A rare case of unilateral left side multicystic dysplastic kidney and contralateral Vesico-ureteric reflux in a male neonate with review of literature

Naila Mazher, Swathi Chacham, Uppin Narayan Reddy, Jillalla Nursing Rao, Janampally Ravi Kiran, Jakkampudi Naga Sravani, Aslam

ABSTRACT

Introduction: Multicystic dysplastic kidney (MCDK) is a rare congenital disorder, resulting from malformation of the kidney during fetal development. It could be unilateral, bilateral or segmental and bilateral MCDK is incompatible with survival. Unilateral MCDK occurs in 1 in 4300 live births and combined incidence of unilateral and bilateral MCDK is 1 in 3600 live births. The malformed kidney is non-functional with multiple irregular cysts of varying size, separated by dysplastic parenchyma along with absent pelvicaliceal system.

Case Report: A 35-week, preterm, male neonate was born to a primigravid by C-Section. There was a history of second degree consanguinity. Antenatal ultrasonography at 17th week of gestation showed left side MCDK with oligohydramnios and without other malformations, which was confirmed by fetal magnetic resonance imaging (MRI). Postnatal ultrasonography also revealed left side MCDK with grade II vesico-ureteric reflux on the right side in micturating cystourethrogram and absent function in the affected kidney in dimercaptosuccinic acid (DMSA) scintigraphy.

Conclusion: We report a preterm, male neonate with antenatally detected non-functional left side multicystic dysplastic kidney with postnatal confirmation and grade II vesico-ureteric reflex on the right side.
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Keywords: Dimercaptosuccinic acid, Multicystic dysplastic kidney, Neonate, Oligohydramnios, Vesico-ureteric reflux

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INTRODUCTION

Multicystic dysplastic kidney (MCDK) is an infrequent congenital cystic malformation of the kidney. It is classified as unilateral, bilateral or segmental and bilateral MCDK is often lethal. The reported incidence of unilateral MCDK is 1 in 4300 live births [1] and that of combined incidence of unilateral and bilateral MCDK is 1 in 3600 live births. The dysplastic malformed kidney consists of non-functional, multiple non-communicating irregular cysts of varying size along with intervening dysplastic parenchyma and absent pelvicaliceal system. Although majority of the cases occur sporadically, autosomal dominant inheritance has also been reported [2]. MCDK results from an abnormal induction of the metanephric mesenchyme by the ureteral bud [3]. With the advances in antenatal screening by ultrasound, majority of these cases are diagnosed antenatally, which can be confirmed by postnatal ultrasound. MCDK can be complicated by hypertension, infection, renal failure and rarely malignant changes.

CASE REPORT

A 35-week, preterm, male neonate was born to a primigravid by C-section with normal APGAR score. There was a history of second degree consanguinity. Antenatal ultrasound at 17th week of gestation showed (Figure 1) left side multicystic dysplastic kidney with oligohydramnios (Amniotic fluid index-5). Fetal MRI confirmed left side MCDK and did not reveal other abnormalities. Fetal echo cardiography was also normal.

After birth, the neonate had respiratory distress requiring ventilatory support and baseline antimicrobials. Initially, pulmonary agenesis was suspected as it is an important accompaniment of renal malformations. However, the chest radiograph showed normal lung expansion and there were few in homogenous radio-opaque shadows, suggesting congenital pneumonia. The respiratory distress subsided after four days and the neonate was weaned from ventilator. Clinically, there were no potter’s facies (Figure 2), there was no mass palpable per abdomen and the urine output was adequate with normal stream. His mean arterial pressure was normal (no evidence of hypertension) and the renal parameters were within normal range. Postnatal abdominal ultrasonography showed MCDK (Figure 3) with a normal right kidney and the postnatal echocardiography was normal. The neonate was started on uroprophylaxis with amoxicillin, in view of MCDK. The micturating cystourethrogram (MCUG) at 40th day of life revealed grade II vesicoureteric reflux (VUR) on the contra-lateral side (Figure 4). Further evaluation with DMSA scintigraphy at second month of life showed absent function in the left kidney (Figure 5). The infant received supportive and symptomatic treatment along with uroprophylaxis and was thriving well in periodic follow-up.

DISCUSSION

The MCDK consists of multiple cysts of various size with small intervening islands of immature glomeruli, primitive tubules, cysts derived from tubular and
glomerular structures. The left kidney is involved more often (55%) than the right kidney (45%). Similarly, the index neonate had MCDK on the left side. As per literature, males are affected more frequently than females (male : female ratio-1.48:1) [5]. Likewise, the index case was also male. Bilateral MCDK is often incompatible with life and it contributes to 20% of prenatally diagnosed cases. It is frequently associated with oligohydramnios, amnion nodosum, pulmonary hypoplasia, and Potter sequence.

The pathogenesis results from abnormal metanephros differentiation resulting from interrupted union of ureteric bud [3] with renal blastema and abnormal division at the stage of metanephros. This might be due to a disruption in the formation of the mesonephric duct, the malformation of the ureteric bud or the degeneration of the ureteric bud at an early stage. The final structure of the dysplastic kidney depends on the timing and degree of the injury to the ureteric bud. Mutations in genes responsible for ureteric bud development have been identified in syndromes with renal dysplasia, including MCDK. Specifically, mutations in EYA1 or SIX1 genes that lead to branchio-oto-renal (BOR) syndrome are associated with renal malformations, including MCDK [6, 7]. Exposure to viral infections and teratogens in-utero has been associated with MCDK.

Dysplastic kidney may persist without any change, increase in size, or might undergo spontaneous involution. Calcification usually occurs when it persists till adulthood, but can be seen as early as three months of age. Most cases of unilateral MCDK undergo spontaneous involution. Index infant had no resolution of MCDK in follow-up and needs close monitoring for its complications. In prenatally diagnosed cases, the
The mass is usually mobile, ballottable, irregular in shape and non-tender with occasional transillumination. It is usually asymptomatic and can remain undetected into adulthood, if not detected antenatally. However, in the index case, there was no palpable mass. MCDK has been reported in association with Alagille syndrome, Beckwith-Wiedemann syndrome, Branchio-oto-renal (BOR) syndrome, Joubert syndrome, Trisomy 18, Waardenburg syndrome type 1, and Renal coloboma syndrome. It is also associated with multiple non-renal and renal malformations including gastrointestinal, central nervous system, cardiovascular, and musculoskeletal system. In approximately 17–43%, there are also abnormalities of the contralateral kidney [8, 9], of which VUR is reported in 4–19% of patients [10,11]. Similarly, the index infant also had grade II VUR on the contra lateral side, though the exact cause of it is unknown. Ipsilateral VUR might also be present in segmental MCDK [12]. Ureteropelvic junction obstruction (UPJO) on the contralateral side has been reported in 7–12% of patients, which was not seen in the index case. Most cases are detected during fetal ultrasonography and can be identified as early as 15 weeks of gestation. Urine for dipstick, microscopic analysis and culture should be obtained in patients with MCDK. Blood tests for creatinine, urea, and electrolytes should be performed. Renal ultrasonography is the recommended preliminary diagnostic imaging study [13] and all these investigations were done in the index infant.

Ultrasonography reveals randomly arranged multiple cysts of variable size, alienated by little or no echogenic parenchyma. Renal pelvis is usually not identified. Voiding cystourethrogram (VCUG) should be performed to look for VUR. No difference in the incidence of urinary tract infections (UTIs) or renal scarring was observed between children with or without VUR in the contralateral kidney [10]. However this case did not have UTI, although there was grade II VUR on the contra lateral side. DMSA scintigraphy is indicated when, ultrasonography fails to reveal the classic features of MCDK and also to quantify the function of the affected kidney. In the current case, the DMSA scan showed absent function in the affected kidney. In the rare situation, where severe UPJO cannot be differentiated from MCDK by ultrasonography and DTPA renal scanning, a diagnostic puncture of a cyst with instillation of radiographic contrast material might help to distinguish these two disorders. The presence of cysts connected by tubular structures and the absence of a collecting system is diagnostic of MCDK. However, this was not required in our case.

Treatment is supportive and symptomatic. The role of nephrectomy is controversial and is indicated to treat or prevent complications like abdominal pain, UTI, hypertension, or renal malignancy. Children with MCDK should undergo renal ultrasonography every 6–12 months until the age of five years or until involution is noted. Also, they should receive antibiotic prophylaxis during infancy and early childhood, as this age group is at highest risk of scarring due to pyelonephritis. Similarly, the index case was started on uroprophylaxis. Blood pressure monitoring is indicated once each year and if hypertension is evident, it should be treated. The index case had normal blood pressure in the follow-up, but requires close monitoring into childhood for detection of hypertension and its complications [14], which is required in this case. Prognosis depends on unilateral or bilateral occurrence of MCDK and the severity of associated anomalies. Most children with isolated unilateral MCDK are less prone for complications as in the index infant with a better prognosis.

CONCLUSION

We report a preterm, male neonate with antenatally detected multicystic dysplastic kidney (MCDK) on the left side with postnatal confirmation. The affected kidney was non functional on dimercaptosuccinic acid (DMSA) scintigraphy and was complicated by grade II vesico-ureteric reflex on the contra-lateral side.

ABBREVIATIONS

- Branchio-oto-renal (BOR) syndrome
- Dimercaptosuccinic acid (DMSA)
- Diethylenetriaminepentaacetic acid (DTPA)
- Intravenous pyelography (IVP)
- Magnetic resonance imaging (MRI)
- Multicystic dysplastic kidney (MCDK)
- Micturatingcystourethrogram (MCUG)
- Mercaptoacetyltriglycine (MAG-3)
- Urinary tract infection (UTI)
- Ureteropelvic junction obstruction (UPJO)
- Voiding cystourethrogram (VCUG)
- Vesico-ureteric reflux (VUR)

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The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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