Extralobar Pulmonary Sequestration With Anomalous Blood Supply From The Subclavian Artery

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Abstract

Pulmonary sequestration is a rare embryonic mass of lung tissue that has no identifiable bronchial communication and that receives its blood supply from one or more anomalous systemic arteries. Systemic blood supply is commonly from the thoracic aorta. Venous drainage is commonly to systemic pulmonary vessels. A healthy, asymptomatic 20 year-old male was admitted to our hospital for evaluation of a left sided mass. The standard chest roentgenograms, a contrast-enhanced chest tomography, a bronchoscopic examination, thoracic magnetic resonance imaging and a selective angiography were performed. With these findings we decided that this heterogeneous lesion between mediastinum and left upper lobe as extralobar pulmonary sequestration. The patient underwent left sided posterolateral thoracotomy. We performed sequestrectomy. Postoperative period was uneventful.

INTRODUCTION

Pulmonary sequestration is an uncommon congenital pulmonary malformation, characterized by a mass of non-functioning lung tissue, which is separated from the normal bronchopulmonary tree. Anatomically, it is classified into two types, intralobar (ILS) and extralobar (ELS). Systemic blood supply is commonly from the thoracic aorta, but arteries may occasionally arise from other sites including the abdominal aorta, intercostal arteries, internal mammarian artery, subclavian artery or from circumflex coronary artery, rarely. Venous drainage is commonly to systemic pulmonary vessels, but variable drainage may be to superior and inferior cava, brachiocephalic or azygous vena. In the ILS, the malformation is incorporated in the normal pulmonary parenchyma of a lobe. The ELS consists of pulmonary parenchyma separated from the rest of the lung by its own pleural envelope and corresponds to a true accessory lung [1,2,3]. This present study aimed to report extraordinary located extralobar pulmonary sequestration arising from the subclavian artery.

CASE REPORT

A 20 year-old male was admitted to our hospital for evaluation of a left sided cystic lesion. He was healthy and asymptomatic and had no previous history of respiratory symptoms. His body physical examination was normal. The standard chest roentgenograms showed a rounded density on the left upper lobe adjacent to aortic arch. A contrast-enhanced chest tomography was performed (Figure 1).

Figure 1

Tomography demonstrated a heterogeneous high-attenuating non-enhancing mass with irregular multi cystic parameter on the anterior mediastinum broad to left upper lobe with dimension 6x4x5 centimeters. This lesion was between manubrium sterni, pericardium and posterior segment of left
upper lob. A bronchoscopic examination was normal. Thoracic magnetic resonance imaging demonstrated no vessel invasion by this lesion. We performed a selective angiography because of suspicion that can be pulmonary sequestration. That showed an arterial supply from left subclavian artery, venous drainage was to superior vena cava and superior pulmonary venous system (Figure 2).

**Figure 2**
Figure 2: Selective angiography; arterial supply was from subclavian artery, venous drainage was to superior vena cava and superior pulmonary venous system.

With these findings we decided that the lesion between mediastinum and left upper lobe as extralobar pulmonary sequestration inspite of the lesion had irregular multi cystic parameter. We performed left posterolateral thoracotomy. The ELS was removed by sequestrectomy. No pulmonary resection was carried out.

At pathological evaluation, the specimen consisted of a 170-gram spongy cystic structure filled with thick yellow-brown fluid. It was covered with an own abnormally visceral pleura and color was brown. Microscopic evaluation revealed dilated bronchioles lined by ciliated pseudostratified columnar epithelium. Amorphous material constitute of basophilic polygonal and rod-shaped calcifications filled the air spaces. Chronic inflammatory cells were identified in the interstitium and alveoli.

The patient was discharged at 7th postoperative day and returned his routine activity.

**DISCUSSION**
Pulmonary sequestration is an unusual congenital malformation of nonfunctioning lung tissue that lacks normal tracheobronchial connection, lacks normal arterial supply, and has anomalous venous drainage. Pulmonary sequestration arises from an abnormal pulmonary development along with pathological vascularisation [1,4]. It has two different anatomical forms. The ILS exists when dysplastic tissue is encased within the lung’s normal visceral pleura, and the ELS exists when the tissue mass has its own pleura [5].

Aortic arteriography is considered the diagnostic gold standard for pulmonary sequestration. Systemic arterial supply arise 50 % from the thoracic aorta and 30 % from the abdominal aorta. However, Sauvanet and colleagues reported that the arteriography was diagnostic in only 28 of 40 cases [6]. Magnetic resonance angiography is useful in delineating the vascular anatomy of the sequestration [7]. Except arteriography, pulmonary angiography, magnetic resonance imaging; computed tomographic scanning, bronchography, and ultrasonographies have all been used in selected cases for preoperative diagnosis. In our case, we used the standard chest roentgenograms, a contrast-enhanced chest tomography, a bronchoscopy, thoracic magnetic resonance imaging and angiography.

The arterial supply from the subclavian artery is extremely rare and seldom has been diagnosed preoperatively. In a review of 464 cases of pulmonary sequestration, Savic et al found only 3 (0.6 %) lesions supplied by the subclavian artery, all of which were intralobar pulmonary sequestration [5]. There have been 4 case reports in the literature of extralobar sequestration with subclavian arterial supply [6-9]. Gamillscheg et al reported an extralobar sequestration between the left lower lobe and diaphragm. Angiography showed a large systemic artery arising from the subclavian artery and the venous return paralleled this anomalous artery and drained into the left subclavian vein. Apical extralobar sequestrations with such an unusual arterial supply have been presented by Zumbo et al and Werthammer et al, but the reports did not include details of preoperative imaging. Ito et al reported a case which was a
newborn boy and had extralobar sequestration on right upper part of the lung field. Aortography showed a large anomalous artery arising from the right subclavian artery. In our case which has extralobar pulmonary sequestration the arterial supply was from the subclavian artery. Angiography which performed preoperatively showed that the arterial blood circulation to sequestration were from the left subclavian artery.

Extralobar pulmonary sequestration occurs in the left hemithorax 80 % of cases. It occurs most commonly between the lower lobe and diaphragm, but intradiaphragmatic, pericardial and retroperitoneal locations have been also reported [14]. Although there may be attachment of the mediastinum or diaphragm, upper mediastinal locations are infrequently. In our case, the mass occurred on left upper mediastinum located retrosternum and adjacent to left upper lobe.

The definitive treatment of pulmonary sequestration is resection, especially in symptomatic cases, although limited success has been reported with the use of a simple ligation technique for the feeding artery [16]. We performed sequestrectomy via left sided posterolateral thoracotomy.

In conclusion; it must be keep in mind that pulmonary sequestration which is a congenital foregut anomaly can be seen anywhere in thoracic cavity. Therefore, despite its rarity, a pulmonary sequestration must be considered in the differential diagnosis of upper thoracic mass or cystic lesion.

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