A 53-Year-Old Man With Hemoptysis*

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A 53-year-old black man presented with recurrent episodes of hemoptysis, with 1.5 cups of bright red blood expectorated over a 2-day period. He denied having fever, chest pain, vomiting, night sweats, shortness of breath, or dizziness. He had had a dry cough for several months before admission to the hospital. He also had lost 40 lb from dieting in the past several years. He was being treated for hypertension and gastroesophageal reflux disease. He had a 30-year history of tobacco use, occasional alcohol use, and no IV drug abuse. Clinical examination revealed a pleasant black man not in acute distress. Vital signs were stable: pulse rate, 79 beats/min; BP, 154/92 mm Hg; and respiratory rate, 21 breaths/min. There was no clubbing or lymphadenopathy. Chest examination revealed scattered crackles at both lung bases. The rest of the physical examination was unremarkable. Laboratory investigation showed a hemoglobin level of 14.2 g/dL, a WBC count of 6.0 × 10^3/μL, and a platelet count of 277 × 10^3/μL. Prothrombin time was 24.2 s, and international normalized ratio was 0.88. Fiberoptic bronchoscopy revealed active oozing of blood from the left lower lobe bronchus with mildly edematous airways.

A chest radiograph (Fig 1) obtained at the time of admission demonstrated a vague airspace opacity in the left lower lobe. Contrast-enhanced CT of the chest was performed. A dilated left lower lobe pulmonary vein was seen draining into the left atrium (Fig 2, top). An enhancing round vascular structure was also seen anterolateral to the descending thoracic aorta (Fig 2, top). Ill-defined airspace opacities in the posterior basal segment of left lower lobe (Fig 2, bottom) were thought to represent focal pulmonary hemorrhage. To evaluate the vascular anomaly, a pulmonary angiogram was obtained, which demonstrated the absence of pulmonary artery to the posterior basal segment of the left lower lobe (Fig 3). There was no evidence of thrombosis, arteriovenous malformation, or pulmonary varix. The patient experienced three more episodes of significant hemoptysis while he was in the hospital. He underwent urgent left thoracotomy.

What is the diagnosis?

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Figure 1. Chest radiograph shows an ill-defined airspace opacity (*) in the retrocardiac region of the left lower lobe.
Figure 2. Top: Contrast-enhanced CT (soft-tissue window) at the level of lower lobe demonstrates an ovoid, enhancing, dilated left inferior pulmonary vein (arrow). Another round, enhancing structure next to the descending aorta was suspected to be an aberrant systemic artery supplying the left lower lobe. Bottom: A CT image (lung window) 1 cm below that image, demonstrates ill-defined airspace opacity and increased vascularity in the posterior basal segment of the left lower lobe.

Figure 3. Selective left pulmonary arteriogram in the right anterior oblique position demonstrates absence of arterial supply to the posterior basal segment of left lower lobe (asterisk). All the visualized pulmonary arterial branches are normal in appearance. There was no evidence of pulmonary thrombosis (acute or chronic), or arteriovenous malformation.
Diagnosis: Aberrant systemic artery supplying the left lower lobe without sequestration (Pryce’s type 1 anomaly)

CT demonstrated an increased number and size of pulmonary vessels in the left lower lobe compared to the right side (Fig 2, bottom). A vascular anomaly, either a complex arteriovenous malformation or abnormal systemic artery supplying the left lower lobe, was suspected. An arteriovenous malformation was considered less likely because only a dilated pulmonary vein is seen, without an accompanying large pulmonary artery.

Based on the CT and pulmonary angiographic findings, the presumptive diagnosis was sequestration of the lung. Since an abnormal arterial supply from the thoracic aorta was identified on CT, an aortogram was not obtained.

At surgery, the left lower lobe was discolored. The superior and inferior pulmonary veins were normal. A large, 2- to 3-cm, hard, calcified, aberrant artery was seen arising from the distal descending thoracic aorta supplying the posterior segment of the left lower lobe. There was a normal pleural lining of the lung. Left lower lobeectomy was performed. Microscopic examination of the aberrant systemic artery demonstrated an elastic artery with extensive calcific atherosclerosis and fibrosis. In the excised lobe, there was extensive intimal thickening and fibrosis of the pulmonary arteries and arterioles consistent with pulmonary hypertension. The lung parenchyma showed accentuation of septae, focal anthracosis, and massive recent as well as old intra-alveolar hemorrhage. Multiple resected hilar nodes showed sinus histiocytosis and anthracosis. No cancerous tissue was seen in the lung parenchyma or the resected nodes.

Discussion

The term sequestration was coined by Pryce1 in 1946 to describe a disconnected bronchopulmonary segment or cyst with an abnormal systemic artery supplying the involved lung. There are many variants of sequestration that do not strictly meet these criteria. Sade et al2 in 1974 suggested the term spectrum of pulmonary sequestration to include the diverse group of pulmonary abnormalities that appear to be related and may have a common embryogenesis. This spectrum includes normal vessels supplying abnormal lung at one end and abnormal vessels supplying normal lung at the other.3

Pulmonary sequestration is an uncommon congenital malformation. Two types are commonly described: intralobar sequestration, which describes the sequestered lung contained within the normal visceral pleura; and extralobar sequestration, in which the abnormal lung is completely separate and enclosed in its own pleural sac. The majority (75%) of cases are of the intralobar sequestration variety. Pryce, in his original article, described three types of intralobar abnormalities based on the distribution of the aberrant artery. In type 1 (our patient), the anomalous artery supplies functional normal lung tissue, which communicated with the tracheobronchial tree. In type 2, the systemic artery supplies both normal lung as well as nonfunctional, noncommunicating lung tissue. In type 3, the anomalous artery supplies lung that is isolated from the tracheobronchial tree.4

More than 50% of patients with intralobar sequestration become symptomatic after the age of 20 years.4 The common symptoms include cough, sputum production, and recurrent pulmonary infections. Hemoptysis is also a common presenting sign. Chest pain, asthma, and pleuritic pain are less common presentations. A small number (15%) of patients may be asymptomatic when the lesion is discovered.4 The less common extralobar sequestration frequently presents in the neonates and early childhood with respiratory distress, cyanosis, and feeding problems. A majority of patients with extralobar sequestration have other congenital abnormalities, including esophagobronchial diverticulum, diaphragmatic hernia, skeletal deformities, cardiovascular defects, and renal anomalies.5,6

Systemic arterial supply to normal lung is a rare but well-recognized variant of the sequestration spectrum.7 The aberrant systemic artery usually arises from the descending thoracic aorta. The basal segments of left lower lobe are more frequently involved, with no radiographic abnormality in the lung parenchyma. Drainage of the involved lung is usually accomplished via the inferior pulmonary vein into the left atrium. Although the etiology of systemic arterial supply to the normal lung is unknown, the most likely explanation is that one or more of the intersegmental arteries from the dorsal aorta retain their original embryonic connection between the aorta and the pulmonary parenchyma.8 These patients may be asymptomatic. However, left ventricular enlargement and congestive heart failure from left-to-right shunt may eventually develop. Hemoptysis, as in our patient, is a rare but recognized presenting feature.4,8 It results from rupture of the abnormal pulmonary vessels in the affected lung, caused by the pulmonary hypertension, resulting from transmission of systemic pressure to the affected lung. Focal pulmonary arterial hypertension occurs since the pulmonary arterial branches of the sequestered lung do not communicate with the rest of the pulmonary vasculature. Cardiac catheterization often does not reveal pulmonary hypertension.

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unless longstanding left ventricular failure occurs from large left-to-right shunt. The term pseudosequestration has been used to describe the combination of systemic arterial supply to lung with normal bronchial connections but with coexistent chronic/recurrent pulmonary infection. In these cases, the chronically inflamed lung and pleura recruit the normally occurring systemic arteries supplying the lung and chest wall (bronchial, intercostal, diaphragmatic, and pulmonary ligament arteries), which result in a systemic arterial blood supply to the inflamed lung tissue. However, these cases are distinct from the anomaly in our patient, in which one or more discrete abnormal anterior branches from descending aorta supply the affected lung. In the former condition (pseudosequestration), the systemic arterial supply is from hypervascularization of normal anterior branches of the chest wall (eg, intercostal and internal mammary arteries).

A chest radiograph may demonstrate a homogeneous opacity in the lung base in uncomplicated intralobar sequestration. Complicated sequestration may present as a cystic lesion that may contain air-fluid levels. Traditionally, diagnosis of pulmonary sequestration is made with aortography. The classic findings are the anomalous systemic arterial supply and anomalous venous drainage, depending on the type of sequestration. The pulmonary arteriogram may be useful in demonstrating the absence of pulmonary blood supply to an area of lung, as in our case. CT is an useful imaging study, especially for evaluation of a patient with hemoptysis. It can show lung parenchymal changes and the aberrant vessels supplying the sequestration. The anomalous systemic artery is visualized in up to 80% of cases after contrast administration and may be seen in cross-section (as in our case) or as an enhancing linear structure adjacent to the aorta in the inferior pulmonary ligament. Use of dynamic scanning technique, thin-section CT, and helical CT substantially improves the delineation of the origin and course of the anomalous systemic artery. MRI can demonstrate the anomalous vessels and the parenchymal abnormalities in multiple planes. Sonography has been used in the diagnostic evaluation of pulmonary sequestration both in utero and in infants.

Preoperative diagnosis of sequestration is helpful by forewarning the surgeon of the anomalous vascularization of the lung, preventing fatal outcome due to exsanguination from accidental division of the systemic artery.

References
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