

Incidental Detection of Congenital Cystic Adenomatoid Malformation After Thoracoscopic Repair of Diaphragmatic Hernia

Torakoskopik Diyafram Hernisi Onarımı Sonrası Rastlantısal Konjenital Kistik Adenomatoid Malformasyonun Saptanması

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Abstract

Congenital pulmonary airway malformation (CPAM) is a rare benign lung lesion, and CPAM combined with congenital diaphragmatic hernia (CDH) is extremely rare. The patient being described was a neonatal boy with a left-sided CDH diagnosed after birth. Thoracoscopic repair was performed on the first postnatal day. On chest X-ray, a suspicious lesion in terms of recurrence was observed after the postoperative follow-up period. Type 2 CPAM was diagnosed based on computed tomography. The child is doing well on postoperative period and the lesion stays stable without any intervention. Congenital lung lesions with CDH are difficult to diagnose before the CDH repair. Although very rare, congenital lung lesions concurrent with CDH should be considered when managing these patients.

Keywords: Cystic adenomatoid malformation of lung, congenital, pulmonary surgical procedures, newborns, surgery, thoracoscopic

Öz

Konjenital pulmoner hava yolu malformasyonu (CPAM) nadir görülen benign bir akciğer lezyonudur ve konjenital diyafragma hernisi (KDH) ile birlikte CPAM oldukça nadirdir. Doğum sonrası sol taraflı konjenital diyafragma hernisi KDH tanısı alan yenidoğan erkek bir olgu sunulmuştur. Doğum sonrası birinci günde torakoskopik onarım yapıldı. Ameliyat sonrası takiplerinde çekilen akciğer grafisinde nüks açısından şüpheli lezyon görüldü. Tip 2 CPAM tanısı bilgisayarlı tomografi ile konuldu. Ameliyat sonrası dönemde çocuğun genel durumu iyi olup, lezyon herhangi bir müdahaleye gerek kalmadan stabil takip ediliyor. KDH ile birliktelik gösteren konjenital akciğer lezyonlarının KDH onarımından önce teşhis edilmesi zordur. Bu hastaların tedavisinde KDH ile birlikte nadir görülen konjenital akciğer lezyonları da göz önünde bulundurulmalıdır.

Anahtar Kelimeler: Akciğerin kistik adenomatoid malformasyonu, konjenital, akciğer cerrahisi işlemleri, yenidoğanlar, ameliyat, torakoskopik

Introduction

There are several theories explaining the development of pulmonary hypoplasia. Most accepted theory is that pulmonary hypoplasia occurs due to the mass effect of abdominal contents present in the thorax during lung development (1).

Structural defects with respiratory abnormalities which include congenital lung lesions such as congenital pulmonary airway malformation (CPAM) and bronchopulmonary sequestration (BPS) are rarely seen concurrent with congenital diaphragmatic hernia (CDH) (2). The presentation of an additional airway malformation rises concerns of morbidity with varying degrees of respiratory distress. There are also

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studies demonstrating no difference in the outcome of CDH patients with concurrent congenital lung lesions (3). However, there is no specific data about the prognosis of CDH and CPAM.

The aim of this study is to present a child who has CDH with concurrent CPAM.

Case Presentation

Informed consent was obtained from the parents. A male newborn was born through cesarean section at the 39th week to a 27-year-old mother who was prenatally followed by an obstetrics and gynecology specialist. He had an apgar score of 6/8 at birth and developed respiratory distress in delivery room. The newborn was admitted to neonatal intensive care unit and nasal intermittent positive pressure ventilation was initiated. Arterial blood gas was evaluated to show normal results (power of hydrogen pH:: 7.34, pressure of carbon dioxide $p\text{CO}_2$: 39.8, lactate: 1.2). A chest X-ray was taken due to respiratory distress (Figure 1). Even though CDH was not prenatally diagnosed, signs of CDH were presented on the X-ray and the patient was rapidly intubated. Thorax ultrasound (US) imaging showed that the intestines and the colon was partially located in the left hemithorax. Echocardiography of the newborn revealed patent ductus arteriosus and pulmonary hypertension. Abdominal and urinary US of the newborn revealed no other anomalies. On the

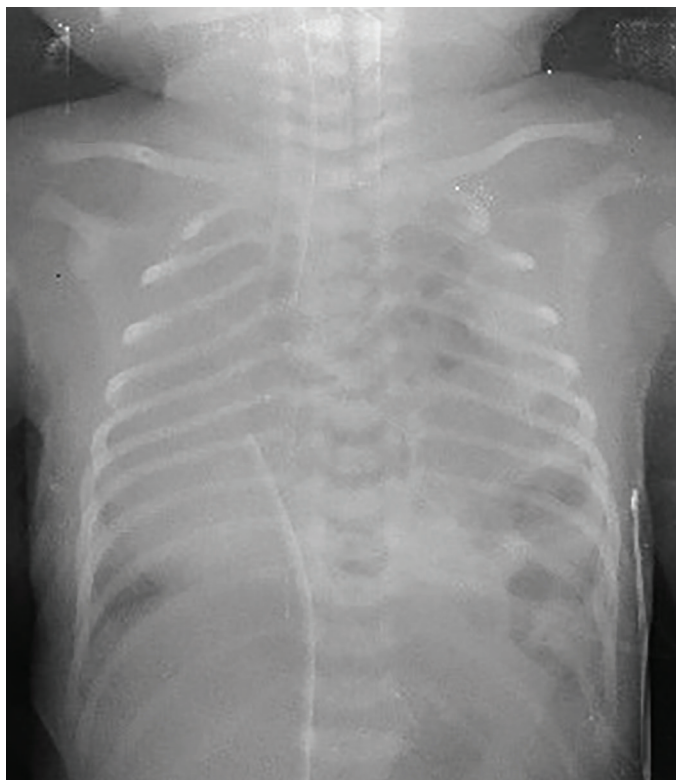


Figure 1: Chest X-ray which demonstrates congenital diaphragmatic hernia on the left side. Right hemithorax looks radiopaque possibly due to mediastinal shift

first postnatal day, thoracoscopic diaphragmatic hernia repair was performed. After the operation the newborn was stayed on mechanical ventilation and extubated on 3rd day of operation. The baby did not require any oxygen or respiratory support afterwards. In the first postoperative day, chest X-ray revealed a suspicious lesion (there is a multiloculated cystic lesion which was thought to be recurrent diaphragmatic hernia) in terms of recurrence was observed while the patient was clinically asymptomatic without any need for oxygen support (Figure 2). Distal colon imaging was performed which showed no recurrence. Thorax computed tomography (CT) was performed, and a suspicious cystic lesion was seen to be compatible with type 2 CPAM (Figure 3). The patient was referred to pediatric pulmonologist for follow-up.

Discussion

We report a rare case of CPAM concurrent with CDH which is diagnosed after thoracoscopic CDH repair. Postoperative chest X-ray showed a lesion suspicious of bowel on the left hemithorax, which supported recurrence of CDH. CT was performed to reveal there was no recurrence but type 2 CPAM in the left lower lobe of the newborn was presented.

CDH is a life threatening pathology in newborns and a cause of death due to pulmonary hypoplasia and pulmonary hypertension (4). On the other hand, CPAM has lower mortality but it is a potential cause of infection and malignancy (5). CDH is a major congenital anomaly caused by failure of closure of the pleuroperitoneal cavity by the fusion of septum transversum and pleuroperitoneal folds (2). In cases of pulmonary sequestration concurrence, it is hypothesized that the formation of sequestration during the embryonic period

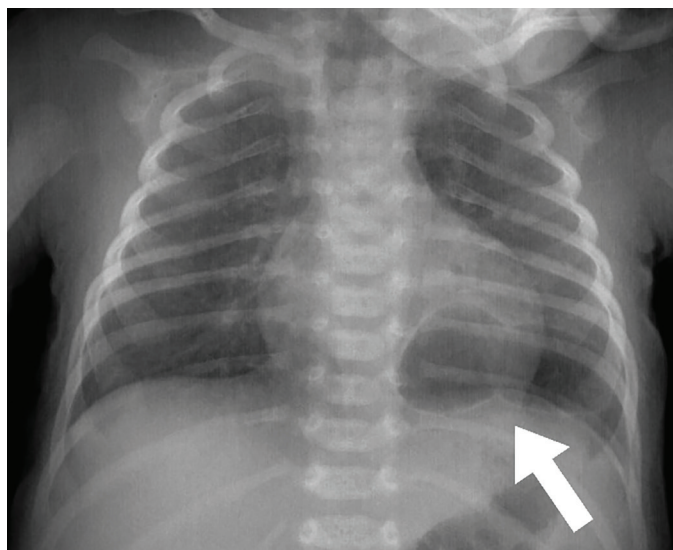


Figure 2: Chest X-ray shows suspicious lesion (there is a multiloculated cystic lesion which was thought to be recurrent diaphragmatic hernia) in the left lower zone

may interfere mechanically with the fusion of the lungs and the diaphragm (3).

In the literature, congenital lung lesions associated with CDH are rarely described and the exact incidence of these lesions concurrent with CDH is uncertain (2). Case series showed that the incidence of BPS with CDH are between 15 and 30% (4). Savic et al. (6) showed that the incidence of CDH with BPS were 3% and 27%. On the other hand, Soni et al. (4) showed that incidence of both CPAM and BPS with CDH was 7.2%. However, CPAM concurrent with CDH are reported very rarely.

CPAM is characterized by hamartomatous lesions of the lungs classified by the size of the cysts (7). Stocker's classification groups CPAM into three groups according to cyst size. Type 1 is the most common and has macrocysts larger than 2 cm. Type 2 which is associated with other congenital anomalies, has multiple cysts smaller than 2 cm. Type 3 has cysts smaller than 0.5 cm and includes solid components (7). In this case, type 2 CPAM was reported on CT following the operation for CDH. When CPAM appears as a single, localized, solid or cyst like lesion, it may be difficult to differentiate it from bronchogenic cysts, bronchial atresia and pulmonary sequestration (8).

The incidence rate of 5–65% for recurrence after CDH repair is reported in literature (9). The postoperative chest X-ray was suspicious of recurrence thus, a CT was performed to confirm diagnosis. CT showed two pulmonary cysts less than 2 cm in the left lower lobe, which was concordant with the lesion on the chest X-ray (Figure 3).

Surgery is a preferred method of treatment for symptomatic CPAM, but postnatal management of asymptomatic lesions are

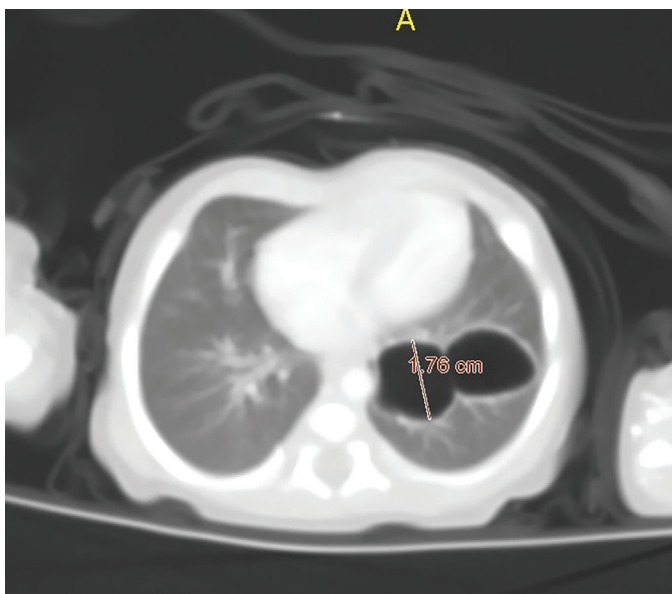


Figure 3: Axial CT scan of chest demonstrates a left lower lobe lesion with two cysts

CT: computed tomography

controversial (10). In the literature, some authors argue that early and elective thoracoscopic surgery prevent infective complications (11). By contrast, some authors argue that surgery is chosen based on patient according to the onset of symptoms (10).

Congenital lung lesions with CDH are difficult to diagnose before the CDH repair. However, observations on the development of the lungs can be made by surgeons during the CDH repair. Although very rare, congenital lung lesions concurrent with CDH should be considered when managing these patients.

Ethics

Informed Consent: Informed consent was obtained from the parents.

Footnotes

Authorship Contributions

Surgical and Medical Practices: D.İ., P.K., E.E., U.A., Concept: D.İ., K.B., P.K., E.E., Design: D.İ., K.B., E.E., Data Collection or Processing: D.İ., E.E., U.A., Literature Search: D.İ., K.B., E.E., U.A., Writing: D.İ., K.B., P.K., E.E.

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