PULMONARY SEQUESTRATION

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A previously healthy 25-year-old non-smoker was admitted to hospital with a three-month history of malaise, sweating and cough productive of clear sputum. He had no relevant past history or obvious risk factors for immunodeficiency.

On examination, he had a pyrexia of 38.5 °C, a sinus tachycardia of 100/min.with a normal blood pressure and heart sounds. Auscultation of the chest revealed crackles at the left base.

Relevant investigations included a neutrophil leucocytosis of 15.7 x 10 9/l, ESR 24, pO_2 7.9 mmHg with normal pCO_2.

The chest X-rays taken on admission are shown as Figures 1a and 1b. There is a retrocardiac shadow in keeping with an area of left lower lobe consolidation. Sputum cultures did not yield any growth; pneumococcal antigen and screen for atypical pneumonia were negative. He was treated with a broad spectrum antibiotic for a presumed bronchopneumonia with symptomatic improvement.

At review, six weeks and three months later, he complained of a persistent non-productive cough. The chest X-ray still showed a retrocardiac area of consolidation. CT scan of the chest revealed a left postero-basal area of consolidation (Figure 2). In view of his recent medical history pulmonary sequestration was suggested as the diagnosis. An aortogram revealed a tortuous solitary aberrant artery supplying the sequestered segment (Figure 3).

The patient was referred to the thoracic surgeons who successfully performed a video-assisted thoracoscopic left lower lobectomy. He made a rapid and uneventful complete recovery.

DISCUSSION

Pulmonary sequestration is one of the rarer congenital anomalies of the adult lung.

Although initially described by Price in 1946[1] as a segment of lung parenchyma separated from the tracheobronchial tree and receiving its blood supply from a systemic artery rather than a pulmonary arterial branch, it is now accepted that there is a spectrum of pulmonary sequestration. This ranges from normal vessels supplying abnormal lung such as in lobar emphysema and cystic adenomatoid malformation, to the more classic form of abnormal vessels supplying normal lung.

There are two types of the classic form of pulmonary sequestration:

a) intralobar sequestration (ILS) – the abnormal segment of lung lies within the normal pulmonary visceral pleura. (Approximately 75%).
b) extralobar sequestration (ELS) – the abnormal segment of the lung is completely separate and enclosed in its own pleural envelope (25%).

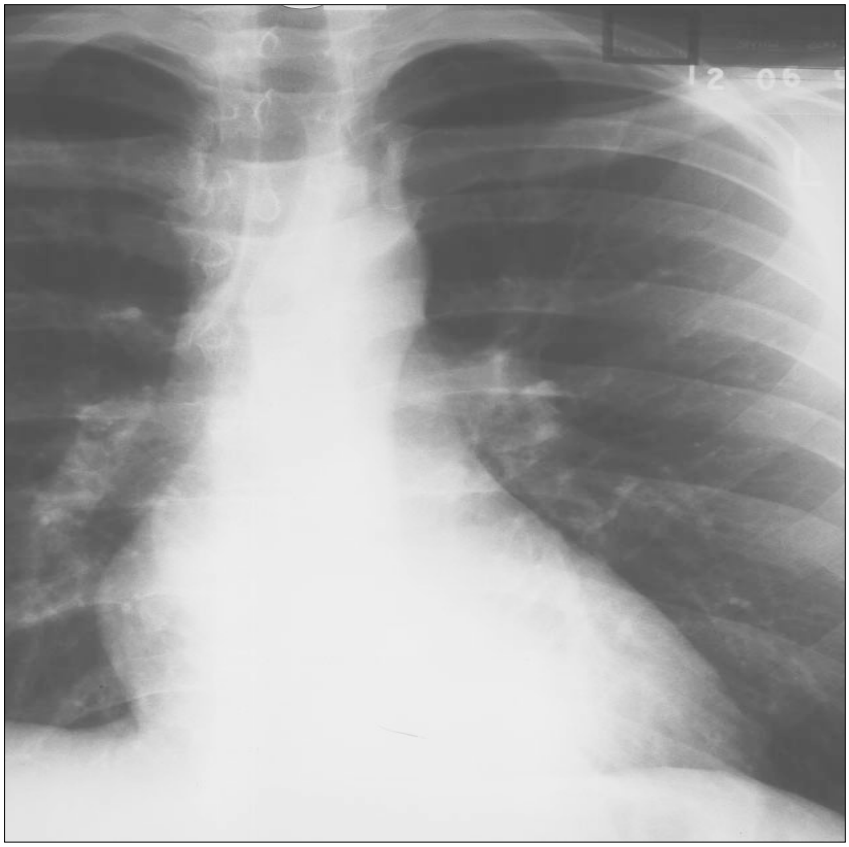
Several theories have been put forward to explain the development of pulmonary sequestration, ranging from the 'accessory bud' theory[2] which proposes that an accessory lung bud develops in the embryo which either becomes incorporated into the normally developing lung (intralobar sequestration) or remains separate (extralobar sequestration), through to the 'acquired theory' which states that the sequestration arises secondary to a localised infectious disease with prominence of normally occurring pulmonary ligament arteries.

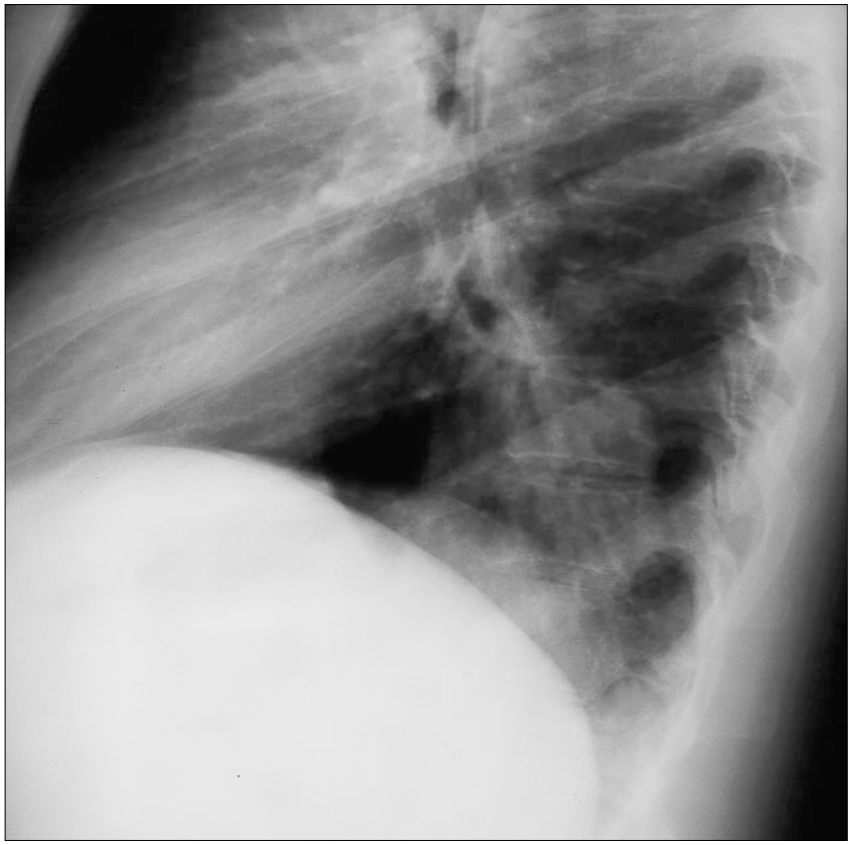
TABLE 1
Diffentiation of extralobar and interlobar pulmonary sequestration.[3]

Parameter	Intralobar sequestration	Extralobar sequestration
Relation to normal lung	Within normal lung and its pleura	Separate, own pleural cover
Venous drainage	Pulmonary	Systemic
Laterality	Left 60%-70%	Left 90%
Assoc. congenital anomalies	Uncommon	Frequent
Age at diagnosis	50% 20 years old	60% 1 year old
Sex ratio	M=F	M:F = 4:1
Infection	Common	Rare

Intralobar Sequestration (ILS) most often occurs in the lower lobes (98%), usually on the left side with all reported cases being above the diaphragm. Patients present with cough, sputum production and recurrent pneumonia with over 50% being symptomatic by the age of 20 years. In 73% of cases the arterial supply is from the descending thoracic aorta; less commonly the arterial supply is from the upper abdominal aorta, coeliac or splenic arteries. Multiple supplying arteries are found in about 16% of cases. Venous drainage is to the left atrium via the pulmonary veins in 95% of cases, with the remainder draining to the systemic circulation via the azygos, hemiazygos, intercostal veins or through the inferior vena cava. In general the anomalous pulmonary vein follows the artery, so if the aberrant pulmonary artery arises above the diaphragm, the vein also drains above the diaphragm.[4]

Extralobar Sequestration (ELS) often (60%) presents in the first six months of life with dyspnoea, cyanosis and feeding difficulties. Only 10% are asymptomatic. They are most often found between the left lower lobe and the diaphragm, but can be found in various other locations such as within the musculature of the diaphragm, in the pleural or pericardial spaces or in the retroperitoneum. 80% of





FIGURES 1A AND 1B
PA and left lateral chest films showing an ill-defined mass behind the heart on the PA and at the level of the diaphragm posteriorly on the lateral.

cases receive arterial supply from the thoracic or abdominal aorta, with 15% receiving their supply from smaller vessels such as splenic, gastric and subclavian arteries. The venous return is in the majority of instances to the systemic circulation via the azygos, hemiazygos and inferior vena cava. In 65% there are associated extrapulmonary anomalies, the most common being a diaphragmatic hernia.[4]

Surgery is the definitive treatment of ILS especially when infection intervenes. The role of imaging is in confirming the suspected diagnosis and displaying the aberrant arterial supply. Pleural involvement and venous drainage are adequately determined intraoperatively. As sequestration is a rare disorder, no comparative study of imaging techniques has been carried out to determine the optimum imaging modality to assess the disorder. Sequential chest X-rays will reveal a persistent, most often left, basal lung opacity. CT, MR imaging and digital subtraction angiography are all performed to display the diagnostic feature of the anomalous artery. In the young child sonography is often useful. Contrast enhanced helical CT with reconstruction not only displays the aberrant vessel but will also reveal any associated lung abnormalities such as localised emphysema or calcification. Breath-holding MR angiography is now replacing conventional contrast angiography to display the aberrant vessel. It is multiplanar and avoids the use of radiation. With the use of Gadolinium-enhanced sequences, there are several reported cases where MR angiography revealed additional aberrant arteries not displayed by conventional digital subtraction angiography.[5] However its role in the sequence of imaging depends on its local availability. It does not give detail of the underlying lung parenchyma. Digital subtraction angiography is still used widely in the pre-operative evaluation.

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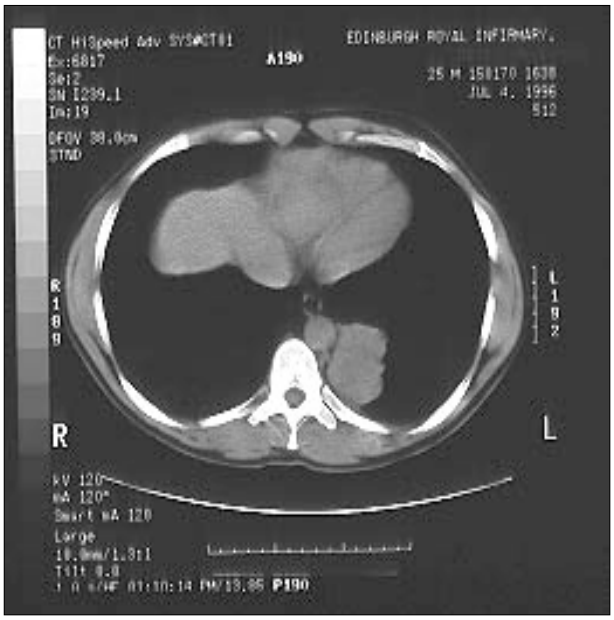

FIGURE 2
Axial CT scan at the level of the apex of the right hemi-diaphragm and lower heart. The abnormal area in the left lung is shown posteromedially adjacent to the aorta and vertebral bodies.

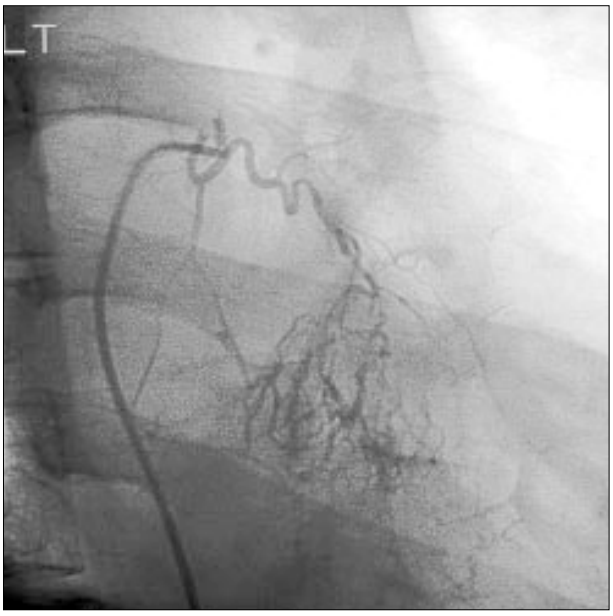

FIGURE 3
An angiogram showing selective catheterization of an aberrant artery supplying the abnormal segment in the lower left lobe.