

Should Come to Mind with a Newborn with Respiratory Distress - Congenital Cystic Adenomatoid Malformation?

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ABSTRACT

Irregular, hamartomatous and adenomatous proliferation of the primary bronchioles characterizes Congenital Cystic Adenomatoid Malformation (CCAM). It constitutes 25% of congenital pulmonary malformations and occurs mostly with respiratory distress syndrome in newborns. In this article, the rarely seen CCAM was aimed to be discussed by presenting a case without prenatal diagnosis and a case with symptoms of respiratory distress in the first hours of life.

Keywords: Congenital, cystic adenomatous malformation, newborn

SOLUNUM SIKINTISI OLAN BİR YENİDOĞANDA AKLA GELMELİ – KONGENİTAL KİSTİK ADENOMATOID MALFORMASYON?

ÖZET

Kongenital kistik adenomatoid malformasyon (KKAM), primer bronşiyollerin düzensiz, hamartamatöz ve adenomatoid proliferasyonu ile karakterizedir. Kongenital akciğer malformasyonlarının %25'ini oluşturur ve yenidoğanlarda en sık solunum sıkıntısı ile ortaya çıkar. Bu yazıda prenatal tanısı olmayan ve yaşamın ilk saatlerinde solunum sıkıntısı semptomları gelişen bir vaka sunularak ender görülen KKAM'ın tartışılması amaçlandı.

Anahtar sözcükler: Kongenital, kistik adenomatoid malformasyon, yenidoğan

Irregular, hamartomatous and adenomatous proliferation of the primary bronchioles characterizes Congenital Cystic Adenomatous Malformation (CCAM) and 1/11000 - 1/35000 is frequently seen (1) It mostly affects male infants. It constitutes 25% of congenital pulmonary malformations and occurs mostly with respiratory distress in newborns (2). The diagnosis of these lesions can be made by prenatal ultrasonography (USG). The radiologist may misdiagnose CCAM as a pulmonary cyst, emphysema, or pneumothorax, surgical monitoring is recommended for symptomatic cases and clinical monitoring is recommended for asymptomatic cases as a general approach for CCAM treatment. In this article, the rarely seen CCAM was aimed to be discussed by presenting a case without prenatal diagnosis and a case with symptoms of respiratory distress in the first hours of life.

Case presentation

According to the period from the fourth pregnancy until the last menstrual period of a 31-year-old mother, it was learned that no pathological findings were found in

the prenatal history of a 36-week-old, 3035 grams, caesarean born male baby. He cried as soon as he was born and that was learned from the natal history. The first and fifth minute of Apgar scores were 9 and 10 respectively. The baby started to reveal respiratory problems after birth during the clinical follow-up and pneumothorax had been detected in the left lung and a cystic lesion in the right one in its chest X-ray (Figure 1). He was brought to our hospital, when he was only one day old, for further examination and treatment by being intubated and by placing a branula. During the physical examination, the general condition was poor; there was cyanosis around the lip and extremities. The peripheral circulation is disturbed, the capillary filling time is more than three seconds. Body weight was 3035 gr (50-90p), the length was 52 cm (50-90p), the head circumference was 34 cm (50-90p), his respiration was tachypneic (70-80/ minute), dyspnoeic and intercostal withdrawal. Oxygen saturation was detected as 80%. The chest diameter increased and the left side of the chest was more prominent than the right one. Less breathing sounds during auscultation. The newborn, who was taken with these complaints to the neonatal intensive care unit, whose oxygen saturation did not improve despite oxygen support, whose blood gas analysis contained respiratory acidosis, was intubated again and the mechanical ventilator settings were made. The ampicillin, gentamicin therapy was initiated. A permanent chest tube was attached to the patient through the left thoracic cage due to the appearance of pneumothorax in the left lung during the follow-up. The complete blood count, biochemical tests, thyroid function tests, abdominal ultrasonography and echocardiographic examination were all normal during the laboratory examination. The chest tube was removed while there was no pneumothorax in the control chest radiographs of the case whose respiratory distress regressed and the general condition improved during the examination. Extubation was done ; however, the patient who started respiratory complaints again was intubated and reconnected to the mechanical ventilator. Vancomycin and ceftazidime treatment was started to the patient who showed an increase in acute phase reactants. Cystic fibrosis gene analysis was reported as normal. High resolution computed tomography (HRCT) of the lung was taken from the patient for the differential diagnosis of the congenital cystic adenomatoid malformation or congenital lobar emphysema. Linear cystic features covering the left upper lobe of the lung and the entire middle lobe were detected in his HRCT and was reported as persistent interstitial emphysema (PIA) (Figure 2). The respiratory distress syndrome did not decline during the examination of the patient, did not benefit from the lateral decubitus position



Figure 1. Chest X-ray



Figure 2. HRCT: Linear cystic features covering the left upper lobe of the lung and the entire middle lobe

directed to the PIA and the right selective intubation. A chest tube was attached to the patient whose left lung a pneumothorax was developed in again. It was decided to implement lobectomy on the patient and lobectomy was performed by the paediatric surgeon to the cystic areas in the upper left lobe of the lung. The patient who had no postoperative problems and whose histopathologic diagnosis was consistent with cystic adenomatous malformation type 3 in the postoperative period was discharged on the seventh postoperative day (Figure 3). Post-discharge controls showed that left lung tissue was well expansive (Figure 4). Informed consent was obtained from the family.

Discussion

CCAM is an embryological developmental disorder resulting in a congenital hamartomatous lung lesion that stops bronchi-alveolar maturation at the fourth to eighth gestational weeks and results in overgrowth of mesenchymal

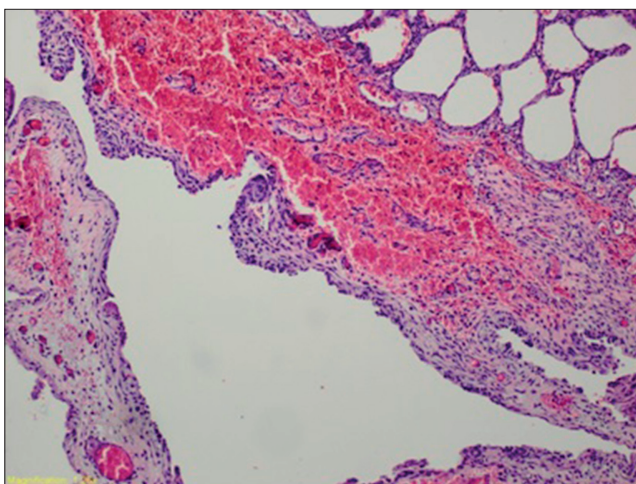


Figure 3. Cuboidal and occasionally pseudostratified epithelial cyst structure (HE, x100)

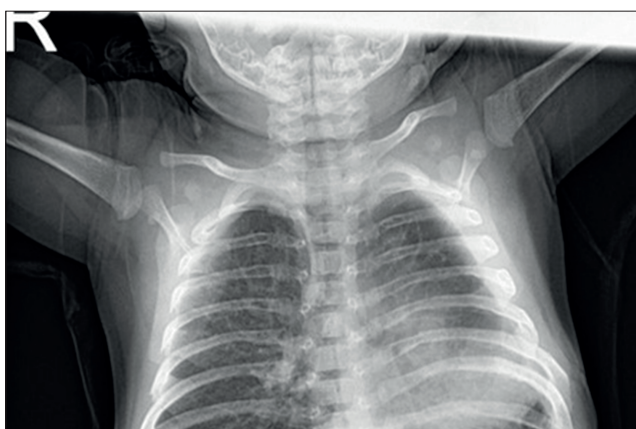


Figure 4. Post-discharge control chest x-ray

cells (3). Cases are seen in less than two years of age due to respiratory distress in the neonatal period or recurrent respiratory tract infections (4). Severe respiratory distress requires urgent surgical resection. There are five types of lesions. Type 0; acinar dysplasia or agenesis and most of them are incompatible with life. Type 1; is the most common type (50-65%), has the best prognosis. Macro-cystic and there is a single or several cysts larger than two centimetre (cm) covered with a ciliated pseudostratified (columnar) epithelium. The cyst wall has smooth muscle and elastic tissue, cartilage is rare. Survival is good. Type 2 has

the worst prognosis (10-40%). Cysts are usually less than one cm and are numerous. These lesions are often associated with stillbirth and prematurity. Common edema, polyhydramnios and associated anomalies (cardiac, renal and chromosomal disorders) are very common. In Type 3, the lung is in the form of a solid mass, consists of multiple small cysts, and is usually less than 0.2 cm (5-10%). The prognosis is poor. Type 4 is rare and its frequency is 2-4%. These are characterized by large peripheral thin-walled cysts covered with type 1 pneumocytes and slightly more round cells representing type 2 pneumocytes (5,6). Prenatal diagnosis is possible with USG, and the best evaluation is done between 16-22 weeks (5). An abnormal high level of amniotic fluid detected in the antenatal USG should be a warning in terms of possible fetal anomalies. Prenatal treatment depends on the presence of symptoms. Fetal intervention is required if fetal hydrops is present. The most commonly used method for diagnosis is lung radiography. On chest radiography, the mediastinum shifts to the opposite side and regional translucence can be observed on the affected side (7). Computed tomography is an adjunctive imaging technique for diagnosis and differential diagnosis together with findings of lung radiography and is also necessary for evaluation of preoperative anatomy. Congenital lobar emphysema, pulmonary sequestration, diaphragm hernia, pneumatocele, bronchogenic cyst, mesenchymal hamartomas should be considered in the differential diagnosis. Treatment is cyst excision. Most important poor prognostic factors in CCAM are, two-sided and/or widespread involvement and the presence of hydrops. In our unit, one patient with CCAM has died as a result of common cystic masses pressure on the lungs by pushing the heart (8).

Conclusion

This article emphasizes the early recognition of CCAM in the prenatal period, possible fetal interventions, the importance of early surgical intervention in terms of prognosis and the importance of coming to mind (considering the CCAM) among the differential diagnoses of a newborn with respiratory distress syndrome.

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