Pediatrik Cerrahi Dergisi, 9:356-358, 1995

Multicystic dysplastic kidney: Prenatal diagnosis and postnatal management

Haluk SARIHAN, Rahmi AKYAZICI, Musa ABEŞ, Şirin GÜLKÖK, Kemal SERÇE

Karadeniz Technical University Faculty of Medicine, Departments of Pediatric Surgery and Radiology, Trabzon, Turkey

Summary

Six infants with multicystic dysplastic kidneys (MCDK) diagnosed prenatally with ultrasonography. One female infant also had a large ovarian cyst. The kidneys were palpable in four newborns at first physical examination. During the first six months, it was established that minimal change occured in kidney morphology. Therefore five patients were operated by the sixth month. One patient was lost to follow up. Prenatal diagnosis of MCDK has provided for a clear line of management and no complications have been encountered with this approach.

Key words: *Kidney, multicystic dysplasia, prenatal diagnosis, surgery*

Introduction

Multicystic dysplastic kidney (MCDK) is the most common renal cyst in the newborn [15,17]. The pathology consists of multiple noncommunicating cysts of variable size, with a central solid core of primitive dysplastic elements. The proximal ureter is atretic or nonpatent [15]. In addition, there is commonly a pelviureteric junction abnormality on the contralateral side [9].

In the past, the disease usually was presented as a unilateral flank mass. Presentley, an increasing number of cases are being detected by antenatal fetal ultrasound (US) examinations performed for obstetric screening. Here we propose to assess the impact of prenatal diagnosis of MCDK and current postnatal management in a small group of patients.

Material and Methods

From June 1992 to October 1994, six cases of multicystic dysplastic kidney were diagnosed in utero at routine ultrasound examinations in compliance with established criteria [5]. All patients had unilateral lesions and the typical features of MCDK. In addition one fetus was found to have a large abdominal cyst which could not be located at that stage. There were no findings of oligohydroamnios and all patients were followed throughout their gestation. The babies were delivered at term per vaginal route.

Work up at birth included US, excretory urography (IVP),voiding cystouretrography in all cases. Isotope examination with Tc-99m DTPA was performed at three newborns. In addition, urinalysis, serum creatinin and electrolytes were checked. The female infant with MCDK was subsequently understood to have a large ovarian cyst and was operated on in the newborn period. The other five patients were followed up for at least six months before an operation was undertaken.

Results

In this group of patients, 4 (%66.7) were boys and 2 (%33.3) were girls. The left kidney was affected more than the right (4 left vs 2 right kidneys). The mean gestational age of the time of diagnosis was 33.4 weeks after the last menstrual period (range 30 to 36 weeks). In utero follow up with US demonstrated no changes in the cysts sizes in kidneys any of the patients. At birth four of the affected kidneys could be palpated. No hypertension, urinary tract infection or uremia were detected prior to operations. Patients with associated anomalies included one patient with a large ovarian cyst and one with a ventricular septal defect.

In all cases IVP revealed nonfunctioning kidney on the affected side and no abnormality was detected in

Address: Haluk Sarıhan, M.D. K.T.Ü. Tıp Fakültesi, Çocuk Cerrahisi Anabilim Dalı, 61080, Trabzon, Turkey

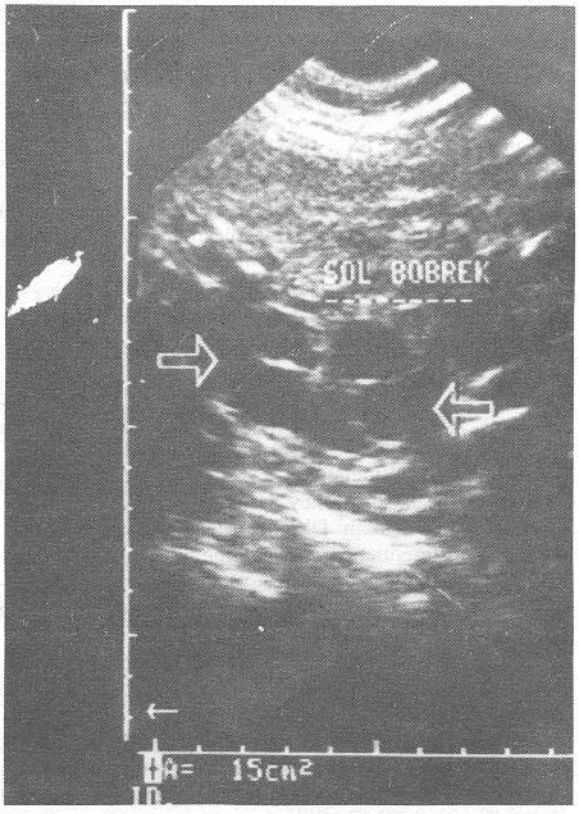

Figure 1. Longitudinal US view of a multicystic dysplasia of left fetal kidney in the third trimester.

the contralateral kidney. Renal scintigraphy in three patients with Tc-99m DTPA demonstrated no uptake in the affected kidneys. Voiding cystourethrography did not indicate vesicoureteral reflux in any of the six patients.

US examination at the time of diagnosis showed multiple cysts randomly arranged and with varying sizes (Fig. 1). At fetal follow up (4-10 weeks) no changes occurred. Postnatal US confirmed the in utero findings. One female newborn with MCDK of right kidney also had a large abdominal cyst thought to be of ovarian or mesenteric in origin. Laparotomy revealed a left ovarian cyst (10 cm in diameter); an oopherectomy and nephrectomy were performed at the same time (Fig. 2). The other five patients were followed up for six months. Serial maesurements of MCDK at US examination in five patients demonstrated minimal reduction in size in two cases and no change in another two patients. There was an increase in one case. The contralateral kidney was also serially measured, and findings were compared

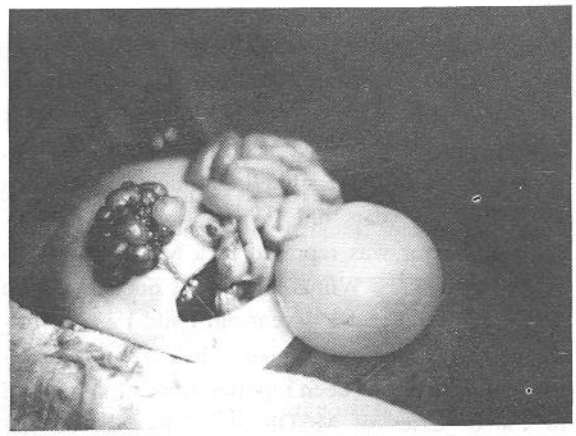

Figure 2. Operative photography demonstrating a large left ovarian cyst, and multicystic dysplasia of right kidney.

with that of standard measurements [10]. Compensatory hypertrophy occurred in four of five infants. These five patients underwent a prophylactic nephrectomy at sixth month of age. The nephrectomy was performed using the extraperitoneal approach and the patients were discharged third to fifth days postoperatively. No mortality or morbidity atributable to the approach occurred.

Discussion

Multicystic dysplastic kidney is the most common abdominal tumor in the newborn. The first case of MCDK was reported in 1864 [15]. Its pathogenesis is still being disputed. It has been suggested that during the migration of the developing kidney from the sacral to the lumbar level, the normal shift of vasculature may be interrupted, leading to an ischemic insult producing both the MCDK and usual ureteral atresia [17].

Prenatal diagnosis of MCDK has become common with the increased use of fetal US. The primary differential diagnosis of MCDK should include obstruction of ureteropelvic junction [2,5]. Most of the early case reports were diagnosed during the third trimester of pregnancy [3]. Today, transvaginal US allows visualization of the normal and anomalous fetal urinary tract during the first and second trimester [7]. The sonographic detection of renal dysplasia has clinical implications, because the presence of renal dysplasia indicates a decreased or absent renal function. Our patients' lesions were detected during the third trimester of pregnancy.

The management of the MCDK remains controversial. A nephrectomy is justified for the risk of potential pathology, particularly malignancy. Six cases of malignancy have been reported in MCDK, including 3 case of renal cell carcinoma in patients 15,26 and 68 years old [4,6,12]. A malignant mesothelial tumor was reported in a MCDK of a 68-year-old man [12]. Wilms tumor was documented in a 10-month-old and a 4-year-old child [11,13]. Furthermore, the premalignant lesion of nephroblastomatosis has been reported to occur in up to 5 to 7% of cases of MCDK [12]. The risk of malignancy is low but it is undeniable.

MCDK have been associated with other complications; notably, hypertension, infection, hematuria and pain [15]. The association of renin-mediated hypertension and MCDK has been documented [8]. On the other hand, abnormalities in the contralateral kidney occur in many patients with unilateral MCDK and hypertension could result from lesions in the noncystic kidney [8]. In the literature, to our knowledge, a total of ten such cases have been reported and only four of these have responded to surgical treatment [1,6,8]. A small risk of hypertension is believed exist in children with MCDK and blood pressure examinations should be a routine part of clinical follow up.

The occurence of hematuria and urinary infection is somewhat difficult to explain, since the MCDK usually is set apart from the lower urinary tract by the ureteral atresia. In the 27 adult patients with MCDK, pain was the most common presenting symptom, which disappears following surgery [15]. Unfortunately, it is difficult to evaluate pain due to MCDK in young. Increasing number of children with MCDK are followed up with serial US rather than early surgical treatment. The pathology in MCDK is progressive and variable [3]. In a number of studies with follow ups in the range of 30 to 35 months, it has been demonstrated that partial or complete involution of the cystic lesion occurs in more than 50% of the cases with no complications [13,14,16].

Conservative management appears to be a sensible option, but the actual risk assumed in the follow up these patients is unknown. Perhaps the real benefit of the conservative approah in MCDK will be to demonstrate the natural history of the disease. We have prefered the surgical approach in order to avoid the demonstrated complications related to MCDK.

References

1. Angermeier KW, Kay R, Levin H: Hypertension as a complication of multicystic dysplastic kidney. Urology 39:55, 1992
2. Avni EF, Thoua Y, Lalmand B, et al: Multicystic dysplastic kidney: Evolving concepts. In utero diagnosis and post-natal follow-up by ultrasound. Ann Rad 29:663, 1986
3. Avni EF, Thoua Y, Lalmand B, et al: Multicystic dysplastic kidney: Natural history from in utero diagnosis and postnatal follow up. J Urol 138:1420, 1987
4. Barrett DM, Wineland RE: Renal cell carcinoma in multicystic dysplastic kidney. Urology 15:152, 1980
5. Beretsky I, Lankin DK, Rusoff JH, Phelan L: Sonographic differentiation between the multicystic dysplastic kidney and the ureteropelvic junction obstruction in-utero using high resolution real time scanners employing digital detection. J Clin Ultrasound 12:429, 1984
6. Birken G, King D, Vane D, Lloyd T: Renal cell carcinoma arising in a multicystic dysplastic kidney. J Ped Surg 20:619, 1985
7. Bronshtein M, Yoffe N, Brandes JM, Blumenfeld Z: First and early second-trimester diagnosis of fetal urinary tract anomalies using transvaginal sonagraphy. Prenatal Diagn 10:653, 1990
8. Chen YH, Stapleton FB, Roy S, Noe HN: Neonatal hypertension from a unilateral multicystic dysplastic kidney. J Urol 133:664, 1985
9. Dewan PA, Goh DW: A study of the radiological anatomy of the multicystic kidney. Pediatr Surg Int 9:368, 1994
10. Han BD, Babcock DS: Sonographic measurements and apprearance of normal kidneys in children. AJR 154:611, 1985
11. Hartman GE, Smolik LM, Shochat SJ: The dilemma of the multicystic dysplastic kidney. Am J Dis Child 140:925, 1986
12. Jung WE, Peters CA, Mandell J, et al: Flow cytometric evaluation of multicystic dysplastic kidneys. J Urol 144:413, 1990
13. Raffensberger J, Abouslemann A: Abdominal masses in children under one year of age. Surgery 63:514, 1986
14. Rickwood AMK, Anderson PAM, William MPL: Multicystic renal dysplasia detected by prenatal ultrasonography. Natural history and results of conservative management. Br J Urol 69:538, 1992
15. Sharp RJ: Developmental anomalies of the kidney. In Ashcraft KW (ed): Pediatric Urology. WB Saunders, Philadelphia 1990, p:77
16. Strife JL, Souza AS, Kirks DR, et al: Multicystic dysplastic kidney in children: US follow-up. Radiology 186:785, 1993
17. Synder III H McCS: Cystic disease of the kidney, dysplasia and agenesis. In Welch KJ, Randolph JG, Ravitch MM, et al (eds): Pediatric Surgery. Chicago. Year-Book Med Publ 1986, p:1124