Case report

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분절형 다낭성 이형성신 영아 보고 1례

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A Female Infant with Segmental Multicystic Dysplastic Kidney

There have only been 35 pediatric cases and one adult case reported on segmental multicystic dysplastic kidney (MCDK) from our search in PubMed, including 19 cases detected antenatally. There is little documentation of segmental MCDK, particularly concerning its natural history. Segmental MCDK can be presented atypically, making diagnosis more difficult. We report an another case with segmental MCDK. Multicystic abdominal mass detected on antenatal sonogram in this infant was diagnosed as segmental MCDK by renal ultrasonography and computed tomography. If a definitive diagnosis of segmental MCDK can be made on imaging, surgery is not required for a diagnostic biopsy.

Key words: Computed tomography, Multicystic renal mass, Renal ultrasonography, Segmental multicystic dysplastic kidney

Introduction

Segmental multicystic dysplastic kidney (MCDK) is a rare subtype of MCDK, found in only approximately 4% of children diagnosed with MCDK [1]. From our search in PubMed, only 35 pediatric cases and one adult case have been reported, including 19 cases detected antenatally. Typical segmental MCDK mostly affects the left kidney and women. Most of the reported cases are associated with duplex collecting systems, of which the upper pole that has atresia of the upper moiety proximal ureter is commonly affected. Vesicoureteral reflux (VUR) is found in 20-33% of cases at the contralateral kidney and in 77% at the ipsilateral kidney [2]. For this paper, we will describe the 37th case of segmental MCDK.

Case report

An 8-day-old female was brought to the nephrology department of our hospital for an evaluation of a cystic mass upon the right kidney. This was evaluated on a postnatal sonogram and performed soon after birth. The mass was detected on prenatal sonogram at the gestational period of 28^{+4} week and it measured 1.7 ×1.2 cm. The infant was born on the 38^{+4} week of pregnancy by vaginal delivery, weighing 3,590 grams at birth. There was no perinatal problem. Maternal disease, her obstetrics history, family history, and neonatal screening test were not remarkable. Vital signs were stable. On physical examination, she was healthy, and her liver, kidneys, and spleen were not palpable. Immediate postnatal abdominal sonogram (US) showed that an approximately 4.3×2.8 cm non-communicating multilocular cystic mass with a small amount of intervening hyperechoic solid portion was located at the right suprarenal fossa (Fig. 1A). There was no connection to the right adrenal gland and a suspicious focal connection with the right upper kidney. The largest cyst measured approximately 2.5×2 cm. The size of both kidneys were normal (right 3.9 cm, left 4.0 cm). There was no hydronephrosis or hydroureter. Abdominal 3D computed tomography (CT) performed immediately after US showed that a non-enhancing cystic mass at the upper portion of the right kidney with a focal connection to the upper portion of the right kidney was detected. The finding on CT resulted in MCDK of the upper pole of the duplex Rt. Kidney (Fig. 1C). According to both imaging studies, segmental MCDK in the upper portion of duplicated right kidney was strongly suggested. At 1weeks of age, the laboratory

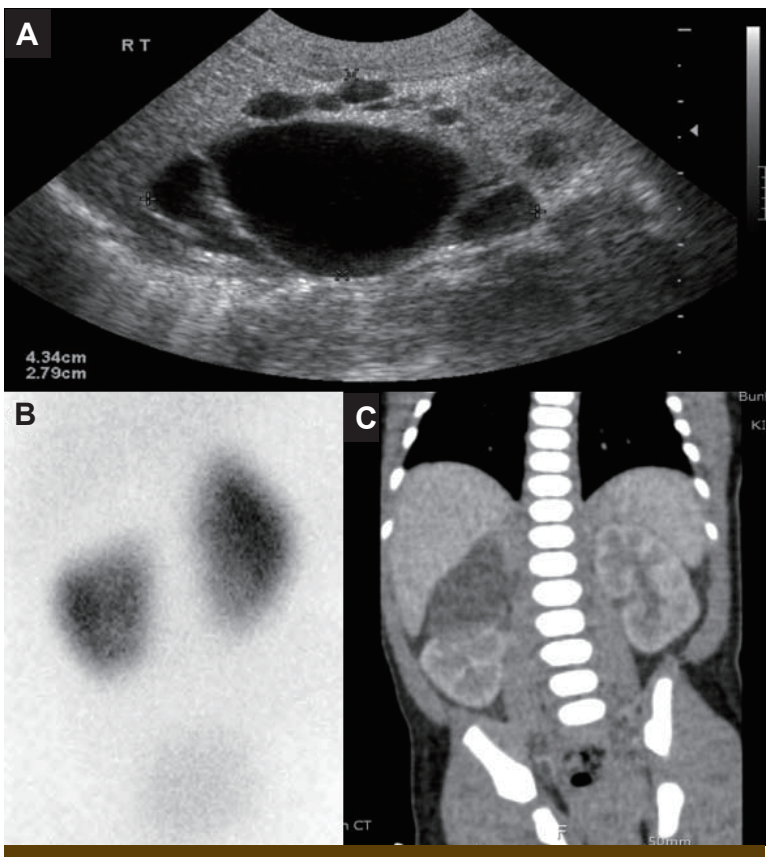

Fig. 1. (A) Non-communicating multilocular cystic mass with small intervening hyperechoic solid portion on renal sonogram; (B) Small right kidney with suspicious cortical defect at the upper pole of the right kidney on 99mTc-dimercaptosuccinic acid renal scan; (C) Mainly cystic non-enhancing lesion at the upper portion of duplex right kidney on CT.

findings were: Hgb 15.7g/dL, WBC 11,290/mm^3, platelet count 397,000/mm^3, CRP 0.15 mg/dL, serum albumin 3.7 g/dL, BUN/cr 7.5/0.9 mg/dL, uric acid 1.8 mg/dlL, OT/PT 37/14 IU/L, serum electrolytes (sodium 136 mEq/L, potassium 4.8 mEq/L, chloride 106 mEq/L, and T-CO2 17.1 mEq/L), LDH 620 U/L, ferritin 551.9 ng/mL, neuron-specific enolase 22.3 ng/mL, α-fetoprotein 266.3 ng/mL, carcinoembryonic antigen 0.69 ng/mL, cystatin C glomerular filtration rate 68 ml/min/1.73m^2, normal urinalysis, and urine microalbumin negative. ^{99m}Tc-dimercaptosuccinic acid (DMSA) renal scan and voiding cystourethrogram (VCUG) were performed after the age of 1 month. A DMSA scan showed that a relatively small-sized right kidney had a suspicious cortical defect in the upper pole with a relative radionuclide uptake of 40.9% (Fig. 1B). The finding of VCUG was normal. US was performed every month until the age of 3 months, and thereafter every three months. The whole shape of the cystic mass did not change. However, the size of the cystic mass at the upper portion of right kidney decreased to 3.1×2.2 cm, measured constantly on supine and longitudinal views. She has been well without unfavorable symptoms or signs through the age of 12 months.

Discussion

Segmental MCDK can be presented atypically, making diagnosis more difficult [3]. There is little documentation of segmental MCDK, particularly concerning its natural history. Especially in cases with a presentation of a cystic mass on antenatal sonogram, differentiation of segmental MCDK with multiloculated renal tumors is necessary because those prognoses are extremely different. The differential diagnosis includes congenital mesoblastic nephroma, multilocular cystic nephroma, cystic nephroma, and neonatal Wilms' tumor. Congenital mesoblastic nephroma is the most common renal neoplasm of infancy. Hematuria is commonly present. The tumor is composed of mature spindle cells that grow by infiltration, resulting in irregular tumor margins. Complete surgical removal is usually curative. However, a cellular subtype of mesoblastic nephroma is more aggressive, and metastatic behavior has been documented in some cases, appearing in larger cystic areas of hemorrhage and necrosis [4]. The sonographic appearance of a multilocular cystic renal tumor includes a sharply circumscribed, multiseptated renal mass with the attenuation value of the cyst contents and thin septa, although the mass may appear solid. After contrast material administration, contrast material does not accumulate within individual loculi, but the septation enhances [4]. Wilms' tumor displays hyperechoic lesions relative to the normal renal parenchyma or hypoechoic regions in the tumor with hemorrhage or necrosis on US, evidence of cystic necrosis less than 10% of the time, or high frequency of the extension of tumor into the renal vein and inferior vena cava on CT scans; this is rarely predominantly cystic [4, 5]. Meanwhile, Peng et al. [6] have reported that both multislice CT and US provide highly accurate diagnoses for the malignant renal cystic masses in children using the Bosniak classification system. Therefore, if a definitive diagnosis can be made on imaging, surgery is not required for a diagnostic biopsy. Although Iscaife et al. [7] have reported that surgery of segmental MCDK was the safest option based on clinical signs of a life threatening condition due to sepsis and the risk of kidney failure, performing surgery on segmental MCDK should be carefully decided on each clinical situation. This is because the frequency of developing complications such as recurrent UTI, hypertension, or Wilms' tumor in patients with segmental MCDK would not be common in childhood as with complete MCDK, and may be dependent upon the presence of associated urinary tract anomalies [8].

In summary, we reported on an another case of segmental MCDK. Multicystic abdominal mass detected on antenatal sonogram in this infant was diagnosed as segmental MCDK by renal US and CT. The size of the cystic mass decreased without changing its shape compared to the first US imaging performed after birth. She has been well through the age of 12 months without unfavorable symptoms or signs. Thus, if a definitive diagnosis of segmental MCDK can be made based on imaging studies, surgery will not be required for a diagnostic biopsy.

한글요약

분절형 다낭성 이형성신은 PubMed의 자료 수집에 따르면 산전에 발견된 19명을 포함해 35명의 소아 환자와 1명의 성인 환자의 증례가 보고 되었다. 분절형 다낭성 이형성신에 대한 기록은 부족하며 비전형적으로 나타날 수 있어 진단이 어렵다. 본 연구에서는 분절형 다낭성 이형성신의 37번째 케이스를 보고하는 바다.

본 연구에서 환아의 산전 초음파에서 발견된 다낭성 복부 종양은 신장 초음파와 컴퓨터 단층촬영에 의해 분절형 다낭성 이형성신으로 진단되었다. 영상 검사를 통해 분절형 다낭성 이형성신의 최종 진단이 내려진다면 진단적 생검을 위한 수술은 불필요할 것이다.

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