

Pulmonary Sequestration with Aberrant Arterial Supply from Celiac Trunk

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Abstract

Case Report

Pulmonary sequestration is a rare anomaly, which does not have a connection with the bronchial system and gets its blood supply, generally, from the aorta or its branches. Anatomically, two forms were described: extralobar and intralobar which is the commoner type of pulmonary sequestration accounting for 75% of all sequestrations. Although 74% of intralobar pulmonary sequestrations get their blood supply from the descending thoracic aorta, they may get their blood supply from different arteries. Furthermore, there is more than one arterial anomaly in 14.8% of cases. We report an intralobar pulmonary sequestration, in, which arterial blood supply is from celiac trunk, with a cardiomegaly at the expense of the right atrium.

Keywords: Sequestration, Intralobar, Extralobar, Chest radiography, Imaging.

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INTRODUCTION

Pulmonary sequestration is a rare congenital malformation. Arterial blood is generally supplied by the aorta or its branches and does not have a connection with the bronchial system. It accounts for 0.15%–6.4% of all congenital pulmonary anomalies. Anatomically, two different types of pulmonary sequestration were identified; intralobar and extralobar forms. Intralobar pulmonary sequestration shares the same pleura with normal lung tissue, but the extralobar form is separated from normal lung tissue with its own pleura. Intralobar pulmonary sequestration is more common than the extralobar form, and it affects the posterior basal segments of lower lobes and involves the left lung inferior lobe more frequently. Venous drainage in pulmonary sequestration is provided by pulmonary veins, in 95% of the cases. It gets its arterial blood supply from the descending thoracic artery, in 74%, and from the abdominal aorta, in 19%, of cases. It may rarely receive its blood supply from intercostal, subclavian, innominate, internal thoracic, pericardiophrenic, celiac, splenic arteries. Furthermore

in 14, 8% of cases arterial anomaly is more than one [1]. Pulmonary sequestration cases, which receive arterial blood supply from more than one anatomical localization, have been reported in the literature [2, 3].

In this article we illustrate the different forms of sequestration and focus on the role of imaging to identify the anomalous arterial supply.

CASE REPORT

A 2-year-old child presented with respiratory distress and fever. The clinical examination shows a dyspneic infant. Chest computed tomography demonstrated a pulmonary condensation of the lung in the right lower lobe (Fig 1). An aberrant artery was seen arising from the celiac trunk and coursing upward to the right thorax to supply this abnormal segment of the lung. The venous drainage of the segment was through the inferior right pulmonary vein related to an intralobar pulmonary sequestration of the right lower lobe. A right lower lobotomy was performed with an uneventful postoperative course.

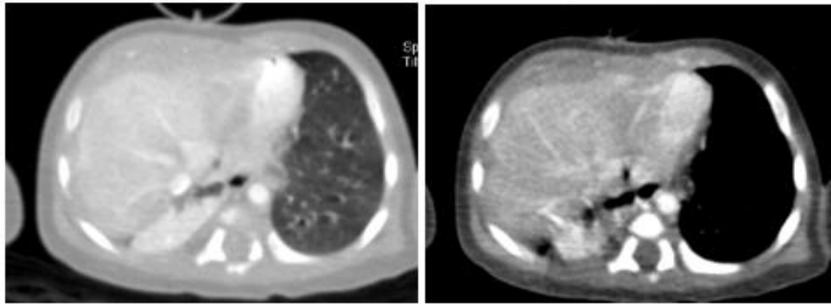


Fig-1: Axial CT scan shows pulmonary condensation of the lung in the right lower lobe



Fig-2: Contrast-enhanced CT showing an aberrant vessel arising from celiac trunk supplying a right lower lobe

DISCUSSION

Bronchopulmonary sequestration (BPS) is a rare congenital malformation of the lower respiratory tract. It consists of a nonfunctioning mass of lung tissue that lacks normal communication with the tracheobronchial tree and receives its arterial blood supply from the systemic circulation, typically from an aberrant artery or arteries that may pass through the inferior pulmonary ligament [4]. It is a rare anomaly representing 0.15% to 6.4% of all pulmonary malformations, reaching 95% of the cases in the left lower lobe[5], and is divided into two types based upon their pleural investment as intralobar or extralobar (Fig. 1). Intralobar sequestration (ILS) is more common (75% of cases) and shares the same visceral pleural covering as the native lung (Fig. 2a) with venous drainage into the pulmonary veins, whereas extralobar sequestration (ELS) (25% of cases) has its own visceral pleural investment outside the normal lung (Fig. 2b) with venous drainage into a systemic vein, and constitutes an accessory lobe, also called a "Rokitanski

lobe". Approximately 10-15% of ELS are found within or below the diaphragm [6].

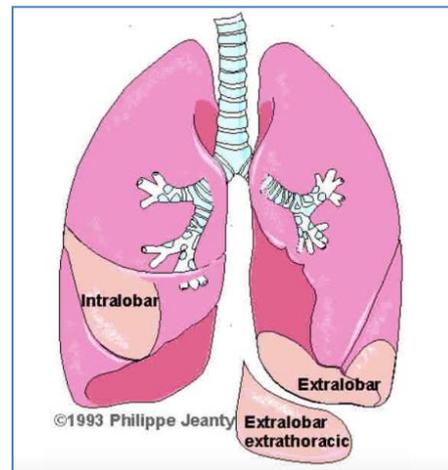


Fig-1: Intra- and extralobar sequestration. The infradiaphragmatic sequestration is not only extralobar (it has its own pleura), but is also located below the diaphragm [10]

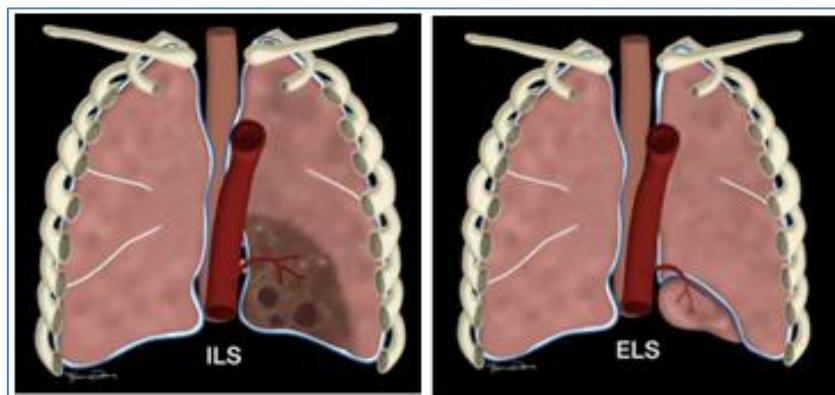


Fig-2: a et b: Graphic illustration of ILS, sharing the same visceral pleural covering as the native lung (a). Graphic illustration of ELS, having its own visceral pleural investment outside the normal lung [4]

The age of presentation depends on the type of sequestration and this in turn determines the clinical presentation. ILS presents late in childhood or adolescence with recurrent pulmonary infection while ELS more commonly presents in newborn with respiratory distress, cyanosis, and infection [7].

Imaging of intralobar sequestration

ILS is a great mimicker with diverse imaging features [4]. The diagnosis may be suggested by US-Doppler in prenatal and new born, by chest radiographic findings alone and should be considered in patients with recurrent pneumonia or localized bronchiectasis, almost always in the posterobasal aspect of a lower lobe.

On chest radiography, there are 3 typical imaging manifestations of ILS: a solitary nodule or mass, a cystic or multicystic lesion, or consolidation.

CT and MRI are both reliable imaging modalities for demonstrating the anomalous artery supplying the ILS. In most cases, a single systemic artery arises from the thoracic aorta (mean diameter 6.3mm), often passing through the ipsilateral inferior pulmonary ligament, to supply the ILS. Occasionally there are multiple systemic arteries supplying the ILS. The supplying artery may also arise from the abdominal aorta, celiac artery, splenic artery, or even a coronary artery [4].

Imaging of extralobar sequestration

The typical appearance of ELS at prenatal ultrasound is a solid, homogeneous, well-circumscribed, hyperechoic mass in the fetal thorax near the medial left costophrenic sulcus. Large lesions are associated with a shift of the mediastinal structures. Cystic areas may be detected within the lesion. Rarely the lesion manifests as a homogeneous or cystic mass in the fetal abdomen or retroperitoneum [8].

Radiographically the lesion most commonly manifests as a well-defined pyramidal, oval, or round mass in the pleural space near the posteromedial aspect of the ipsilateral hemidiaphragm. About two-thirds of ELSs occurs in the left hemithorax. ELS uncommonly occur outside the pleural space in the mediastinum, embedded in the diaphragm, or in the upper abdomen or retroperitoneum. Because of its separate pleural investment from aerated lung, ELS almost never contains air [4].

On CT or MRI, ELS appears as a well-defined mass of uniform soft-tissue attenuation. There is often a single anomalous artery arising from the thoracic or abdominal aorta. The artery may be not being detectable in cases of infarction. In 15% of cases, the lesion is supplied by smaller arterial branches or multiple arteries. In 80% of cases, venous drainage is to

the azygos or hemiazygos vein or to the inferior vena cava [4].

Management of symptomatic or complicated pulmonary sequestration can be surgical isolation and division of anomalous systemic feeding arteries, or endovascular with the embolization of the aberrant arteries with coils or Vascular Plug, for reducing the complications and risks of surgery.

CONCLUSION

Pulmonary sequestration is a mass of lung tissue disconnected from the bronchial tree that receives its blood supply from the systemic circulation. CT and MR are the mainstay for imaging diagnosis of pulmonary sequestration and typically reveal a homogeneous soft tissue mass often having internal cystic areas that show contrast enhancement at the same time as the aorta. Both techniques usually allow to identify the anomalous arterial supply, but are often unable to show the venous drainage.

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