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Extralobar Pulmonary Sequestration in a Neonate

Bir Yenidoğanda Ekstralober Pulmoner Sekestrasyon

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Abstract / Özet

Congenital pulmonary sequestration (CPS) is a type of thoracic malformation that may be represented as a solid or cystic mass composed of nonfunctioning primitive tissue. It does not interact with the tracheobronchial tree and has unusual systemic blood supply. Various congenital malformations may be presented with CPS. More than one-half of CPS cases are diagnosed in later childhood or even in adulthood. Neonates and infants are usually asymptomatic, and they are usually diagnosed due to the presence of other congenital anomalies. Here, we report a 5-day-old neonate who was admitted to our clinic with respiratory distress. CPS was diagnosed without any other malformation.

Keywords: Extralobar lung sequestration, lung, neonate, sequestration

Konjenital Pulmoner Sekestrasyon (KPS) fonksiyon görmeyen primitif dokudan meydana gelen solid veya kistik kitle şeklinde görülebilen bir torasik malformasyondur. Trakeobronşiayal ağaçla bağlantısı yoktur ve alışılmadık bir sistemik kan dolaşımına sahiptir. Çeşitli konjenital malformasyonlar KPS'a eşlik edebilir. Olguların yarıdan çoğuna ileri çocukluk dönemi hatta erişkinlik döneminde tanı konulmaktadır. Yenidoğanlar ve çocuklar genellikle asemptomatiktir. Bu grupta tanı genellikle diğer anomalilerin varlığından dolayı konmaktadır. Burada solunum sıkıntısı ile kliniğimize kabul edilen 5 günlük yenidoğan sunulmaktadır. Başka hiç bir malformasyonu bulunmayan hastamıza KPS tanısı konulmuştur.

Anahtar Kelimeler: Ekstralobar akciğer sekestrasyonu, akciğer, yenidoğan, sekstrasyon

Introduction

Congenital pulmonary sequestration (CPS) is an infrequently occurring congenital pulmonary malformation, which features nonfunctioning tissue as a remnant of a diverticular outgrowth of the foregut (1, 2). It has no normal relationship with the bronchial structure and the pulmonary arteries. The arterial blood supply originates from the systemic circulation, and venous drainage occurs via the pulmonary veins (3-5). Extralobar pulmonary sequestration (EPS) is seen more often in males, and in a number of patients, pulmonary sequestration may include some malformations, such as a congenital diaphragmatic hernia (6, 7). Although the presenting symptoms are variable, most newborn patients can be found with respiratory distress (8).

Herein, we present a case of extralobar pulmonary sequestration in a newborn with respiratory distress symptoms.

Case Report

A 5-day-old male neonate, born at full term via normal vaginal delivery following an uneventful pregnancy to a healthy mother, was brought to the emergency room with respiratory distress symptoms. A radiograph of the chest revealed a left lower lobe opaque shadow (Figure 1), and left lower lobe pneumonia was diagnosed. The neonate was treated with a presumptive diagnosis of pneumonia, and complete resolution of the clinical features was observed. Laboratory tests showed a normal leukocyte count, and a computed tomography (CT) scan of the thorax showed a heterogeneous mass in the posterior basal segment of the left lower lobe after contrast enhancement (Figure 2).

Upon exploration, a left lower lobe sequestration was found to be attached to the left posterior pleura only by the vascular pedicle. It had an independent artery and vein but no bronchi. The excised specimen showed a piece of pyramidal lung tissue that was completely invested with the pleural lining, measuring 6x4x3 cm (Figure 3a, b). The histological examination revealed the bronchus in the pedicle attached to the left posterior pleura. Additionally, it showed immature lung tissue with many of the alveoli and bronchial lumina expanded into the interstitial areas and filled with eosinophilic proteinaceous fluid, neutrophils, and histiocytes (Figure 4). Post-operatively, the patient made an uneventful recovery.

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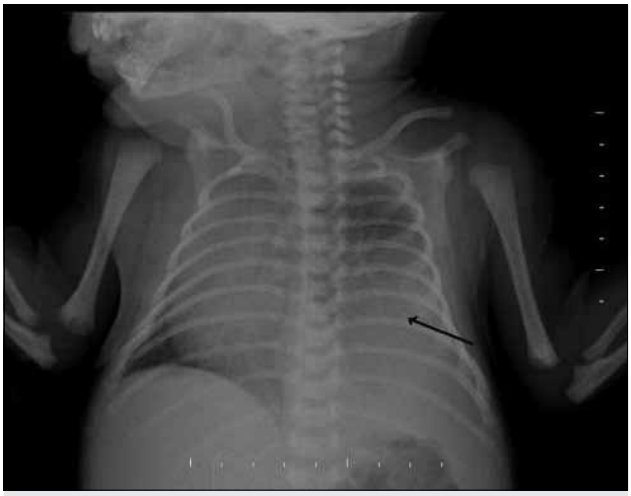

Figure 1. Chest radiograph with opacity in left lower zone (arrow)

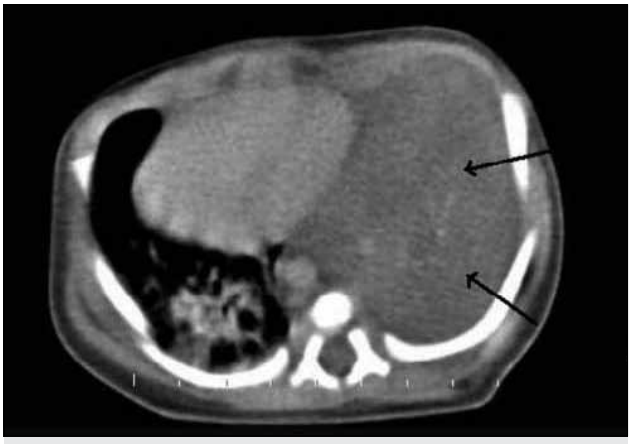

Figure 2. Contrast CT scan of thorax showing heterogenous mass (arrow) in left lower lobe

Discussion

Congenital pulmonary sequestration effects approximately 0.15%-1.7% of all infants. It is characterized by a mass of nonfunctioning, embryonic, cystic tissue that has no relationship with the bronchial tree or the pulmonary arteries. The arterial blood supply is from the thoracic or abdominal aortic branch, and the venous drainage is through the azygos system, pulmonary veins, or inferior vena cava. CPS is divided into two types: extralobar and intrapulmonary. Both sequestrations occur via the same embryological evolution as a remnant of a diverticular outgrowth of the foregut. Therefore, gastric or pancreatic tissue may be found inside of the sequestration (1, 3, 9, 10). The case presented in the present study was an extralobar pulmonary sequestration, and the arterial blood supply was from the abdominal aortic branch.

Extralobar pulmonary sequestration (EPS) is seen in 15%-25% of all pulmonary sequestration patients and creates 0.5% to 6% of congenital lung disease. EPS is encountered more frequently in males, with a ratio of 4:1. There is usually a supernumerary number of lung buds in the pathogenesis, and in as many as 80% (majority) of EPS patients, it occurs posteriorly on the left side. EPS settles generally within the thorax, but cases of EPS have been described within the mediastinum and pericardium and below the hemidiaphragm. It is a sequestered lung that arises after the

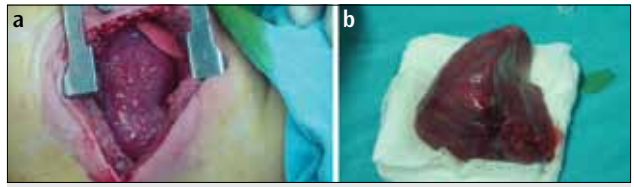

Figure 3. a, b. Sequestering lung tissue view before (a) and after (b) excision

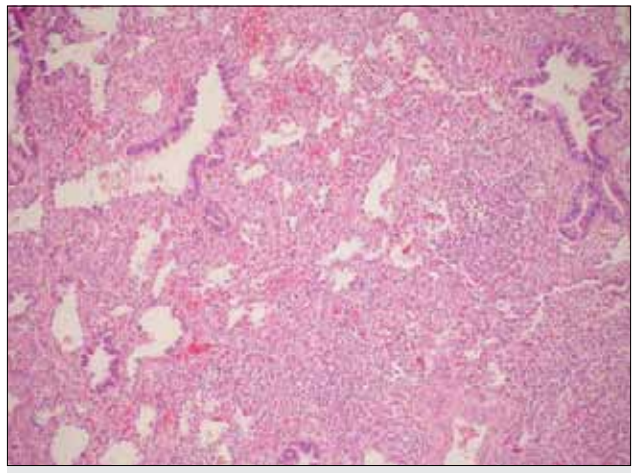

Figure 4. Sequestered lung tissue filled with neutrophils and showing cystic dilatation of bronchioles and alveoli (H&E 100x)

formation of the pleura and thus has its own pleural covering (6, 11-13). Herein, our case was a male infant, and the mass was in the left thorax.

EPS has many related anomalies at a ratio of approximately 65%, such as an accessory spleen, complex heart disease, and diaphragmatic hernia (14). Other associated abnormalities include communication with the esophagus or stomach (T-E fistula), bronchogenic cysts, vertebral abnormalities, megacolon, and cystic adenomatoid malformation of the lung. In about 15%-25% of the cases with type II EPS, cystic adenomatoid malformation of the lung has been reported. In our case, there were no additional anomalies.

The presentation can be variable, ranging from no symptoms to hemoptysis. Usually, it presents as an asymptomatic intrathoracic mass associated with respiratory distress because of airway compression. A fistula between the sequestration and the digestive tract or direct invasion from pneumonia in the surrounding lung may occur in patients with pulmonary sequestration. Heart failure and hemoptysis are other known complications (14, 15). The diagnosis of pulmonary sequestration is confirmed using imaging modalities, such as angiography, computed tomography (CT), MRI, ultrasonography (US), and chest radiographs (CR) (16). CR will often show an opacity in the affected segment, while CT shows an irregular cystic component in the mass. US may show the lesion to be of mixed echogenicity and hyperechogenicity (17). In infants with respiratory failure associated with pulmonary sequestration, early diagnosis and surgical treatment remain the approaches of choice to achieve an optimum outcome (15). On the other hand, surgical treatment in infants with significant respiratory symptoms may cause morbidity and mortality.

Conclusion

In this paper, we presented a case of pulmonary sequestration in an infant with respiratory distress. In the chest radiograph, an opacity was found in the left lower zone. Additionally, the contrast CT scan of the thorax demonstrated a heterogeneous mass in the left lower lobe. The patient was treated for pneumonia, and a CPS resection was performed through a left thoracotomy. After surgery, the patient was discharged from the hospital with no further complications.

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