Case Reports

Multicystic renal dysplasia

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ABSTRACT

Multicystic renal dysplasia, the most common form of cystic renal disease in the newborn period, is a clinically important consequence of abnormal nephrogenesis. It usually presents as an abdominal mass. The dysplasias are usually unilateral, but it can be bilateral, segmental or focal. The clinical presentation usually depends on the extent of the dysplastic involvement and the degree of the associated urinary obstruction. Here, we present a case of histologically multicystic renal dysplasia, which is ?bilateral. The left kidney showed typical radiological, gross and histopathological features of multicystic dysplasia, but the right kidney showed only radiological features of dysplastic cystic kidney.

Keywords: Dysplasia, kidney, multicystic.

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Dysplasia is a developmental disorder and may affect several organs in embryonic life. The most common form of cystic renal disease in the newborn period is multicystic renal dysplasia, others being, multilocular cysts of the kidney, polycystic disease, solitary renal cyst and multiple simple cysts of the kidney. Multicystic renal dysplasias are usually sporadic, but when associated with other genetic syndromes, it can be either autosomal recessive or autosomal dominant. Anomalous differentiation of the metanephros results in the disorganized development of the kidney. The clinical presentation usually depends on the extent of the dysplastic involvement and the degree of the associated urinary obstruction.

Case Report. A 7 month old female of a non-consanguinous couple, delivered at the end of full term presented with abdominal mass. There was a prenatal diagnosis of polycystic disease of the kidney at the 28th week of gestation and the patient was kept on antibiotics. There was no family history of similar illness or developmental defects. There were no urinary symptoms. Radionuclei scan (MAG-3) was performed to asses the renal functions which, showed poor functioning of the left kidney with preserved renal function in the right kidney. Biochemical profile of the kidneys was within normal limits (Table 1). Failure to thrive was noticed. Ultrasonography showed enlarged kidneys bilaterally, with multiple cysts on the left side. The right kidney was hyperechoeic and the size increased over a 6 month period, suggesting multicystic renal dysplasia (bilateral). The right kidney measured 9cms and the left 6cms. Other organs were normal with no hepatosplenomegaly. Micturition cysto-urethrogram showed bladder capacity of 50ml with smooth outline and patient could evacuate the bladder completely with no vesicoureteral reflux. At operation, the left kidney was ectopic in location, having atrophic ureter, pelvis and renal vessels. There was no renal contour and the kidney was entirely replaced by multiple cysts giving an appearance of "bunch of grapes" (Figure 1). The right kidney was apparently normal. Left nephrectomy was performed. The post-operative period was uneventful. The patient is improving with increasing weight till date. The nephrectomy specimen was subjected to histological examination. The left kidney was irregularly bossellated in shape

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Table 1 - Pre and postoperative biochemical profiles.

Parameters/ Periods	Weight (kgs)	Hb (gm%)	MSU	Urea (mmol/L)	Na (mmol/L)	K (mmol/L)	Cl (mmol/L)	HCo3 (mmol/L)	Creatinine (mcmol/L)
3rd PND	2.6	15	PH neutral RBC+++ WBC 3-5/hpf *E.coli*+	2.4	138	4.7	101	24	61
Preoperative	7.5	11.5	PH Acidic WBC 6-8/hpf	2.7	137	5.5	104	23	29
1st Postoperative	NA	12	NA	3.9	137	4.2	107	22	34
4th month Postoperative	8.1	12	PH Acidic WBC 15-20/ hpf	3.4	134	4.5	101	22	34

MSU-Microscopic urine, Na-Sodium, K-Pottasium, CI-Cloride, HC03-Bicarbonate, PND-Post natal day, hpf-high power field, NA-not available, Hb - Hemoglobin, RBC - Red blood cell count, WBC - White blood cell count, *E.coli - Escherichia coli*

and measured 6x4x2.5 cms with a ureter measuring 3.5 cms long. The outer surface was uneven and cystic. On sectioning, it showed multiple cysts of varying sizes measuring from 2.2 to 0.75 cms containing clear fluid. The wall was smooth in texture. Microscopically, multiple sections studied showed several cysts of varied dimensions. These are lined by low cuboidal epithelium with loosely cellular, fibroadipose stroma in between. In addition, there are immature tubular elements, sclerotic, premature glomeruli embedded in primitive mesenchyme (Figure 2). There are several blood vessels and nerve bundles but no collecting system including development of calyceal and pelvic components noticed in the specimen. The ureter is obliterated subtotally.

Discussion. In 1936, Schwartz[1] first described multicystic kidney disease in a 7-month old child. The main characteristic of this lesion was the complete replacement of the kidney by multiple cysts that were held together by loose connective tissue, similar to a bunch of grapes. Ultrasound criteria for the diagnosis of multicystic renal dysplasia included, absence of renal parenchyma, absence of normal renal sinus echoes, multiple cysts of varying sizes that did not communicate and nonmedial location of the largest cyst. Spence first described the characteristics of this disease.[1] He believed that multicystic kidney should not be confused with multilocular cysts of the kidney, polycystic disease, solitary renal cyst or multiple simple cysts of the kidney. Renal dysplasia is usually, but not

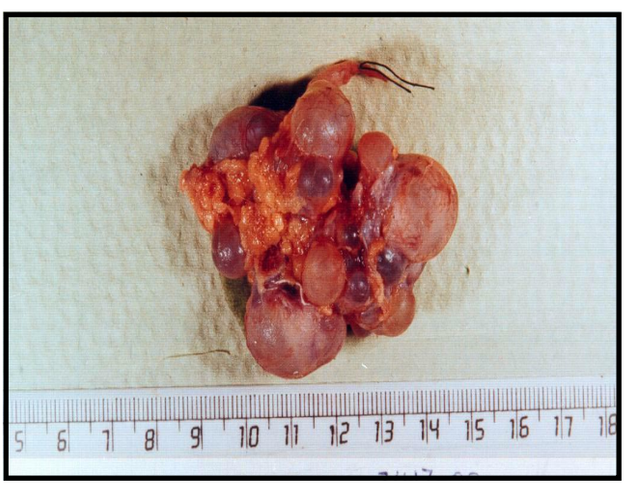

Figure 1 - Kidney with loss of renal contour and replaced by multiple cysts giving an appearance of bunch of grapes.

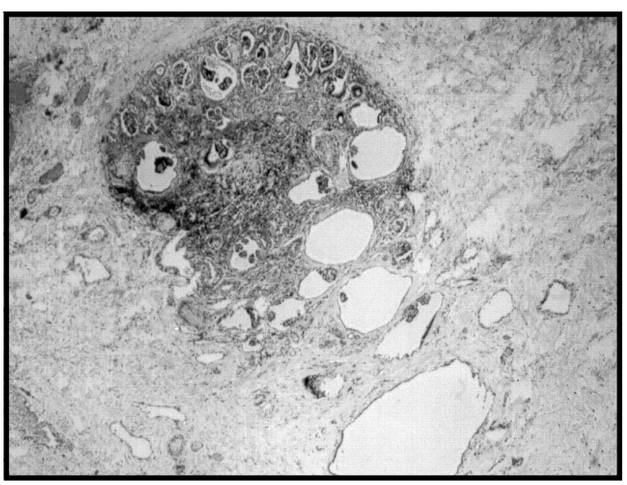

Figure 2 - Several cysts lined by low cuboidal epithelium, loose fibroadipose tissue (single arrow) and immature cellular stroma with tubular elements, sclerotic, premitive glomeruli (double arrow). (100X, Hematoxylin & Eosin stain).

invariably, a unilateral disease associated with ipsilateral atresia or hypoplasia of the ureter and renal pelvis.[5] However, neither the nature, nor the prevalence of the contralateral lesion has been delineated.[3] Renal dysplasia is a congenital lesion believed to have been acquired in utero as the result of failed coordination of development of the metanephros and the branching ureteric bud.[6] Dysplastic kidneys may be small and solid or large with a variable number of cysts, the later condition being referred to as multicystic dysplastic kidney disease (MCDK). Gorden et al[7] estimated that the incidence of unilateral MCDK disease was 1 in 4300 live births. Atiyeh et al[3] found associated urologic abnormalities in 51% of the patients of which the abnormality was contralateral to the MCDK in 39%, ipsilateral in 6% and involved the bladder wall in 6%. The most common genitourinary tract abnormality was vesicoureteral reflux (18%) followed by ureteropelvic junction obstruction (12%). Dysplasia implies misguided development and differentiation of kidney precursor cells. The common features include distortion of the renal architecture, the presence of immature forms such as primitive glomeruli and ductules surrounded by collors of fibromuscular cells, and the formation and expression of ectopic tissue such as cartilage.[4] It is the most common cause of a palpable abdominal mass in the newborn infant. Grossly, the kidneys are reniform in shape and architecture and in the majority of cases associated with an atretic ureter. Postnatally, the histopathology of multicystic dysplastic kidneys is extremely variable, which includes cystic transformation of primitive tubules, an expanded and disorganized supporting interstitium with marked peritubular reaction and the formation of such elements such as cartilage, and variable development of primitive glomeruli and the paucity of proximal tubular formation.[4] Usually these kidneys are considered to be non-functioning.

Urinary tract infections have been associated with MCDK disease.[3] Flack and Bellinger[2] consider that contralateral vesicoureteral reflux in association with multicystic renal dysplasia puts solitary kidneys at risk of pylonephritic scarring and recommends prophylactic antibacterial therapy in children with proved contralateral vesicoureteral reflux. In their series, they found all non-operated children showed spontaneous resolution of vesicoureteral reflux or diminution to grade 1 by age 20 months. Out of 441 cases reported to The American Academy of Pediatrics Multicystic Kidney Registry (1986), 260 were followed non-operatively.[1] Among those, no infant required surgery for abscess or recurrent urinary infection, or no cases of Wilm's tumor have been reported. Since the risk for significant complications in multicystic kidney is extremely low, it is believed reasonable to follow these patients non-operatively and recommend ultrasound every 3-6 months during the first year of life, and repeat ultrasound every 6-12 months until age 5 years, since it appears that most multicystic kidneys may take as long as 20-25 years to resolve or resorb, families and physicians should be prepared for long term follow up.[1] In the isolated multicystic dysplastic kidney, altered growth factor gene expression has been described both in the cystic epithelia and the disorganized interstitium.[4] Over expression of factors that promote the formation of cystic changes in the epithelia of the tubular elements including Pax2 and bcl-2[4,8] as well as underexpression of bcl-2 and increased apoptosis in areas of mesenchymal dysplasia have been demonstrated.[4,9,10]

In conclusion, multicystic disease of the kidney is a frequently encountered renal cystic disease, often unilaterally. In this report, MCDK has been diagnosed in a newborn with bilaterality. Current understanding in the pathogenesis of MCDK, especially unilateral may not be totally incompatible with long term survival.

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