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CASE REPORT / OLGU SUNUMU

Large congenital cystic asdenomatous malformation of the lung in a newborn

Yenidoğanda akciğerin büyük konjenital kistik adenomid malformasyonu

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ABSTRACT

Congenital cystic adenomatous malformation (CCAM) of lung is a rare form of congenital hamartomatous lesions of the lung consisting of cysts filled with air. The general clinic presentation of CCAM is dyspnea in newborns. CCAM may mimic congenital pneumonia or respiratory distress syndrome. After the delivery, the newborn male who had low Apgar score and severe respiratory distress was intubated and admitted to neonatal intensive care unit. Patient was ventilated for 50 days and weaned from the mechanical ventilator at 50th day. Type II CCAM of the lung was diagnosed according to the chest radiographs and computed tomography scan signs. Although the surgeons suggested lobectomy considering the patient's not completely asymptomatic, family did not accept this operation due to the risk of death. The patient was discharged from the hospital until the next control.

Key word: Congenital cystic adenomatous malformation of lunch, newborn, conservative treatment

ÖZET

Akciğerin konjenital kistik adenoid malformasyonu akciğerin hava ile dolu kistlerinden oluşan bir hemartöz lezyondur. Genelde doğumdan sonra solunum sıkıntısı ile bulgu verir. Konjenital pnömoni, respiratuvar distress sendromu ile ayırıcı tanısı zordur. Doğumdan sonra şiddetli solunum sıkıntısı ve düşük Apgar olan bir erkek yenidoğan entübe edildikten sonra yenidoğan yoğun bakım ünitemize yatırıldı. Hasta 50 gün mekanik ventilatörde takip edildikten sonra 50. gün mekanik ventilatörden ayrıldı. Tip II Akciğerin konjenital kistik adenoid malformasyonu tanısı akciğer tomografisi ve akciğer grafi bulguları ile kondu. Cerrahi lobektomi önermesine rağmen hastanın tümüyle aseptomatik olmaması ve ölüm riski nedeniyle aile operasyonu kabul etmedi. Hasta kontrole gelmek üzere taburcu edildi.

Anahtar kelimeler: Akciğerin konjenital kistik adenoid malformasyonu, yenidoğan, konservatif tedavi

INTRODUCTION

Congenital cystic asdenomatous malformation of lung (CCAM) is a rare form of congenital hamartomatous lesions of the lung consisting of cysts filled with air. CCAM may mix with congenital pneumonia, respiratory distress syndrome. It was first described by Ch'in and Tang in 1949 [1]. The incidence of CCAM was estimated as 1 per 25000 to 35000 births. It accounts for 25% of all congenital lung malformations and 95% of congenital cystic lung malformations [2]. The general clinic presentation of CCAM is dyspnea in newborns [3], it can also cause bronchiolo alveolar carcinoma and rhabdomyosarcoma [4], and usually diagnosed by using lung X-ray, antenatal ultrasonography and computed tomography or lung biopsy [5]. Stocker and al. classified three types of CCAM in 1977, type I consists large cysts greater than 2.0 cm diameter; type II consists small cysts and type III consists a resembling homogeneous mass lesions with cysts only seen on microscopy [6]. The aim of this study is to present a newborn with CCAM.

CASE REPORT

A newborn male from healthy parents was born at 38 weeks of gestation by elective cesarean section due to severe toxemia and polyhydramnios. Thirty-eight years old mother had no history of pulmonary disease in her family and the antenatal ultrasonography (USG) was shown only polyhydramnios.

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After the delivery, a newborn male who had poor Apgar score and severe respiratory distress was intubated successfully using a direct laryngoscope and admitted to neonatal intensive care unit. The newborn patient needed to be ventilated for 50 days and weaned from the mechanical ventilator at 50th day of his life. Type II CCAM of the lung was diagnosed according to the chest radiographs and computed tomography scan. The first chest radiographs and computed tomography scan revealed multiple small cysts filled with air in the middle lobes of the lungs and the heart was pushed to the left of the chest (Figure 1, Figure 2).

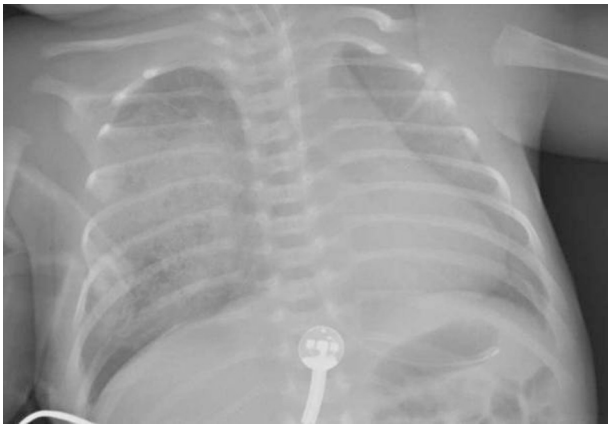

Figure 1. First days chest radiograph; multiple small cystic filled with air in the middle lobe of the lung and the heart was pushed to left of chest

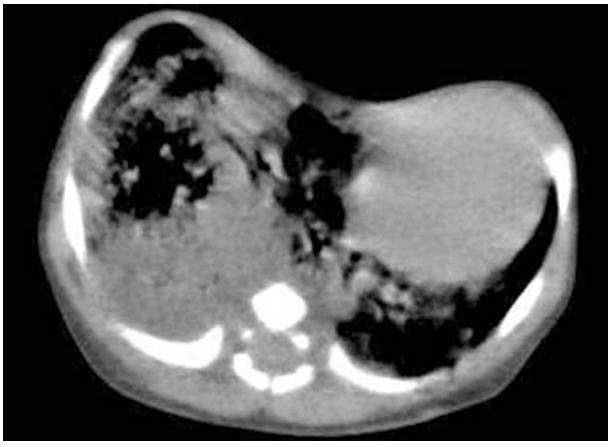

Figure 2. First computed tomography scan; multiple small cystic filled with air in the middle lobe of the lung and heart was pushed to left of chest

In addition, the chest radiographs and computed tomography scan designated a better sign, which is the normal position of the heart (Figure 3, Figure 4). Although the surgeons suggested lobectomy considering the patient is not completely asymptomatic, family did not accept this operation due to the risk of death. The patient was discharged from the hospital until the next control.

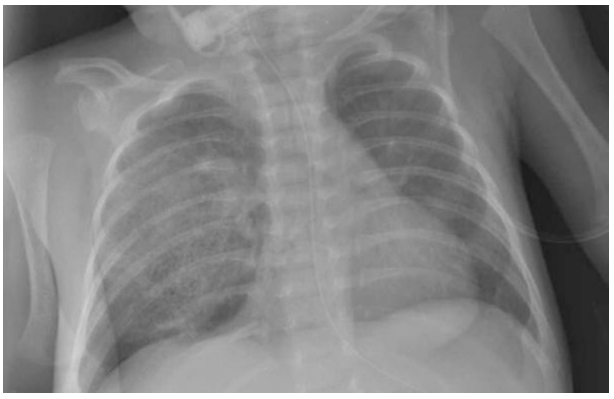

Figure 3. The last days chest radiograph; multiple small cystic filled with air in the middle lobe of the lung and position of the heart was normal

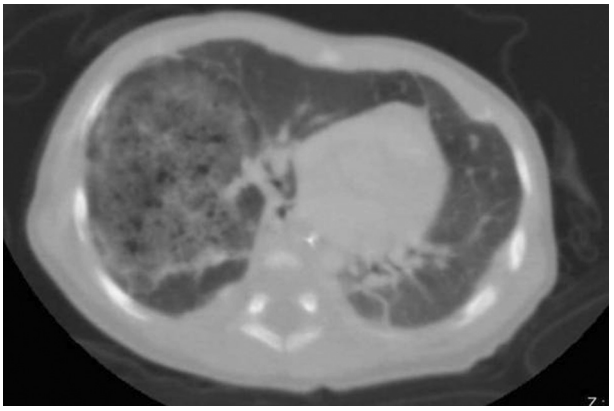

Figure 4. Second contrast-enhanced computed tomography scan; multiple small cystic filled with air in the middle lobe of the lung and position of the heart was normal

DISCUSSION

This case may be the first in the literature that the patient was ventilated for 50 days and had weaned from the mechanical ventilator at 50th day of his life.

Chen et al. reported seven patients with respiratory distress begins immediately after the birth were subsequently diagnosed with CCAM (%50 cases), 62% of patients had type II CCAM and 56% of the lesions were on the right lung [7]. Parikh et al. found all of their 22 patients were asymptomatic

[8]. Some studies reported that 50-72% of the patients with CCAM were asymptomatic at birth and 5-60% of radiographs taken after the birth were normal [9]. Our case was symptomatic type II CCAM with abnormal lung radiographs at birth and then symptoms of the patient regressed slowly.

18-20% of CCAM patients can born with Prune Belly Syndrome, pulmonary sequestration, diaphragmatic hernia, agenesis of a lung, jejunal atresia, cardiac and renal anomalies [10]. Our case had only pectus excavatum abnormality.

CCAM is usually diagnosed with lung X-ray, antenatal ultrasonography and computed tomography or lung biopsy [5]. Our case was diagnosed with images of the chest radiographs and computed tomography scan.

Polyhydramnios and microcystic type of lesions in patients with CCAM are associated with a poor prognosis [11]. The major cause of morbidity is respiratory distress caused by compromised pulmonary function or pulmonary hypoplasia. Surgery should be considered in patients with large CCAM and in the case of poor prognosis11. Dommergues at al. treated 17 patients with CCAM without acute polyhydramnios or hydrops with conservative treatment [12]. Dommergues at al. also treated 17 patients with CCAM without acute polyhydramnios or hydrops by conservative treatment [12]. In our case, although the surgeons suggested lobectomy considering the patient had polyhydramnios so was not completely asymptomatic, family did not accept this operation due to the risk of death. Therefore, we treat the patient with conservative treatment, then the patient's symptoms regressed.

In summary, CCAM should be considered among the causes of the respiratory distress in neonates, and it may mix wit with congenital pneumonia, RDS.

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