Pulmonary Sequestration in an Infant

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Abstract
Pulmonary sequestration is a congenital anomaly of lung parenchyma without a normal connection to the tracheobronchial tree and an anomalous systemic arterial supply. We report an 11-month-old boy with extralobar pulmonary sequestration. Magnetic resonance angiogram identified the arterial supply from aorta and venous drainage to azygous vein. Various imaging techniques for diagnosis of pulmonary sequestration are discussed.

Key words Lung; Magnetic resonance angiogram; Magnetic resonance imaging; Sequestration

Introduction
Pulmonary sequestration refers to region of lung parenchyma which lacks a normal connection to the tracheobronchial tree and possesses an anomalous systemic blood supply, usually from the aorta or its major branches.\(^1\) It was estimated that 6.4% of all congenital pulmonary anomalies were pulmonary sequestration.\(^2\) Pulmonary sequestration is classified into intralobar and extralobar according to the absence or presence of investing pleura. We report a case of extralobar pulmonary sequestration in an infant.

Case Report
An 11-month-old boy was admitted to the Department of Paediatrics of Kwong Wah Hospital in May 1998. He initially presented with high fever and cough. Chest radiograph showed a left lower zone consolidation (Figure 1). Left lower lobe pneumonia was diagnosed. His symptoms subsided after treatment with cefuroxime but repeated chest radiographs showed a persistent left lower zone consolidation. Laboratory tests showed normal leukocyte count and Mantoux test was negative. Computer tomography (CT) scan of thorax showed a heterogeneous mass at posterior basal segment of left lower lobe after contrast enhancement (Figure 2). Flexible bronchoscopy was performed to exclude foreign body. Ultrasound demonstrated a solid left lower lobe lesion containing vascular structure likely representing sequestration but its systemic supply was inconclusively demonstrated by Doppler study. Magnetic resonance imaging (MRI) confirmed an abnormal triangular area in basal region of left hemithorax. On magnetic resonance angiogram (MRA), it was supplied by aorta and drained into azygous vein, consistent with extralobar sequestration (Figures 3a & 3b). The patient was then transferred to Queen Mary Hospital for left lower lobe lobectomy. The operation was uneventful. Intra-operatively, the left lower lobe sequestration was found to be attached to the left posterior...
Figure 1  Chest radiograph showing left lower lobe consolidation (arrow).

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

Figure 3a  Contrast enhanced magnetic resonance angiogram in arterial phase showing the systemic arterial supply (arrow) to the sequestration from the descending aorta.

Figure 3b  Contrast enhanced magnetic resonance angiogram in venous phase showing the venous drainage (short arrow) of the sequestration, crossing behind the aorta (arrow head) to reach the azygous vein (long arrow) on the right side.
pleura by the vascular pedicle only (Figure 4). It had an independent artery and vein but no bronchus was found in the pedicle attached to the left posterior pleura. The excised specimen showed a piece of pyramid shaped lung tissue completely invested by pleural lining, measured 6.5 x 4 x 2 cm. Section of the lung tissue showed areas of consolidation, fibrosis and bronchial dilatation (Figure 5). Histological examination showed lung tissue with many of the alveoli and bronchial lumina filled with eosinophilic proteinaceous fluid. Post-operatively, he made an uneventful recovery.

Discussion

Pulmonary sequestration or accessory lung is a congenital abnormality in which a portion of the lung shows separation from the normal bronchial tree and blood supply. The term "sequestration" was coined by Pryee in 1946 to describe a disconnected bronchopulmonary mass or cyst with an anomalous pulmonary artery. The estimated incidence was 0.15% to 1.7% in general population. There are two main groups. Extralobar sequestrations (ELS) are enclosed in their own pleura, typically receive arterial flow from small branches of the thoracic or abdominal aorta and drain into azygous or hemiazygous system. They are more common in left posterior costophrenic sulcus. Intralobar sequestrations (ILS) are enclosed in the pleura of the adjacent normal lobe. Their arterial supply is usually aortic branches and drainage into pulmonary veins. About 65% of ELS have associated anomalies ranging from innocuous abnormalities like accessory spleen to complex heart disease, with diaphragmatic hernia being the most common abnormality. The most common presentation of pulmonary sequestration is recurrent pneumonia (54%). Pneumonia occurs if fistula forms between the sequestration and airway/digestive tract or direct invasion from pneumonia in the surrounding lung. Other presentations include heart failure, haemoptysis and respiratory distress.

The plain X-ray often shows a triangular or oval-shaped, basal, posterior lung mass, more on the left with fairly well defined margin. Other locations may be possible such as upper half of the thorax in 2% and extralobular sequestration may sometimes be in pericardial space, mediastium and retroperitoneum. In Laurin and Hagerstrand's series, all of their 7 intralobar sequestrations had plain chest X-ray abnormalities identified before any clinical symptoms developed. Hence, reference to previous chest radiographs taken for some other reasons might show the pulmonary abnormality that suggested sequestration. The abnormality

![Figure 4](image_url) Extra-lobar sequestration (solid arrow) with vascular pedicle (arrow head) attached to the left posterior pleura (open arrow).

![Figure 5](image_url) Section of sequestration showed bronchial dilatation (arrow), fibrosis and consolidation.
is usually solid opacity on plain film but occasionally it may be air filled with or without air fluid level. This may be attributed to fistula formation to adjacent bronchi or air drift through the channels of Lambert.

Traditionally, the diagnosis of pulmonary sequestration is confirmed with angiography demonstrating the anomalous systemic arterial supply. Nowadays, other imaging modalities now allow non-invasive demonstration of anomalous artery in the majority of cases, including spiral CT, MRA and colour duplex sonography. Computed tomography usually shows irregular cystic component in the lesion and there is high incidence of emphysema in the adjacent lung due to collateral air drift and air trapping. Ultrasound may show the lesion to be of mixed echogenicity with predominately hyperechogenicity. The most specific pre-operative diagnosis rests on the identification of a systemic arterial supply. Although this can be done precisely with a catheter angiogram, it is invasive and not without complications. Smart and Hendry found systemic supplying artery by duplex Doppler scanning in 4 out of 7 patients of their series. However, there was much false negative in the series and ultrasound examination is operator dependant. Magnetic resonance imaging is able to identify the vascular supply as flow void in cross section. It may also show the supply with conventional magnetic resonance angiography using mainly the time-of-flight technique. There are many shortcomings of this technique that depends very much on the positioning of the acquisition slab with the direction of flow. The sequence is in term of minutes and this long time may not be applicable or reliable in children. With modern equipment, the contrast-enhanced magnetic resonance imaging overcomes much of the problem. The vascular signal offered by the Gadolinium coupled with modern hardware and software allows a sequence to be acquired in term of 2 to 20 seconds alleviating many problems of motion. It also allows the possibility of time resolution simulating a catheter angiogram in MR machine with high gradient. This may help to distinguish whether the venous drainage of the sequestration is systemic or pulmonary. However, one must be aware of the fact that systemic arterial supply to pulmonary parenchyma may also be seen in arteriovenous fistula, pulmonary artery aplasia, chronic pulmonary infection or post-operative lung. In appropriate clinical setting coupled with the non-invasive magnetic resonance image finding of systemic arterial supply to a lung segment, accurate pre-operative diagnosis of sequestration can be made. Angiography is now reserved for those cases in which non-invasive imaging methods have failed to confirm the diagnosis.

References