An 18-year-old man presented with a history of recurrent chest infections over the past 4 years. The patient had been treated intermittently with several courses of antibiotics. Apart from his chest infections, he had no significant medical or surgical history. On examination, the only significant finding was an area of decreased air entry in the right lower lobe. Laboratory data revealed no abnormality. A chest radiograph and, subsequently, a CT were requested.

The chest radiograph showed an area of ill-defined consolidation involving the medial segment of the right lower lobe (Fig 1). The remainder of the lung appeared normal. The radiograph was otherwise unremarkable. A repeat radiograph after a 2-month period, during which time the patient received a further course of antibiotics, revealed no significant change. A CT scan of the chest was obtained.

What is the diagnosis?

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**FIGURE 1.** Posteroanterior (top, A) and lateral (bottom, B) chest radiographs demonstrate an area of ill-defined consolidation involving the medial segment of the right lower lobe. The adjacent lung appears normal.
Diagnosis: Pulmonary sequestration

A spiral CT scan of the chest obtained during the administration of IV contrast revealed an inhomogeneously enhancing mass involving the medial aspect of the right lower lobe. Focal areas of low density in keeping with necrotic regions were seen within the mass. No air bronchogram or area of cavitation was identified (Fig 2). A close assessment of the descending aorta, confirmed by a three-dimensional surface shaded display, showed an anomalous artery arising off the anterior aspect of the distal descending aorta, running down in front of the aorta for 3 cm, and eventually swinging laterally to supply the mass described above (Fig 3). This systemic supply confirms the diagnosis of pulmonary sequestration.

Pulmonary sequestration is a congenital abnormality in which a portion of the lung shows separation from the normal bronchial tree and blood supply, though retaining some characteristics of lung tissue. The term “sequestration” was coined by Pryce in 1946 to describe a disconnected bronchopulmonary mass or cyst with an anomalous pulmonary artery. Cases can be divided into two main groups: extralobar sequestration (ELS), when the sequestrated segment develops enclosed in its own pleura; and intralobar sequestration (ILS), when it is lying within the lung and therefore without its own pleura. ELS is the least common type, accounting for approximately 25% of all cases. With ELS, the arterial supply is frequently from the abdominal aorta or one of its branches. Venous drainage is usually via the inferior vena cava, azygos, or portal venous systems. ELS tends to present at an earlier stage, with a majority of cases (61%) presenting in the first 6 months of life with dyspnea, cyanosis, and feeding difficulties. Most cases of ELS occur on the left side. A majority of ELS cases (65%) have associated anomalies ranging from innocuous abnormalities (such as accessory spleen) to complex heart disease, with diaphragmatic hernia being the most common abnormality.

ILS accounts for approximately 75% of all cases. Recurrent chest infection is the most common presenting complaint, with > 50% of ILS becoming symptomatic by the age of 20. The vast majority of cases (98%) occur in the lower lobes, with the left side again being more commonly involved. Typically, the segment is not connected with the normal bronchial tree; if communication is established, it is usually associated with infection. With ILS, the...
arterial supply is usually from the descending thoracic aorta but may originate from the abdominal aorta. Venous drainage is generally via the pulmonary veins, occasionally via the inferior vena cava or azygos system.

The role of imaging is not only to detect the sequestered or dysplastic lung itself, but also to identify the aberrant arterial supply. Along with an appropriate history, plain radiographs showing a persistent area of consolidation that does not improve following antibiotic therapy will suggest the diagnosis. Occasionally, the sequestration may cavitate or appear cystic. Classically, there is no communication of the sequestered segment to the normal bronchial tree. Traditionally, the diagnosis of pulmonary sequestration was confirmed with angiography demonstrating the anomalous arterial supply. However, other imaging modalities now allow noninvasive demonstration of the anomalous artery in the majority of cases. Spiral CT obtained during the administration of IV contrast has the ability to visualize not only the sequestered segment but also the aberrant vascular supply in the majority of cases in a noninvasive manner, making it the imaging investigation of choice. Magnetic resonance angiography (MRA) has also been utilized in the demonstration of the aberrant vasculature of the sequestered segment. MRA also allows multiplanar reconstruction with the added advantage of not requiring ionizing radiation. Color duplex sonography has occasionally allowed the diagnosis to be made not only in children and adults but also in utero. Angiography is now reserved for those cases in which noninvasive imaging methods have failed to confirm the diagnosis.

REFERENCES