MULTICYSTIC DYSPLASTIC KIDNEY WITH CONTRALATERAL RENAL HYPOPLASIA PRESENTING WITH RENAL FAILURE IN A NEWBORN: A CASE REPORT

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Multicystic dysplastic kidney (MCDK) is the most common cystic kidney disease in children. Renal abnormalities occur in the contralateral kidney in 20-57% of cases. The prognosis of MCDK depends primarily on the status of the opposite kidney. We describe a case of unilateral MCDK and contralateral hypoplasia presenting with renal failure during the neonatal period. Abdominal ultrasound showed a multiple non-communicating cystic mass measuring 3.5 × 1.8 cm in the left kidney. The right kidney was small measuring 2.5 × 1.3 cm with increased echogenicity. $^{99m}$Tc–mercaptoacetyltriglycine renal scintigraphy showed an absence of uptake and flow of the left kidney and a decreased uptake of the right kidney. At 1.5-year of follow up, the MCDK had a partial involution, and the patient had chronic renal failure with growth retardation. In conclusion, children with unilateral MCDK and contralateral renal hypoplasia may develop renal failure early in their lives. The optimal conservative treatment of MCDK patients with chronic renal failure requires close monitoring of each patient's clinical and laboratory status to slow the disease progression and to induce healthy growth. (Acta Nephrologica 2007; 21: 138-142)

Key words: contralateral renal anomalies, multicystic dysplastic kidney, newborn, renal failure, renal hypoplasia

INTRODUCTION

Multicystic dysplastic kidney (MCDK) is the most common form of cystic renal disease in children. The anomaly is thought to represent the end result of hydrenephrosis developing secondary to ureteric maldevelopment early in gestation.1-4 MCDK is usually a unilateral anomaly that involves the entire kidney with the left side more commonly involved. Bilateral MCDK is invariably fatal. The incidence of unilateral MCDK is estimated between 1 in 2700 and 1 in 4300 live births.5,6 Boys appear to be more frequently affected. Renal anomalies contralateral to the MCDK are common, and have been reported in 20-57% of all MCDK cases.7-11 Recently, Damen-Elias et al.12 reported when cystoscopy and colposcopy were added to the routine investigations the rate of detection of ipsilateral and contralateral anomalies increased to 75% of cases, twice the rate reported in the literature. The abnormalities of the contralateral kidney most often include vesicoureteral reflux (VUR) and ureteropelvic junction obstruction. The prognosis for unilateral MCDK is more favorable, and depends primarily on the status of the opposite kidney. Children with unilateral MCDK and renal failure during the neonatal period are uncommon. We report a case of unilateral MCDK with contralateral renal hypoplasia presenting with renal failure during the neonatal period and she had a chronic renal failure with growth retardation at the 1.5-year follow-up.

CASE REPORT

A 9-day-old female newborn was admitted due to poor activity, poor feeding, and a cystic renal mass. She was born at 39 weeks’ gestation with a birth weight of 2980 g (25-50 percentile) and a body length of 48 cm (25-50 percentile). The pregnancy was uneventful. There was no oligohydramnios or fetal distress during...
maternal gestational period. An abdominal cystic mass was identified antenatally during the prenatal echography. Both the mother’s personal history and her family history were noncontributory. The patient suffered from tachypnea after delivery and received oxygen therapy for 5 days with a diagnosis of transient tachypnea of the newborn at a local hospital. Then she developed poor activity and poor feeding and was referred to our hospital for further management.

On physical examination, the patient appeared acutely ill. Her vital signs were blood pressure of 85/52 mm Hg, temperature of 36.2°C, heart rate of 144/min, and respiratory rate of 42/min. Body weight was 2700 g and body length 49 cm. Gross appearances were normal. Abdominal examination showed no palpable masses.

On the day of admission, her hemoglobin level was 16.3 g/dL and hematocrit was 52.6%. The white blood cell count was 15190/mm$^3$ with 59.3% segmented neutrophils and 30.1% lymphocytes. The platelet count was 410000/mm$^3$. The initial laboratory data were as follows: sodium, 139 mEq/L; potassium, 4.4 mEq/L; Cl, 110 mEq/L; blood urea nitrogen (BUN), 45.8 mg/dL; creatinine, 2.2 mg/dL; albumin, 3.6 g/dL; calcium, 10.6 mg/dL; phosphorus, 4.5 mg/dL; and uric acid, 7.7 mg/dL. Blood gas showed a mild metabolic acidosis with pH of 7.324, PCO$_2$ of 34.5 mm Hg, and HCO$_3$– of 16.8 mmol/L. Urinalysis results were 2+ for occult blood and 1+ for proteinuria; the sediment contained red blood cells 2-3 and white blood cells 0-1 per high-power field. A chest radiogram revealed a normal lung field. Abdominal ultrasound showed a cystic mass measuring 3.5 × 1.8 cm over the left renal fossa. The cystic renal mass contained multiple non-communicating various sized cysts, oval to round, with echogenic septa (Fig. 1). The right kidney was small measuring 2.5 × 1.3 cm with increased echogenicity. There were no lesions in the liver or spleen. $^{99m}$Tc–mercaptoacetyltriglycine renal scintigraphy showed an absence of uptake and flow of the left kidney and a decreased uptake of the right kidney. Voiding cystourethrography disclosed a normal bladder and no VUR.

The findings of the imaging studies revealed a left unilateral MCDK with hypoplasia of the contralateral kidney. The renal function, as assessed as serum creatinine (2.2 mg/dL, normal range for this age < 0.3 mg/dl) and estimated glomerular filtration rate (Schwartz formula: 10.2 mL/min/1.73 m$^2$, normal range for this age 46.6 ± 5.2 mL/min/1.73 m$^2$), was markedly impaired. A non-surgical approach was advised, and the MCDK was managed conservatively. The patient received supplementation with sodium bicarbonate to correct metabolic acidosis. She was followed up with serial ultrasound scans, blood pressure measurements and laboratory analyses. At 1.5-year after discharge, follow-up blood laboratory data revealed hemoglobin, 11.1 g/dL; hematocrit, 33.1%; BUN, 35 mg/dL; creatinine, 2.7 mg/dL; Na, 140 mEq/L; K, 4.1 mEq/L; Cl, 109 mEq/L; albumin, 3.1 g/dL; calcium, 8.2 mg/dL; phosphorus, 5.6 mg/dL; intact parathyroid hormone, 156 pg/mL; alkaline phosphatase, 278 IU/L; pH, 7.334; PCO$_2$, 41.8 mm

Fig. 1. Abdominal sonogram shows a multiple non-communicating various sized cystic mass with echogenic septa.
containing multiple non-communicating cysts of varying sizes. The diagnostic imaging study. Ultrasound reveals a kidney that is smaller than that in a normal kidney. It is thought that the size of MCDK in our patient was abnormally shaped, and often resembles a bunch of grapes. However, the size of MCDK in our patient was smaller than that in a normal kidney. It is thought that the MC

**DISCUSSION**

MCDK, the most common cystic form of kidney disease in children, is a nonhereditary developmental anomaly characterized by varying numbers and sizes of cysts with no or little discernible normal renal tissue. It is possible that many cases of congenital absence of the kidney diagnosed in adults are due to MCDK which has regressed to the point become radiographically undetectable. The pathogenesis of this disorder remains poorly understood. It has been proposed that the development of MCDK results from atresia of the ureteral bud system at the level of the upper ureter during the first trimester. However, a high incidence of urogenital and other nonrenal abnormalities suggests aberrations in shared common developmental programs rather than antenatal obstruction of the urinary tract as the primary etiology of MCDK.

The affected kidney of MCDK is usually enlarged, abnormally shaped, and often resembles a bunch of grapes. However, the size of MCDK in our patient was smaller than that in a normal kidney. It is thought that the MCDK progressively involutes during gestation. If the disease is bilateral, then it is usually associated with anhydramnios and pulmonary hypoplasia during gestation which inevitably leads to death either antenatal or early neonatal in the life. Although our case had unilateral MCDK with contralateral hypoplasia, no oligohydramnios or pulmonary hypoplasia occurred during pregnancy.

In the past, nearly all MCDK were detected in newborns following detection of abdominal masses. The widespread use of antenatal ultrasound has resulted in the majority of MCDK being detected before birth. However, at times MCDK cannot be distinguished from an obstructed hydronephrotic kidney prenatally. Therefore, prompt postnatal evaluation is necessary. Renal ultrasonography is the recommended preliminary diagnostic imaging study. Ultrasound reveals a kidney containing multiple non-communicating cysts of various sizes with echogenic septa. Nuclear renal scanning may be necessary if ultrasonography does not reveal the classic features of MCDK. Renal scintigraphy demonstrates a non-functional kidney in cases of MCDK. The combination of ultrasonography and nuclear renal scan has provided a higher diagnostic accuracy for identifying MCDK. The findings on imaging studies of our case are consistent with the classic characteristics of MCDK with contralateral renal hypoplasia.

Complications of MCDK might be categorized as those due to MCDK per se and those due to associated urinary malformations. The complications of MCDK include abdominal or flank pain, hypertension, renal malignancy, and respiratory depression due to mass compression. The role of nephrectomy for management of unilateral MCDK remains controversial. Prior to the era of ultrasonography, nephrectomy was often required to establish the diagnosis. Now, it has been recommended that in the absence of complications such as hypertension, infection and malignant transformation, the affected kidney can be managed conservatively. A non-surgical approach for our patient was advised. She received conservative treatment and serial follow-up examinations with laboratory analysis and ultrasonography.

Children with unilateral MCDK have increased risks of the contralateral renal anomalies. It has been reported that the contralateral kidneys were abnormal in 20% to 57% of all MCDK cases. The abnormalities of the contralateral kidney include VUR, ureteropelvic junction obstruction, agenesis, megareter, duplex kidney, ureterocele, and hypoplasia. VUR, the most common contralateral anomaly identified, was reported in 14.5-43% of patients. The incidence of the contralateral renal hypoplasia, however, is as low as 1-5% of cases. Prognosis of MCDK depends on whether the involvement is unilateral or bilateral and on the presence and severity of associated contralateral anomalies. Most children with isolated unilateral MCDK do not experience any problems or complications, while bilateral MCDK and unilateral MCDK with contralateral agenesis are inevitably fatal. Recently researchers have reported that serial ultrasound examinations of MCDK demonstrated the rate of partial or complete involution with time ranging from 76% to 89% of cases. Compensatory hypertrophy in the contralateral kidney was also observed in 77-100% of patients. The size of MCDK in our case was involuted (2.9 × 1.4 cm vs. 3.5 × 1.8 cm) at 1.5-year of follow up examination, while the contralateral kidney showed no change in size.

Failure of compensatory growth might be related to the contralateral dysfunction. Evidences for this conclusion were based on previous observations that these
children with a MCDK and renal failure usually associated with a reduced renal mass in the contralateral kidney which was regarded as hypoplastic or dysplastic.\textsuperscript{20,21} Selzman et al’s study of children with a MCDK during a mean follow-up of 3.1 years showed that of the 65 children with MCDK renal insufficiency only developed in 2 (3%). They found that renal insufficiency was not noted in any child with a MCDK and contralateral VUR, but it developed in 2 children both had a contralateral small echogenic kidney. These 2 children presented with renal insufficiency at the age of 1 year and 12 years, respectively. Our report is in agreement with the previous studies that children with a MCDK and contralateral hypoplasia may progress to renal failure early in their lives even during the neonatal period.

Chronic renal failure ultimately progresses to end-stage renal disease (ESRD). Several types of treatment have been proposed to slow this progression, including modification of dietary protein, lowering of blood pressure, correction of acidosis and anemia, and control of renal osteodystrophy. Our patient was a case of MCDK and chronic renal failure with a mild metabolic acidosis and secondary hyperparathyroidism, which is under control with supplementation of sodium bicarbonate, phosphate binder and vitamin D. Growth failure is a unique and recognized feature of chronic renal failure in children. Previously, researchers have detailed the effects of recombinant human growth hormone (rhGH) therapy on children who have ESRD or on those who are on dialysis or on post renal transplant.\textsuperscript{22-25} However, the cost of rhGH is very expensive, our case was managed with conservative treatment and was not treated with rhGH due to economic restrictions.

The occurrence of MCDK in several members of a family has recently been reported.\textsuperscript{26,27} This would support the possibility that the development of MCDK is, at least in part, may be regulated by genetic factors. Although MCDK may occasionally occur on a familial basis, most cases of MCDK are a sporadic anomaly. Belk et al.\textsuperscript{26} indicated that the overall prevalence of renal anomalies in first-degree relatives of children with a MCDK was low. They suggested that extensive family screening is not justified. However, screening for MCDK is easily performed by using ultrasonography, and renal evaluation of relatives (at least parents and siblings) is recommended. Prenatal diagnosis of most familial cases of MCDK may be identified by routine prenatal ultrasonography.

In summary, a MCDK is usually unilateral, but other genitourinary tract abnormalities are frequently present. Associated anomalies of the contralateral kidney play an important role for the prognosis of children with a MCDK. If unilateral MCDK with contralateral renal hypoplasia occurs in children, they may develop to renal failure early in their lives. We recommend that these children with chronic renal failure and growth retardation need optimal conservative management to slow disease progression and to induce healthy growth.

REFERENCES