



Case Report

Two Cases of Bronchopulmonary Sequestration in Adult Life: Cross-sectional Imaging with Emphasis on Angio-CT

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INTRODUCTION

Pulmonary sequestration is an uncommon congenital anomaly of the primitive foregut. It is defined as a non-functioning mass composed of dysplastic lung parenchyma, embryologically detached from the tracheobronchial tree and receiving its own blood supply from a systemic artery, usually the thoracic or abdominal aorta. It may be intra or extralobar, depending on the presence of an independent pleural envelope. The venous drainage is variable [1].

Identification of an anomalous blood supply is key to the diagnosis, and its demonstration was traditionally made by intra-arterial DSA. Currently, minimally invasive angiographic techniques such as Computed Tomography (CT) and Magnetic Resonance (MR) angiography are able to demonstrate the aberrant arterial vessel feeding the sequestration.

CASE REPORT 1

A 55-year-old male presented with a 2 month history of right thoracic pain, cough, anorexia and weight loss, but no fever. Physical examination and laboratory data were unremarkable. Chest radiography showed obliteration of the right costophrenic angle and loss of clarity of the adjacent hemidiaphragm. CT disclosed a 6 cm well-circumscribed solid mass, located between the posterior chest wall and the liver, displaying moderate contrast enhancement with a central, non-enhancing, hypodense area (Fig. 1a). The lesion was thought to be non-thoracic. Since its precise location, i.e. intra or infradiaphragmatic, was impossible to ascertain, MRI was performed using a 1.5T magnet. On spin echo sequences (SE), the lesion was seen adjacent to the posterior aspect of the right lobe of the liver, being hypointense on T1-weighted images and moderately hyperintense on T2-weighted images. On both gradient-echo (GRE) FLASH and axial SE T1-weighted images, the hypointense curvilinear structure corresponding to the image of the right diaphragm seemed to “open” to contain the mass (Fig. 1b). A primary diaphragmatic neoplasm was suspected and digital subtraction angiography (DSA) was requested to determine its vascular supply. Selective injection of the right diaphragmatic artery

disclosed a large arterial vessel without malignant features entering the lesion with subsequent venous return by the pulmonary veins towards the left atrium (Fig. 1c). Due to the type of venous return, angiographic findings were thought to be consistent with the diagnosis of intralobar pulmonary sequestration, although contradictory to the location within the diaphragm that more compatible with an extralobar sequestration.

The patient underwent surgery, and a hard mass located within the diaphragm was resected. A 1 cm diameter arterial vessel supplied the lesion and venous drainage via the pulmonary veins to the left atrium was confirmed.

Pathologic examination revealed a mass composed of dysplastic pulmonary tissue with an independent pleural envelope, thus confirming the diagnosis of intra-diaphragmatic pulmonary sequestration of the extralobar type.

CASE REPORT 2

Routine chest radiography performed in an asymptomatic 27-year-old female, disclosed a large well circumscribed opacity located at the lower right pulmonary lobe. CT revealed a 5-cm well defined solid mass surrounded by hyperaeration of the adjacent lung (Fig. 2a). Single-slice helical CT angiography (Hi-speed, General Electric[®], Milwaukee, U.S.A.) was performed after injection of 2 ml per kg/body weight of iodinated contrast material with a concentration of 300 mg I/ml and using an automatic injector at a flow rate of 4 ml/sec. Angiographic images were created using three-dimensional reconstructions of overlapping slices applying maximum intensity projection and volume rendering techniques. The collimation was 3 mm and pitch 1.5, reconstructed with 50% overlap. An aberrant arterial vessel was depicted arising directly from the abdominal aorta feeding the pulmonary lesion (Fig. 2b). Pathologic examination revealed a mass within the resected right lower lobe, composed of dysplastic pulmonary tissue without an independent pleural envelope, thus confirming the diagnosis of intralobar pulmonary sequestration.

DISCUSSION

The anomalous blood supply of a pulmonary sequestration is the clue to its diagnosis. Identification was traditionally made, as in case report 1, by intra-arterial DSA. Currently, minimally invasive angiographic techniques such as CT and MR angiography are also able to demonstrate the

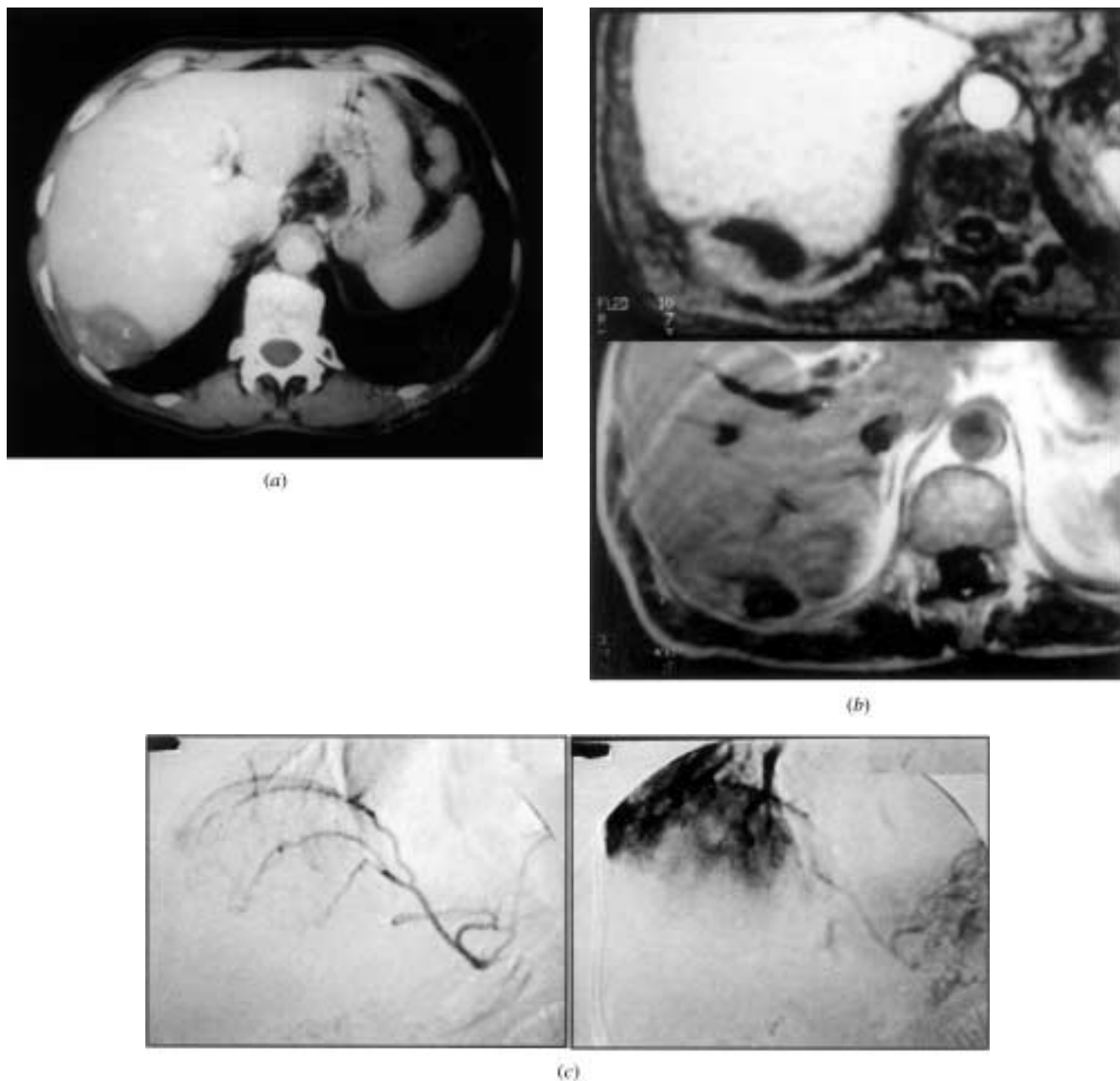


Fig. 1 – (a) CT: 6 cm well-circumscribed solid mass between chest wall and liver, displaying moderate enhancement with a non-enhanced hypodense area. (b) MRI (gradient-echo and spin-echo axial T1 weighted images): Hypointense curvilinear structure corresponding to the right diaphragm, seemed to “open” and contain the mass. (c) Digital Subtraction Angiogram: Mass vessel arising from the diaphragmatic artery; venous return by the pulmonary veins.

aberrant arterial vessel feeding the pulmonary sequestration, as described in the second case. Three-dimensional reconstruction techniques such as maximum-intensity-projection (MIP) or more recently volume rendering (VR) are useful, with the last method having the advantage of displaying simultaneously several ranges of tissue densities. It is now accepted that intra-arterial angiography is no longer mandatory to achieve the diagnosis of these malformations [2] but instead should be reserved for the few cases in which embolization is contemplated [3].

The differential diagnosis of pulmonary sequestrations is extensive, and is modified by the age at presentation. In the newborn other entities should be ruled out, such as a congenital diaphragmatic hernia, intrapulmonary bronchogenic cyst, cystic adenomatoid malformation or bronchial atresia. Later on life, several tumours and tumour-like conditions must be excluded including bronchopulmonary neoplasia, bronchiectasis or arteriovenous fistula [1,4].

Two forms of pulmonary sequestration are typically described: the intralobar and extralobar types. The intralobar



Fig. 2 – (a) CT: 5 cm well circumscribed solid mass, in the right lower lobe, surrounded by abnormally modelled low attenuation lung parenchyma. (b) CT angiography (MIP and VR): Abnormal vessel arising from the abdominal aorta feeding the pulmonary mass at the right inferior lobe.

sequestration, which is the most common (75–86% of all cases), has no gender predominance and is frequently diagnosed during the first decade of life, due to the frequency of respiratory symptoms. Usually located in the left lower lobe, the sequestered portion of lung is contained by the normal visceral pleura [4]. It has its own systemic arterial supply, most commonly arising from the thoracic aorta showing venous drainage by the pulmonary veins towards the left atrium in 95% of cases [1]. A variety of radiologic presentations have been described, including solid homogeneous or heterogeneous lesions with or without cystic components, sometimes with air-fluid levels. Frequently, adjacent parenchymal changes such as inflammatory infiltrates can be seen [4]. The intralobar sequestration is associated in 10% of cases with other congenital abnormalities.

Extralobar sequestration is a much rare-entity and the diagnosis is usually made in early life [5]. Some cases have been reported to be diagnosed during antenatal ultrasonography. The dysplastic lung tissue, covered by its own separate pleural envelope, can be found in various locations, above or below the diaphragm, but more often between the lower left lobe and the diaphragm [4]. The location found in case report 1, and the age of presentation are extremely rare. Its arterial blood supply is similar to the intra-lobar variant with the vessel arising directly from the aorta, with venous drainage to the systemic circulation, via inferior vena cava, vena porta, or azygos system, ultimately reaching the right atrium. Occasionally, as in case report 1, the venous drainage is made directly to the pulmonary veins towards the left atrium [1].

Some authors claim that the demonstration of the different venous drainage (pulmonary or systemic) is the key feature to diagnose extra- versus intra-lobar sequestration and conclude that angiography remains an essential diagnostic tool [6]. However, since the venous drainage of an extra-lobar sequestration can be made by the pulmonary veins towards the left atrium as in our case report 1, the differentiation between the two types can be difficult from imaging studies alone.

In conclusion, we think that the diagnosis of pulmonary sequestration relies on the demonstration of the aberrant systemic arterial supply to a pulmonary or justa-pulmonary mass, which can be achieved by means of non-invasive CT or MR angiographic techniques. However, only pathological examination is capable of differentiating with certainty between the two variants based on the demonstration of independent pleura enveloping the dysplastic lung in the case of extralobar sequestration.

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