Intralobar Sequestration with Pulmonary Vascular Anomaly and Pulmonary Hypertension

Pulmoner Vasküler Anomali ve Pulmoner Hipertansiyon ile Birlikte Olan İntralober Sekestrasyon

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ABSTRACT

Pulmonary sequestration is a portion of pulmonary tissue that does not have communication with the tracheobronchial system and is supplied by anomalous systemic arteries. Association of the pulmonary sequestration with congenital anomaly is not rare. However, to date, pulmonary vascular anomaly association has not been reported in the literature. Herein, we presented a 20-year-old man having intralobar sequestration associated with pulmonary vascular anomaly and high pulmonary artery pressure which decreased significantly after right lower lobe resection. *(Tur Toraks Der 2009; 11: 124-6)*

Key words: Lung, congenital lesion, pulmonary hypertension, intralobar sequestration

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ÖZET

Pulmoner sekestrasyon, trakeobronşiyal damar sistemi ile bağlantısı olmayan ve anormal bir sistemik damarlanması olan pulmoner dokulardır. Konjenital anomaliler ile ilişkisi ise nadir değildir. Ancak bugüne kadar pulmoner vasküler anomali ile olan ilişki henüz literatürde tanımlanmamıştır. Bu yazıda, 20 yaşında pulmoner vasküler anomali ve artmış pulmoner arter basıncı ile ilişkili intralober sekestrasyonu olan hastanın pulmoner arter basıncının, sağ alt lob rezeksiyonu sonrası anlamlı bir şekilde düştüğünü gösterdik. *(Tur Toraks Der 2010; 11: 124-6)*

Anahtar sözcükler: Akciğer, konjenital lezyonlar, pulmoner hipertansiyon, intralober sekestrasyon

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INTRODUCTION

Pulmonary sequestration (PS) is defined as a congenital malformation consisting of aberrant lung tissue mass that does not have a normal connection with the tracheobronchial tree and has a systemic arterial blood supply. The sequestration term was first mentioned by Price in 1946. PS's are divided into two subgroups; intralobar sequestration (ILS), which is situated within normal lung parenchyma; and extralobar sequestration (ELS), which is separated from the normal lung and has its own visceral pleura. Pulmonary sequestration is usually seen in the left lower lobe. The arterial supply to PS is commonly from a systemic artery and venous drainage is to the pulmonary vein in ILS and to a systemic vein in ELS [1]. ILS consists of 75% of all PS's. Herein we presented a 20-year-old man having ILS who was diagnosed while associated cardiac disease was being evaluated. The patient's pulmonary artery pressure was 56 mmHg and three months after surgery it was 47 mmHg. For our case, it was interesting that it was not established that the superior pulmonary vein and the middle lobe venous drainage was to the inferior pulmonary vein.

CASE

A 20-year old man presented to the cardiology clinic complaining of atypical chest pain. He was hospitalized to evaluate atrial septal defect and other differential diagnosis. At presentation, the vital findings were normal, excluding the decreased respiratory sounds in the lower zone of the right hemithorax. Electrocardiography was normal. Pulmonary artery pressure was measured as 56 mmHg and first degree tricuspid insufficiency was discovered by Echocardiography. Other findings were normal.

A chest radiograph revealed an ill-defined opacity in the right lower lung field. Contrast-enhanced computed tomography (CT) of the thorax was carried out for further evaluation. It revealed a multiloculated cystic lesion within the medial right-lower lobe. A 9-mm caliber supplying artery from the thoracic aorta to this cystic field was determined (Figure 1). After determining these imaging features characteristic of ILS, the patient was referred to our clinic.

He underwent right lower-lobe resection via right thoracotomy. During surgery, the inferior pulmonary ligament was carefully dissected and the aberrant artery within it was not injured (Figure 2). After right hiler dis-

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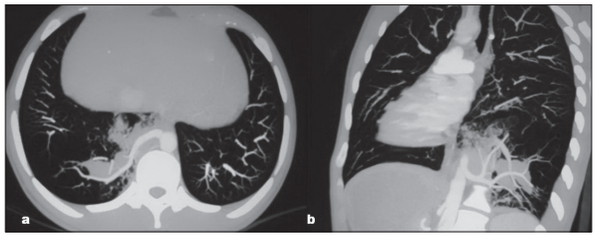

Figure 1. Contrast-enhanced computed tomography (CT) of the thorax, axial (a) and coronal (b) plane, show the 9-mm caliber feeding vessel of the sequestration arising from the thoracic aorta

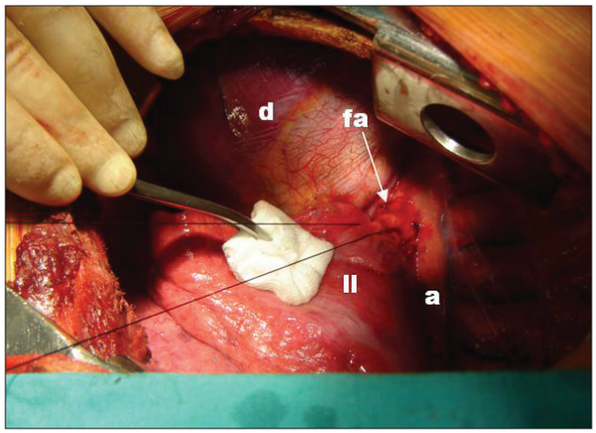

Figure 2. Intraoperative view of the feeding artery of the right lower lobe sequestration after dissection of inferior pulmonary ligament. fa: feeding artery, a: aorta, d: diaphragm, ll: lower lobe

section it was seen that inferior pulmonary venous drainage was to the middle lobe vein, not to the right atrium. The postoperative course was uneventful. A 2-month follow-up examination was carried out and the pulmonary artery pressure was measured as 47 mmHg, which was a significant decrease. Histopathological examination had suggested ILS.

DISCUSSION

Pulmonary sequestration indicates an abnormal pulmonary tissue that does not have a connection with the tracheabronchial tree and its blood supply is usually from an aberrant artery arising from the aorta or one of its branches.

Sequestration is a rare congenital anomaly and comprises 0.15-6% of all pulmonary malformations [2]. The etiology of PS has not yet been clarified. Numerous theories of the pathogenesis of PS have been suggested in the literature and have been summarized by Corbett and Humphy [3]. The most widely accepted theory is that it results from formation of an accessory lung bud inferior to the normal lung buds during development. During embryogenesis, this accessory lung bud develops and an independent vascular supply, usually from the aorta, remains independent from the normally developing tracheabronchial tree [3].

On the other hand, in the study of Stocher and Makzak, arteries from the thoracic aorta branching into the visceral pleura transversing the ligamentum pulmonale have been found. They proposed a sequence of events, including a chronically infected lesion obstructing a bronchus and occluding the pulmonary artery that could cause the acquired ILS [4].

Because of the high frequency of ELS with other congenital abnormalities, including bronchogenic cyst, diaphragmatic hernia, adenoid cystic malformation and lobar emphysema, ELS is confined to neonates. Diaphragmatic hernia is the most frequently associated congenital anomaly in 30% of all ELS cases [5]. In contrast, ILS is rarely associated with congenital abnormalities. The majority of patients with ILS are asymptomatic and they are usually diagnosed accidentally during investigation for unrelated symptoms in a routine chest radiography and half of them present after the age of 20 years [6]. Our case was diagnosed during evaluation for etiology of high pulmonary artery pressure in the cardiology clinic.

Recurrent episodes of pneumonia, including the signs of fever and productive cough, are the most common, but nonspecific symptoms including chest pain, pleuritic pain, shortness of breath and wheezing could be found at presentation. PS's are not completely isolated from the normal lung and they have connections via the alveolar pores of Kohn by which bacteria can invade. The bacterial colonization of the PS results with frank infection because of the lack of normal bronchial drainage.

The vascular changes, which included medial and intimal thickening, angioblastic proliferation, plexiform lesions, and dilation lesions, occur in a setting of hypoxia, chronic inflammation, and high pressure and flow via a systemic arterial supply [7]. An increase in cardiac output attributed to left to left? shunting in ILS may lead to cardiac failure. Left ventricular function may also be impaired due to continuous enlargement of the shunt. Thus, ILS can be masked as a cardiac malformation for years. Our case was evaluated for the etiology of high pulmonary artery pressure and he was diagnosed after a careful physical examination and accurate diagnostic tool choice.

Plain chest radiograph is the first step and reveals an ill-defined consolidation such as pneumonia or a soft tissue mass with well or ill-defined borders which is usually nonspecific but warrants further evaluation [8,9]. Thorax CT usually reveals a discrete mass in the posterior or mediobasal segment of the lower lobe, with or without cystic changes [9]. Emphysematous changes in the surrounding lung parenchyma are characteristic CT findings of sequestration [8,9]. Thorax CT may also show the mass, with an abnormal vessel originating from the thoracic aorta and associated congenital anomalies, if present.

The differential diagnosis of ILS includes pneumonia, abscess, tuberculosis, bronchiectasis, bronchogenic cyst, pericardial cyst and tumoral lesions. The accurate diagnostic tool in PS is selective angiography but less invasive radiographic methods can be used for identifying the supplying artery. Pre-operative identification of the supplying artery is important to avoid unexpected bleeding during surgery. However, Suat Gezer at al. identified only three

of 27 patients with angiography preoperatively and they reported that they encountered no difficulty such as major bleeding because of careful dissection [10].

In the literature there are many case presentations concerning the association of PS with bronchogenic cyst, diaphragmatic hernia or adenoid cystic malformation. However, we did not find any PS cases associated with pulmonary vascular anomaly. Also pulmonary hypertension was another interesting association with PS, which was significantly decreased after surgical resection.

The treatment choice in PS is surgical removal. Severe complications such as massive hemoptysis from ILS, which can be fatal, makes surgical removal mandatory at the time of diagnosis. The optimal treatments are mass resection for ELS and lobectomy for ILS [11]. Surgical removal via video assisted thoracoscopic surgery (VATS) is now the popular method, but inflammation and fibrosis due to recurrent infection cause difficulties in carrying out this method. However, Kestenholz and associates published the successful outcomes of 14 patients with PS in whom VATS resection was performed. In only one of the 14 patients was it necessary to convert to a thoracotomy because of bleeding from an aberrant artery [12]. In addition, angiographic embolization of the feeding systemic vessel is an alternative treatment option for the cases where the risk of surgery is too great.

This case is an interesting presentation of an ILS because of the association with the pulmonary vascular anomaly and pulmonary hypertension. The association of pulmonary vascular anomaly with pulmonary sequestration is infrequent. It is important to be aware of the coexistence of these anomalies in order to achieve uncomplicated surgery and evaluation of patients with pulmonary hypertension.

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