Wilms' Tumor Complicating Multicystic Dysplastic Kidney (MCDK)

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Abstract

A 3-year old girl was diagnosed to have bilateral vesicoureteric reflux since the age of 1 year, proved to have Wilms' tumor complicating multicystic dysplastic kidney (MCDK).

Introduction

MCDK is a congenital maldevelopment in which the renal cortex is replaced by numerous cysts of multiple sizes; a dysplastic parenchyma anchors the cysts, the arrangement of which resembles a bunch of grapes. The calyceal drainage system is absent. Typically, MCDK is a unilateral disorder; the bilateral condition is incompatible with life (1). Various types of MCDK had been recognized; the classic, the hydronephrotic and the solid cystic dysplasia types. MCDK should not be confused with polycystic kidney disease (PCKD) or other renal cystic diseases (2).

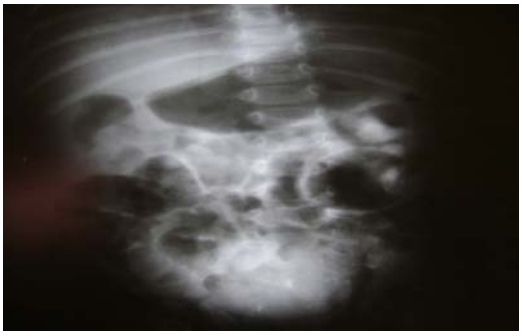
MCDK was recognized as a distinct entity in 1955 (3). The incidence of MCDK ranges from 1:2400 – 1:4300 live births (4, 5). Prenatal ultrasound scans detects 77 – 88% of cases of MCDK (6). The condition is more commonly diagnosed in boys (7) and the left side (8). In over 50 % of cases other urinary tract defects are also detected; uretropelvic junction obstruction (UPJO), vesicoureteral reflux (VUR) and ureterovesical junction obstruction (UVJO) are the most common defects in the contralateral kidney (9, 10).

Case Report

H. M. is a 3-year old girl presented to the maternity and children teaching hospital in Diwaniyah on 12.4.2008 with 1-week history of pallor, rapid breathing and anorexia. Her condition dated back to the age of 8 months when she started to have recurrent attacks of fever, vomiting and crying during micturition. The diagnosis of recurrent urinary tract infections was presumed. At the age of one year, investigations were done for

her and the diagnosis of bilateral hydronephrosis and hydroureter due to vesicoureteral reflux (VUR) was suggested. She was maintained on urinary (Foley's) catheter and frequent courses of antibiotics. Since that time she had frequent urinary tract infections and failure to thrive.

On examination: a pale, malnourished girl with acidotic breathing. She had neither finger clubbing nor cyanosis. Her heart rate was 110/minute, the respiratory rate was 35/minute, and her temperature was 38.2 c˚ (axillary). Her measurements were: weight 9.250 kg, height 80 cm, OFC 45 cm; all were below the 3rd centile. She had no other abnormality detected in the examination of systems apart from a bimanually palpable right kidney.



Investigations

Hemoglobin 7.5 gm/dl, WBC 8000/cmm, normal platelets count. The RBCs are hypochromic microcytic, the reticulocyte count 1%. Urinalysis revealed an acidic reaction with pus cells of 20/HPF, the random blood sugar was 55 mg/dl, blood urea 4 m mol/l (normal 3.3 – 7.5 m mol/l), serum creatinine 68 m mol/l (normal 62–124 m mol/l), and urine culture: growth of Pseudomonas aeroginosa sensitive to cefotaxime.

H.M. was admitted to the hospital & received parenteral antibiotics with sodium bicarbonate, oral fluids, oxygen and antipyretics. She received blood transfusion, the Foleys catheter was reviewed. Abdominal ultrasound showed bilateral hydronephrosis and hydroureter, the parenchymal thickness was between 6–8 mm. Voiding cystourethrography (VCUG) and renal scintigraphy (99m Tc-DMSA) are not available in this hospital. Intravenous urography (IVU) revealed a completely non-functioning right kidney, the delayed films showed hydronephrosis and hydroureter of the left kidney (Figure 1).

The diagnosis of severe right hydronephrosis and non-functioning right kidney with moderate left hydronephrosis was established and the decision of right nephrectomy was discussed and made. At operation, the right kidney was found large and containing multiple cysts and masses (Figure 2)

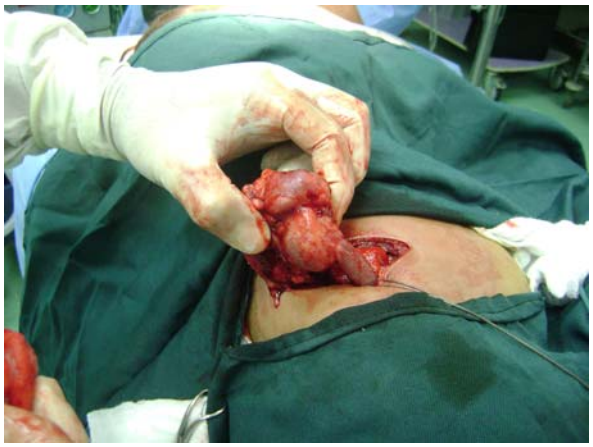


Figure 2

The histopathology examination report showed that the right kidney is affected by Wilms' tumor stage I (Figure 3).

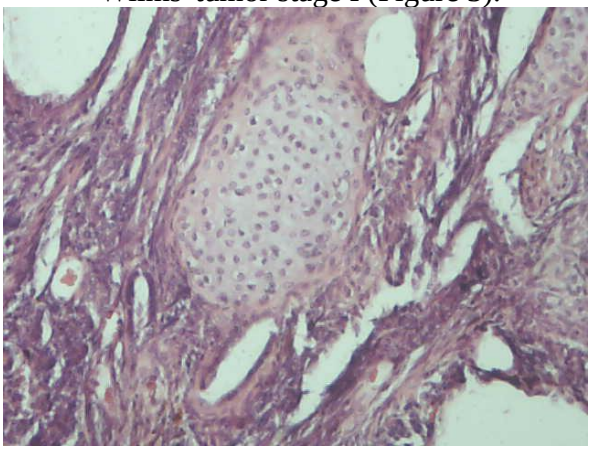


Figure 3

Discussion

The risk of malignant degeneration complicating MCDK is a controversial issue. The risk of Wilms' tumor is 1:10000 in the general population, compared with 1:1000 to 1:3300 in children with MCDK (11), other reports mentioned a risk of Wilms' tumor of 1 in 333 (12). Cystic Wilms' tumor and UPJO can mimic the hydronephrotic form of MCDK, and radionuclide scanning is necessary to confirm the diagnosis. CT scan and MRI are not part of the diagnostic investigation. Renal scarring in patients with MCDK may result from hypoplasia of the contralateral kidney, reflux nephropathy, obstructive uropathy or late postinflammtory lesions complicating acute pyelonephritis.

VCUG is indicated in patients with MCDK to evaluate the urinary tract for VUR and other anomalies (13, 14). Six children with MCDK developed an ipsilateral Wilms tumor, and 5

of 6 adults developed ipsilateral renal cell carcinoma. Embryonic nephrogenic rests, which can be premalignant, are more prevalent in MCDK than in other conditions. The management of MCDK is a controversial topic, few authors have recommended routine nephrectomy to prevent potential complications but the rarity of such complications does not justify the surgical and anaesthetic risk (15, 16). This opinion is not definitely applied to our patient who had a complicated MCDK, both in the ipsilateral and contralateral kidneys.

References

1. Hussein S, Bengum N. Multicystic dysplastic disease of kidney in fetus. J Ayub Med Coll Abbottabad 2007; 19(2):68-9.
2. Levine E, Hartman DS, et al. Current concepts and controversies in imaging of renal cystic diseases. Urol Clin Noth Am 1997; 24(3):523-43.
3. Spence HM. Congenital unilateral multicystic kidney: an entity to be distinguished from polycystic kidney disease and other cystic disorders. J Urol 1955; 74(6):693-706.
4. Belk RA, Thomas DF, et al. A family study and the natural history of prenatally detected unilateral multicystic dysplastic kidney. J Urol 2002; 167:666-9.
5. Feldenberg LR, Siegel NJ. Clinical course and outcome for children with multicystic dysplastic kidneys. Pediatr Nephrol 2000; 14:1098-1101.
6. Ylinen E, Ahonen S, et al. Nephrectomy for multicystic dysplastic kidney: if and when? Urology 2004; 63:768-771.
7. Cascio S, Paran S, Puri P. Associated urological anomalies in children with unilateral renal agenesis. J Urol 1999; 162:1081-1083.
8. Ranke A, Schmitt M, et al. Antenatal diagnosis of multicystic renal dysplasia. Eur J Pediatr Surg 2001; 11:246-254.
9. Cambio AJ, Evans CP, Kurzrock EA. Non surgical management of multicystic dysplastic kidney. BUJ Int Jan 2008(medline).
10. Abidari JM, Park KH, et al. Serial follow-up of the contralateral renal size in children with multicystic dysplastic kidney. J Urol 2002; 168:1821-1825.
11. Beckwith JB. Should asymptomatic unilateral multicystic dysplastic kidney be removed because of furure risk of neoplasia? Pediatr Nephrol 1992; 6:511.
12. Narchi H. Risk of wilms tumor with multicystic kidney disease: A systematic review. Arch Dis Child 2005; 90:147-149.
13. Farnham SB, Adams MC, et al. Pediatric urological causes of hypertension. J Urol 2005;173:697-704.
14. Kuwertz-Broeking E, Brinkmann OA, et al. Unilateral multicystic dysplastic kidney:experience in children. BUJ Int 2004; 93:388-392.
15. Blew B, Carpenter B, Leonard MP. Incidentally detected nephrogenic rests in the setting of congenital obstructive uropathy. Can J Urol 2002; 9(4):1595-8.
16. Wiener JS, Maloney ME. Multicystic dysplastic kidney. eMdicine Reference Feb 19, 2008.