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Congenital giant megaureter associated with ipsilateral multicystic dysplastic kidney in newborn

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Abstract

Congenital giant megaureter presents as abdominal mass and impose diagnostic difficulties. It can be associated with other upper urinary tract anomalies. A female newborn with antenatal diagnosis of polycystic kidneys was admitted at birth due to lower abdominal mass. Ultrasound and CT scans diagnosed a multiloculated cystic lesion in the mid and lower abdomen along with right side multicystic kidney. At laparotomy, an extaperitoneal, lobulated cystic swelling was found due to rightside giant megaureter. Its lower end was of normal caliber and orthotopic. End cutaneous ureterostomy was done. Intravenous urogram and isotope renograms showed nonfunctioning right kidney. She also had grade II vesicoureteral reflux on left side. Child suffered urinary infection twice. At 9m age, right nephroureterectomy was done. Histopathologic examination was consistent with cystic renal dysplasia and dilated ureter. This is the first case report of giant megaureter associated with ipsilateral multicystic dysplastic kidney in newborn.

Keywords

Congenital giant megaureter; abdominal mass; newborn; multicystic dysplastic kidney, obstructing megaureter; ureterostomy.

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Introduction

Congenital giant megaureter (CGM) is characterized by giant ureteral dilatation producing elongated and tortuous ureter. Giant megaureter presents as protuberance of abdomen or lower abdominal mass [1]. CGM can be associated with other upper urinary tract anomalies [1]. To our knowledge, here we report the first case of CGM associated with ipsilateral multicystic dysplastic kidney (MDK).

Case Report

Female newborn, fullterm, having 2900g birthweight, with maternal polyhydramnios and antenatal diagnosis of polycystic kidneys, was admitted at birth due to abdominal mass. Two soft, cystic masses were palpable. One was in hypogastrium and right iliac region, with well-defined borders, and movable transversely. Its compression did not evacuate urine. The upper mass was in right hypochondrium and not movable. Ultrasound (US) scan of abdomen on 3rd day of birth showed enlarged right kidney (RK) with increased echogenicity and cysts of variable size suspicious of multicystic dysplastic kidney (MDK). Scan also showed anechoic lesions in lower abdomen, largest one measuring 40x32mm, with moving debris suggestive of cystic lesions. Renal function tests (RFT) and serum electrolyte values were normal. CT scan of abdomen at 35d age showed multiloculated lesion 85x65x50mm in the pelvis and midabdomen [Fig.1, Fig.2].

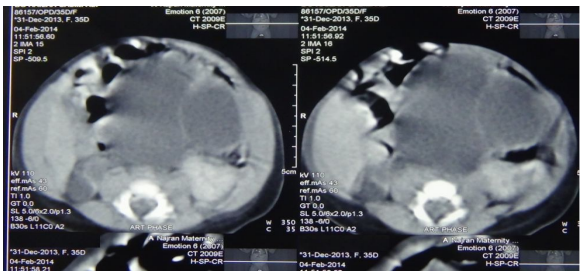

Fig. 1. Abdominal CT scan showing large low attenuation multiloculated lesion (?multiple low attenuation lesions) in the midabdomen with no enhancement after injection of intravenous contrast.

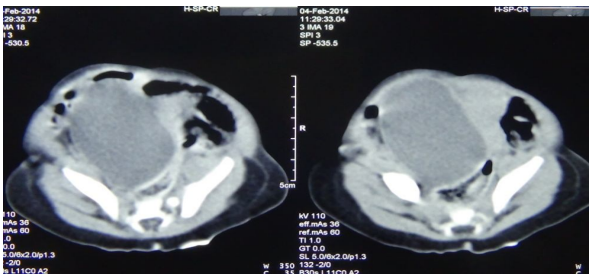

Fig. 2. Pelvic CT scan showing the cyst arising mostly from right side of pelvis and pushing the urinary bladder to left. The cyst was extending upwards to midabdomen.

It pushed urinary bladder (UB) to left side [Fig. 3].

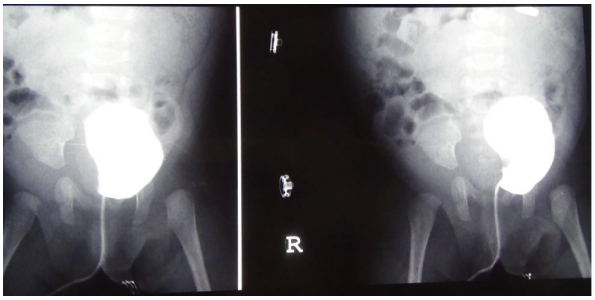

Fig. 3. Retrograde cystogram showing the urinary bladder pushed to left by the lesion in pelvis.

RK had multiple small cysts (largest one measuring about 15x12mm), doubtful extrarenal pelvis (?hydronephrosis) and proximal hydroureter. Diagnosis was large right ovarian cyst (vs. lymphangiomatous mesenteric cyst) and right renal cysts. At the age of 81d, laparotomy was done by right lower transverse incision. An extaperitoneal, lobulated cystic swelling was seen in right side of pelvis, pushing UB to left. After partial emptying the cyst of clear fluid, it was dissected to find that it was giant megaureter [Fig. 4A, B] of primary obstructing type with orthotopic insertion into bladder.

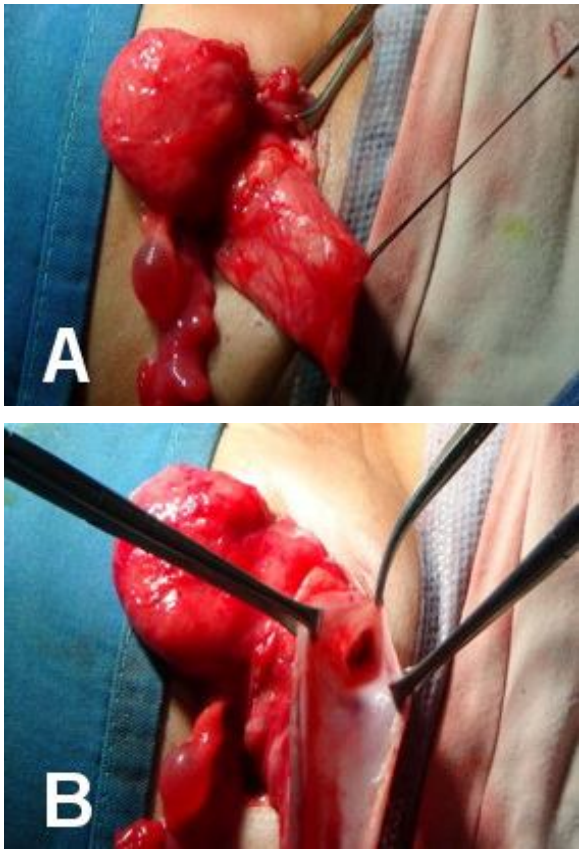

Fig. 4 Peroperative pictures: (**A**) Giant megaureter seen after partial evacuation of fluid from the lobulated cystic swelling. **(B)** Picture showing the normal caliber of terminal ureter after splitting open the lower part of giant megaureter. It also had orthotopic insertion into urinary bladder.

End-cutaneous ureterostomy, UB repair and urethral-catheter drainage were done. There was scanty urine output from ureterostomy and normal output from UB. Retrograde ureterogram (RGU) from ureterostomy showed dilated ureter without showing pelvicalyceal system (PCS).

Intravenous urogram done at 90 days of age showed nonvisualised RK and normal left kidney (LK) and UB. At 5m age, US showed atrophic (25x 14mm) RK, normal (57X32mm) LK and no left ureteric dilation. ^{99m}Tc-dimercaptosuccinic acid static renal scan did not show RK. LK showed uniform tracer uptake, without any scarring. ^{99m}Tc-diethylene triamine pentaacetic acid dynamic renal scan showed no tracer handling in RK. LK was well perfused with normal uptake, transit & drainage, showing good response to lasix injection. Left renogram was normal. Split renal function was: LK=100%, RK=0%. Estimated total glomerular filtration rate (GFR) = GFR of LK= 93.3ml/mt. At 6m age, voiding cystourethrogram (VCUG) showed grade II vesicoureteral reflux (VUR) on left side. She was started on prophylactic dose of trimethoprim + sulphamethoxazole.

She had urinary tract infection (UTI) twice. At the age of 9 months RFT results were normal. Right nephroureterectomy was done [Fig. 5].

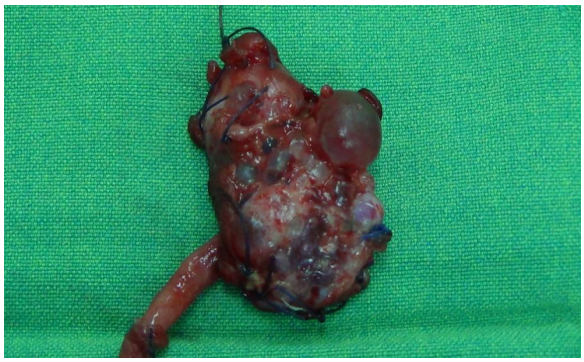

Fig. 5. Nephroureterectomy specimen showing atrophied and multicystic kidney (size: 35x20x10mm) and dilated ureter.

Histopathologic examination revealed disorganized renal parenchyma disrupted by cysts of various size lined by flat epithelium which was consistent with cystic renal dysplasia. The report was consistent with cystic renal dysplasia. At the age of 15 months, VCUG showed complete disappearance of VUR.

Discussion

CGM is a very rare unilateral urinary anomaly, which is defined as "ureter whose lumen is congenitally, focally and segmentally dilated to more than 10 times the normal diameter" [2]. The definition of CGM is purely descriptive; it can be secondary to several different diseases. Huang [1] reported 21 patients aged 2m to 8y with CGM. These cases presented with marked protuberance of the mid and lower abdomen. These ureters usually have a dysplastic and disorganized muscular coat, lined with a columnar epithelial mucosa rather than the usual transitional epithelium. The associated kidneys were hypoplastic, dysplastic, or had mild hydronephrosis and atrophic renal parenchyma. Function and volume of the bladder was normal. Twelve of 21 patients had duplex collecting system on the affected side. Associated congenital anomalies of urinary tract such as ureterocele, ectopic ureter [3,4], crossed ectopic ureter, ureteral atresia [5], supernumerary kidneys [6] and duplex urinary system are very common. Giant megaureter is most commonly associated with duplex upper urinary system [1,5-8].

Giant megaureter can present as huge cystic intraabdominal mass and should be differentiated from other retroperitoneal cysts such as hydronephrosis, teratoma, duplication cyst and lymphatic cyst and also adnexal masses in females. These masses are usually round to oval in contrast to the tubular and tortuous shape of the megaureters. Ureteral dilatation can be recognized sonographically by its tubular appearance and anatomic course. However,

when the megaureter became so enlarged and folded upon itself, it was no longer tubular but appeared as a cystic, partially septated mass in US scan and CT scan of our case. Giant megaureter was our unexpected operative finding. It was primary obstructing type of megaureter, because there was no ipsilateral VUR, it was inserted in orthotopic position, there was no lesion in terminal ureter such as ureterocele and there was no vesical dysfunction. Primary obstructing megaureter is due to abnormal terminal ureter referred to as 'adynamic segment' which is aganglionic [9].

MDK is the most common form of renal cystic disease and is one of the most common causes of abdominal mass in infancy. It is usually associated with atretic ureters [10]. Moralioğlu [11] published data of 68 patients with MDK. Ten patients (14.7%) had additional urological anomalies in contralateral kidney such as VUR, megaureter, cortical renal cyst, ureteropelvic junction obstruction, and renal ectopy. No patient had ipsilateral megaureter. Recurrent UTI was the most common indication for nephrectomy. Contralateral VUR is seen in 18% to 43% cases of MDK [10].

Ureterostomy was done as the primary surgery in our case to allow the megaureter to recover and for the kidney function to improve following free outflow of urine. RGU demonstrated improvement of diameter and tortuosity of giant megaureter; more recovery was evident from nephroureterectomy specimen. Renal function did not improve, but deteriorated to zero and child got repeated UTI. These were the indications for nephroureterectomy. CGM were reported in newborns [2,4,5] or there was history suggestive of mass since birth in other cases of CGM [7,8]. Giant megaureter associated with polar dyplastic kidney has been reported [7,12,13]; one of these had upper pole multicystic dysplasia [13]. MDK associated with ipsilateral CGM has not been reported so far in English literature; this is the first such case.

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