□ Case Report □

A case of multicystic dysplastic kidney and cystic adenomatoid malformation of the lung identified as incidental findings

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Multicystic dysplastic kidney and congenital cystic adenomatoid malformation of the lung are independent disorders, but both result from abnormal morphogenesis during embryogenesis. Congenital cystic adenomatoid malformation of the lung is associated with renal anomalies as well as other extrapulmonary anomalies and almost all cases with these anomalies are stillborn. We report a case of a 21-month-old male who was admitted with the impression of acute infectious gastroenteritis; multicystic dysplastic kidney with congenital cystic adenomatoid malformation of the lung was detected incidentally during evaluation. **(Korean J Pediatr 2006;49:796-799)**

Key Words : Congenital cystic adenomatoid malformation, Multicystic dysplastic kidney

Introduction

Pediatric renal malformations are common congenital anomalies and they affected fewer than 10% of all births[1]. Pediatric cystic and dysplastic lesions of the kidney are classified into 3 major categories; developmental, hereditary and acquired[2]. Renal dysplasia is a developmental disorder and is categorized as either hypoplastic dysplasia, multicystic dysplasia, agenesis, segmental dysplasia, or congenital hydronephrosis with or without dysplasia[3]. Multicystic dysplastic kidney is the most common dysplasia variant and is a common form of cystic disease in the newborn. The unilateral form, however, is the most common cystic disease identified throughout childhood[4]. Multicystic dysplastic kidney may be one component of a generalized association of anomalies such as the vertebral defects, imperforate anus, tracheoesophageal fistula, radial and renal dysplasia (VATER) syndrome, Zellweger syndrome, or the branchio-oto-renal (BOR) syndrome[5].

Congenital cystic adenomatoid malformation of the lung is rare. It was first reported in 1949 by Ch'in and Tang[6]. Eighty percent of cases are diagnosed in the neonatal period due to respiratory distress at birth. Other congenital malformations have been associated with congenital cystic adenomatoid malformation of the lung in 18-20% of cases. These malformations include prune belly, renal agenesis, pulmonary sequestration, diaphragmatic hernia, and agenesis of the contralateral lung[7].

We report here the coincident case of multicystic dysplastic kidney with congenital cystic adenomatoid malformation of the lung diagnosed as incidental findings.

Case Report

A 21-month-old male was admitted to the hospital for treatment of acute infectious gastroenteritis with dehydration. His birth and family history were unremarkable. His vital signs on admission were temperature 37.5℃, pulse 110/min and respiratory rate 30/min. His systolic blood pressure was 90 mmHg, diastolic pressure was 60 mmHg. Laboratory findings showed the following results : blood urea nitrogen 13 mg/dL, creatinine 0.3 mg/dL, AST/ALT 179/269 U/L, hemoglobin 12.9 g/dL, hematocrit 38.6% and WBC 14,900/mm^3. Urinalysis showed a specific gravity of 1.015 and 1-4 WBC/HPF microscopically. Stool examination

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and culture study were all unremarkable. An abdominal ultrasonogram was obtained for elevated liver enzymes; it showed hypertrophy of the right kidney and absence of left kidney. Further evaluation with abdominal computed tomography revealed a multicystic dysplastic kidney in the left kidney and multiple cystic lesions of the right lower lobe of the lung which was detected incidentally (Fig. 1). The chest computed tomography showed an uncomplicated congenital cystic adenomatoid malformation of the lung, type II (Fig. 2). The voiding cystourethrogram showed no vesicoureteral reflux. A diuretic renal scan detected no left kidney activity. The patient's chromosome study revealed a normal male karyotype, 46, XY.

The acute infectious gastroenteritis and elevated liver enzymes improved and the patient was discharged on hospital day 8. The multicystic dysplastic left kidney and congenital cystic adenomatoid malformation of the lung are all asymptomatic. Follow up was arranged with the urology and pediatric surgery departments. We plan to follow up computed tomography studies every 12 months for surveillance of the development of hypertension or other complications.

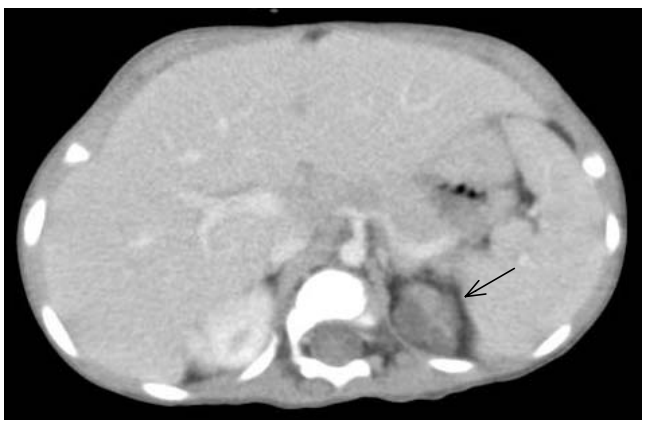

Fig. 1. The abdominal computed tomography reveals a multicystic dysplastic kidney in the left.

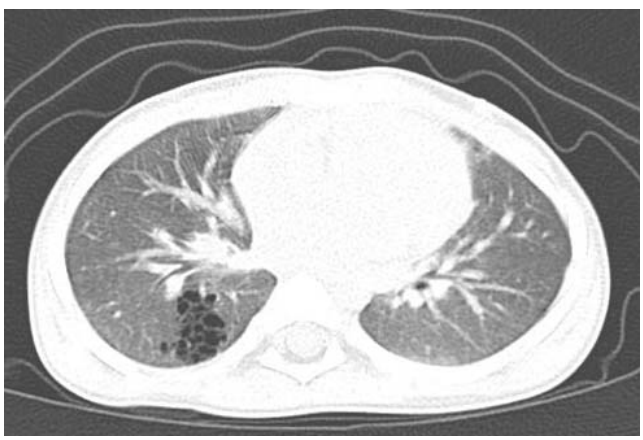

Fig. 2. The chest computed tomography shows an uncomplicated congenital cystic adenomatoid malformation of the lung, type II.

Discussion

Multicystic dysplastic kidney is a congenital renal malformation characterized by the presence of multiple cysts in dysplastic kidneys. Multicystic dysplastic kidney is not one of the inherited renal cystic diseases. It is a form of renal dysplasia, where cystic elements are found in the kidney along with immature, undifferentiated, primitive tissue [8]. Embryologically, multicystic dysplastic kidney may result from abnormal renal morphogenesis, likely consequent to abnormalities of developmentally expressed genes as well as some degree of ureteropelvic junction obstruction[9]. The majority of cases of multicystic dysplastic kidneys are identified as sporadic anomalies, with no current genetic explanation. Bilateral multicystic dysplastic kidney malformations are frequently associated with the fatal Potter anomaly. The incidence of unilateral multicystic dysplastic kidney is reported to be 1 in 4,000 live births[10]. Almost all known cases are sporadic. The primary etiology of multicystic dysplastic kidney is thought to be an early obstruction of the fetal urinary tract during the first trimester of pregnancy[11]. A high incidence of genitourinary abnormalities has been reported in patients with multicystic dysplastic kidney[12]. It has been reported that the prevalence of contralateral associated urologic anomalies in patients with multicystic dysplastic kidney is 39% and that vesicoureteral reflux is the most common anomaly identified (18%)[13]. Associated extrarenal anomalies include esophageal atresia, tracheoesophageal fistula, ventricular septal defect and patent ductus arteriosus[14].

The standard management of patients with multicystic dysplastic kidney is nephrectomy. Surgery is generally performed to confirm the diagnosis, to relieve symptoms, and to eliminate the risk for hypertension and malignant changes. However, this approach has recently been replaced with non-surgical management[15]. Recent information suggests that the true prevalence of multicystic dysplastic kidney, in the general population, is far greater than previously suspected, and that the majority of affected individuals are completely asymptomatic[16]. However, the best manage-

ment of patients with multicystic dysplastic kidney continues to be debated.

Congenital cystic adenomatoid malformation of the lung is considered a hamartomatous lesion of the bronchial tree by some. However, others have suggested that this congenital anomaly results from an arrest in the development of the fetal bronchial tree[17]. This anomaly was initially classified into 3 types by Stocker; the incidence has been estimated to be 1 : 25,000 to 1 : 35,000 pregnancies[18]. Type I lesions consist of single or multiple cysts larger than 2 cm in diameter with few or no adenomatoid areas. Cysts are often filled with air, fluid, or both, and are lined by a ciliated pseudostratified columnar epithelium. Type II lesions are characterized by a mixture of cysts smaller than 1 cm in diameter with adenomatoid areas. These cysts are lined by a ciliated cuboidal or columnar epithelium. Type III lesions are entirely adenomatoid. These lesions have small, evenly spaced cysts resulting in a firm, bulky mass leading to a shift in the mediastinum. Congenital cystic adenomatoid malformation of the lung is diagnosed within the first month of life in the majority of cases. However, there are rare cases in which the lesion is first detected after infancy as in the case presented here. Children with this congenital anomaly usually present with persistent infection, pneumothorax, unilobar cysts, or lung abscess[19]. The treatment of choice for patients with this finding is surgical resection. However, there are many advocates for conservative management of these patients including clinical follow up with delay of surgery until absolutely necessary.

Our case may be viewed as one that would support conservative management as the findings of multicystic dysplastic kidney with congenital cystic adenomatoid malformation of the lung diagnosed were diagnosed as only incidental findings during management of a 21-month-old with acute infectious gastroenteritis.

한 글 요 약

우연히 발견된 편측성 다낭성 신 이형성증과 폐의 선천성 낭성 선종양 기형이 합병된 증례

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편측성 다낭성 신 이형성증과 폐의 선천성 낭성 선종양 기형은 각각 독립된 질환으로 배아 시기에 발생 과정의 문제로 생긴다고 알려져 있다. 폐의 선천성 낭성 선종양 기형에서 폐 외의 기형으로 신장 기형이 동반될 수 있으나 생존 가능성이 적어 사산아로 출생되는 경우가 많다. 저자들은 위장관염으로 입원한 21개월 남아에서 우연히 편측성 다낭성 신 이형성증을 발견했고, 이 과정에서 폐의 선천성 낭성 선종양 기형도 발견하였기에 보고하는 바이다.

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