The Turkish Journal of Pediatrics 35: 299-303, 1993

CONGENITAL CYSTIC ADENOMATOID MALFORMATION*
A report of a case and review of the literature

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SUMMARY: Göçmen A, Kiper N, Tanyel C, Göğüş S, Özçelik U, Büyükpamukçu N. (From the Pediatric Chest Disease Unit, Department of Pediatrics, and the Department of Pediatric Surgery, Hacettepe University Faculty of Medicine, Ankara, Turkey). Congenital cystic adenomatoid malformation. Turk J Pediatr 1993; 35: 299-303.

A ten-month-old boy was admitted to our hospital with fever, cough and dyspnea. His history was remarkable for mild respiratory problems from the early neonatal period. Although medical treatment was administered several times in a local hospital, his symptoms recurred. Chest x-ray revealed multiple cystic lesions on the right side. The diagnosis of congenital cystic disease was considered preoperatively, to our knowledge for the first time in our hospital. The patient's postoperative course was uneventful, and no respiratory problems have recurred in 24 months of follow-up. *Key words: lung, cystic adenomatoid malformation, congenital.*

Congenital cystic disease of the lung is an uncommon malformation accounting for about 25% of lung anomalies. Lobar emphysema, bronchogenic cysts, pulmonary lymphangiectasia, and congenital cystic adenomatoid malformation of the lung (CCAM) are the four pathologic categories of congenital cystic disease of the lung[1,2]. CCAM was first described as a separate entity by Chin and Tang[3]. Most cases have been reported to occur in the neonatal period[2,3]. In this period, the respiratory symptoms result from the compression of normal lung and mediastinum by the cysts. Later in life, CCAM causes recurrent lower respiratory tract infections[3]. As in our case, CCAM should be considered when recurrent respiratory infections and cystic radiological findings are persistent.

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Case Report

A 10-month-old boy was admitted to our hospital with fever, cough and respiratory distress. The pregnancy and delivery were uncomplicated. The family history was unremarkable. In the early neonatal period, his mother noted that he was slow to feed. At six months of age, he was admitted to a local hospital with fever and cough, and his chest x-ray revealed multiple cystic lesions on the right side. Intravenous antibiotics were administered for 15 days for suspected but not confirmed staphylococcal pneumonia. Two weeks after treatment, his chest x-ray findings remained unchanged and his symptoms recurred, so he was referred to Hacettepe Children's Hospital. The patient did not have contact with individuals known to have tuberculosis.

On physical examination, the patient's body temperature was 38.8°C, pulse rate was 110/minute and respiratory rate was 56/minute. Blood pressure was 100/50 mmHg. His weight was 9 kg (50th percentile) and his height was 77 cm (90th percentile). The child appeared well. On chest examination, a mild pectus carinatum and diminished breath sounds were noted on the right side. The liver was palpable 5 cm below the right costal margin, and the spleen tip was palpable at the lower left costal margin. Physical examination was otherwise negative.

The results of laboratory studies included a hemoglobin of 10.25 g/dl, white blood cell count of 9600/mm^3 with 34% polymorphonuclear leukocytes, 2% stub and 64% lymphocytes. Urinalysis, serum proteins, blood urea and electrolytes were normal. Sweat chloride concentration was 22 mEq/L. Serum immunoglobulins were within normal levels. A chest x-ray displayed multiple cystic structures with areas of surrounding atelectasis on the right (Fig. 1a). A computed tomographic (CT) scan of the thorax disclosed a normal appearance of the left lung and a solid mass with surrounding multiple cystic structures on the right middle and lower lobes. Radionuclide scanning revealed a generalized perfusion defect on the right side and non-specific signs on the left.

Antimicrobial therapy was administered intravenously for secondary infection. On the 20th day of the patient's hospitalization, he was afebrile and his condition was stable. With his history and positive chest x-ray, our preoperative diagnosis was CCAM. A right posterolateral thoracotomy was performed and revealed multiple cystic structures in the lower lobe, which were removed. The child had an uneventful postoperative course. Gross and microscopic findings confirmed the diagnosis of type I CCAM of the lung (Fig. 2). Eighteen months later his physical examination and x-ray findings were normal (Fig. 1b).

Discussion

Congenital cystic adenomatoid malformation (CCAM) of the lung may result from an embryonic defect occurring within the first fifty days of gestation[1]. The lesion

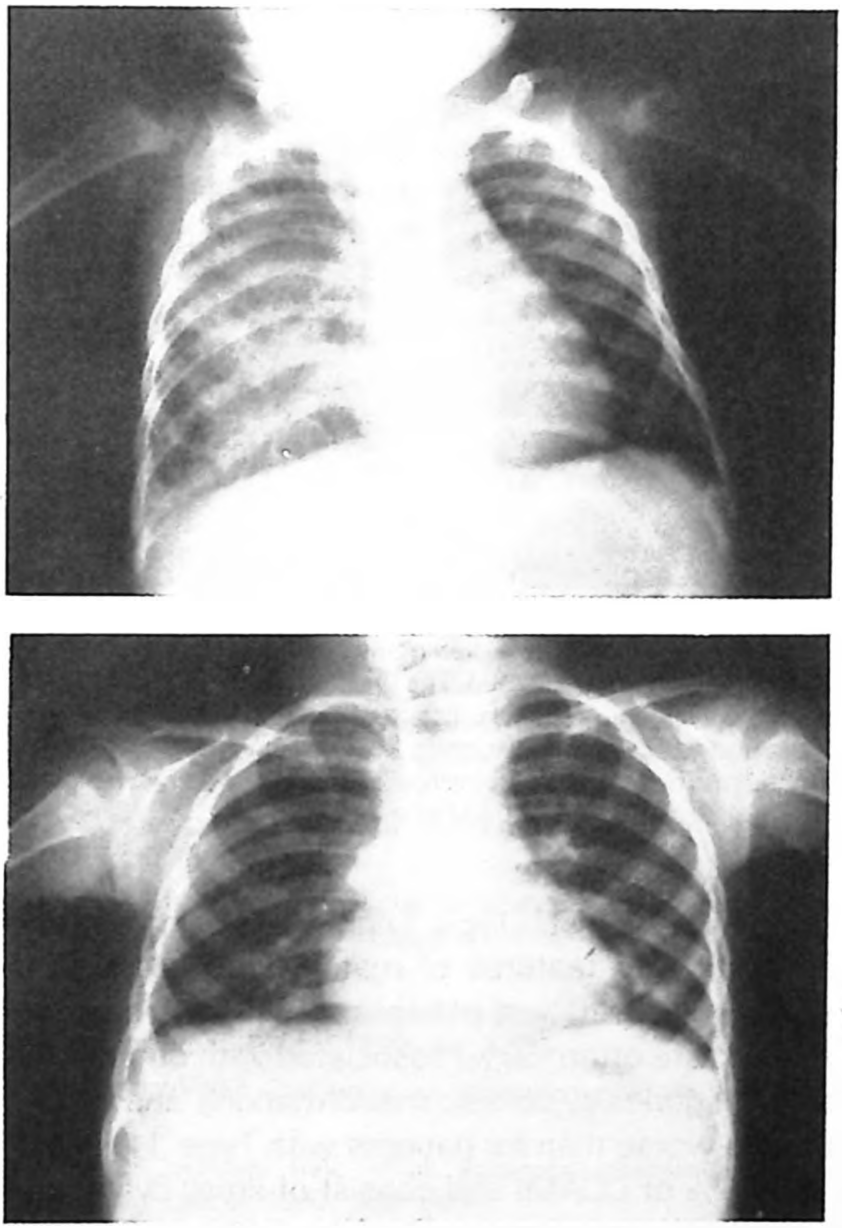

Fig. 1a: Chest radiography of an infant with CCAM of the right lung, showing multiple cysts of varying sizes and shapes; mediastinum is shifted to the left side. b: Postoperative chest-radiography.

is considered to be a "hamartoma" due to excessive growth of terminal bronchiolar structures. However, Buntain and colleagues[4] have suggested that it is "a focal pulmonary dysplasia" because in many cases there is skeletal muscle in the cyst walls. Microscopic examination reveals the following: 1-cysts of varying sizes, lined by cuboidal to ciliated pseudostratified columnar epithelium; 2-polypoid configuration of the mucosa of the cysts; 3-absence of cartilage plates except as components of "entrapped" normal bronchi; 4-presence of a group of mucogenic cells lining the cyst wall; and 5-absence of inflamation. These are the generally accepted histopathological criteria[4,5].

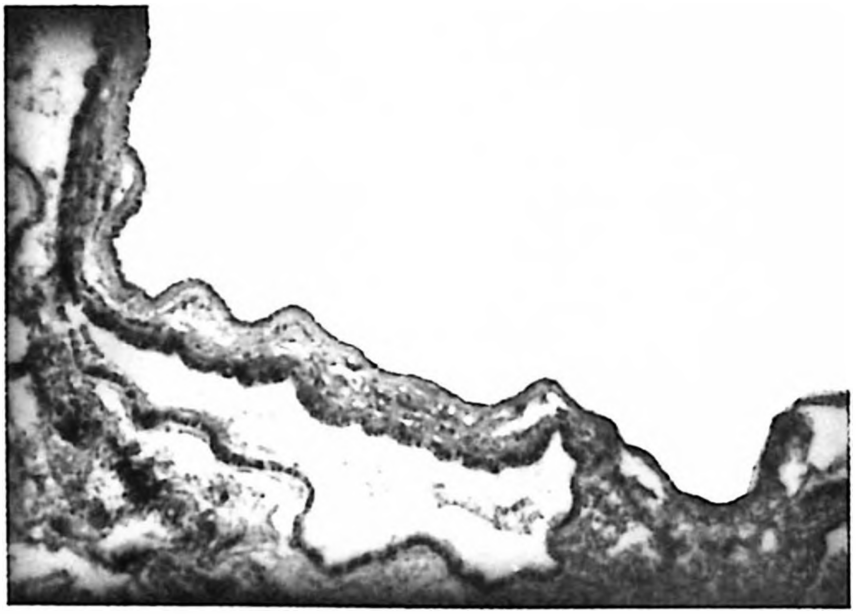

Fig. 2: Right lower lobectomy material revealed many cystic structures which measured 0.5-2.5 cm in diameter and occupied a portion of the lobe. The walls of the cysts were lined by pseudostratified columnar epithelium overlying fibromuscular tissue of variable thickness. Relatively normal alveoli were present between and adjacent to these cysts. A chronic inflammatory reaction was also noticed in the lobectomy material. These gross and microscopic findings confirmed the diagnosis of type I CCAM of the lung.

There are three types of CCAM[6]. Type I consists of single or multiple, large (1-5 cm in diameter) cysts with features of mature lung tissue between the cysts. Type II lesions, occurring in 20% of patients with CCAM, consist of multiple small (0.5-1.5 cm) cysts and are often (60%) associated with other anomalies, especially renal agenesis or dysgenesis, cardiac malformations and intestinal atresia. The survival rate (40%) is worse than for patients with Type I lesions. Type III lesions are the rarest (only 5% of CCAM) and consist of small cysts, less than 0.5 cm in diameter. The survival rate is reported to be 50%.

Type I is the most common, and 80 percent of cases present in the neonatal period with respiratory distress[2,5]. After the first year of life, cysts are very prone to secondary infection. In this period, chest x-ray differentiate staphylococcal pneumatoceles. In our case, the diagnosis of CCAM had been made with the help of the history and serial radiography, and was confirmed by histopathological examination. As a presenting symptom, recurrent respiratory infection is rare[2,5].

The radiological findings are usually sufficiently specific to distinguish CCAM from other forms of pulmonary disease. Approximately 75% of type I lesions are on the right side, and the most typical radiologic findings are sharply outlined radiolucent areas of irregular sizes and shapes and areas of surrounding atelectasis. Our patient had findings similar to those reported in the literature. Bronchoscopy, bronchography, and radionuclide scanning are of limited value for

diagnosis. Angiography and computed tomography of the chest are recommended[2,4]. This abnormality may also be diagnosed antenatally by ultrasound[7].

Definitive diagnosis is based on histological examination of the excised lobe, and definitive treatment of CCAM of the lung is by thoracotomy and excision of the affected lobe[1-3]. Long-term follow-up has not shown any restriction of pulmonary function, and radioisotopic scanning shows that the remaining lung grows normally and expands to fill the thoracic cavity[1].

As in our case, this uncommon malformation should be considered in the differential diagnosis of recurrent respiratory infection. Early diagnosis and treatment of this condition will permit the salvaging of an otherwise normal child.

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