Bronchopulmonary sequestration is an uncommon pulmonary disorder that affects both sexes equally and has been described in all age groups. It is defined as an area of nonfunctioning abnormal lung tissue which receives its blood supply from a systemic artery and characteristically has no connection with the tracheobronchial tree. About 70 percent of all the reported cases are of the intralobar variety. The abnormal lung tissue in the intralobar type is located within the visceral pleura of a pulmonary lobe, whereas the extralobar form is like an accessory lobe because it has its own visceral pleura. The venous drainage of the extralobar type is usually into the systemic venous system, whereas the intralobar type drains into the pulmonary venous system. All reported intralobar sequestrations are located above the diaphragm and are almost always in the lower lung zones (98 percent). Most of the reported extralobar sequestrations are in the lower lung zones as well (77 percent) with a predilection for the left hemithorax (83 percent).

The etiology of the extralobar form is widely accepted to be congenital. The primary defect is believed to be insufficient pulmonary arterial vascularization during fetal lung development. The consequence is persistent systemic arterial vascularization after birth, which leads to cystic degeneration and fibrosis of the sequestered lung tissue. The etiology of the intralobar type remains controversial. The most popular belief is that it is also a congenital defect; however, it is intriguing that intralobar sequestration is rarely found in neonatal autopsies. Recent studies by Stocker et al suggest that some, if not all, of the intralobar sequestrations may be acquired after birth. The proposed mechanism is that bronchial obstruction in early childhood leads to secondary vasoconstriction in the affected region. This is followed by compensatory systemic arterial vascularization by one or more of the pulmonary ligament arteries arising from the descending aorta. Eventually, cystic degeneration and fibrosis develop in the sequestered lung.

CASE REPORT

A 30-year-old Chinese man presented in January 1984 with a one-week history of fever, chills and nonproductive cough. His past history included exposure to tuberculosis in 1981 and his tuberculin skin test was positive at that time. A chest roentgenogram apparently showed a "scar" in his right lung in 1983 (Fig 1), but he never received isoniazid prophylaxis. Physical examination (in 1984) was unremarkable, but his x-ray film on this occasion showed a right lung infiltrate. A ten-day course of ampicillin therapy improved his pulmonary condition.

FIGURE 1. This PA chest film was interpreted as normal except for a small scar in the right lung base. The arrow points to the systemic feeding artery.
symptoms; however, he suffered relapse one week later with cough, hemoptysis, night sweats and dyspnea on exertion. A repeat chest film showed extension of the right lung infiltrate (Fig 2). Bronchoscopic examination revealed inflammation and blood at the orifice of the superior segment of the right lower lobe. Sputum, bronchial washings and brushings, and percutaneous transthoracic needle aspiration were all negative for microbiology and cytology.

Due to the lack of a specific diagnosis and the unusually slow resolution of the pulmonary infiltrate, a chest CT scan was performed in search for “unusual anatomy” or localized bronchiectasis. The chest CT scan showed multiple cystic cavities in the right lower lobe region (Fig 3). A vessel-like structure that appeared to arise from the descending aorta was seen on a lower cut (Fig 4). In retrospect, the feeding artery was present on the 1983 film (Fig 1). The chest CT findings are consistent with bronchopulmonary sequestration. A bronchographic study performed two months after resolution of the infiltrate showed anterior displacement of the lower lobe bronchi. The full complement of right middle and lower lobe segmental bronchi were present, but the lower lobe bronchi were displaced anteromedially. Numerous attempts failed to demonstrate any bronchial connection with the cystic-cavitary structures seen on the chest CT scan. An aortogram demonstrated a systemic feeding artery arising from the descending aorta (Fig 5). The venous phase showed a vein draining into the pulmonary vein (Fig 6) which is the angiographic hallmark of an intralobar sequestration. The diagnosis of intralobar bronchopulmonary sequestration was confirmed at thoracotomy.

**DISCUSSION**

A variety of plain film findings may be encountered in pulmonary sequestration. The spectrum ranges from homogeneous opacities to multiple cystic areas. Less specific findings include recurrent pneumonias and focal bronchiectatic changes. In fact, any persistent abnormality in the posteromedial basal segment of a young adult should suggest the diagnosis. Extremely subtle findings may be present in uninfected intralobar pulmonary sequestration with only the faint shadow of the aberrant artery being detectable. Rarely, sequestrations may be calcified. Tomography may accentuate the plain film findings of the abnormal artery and may demonstrate bronchiectatic changes or bronchi draped around the abnormal area. In children with extralobar pulmonary sequestration, barium studies may demonstrate communication between the gastrointestinal tract and the pulmonary sequestration.

Historically, bronchography has been important in suggesting the diagnosis. Most cases show normal bronchi draped around, but not communicating with, the involved segment of lung, although rarely contrast media may enter the abnormal bronchial tree of the
pulmonary sequestration.\textsuperscript{10}

Definitive diagnosis has generally been made by angiographic demonstration of an abnormal artery arising from a systemic artery and supplying the lung. Most of these arteries arise from the thoracic or abdominal aorta. In rare cases the abnormal vessel may arise from the celiac, innominate, subclavian or bronchial arteries.\textsuperscript{1} Angiography also shows the venous drainage, allowing classification into intralobar (drainage to pulmonary veins) and extralobar (drainage to systemic veins) pulmonary sequestration. Rare variants of arterial and venous drainage can be identified helping the surgeon to avoid operative complications. It must be remembered that aberrant systemic arteries to the lung can also be seen in chronic lung infections, arteriovenous fistulae, and rarely to normal lung.

With high quality CT scans the anomalous artery can sometimes be demonstrated.\textsuperscript{16} Contrast enhancement can confirm that the linear structure in the lung base is a blood vessel and that it arises from the aorta. Dynamic scanning and time density curves can confirm that the arterial filling and enhancement occur during the systemic opacification phase.\textsuperscript{11,12} CT scan also defines the exact location and extent of the abnormality, particularly in unusual circumstances, such as retroperitoneal pulmonary sequestration.\textsuperscript{13} Associated abnormalities, which occur in 15-40 percent of extralobar pulmonary sequestration, may also be detected.\textsuperscript{14} If surgery is not being considered, CT scanning may obviate the need for angiography. Failure to demonstrate the ectopic artery on CT scans does not exclude the diagnosis.

Long-term followup of asymptomatic patients has been reported. However, once infected, patients are at high risk of abscess formation in the sequestered lung due to poor bronchial drainage. The available literature seems to support the following pragmatic approach: (1) observe the asymptomatic cases, (2) identify the offending micro-organism(s) in the infected cases and treat with appropriate antibiotic(s), and (3) follow with surgical resection (ideally when the infection is quiescent).

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