Multicystic Dysplastic Kidney Complicated by Recurrent Urinary Tract Infection: A Case Report

Tekrarlayan Idrar Yolu Enfeksiyonu ile Komplike hale Gelen Multikistik Displastik Böbrek: Olgu Sunumu

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ABSTRACT

Multicystic Dysplastic Kidney is one of the most common renal diseases presented with abdominal mass in the pediatric age group. The natural course of the disease is full or partial involution starting in the early stages of the fetal period and progressing through the postnatal period. In symptomatic cases, ultrasonography is the primary diagnostic method. Today, conservative treatment is preferred instead of surgery. Surgical treatment is preferred in selected cases or cases with recurrent urinary tract infections, hypertension, and suspected malignancy. In this study, we presented a one-year-old girl case with a multicystic dysplastic kidney, complicated by recurrent urinary tract infection and undergoing nephrectomy.

Keywords: Multicystic dysplastic kidney, urinary tract infections, neonate, recurrence, nephrectomy

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ÖZET

Multikistik Displastik Böbrek pediatrik yaş grubunda en sık görülen abdominal kitle ile prezente olan renal hastalıklardan biridir. Hastalığın doğal seyri, fetal dönemin erken evrelerinde başlayan ve postnatal dönem boyunca ilerleyen tam veya kısmi involüsyondur. Semptomatik hale gelen vakalarda ise ultrasonografi primer tanı yöntemidir. Günümüzde tedavide cerrahi yerine konservatif tedavi tercih edilmektedir. Cerrahi tedavi seçilmiş veya tekrarlayan idrar yolu enfeksiyonu, hipertansiyon veya malignite gelişiminden şüphe edilen vakalarda uygulanmaktadır. Bu çalışmada bir yaşında kız hastada tekrarlayan idrar yolu enfeksiyonu ile komplike hale gelen ve nefrektomi uygulanan multikistik displastik böbrek olgusu sunulmuştur.

Anahtar Sözcükler: Multikistik displastik böbrek, idrar yolu enfeksiyonu, yenidoğan, rekürrens, nefrektomi

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INTRODUCTION

Multicystic dysplastic kidney (MCDK) is among the most prevalent congenital anomalies of the urinary system. It occurs approximately 1 in 1000-4300 live births. Prevalent use of antenatal and postnatal ultrasonography in recent years has manifested the increased incidences of MCDK. It is often associated with urinary malformations: vesicoureteral reflux (VUR) followed by ureteropelvic junction obstruction (UPJO) (1, 2). These anomalies cause recurrent urinary tract infections (UTI), subsequent renal scar formation, and chronic kidney failure (3). In this study, we presented a case of a multicystic dysplastic kidney complicated by vesicoureteral reflux and recurrent urinary tract infection.

CASE REPORT

A 1-year-old girl who was followed up for multicystic kidney (right), grade 2-3 vesicoureteral reflux (VUR), and recurrent urinary tract infection (UTI) was admitted to the pediatric surgery clinic. She was using prophylactic sulphamethoxazole/trimethoprim for recurrent UTI. In the laboratory examination, we observed mild leukocytosis (12.1 K/uL, reference 4.5-11 K/uL). No pathological finding was detected in urine microscopy, and there was no growth in urine culture. USG for the urinary system revealed a multicystic structure consisting of 12x9 mm multiple cysts in the right kidney lodge, which was primarily evaluated in favor of a multicystic dysplastic kidney. Left kidney localization and size (60 mm) were normal; parenchyma thickness (10 mm) was normal. The patient underwent radical nephrectomy, and we sent the sample to the pathology laboratory. The informed consent was taken from the parents.

In the macroscopic examination, we observed a 3.5x3.2x1.5 cm nephrectomy material with a 3 cm long and 0.4 cm diameter ureter. On the material, we detected a multilobular cystic structure with a 1.6x1.5 cm surface with a 0.6 cm swelling from the parenchyma (Figure 1a). The ureter was dilated. We noted cystic structures on the kidney section surface (Figure 1b).

Figure 1a

Figure 1b

Figure 1: We observed a 3.5x3.2x1.5 cm nephrectomy material, with a ureter of 3 cm in length and 0.4 cm in diameter (1a). We detected numerous cystic structures on the section surface (1b).

In the microscopic examination, we observed numerous cyst structures lined with cuboidal epithelium (Figure 2a) and found disorganized immature tubular and glomerular structures in the fibrous stroma (Figure 2a-2b). In addition, we observed cartilage tissue in a focal focus (Figure 2c). Regarding to these findings, we reported the case as the multicystic dysplastic kidney (cystic renal dysplasia).

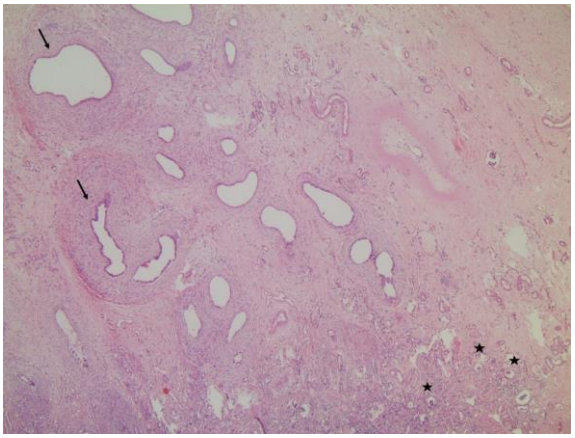

Figure 2a

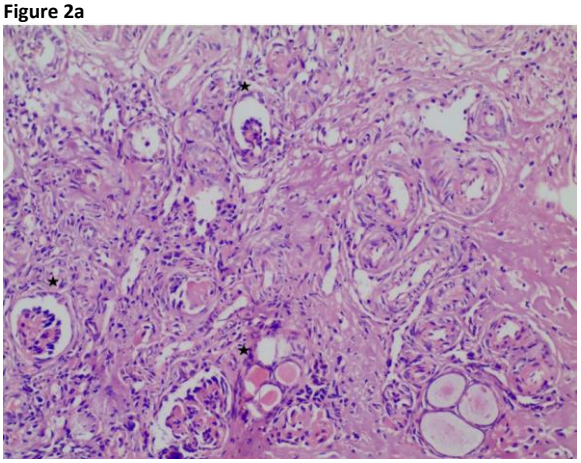

Figure 2b

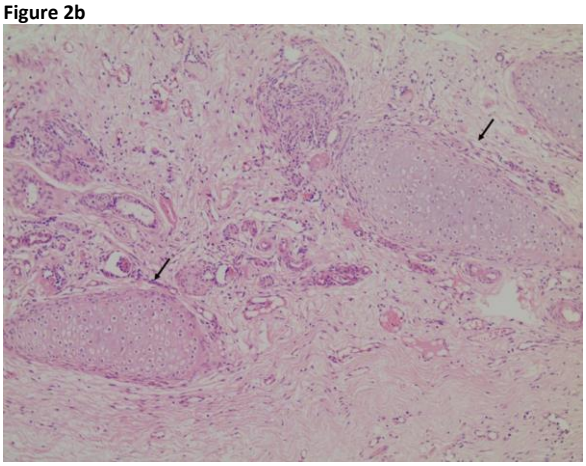

Figure 2c

Figure 2: We observed cystic structures (arrows) lined by a single-layered cuboidal epithelium in a fibrous stroma (2a,HEX40) and immature tubular and glomerular structures in disorganized appearance (asterisks) (2a,HEX40; 2b,HEX200). We detected cartilage tissue in focal areas (arrows)(2c, HEX100).

DISCUSSION

MCDK is the most common cause of abdominal masses in the neonatal period. It is seen unilaterally; involvement in both kidneys often leads to mortality (4). It is a type of renal dysplasia characterized by the presence of independent cysts of different sizes separated by dysplastic parenchyma on ultrasonography and the absence of a normal pelvicalyceal system (1). Abdominal pain or pain in the flank region and respiratory distress are rare symptoms developing due to the compression effect of the abnormal kidney (1). It is often seen in boys and is located on the left side (1, 5). What differed our case from the cases in the literature were female sex and right-sided dysplastic kidney.

Urinary system ultrasonography is sufficient for the diagnosis of MCDK and the detection of anomalies in the contralateral kidney. In addition, DMSA scintigraphy is performed to confirm the presence of a non-functioning kidney, while voiding cystourethrography (VCUG) is performed to show the presence of VUR. It is controversial to use all these imaging methods simultaneously (3). MCDK can be found in isolation, but it is often accompanied by renal malformations: VUR, UPJO, renal calculus, and hypospadias. Contralateral VUR is the most common urological anomaly, and one can also observe ipsilateral VUR (1).

Contralateral VUR causes increased infection, renal scarring development, and, eventually, hypertension and chronic renal failure. VUR is generally low grade (grade 1-3), and its regression in the follow-up is promising (3). Our case underwent DMSA scintigraphy and VCUG in an external center; consequently, multicystic kidney on the right and grade 2-3 ipsilateral VUR were detected.

While surgery is considered a standard treatment method for MCDK, it has become controversial today. In the past, surgery was used for definitive diagnosis and prevention of complications such as UTI, hypertension, and malignancy. Recent studies have shown that the dysplastic kidney has an overall excellent prognosis with spontaneous involution and a very low incidence of complications (6). Therefore, conservative treatment is preferred over surgical treatment today. Besides, kidney size is important in determining involution. If the kidney size is 50 mm or more, complete involution and conservative treatment are not in the subject (7).

The diagnosis can be confirmed histopathologically in cases with nephrectomy. On microscopic examination, the most common features of cystic renal dysplasia are primitive duct formation surrounded by fibromuscular tissue, lobar disorganization, and cysts, as well as metaplastic cartilage formation in approximately half of the cases. Cysts characteristically replace the renal parenchyma. Large cysts are lined by flattened cuboidal epithelium. Immature and aberrant glomerular formations can rarely be seen in the parenchyma (5, 8). Radical nephrectomy was also performed in our case. In the microscopic examination, we observed many cystic structures between immature tubular and glomerular structures. There was also a small focal cartilage tissue.

There are some cases where USG cannot help diagnose MCDK or which raise clinical suspicion and require a differential diagnosis. These cases may be hydronephrotic kidney, benign multiloculated cyst, localized renal cystic disease, cystic Wilm's tumor, or cystic renal cell carcinoma. DMSA scintigraphy is generally used for a differential diagnosis. While uptake is not observed in MCDK, it may be seen in other diseases due to normal renal parenchyma. In cases where Wilm's tumor or RCC distinction cannot be made definitively, nephrectomy is necessary for definitive histopathological diagnosis. (4).

As a conclusion, ultrasonography is sufficient for the diagnosis of MCDK. In recent years, the conservative method is preferred instead of the surgical method due to the low incidence of complications and the regression of the dysplastic kidney. In cases complicated by the development of recurrent UTI, radical nephrectomy can be performed. In the postoperative period, the patient should be followed up at regular intervals for the functionality of the contralateral kidney.

Conflict of interest

No conflict of interest was declared by the authors.

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