the same time, in the cases of segmental megaureter, the patient should be investigated for possible associated urinary system anomalies such as megacalycosis, duplication of collecting system and hypoplastic, dysplastic or nonfunctioning kidney.

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