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Multicystic dysplastic kidney: Ten-year evaluation

B U Sowmya[1], V S Nandish[2,*], Prema Saldanha[1]

[1]*Dept. of Pathology, Yenepoya Medical College, Mangalore, Karnataka, India*
[2]*Dept. of Pathology, SSIMS&RC, Davanagere, Karnataka, India*

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ABSTRACT

Introduction: Multicystic dysplastic kidney (MCDK) is the most common form of developmental abnormality seen in infants and children. It is almost always unilateral and slightly more frequent in boys and on the left side. The other associated extra-renal abnormalities include esophageal atresia, tracheoesophageal atresia, ventricular septal defect and patent ductus arteriosus. The routine widespread use of antenatal ultrasonography has led to early detection of this abnormality.

Materials and Methods: We retrospectively assessed fourteen cases of Multicystic dysplastic kidney between January 2004 and December 2014. All the cases underwent total nephrectomy and the specimens were sent to the department of Pathology with their respective clinical details and pathological findings were analyzed.

Results: A total of fourteen (14) patients were diagnosed with unilateral MCDK over a period of 10 years with no sex predilection. Majority were on left side and antenatally diagnosed. Some patients had associated hypertension, vesicoureteral reflux and ureteropelvic junction obstruction. Microscopy revealed renal parenchyma replaced by cysts of variable sizes lined by flattened cuboidal epithelium. Intervening fibrotic stroma with mesenchymal components, primitive glomeruli, nerve bundles and dysplastic ducts.

Conclusion: MCDK is mostly a unilateral sporadic anomaly usually found on the left side, however can be familial. It may be associated with hypertension, vesicoureteral reflux and ureteropelvic junction obstruction. Most of the patients can be diagnosed antenatally and confirmed on histopathology.

1. Introduction

Pediatric renal malformations are common congenital anomalies affecting less than 10% of all births. Pediatric cystic and dysplastic lesions of the kidney are classified into three major categories; developmental, hereditary and acquired.[1,2] Multicystic dysplastic kidney (MCDK) is the second most common congenital anomaly of urinary tract after hydronephrosis and variant of renal dysplasia.[3–5] The incidence ranges between 1 in 3640 to 4300 live births.[3,5,6] It is almost always unilateral and slightly more frequent in boys and on the left side. MCDK most often occurs sporadically, but familial cases sometimes occur but no significant recurrence risk has been observed for future pregnancies.[7,8] Exposure to teratogens and some genetic mutations have also been reported.[9] It is most commonly seen unilaterally with predominance in boys and on left side.[7,10,11]

During embryogenesis, the renal tissue fails to undergo normal process of differentiation to mature functioning nephrons as proposed by Mackie and Stephen.[9] The ureteric bud undergoes a series of division to form collecting system of the kidney. However, in MCDK the ureteric bud is thought to have abnormal branching into metanephric blastema resulting in cystic dilatation resembling bunch of grapes.[3,12] The contralateral kidney exhibits some anomalies like vesicoureteral reflux and obstruction of the ureteropelvic junction.[13,14] Associated involvement of other organs include esophageal atresia, patent ductus arteriosus, vertebral defects, imperforate anus, tracheoesophageal fistula, radial and renal dysplasia (VATER) syndrome, Zellweger syndrome or branchio-oto-

* *Corresponding author*.
E-mail address: nandishvs@gmail.com (V. S. Nandish).

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renal syndrome.[1,11,13,15,16] It usually undergo involution during first five years of life.[10,17–19]

Conservative approach is advocated to children with MCKD, but some have suggested surgical removal on the basis of risk of hypertension, mass effect, potential for malignant transformation and cost of repeated ultrasound examination.[4,10,20] Based on the gross appearance it is of two subtypes i) The Classic type and ii) The Hydronephrotic type. The former has random configuration of cysts, whereas the latter presents with discernible dilated pelvis surrounded by cysts.[4,21]

With the extensive and routine use of antenatal ultrasound scans *in utero* diagnosis as early as 15 weeks of gestation has been possible. In a new born, unilateral renal dysplasia presents as a lump in the abdomen. Whereas, asymptomatic in case of adults and is an incidental diagnosis.[9,22,23]

2. Materials and Methods

We retrospectively assessed fourteen patients of Multicystic dysplastic kidney between January 2004 and December 2014. These included patients with diagnosis of MCDK based on ultrasound findings of a kidney with multiple non-communicating cysts of varying sizes, loss of reniform configuration and parenchymal tissue. The other associated findings like vesicoureteral reflux (VUR) and ureteropelvic junction obstruction (UPJO) were also documented. All the patients were subjected to total nephrectomy and the specimens were sent to the department of Pathology with their respective clinical details. For histopathological evaluation, the tissue samples were processed routinely and fixed in 10% formalin solution and embedded in paraffin. Tissue sections of 3 microns were obtained and stained with hematoxylin and eosin (H&E). Histopathological examinations were performed under the light microscope. The data was analyzed with respect to patient characteristics, including age, sex, side of MCDK, time of diagnosis (antenatal or postnatal), hypertension, family history and other associated anomalies.

3. Results

A total of fourteen (14) patients were diagnosed with unilateral MCDK over a period of 10 years. Patient characteristics are given in Table 1 and associated anomalies in Table 2. No clear sex predilection (incidence of 50% each in males and females) was observed and the mean age of presentation was 104.78 months (0 to 37 years). MCDK was seen predominantly in left side 9 (64.29%) when compared to right side 5 (35.71%) patients. Majority of the patients 10 (71.42%) were diagnosed antenatally. Postnatal diagnosis of MCDK was seen in 4 (28. 58%) patients. The associated VUR and UPJO was seen in 4 (28. 58%) and 2 (14.28%) patients respectively.

All patients with MCDK underwent total nephrectomy and were subjected to histopathological examination. Microscopy revealed renal parenchyma replaced by cysts of variable sizes lined by flattened cuboidal epithelium. Intervening fibrotic stroma with mesenchymal components, primitive glomeruli, nerve bundles and dysplastic ducts.

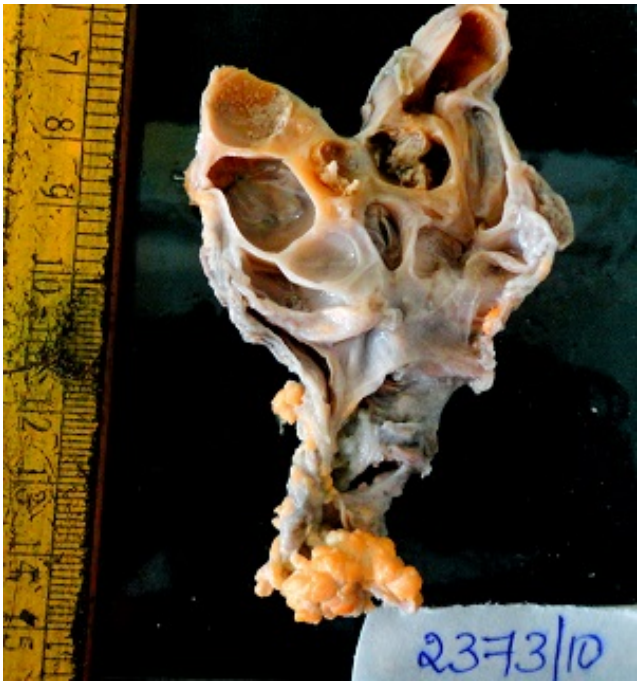

Fig. 1: Cut section of nephrectomy specimen with multiple cysts of varying sizes

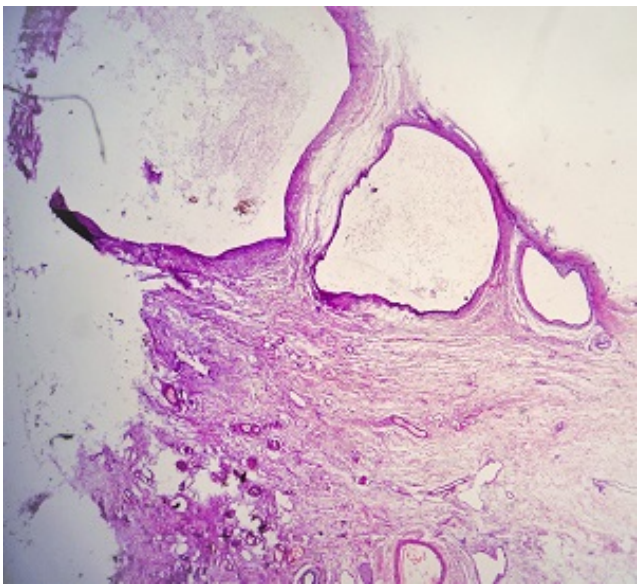

Fig. 2: Cystically dilated spaces lined by flattened cells. H&E, x4

Table 1: Patient characteristics

Total number of patients	**14**
Sex: Males Females	7 (50%) 7 (50%)
Side of MCDK: Right Left	5 (35.71%) 9 (64.29%)
Time of diagnosis: Antenatal Postnatal	10 (71.42%) 4 (28. 58%)
Mean age at diagnosis	104.78 months (0 – 37 years)

Table 2: Incidence of associated anomalies

Associated anomalies	**Number of patients (%)**
VUR	4 (28. 58%)
UPJO	2 (14.28%)

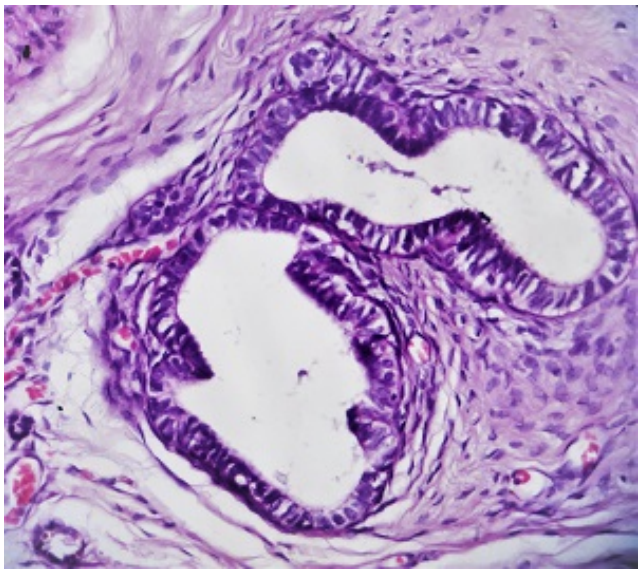

Fig. 3: Primitive appearing tubules. H&E, x40

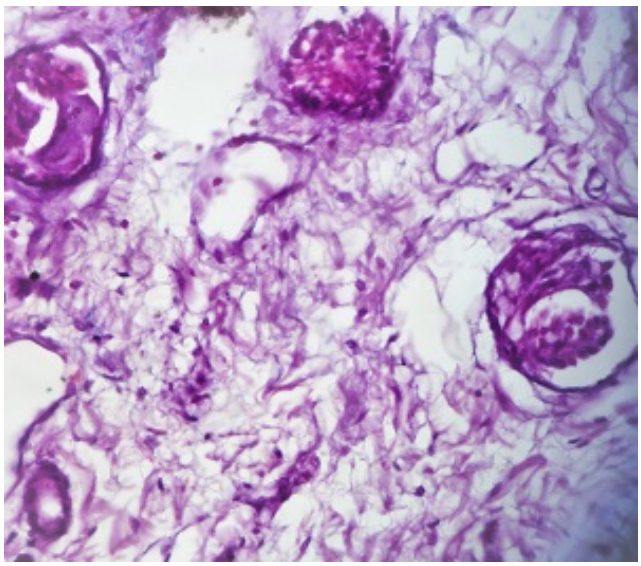

Fig. 4: Immature glomeruli. H&E, x10

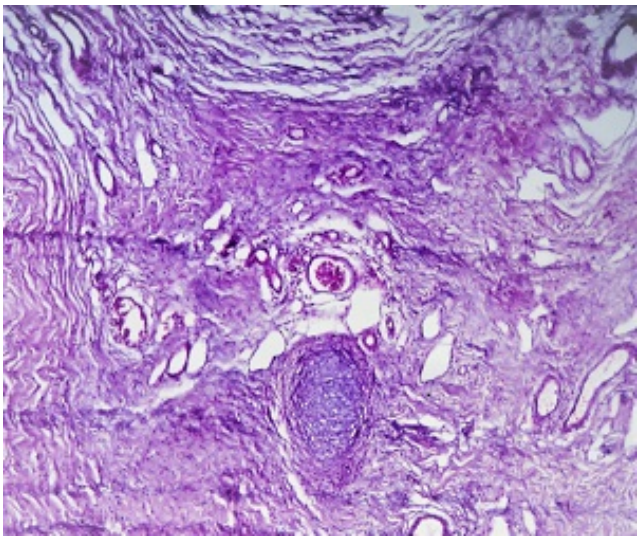

Fig. 5: Mesenchymal component within dysplastic kidney

4. Discussion

Multicystic dysplastic kidneys are non-functional and contain non-communicating cysts with intervening dysplastic renal parenchymal tissue, including primitive glomeruli, primitive tubules and cysts derived from tubular and glomerular structures.[24–26] MCDK is believed to be due to abnormal branching of ureteric bud into the metanephric mesenchyme during development.[4,12] MCDK was divided into simple (unilateral MCDK with no other associated genitourinary abnormalities) and complex (unilateral MCDK with other genitourinary abnormalities or bilateral renal dysplasia with or without other genitourinary abnormalities) by Feldenberg and Seigel.[4,27]

In the present study, unilateral MCDK was seen equally in both sexes without any sex predilection and left kidney was more affected than right similar to that reported in the literature.[28] Among 14 cases 10 (71.42%) were diagnosed antenatally which was comparable to the cases reported in the western countries describing antenatal diagnosis of 80-85% cases.[29,30] Among the 14 patients two presented

with ipsilateral ureteropelvic junction obstruction and four each presented with vesicoureteral reflux and hypertension respectively. As patients with MCDK have only one functional kidney, the anomalies in the contralateral kidney and functionality play a crucial role in the prognosis.[6,13]

The frequency of diagnosis of MCDK has increased due to the widespread use of both antenatal and postnatal ultrasonography that gives a characteristic picture at 24-28 weeks.[31,32] During fetal growth, transient dilatation of the urinary tract is normal, occurring in 1 in every 100 fetuses. In fetus younger than 24 weeks a pelvic diameter greater than 10 mm was 76% sensitive identifying obstruction.[31,33]

To conclude, MCDK is mostly a unilateral sporadic anomaly usually found on the left side, however can be familial. It may be associated with hypertension, vesicoureteral reflux and ureteropelvic junction obstruction. Most of the patients can be diagnosed antenatally and confirmed on histopathology.

5. Source of funding

Nil

6. Conflict of interest

The authors declare no conflicts of interest.

References

1. Lee S, Lee J, Kim H, Kim S, Hahn S, Hwang J. A case of Multicystic dysplastic kidney and cystic adenomatoid malformation of the lung identified as incidental findings. *Korean J Pediatrics*. 2006;49:796–796.
2. Glassberg KI, Stephens FD, Lebowitz RL, Braren V, Duckett JW, Jacobs EC. Renal dysgenesis and cystic disease of the kidney : a report of the Committee on Terminology, Nomenclature and Classification, Section on Urology. *Am Acad Pediatrics J Urol*. 1987;138:1085–1092.
3. Nepple K, Cooper C. Multicystic Dysplastic Kidney. *J Pediatr Urol Prim Care Physician*. 2011;7:11–17.
4. Singh J, Kanojia R, Narasimhan K. Multicystic dysplastic kidney in children - A need for conservative and long term approach. *Indian J Pediatr*. 2009;76:809–821.
5. Aytac B, Sehitoglu I, Vuruskan H. Multicystic dysplastic kidney: four-year evaluation. *TurkJ Pathol*. 2011;27:210–210.
6. Sharada S, Vijayakumar M, Nageswaran P, Ekambaram S, Udani A. Multicystic dysplastic kidney: A retrospective study. *Indian Pediatrics*. 2014;51:641–644.
7. Rukmangadha N, Chowhan AK, Patnayak R, Radhika K, Phaneendra BV, et al. Multicystic dysplastic kidney-a retrospective study with clinicopathological correlation. *J Clin Sci Res*. 2013;2:88–93.
8. Belk RA, Thomas DF, Mueller RF, Godbole P, Markham AF, Weston MJ. A family study and the natural history of prenatally detected unilateral Multicystic dysplastic kidney. *J Urol*. 2002;167:666–675.
9. Joshi N, Sahu S, Yadav V, Jadhav R. Acute pyelonephritis mimicking acute abdomen: An atypical presentation of unilateral dysplasia of the kidney in an adult. *Ann Nigerian Med*. 2012;6:44–50.
10. Unilateral multicystic dysplastic kidney: long term outcomes. *Arch Disease Childhood*. 2006;91:820–823.
11. Lavanya M, Premaleela K, Gulia SP. Bilateral Multicystic Dysplastic Kidneys in Association with High Ano-Rectal Anomaly - A Case Report. Peoples. *J Sci Res*. 2014;7:37–46.
12. Bisceglia M, Galliani CA, Senget C, Stallone C, Sessa A. Renal cystic diseases: a review. *Adv Anat Pathol*. 2006;13(1):26–56.
13. Rios H, Santos R, Gomes C, Correia A. Multicystic dysplastic kidney: a review of eleven years (2000 - 2010). *Port J Nephrol Hypert*. 2012;26:41–47.
14. Mansoor O, Chandar J, Rodriguez MM. Long-term risk of chronic kidney disease in unilateral multicystic kidney. *Pediatr Nephrol*. 2011;26:597–603.
15. Badeli HR, Sajedi SA, Tangestaninejad A, Ahmadian MR, Kanfi KA. Multicystic Dysplastic Kidney in Association with Congenital Ichthyosiform Erythroderma. *Iran J Kidney Dis*. 2007;1(2):102–106.
16. Wang RY, Earl DL, Ruder RO, Graham JM. Syndromic ear anomalies and renal ultrasounds. *Pediatrics*. 2001;108:32–32.
17. Sukthankar S, Watson AR. Unilateral multicystic dysplastic kidney disease: defining the natural history. *Anglia Nephrourol Group*. 2000;89:811–814. Acta Paediatr.
18. John U, Rudnik-Schoneborn S, Zerres K. Kidney growth and renal function in unilateral multicystic dysplastic kidney disease. *Pediatr Nephrol*. 1998;12:567–71.
19. Rottenberg GT, Gordon I, Bruyn RD. The natural history of the Multicystic dysplastic kidney in children. *Br J Urol*. 1997;70:347–50.
20. Sharma AK, Rattan KN, Sharma N. Congenital unilateral Multicystic dysplastic kidney in children: a clinical study. *Indian J Urol*. 2000;17:32–38.
21. Beckwith. Wilms tumor and Multicystic dysplastic kidney disease. Editorial comment. *J Urol*. 1997;158:2259–2260.
22. Longino LA, Martin LW. Abdominal masses in newborn infant. *Pediatrics*. 1958;21:596–604.
23. CY WU, Chiou YH, Sung PK. Left multicystic dysplasia kidney with contralateral ectopic kidney and ureterovesicle junction obstruction. *Zhonghua Yi Xue Za Zhi (Taipei)*. 2001;64:129–132.
24. Alam K, Varshney M, Aziz M, Maheshwari V, Basha M, Gaur K. Multicystic renal dysplasia: a diagnostic dilemma. *Case Reports*. 2011;p. 1–5.
25. Belk RA, Df, Mueller RF. A family study and the natural history of prenatally detected unilateral multicystic dysplastic kidney. *J Urol*. 2002;167:666–675.
26. Caglayan A, Gumus H, Erdogan I. Case Report A female case with multicystic dysplastic kidney: new findings, genetic counseling, and literature review. *Central Eur J Urol*. 2010;63:151–153.
27. Feldenberg IR, Siegel NJ. Clinical course and outcome for children with Multicystic dysplastic kidneys. *Pediatr Nephrol*. 2000;14:1098–101.
28. Cambio AJ, Evans CP, Kurzrock EA. Non-surgical management of multicystic dysplastic kidney. *BJU Int*. 2008;101:804–812.
29. Onal B, Kogan BA. Natural history of patients with multicystic dysplastic kidney-what followup is needed? *J Urol*. 2006;176:1607–1618.
30. Broeking EK, Brinkmann OA, Lengerke V, Sciuk HJ, Fruend J, et al. Unilateral multicystic dysplastic kidney:experience in children. *BJU Int*. 2004;93:388–92.
31. Hussain S, Begum N. Multicystic dysplastic disease of kidney in fetus. *J Ayub Med Coll Abbottabad*. 2007;19:68–77.
32. Irsutti M, Puget C, Duga I, Sarramon MF, Guitard J. Mesoblastic Nephroma: prenatal ultrasonographic and MRI features. *Pediatr Radiol*. 2000;30:147–50.
33. Peters CA, Carr MC, Lais A, Retik AB, Mandell J. The response of fetal kidney to obstruction. *J Urol*. 1992;148:503–512.

Author biography

B U Sowmya Assistant Professor

V S Nandish Assistant Professor

Prema Saldanha Professor

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