A 66-Year-Old Man With Dyspnea, Left Lower Lobe Infiltrate, and Abnormal Imaging*

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A 66-year-old nonsmoking man was referred to the pulmonary clinic for workup after abnormal findings on chest radiograph were discovered during his evaluation for cardiac disease. The patient had a history of mitral regurgitation and atrial fibrillation, and he had lived in China at an altitude of 1,900 m all of his life. He traveled to the United States for evaluation of dyspnea.

Over the past 1 year, he had been hospitalized three times for dyspnea, which was thought to be secondary to cardiac disease. His exercise tolerance included two flights of stairs or brisk walking. He had a chronic cough productive of approximately 50 mL of white sputum per day. He denied hemoptysis, fever, chills, or sweats.

At age 20, the patient was treated for a chest infection. Since that time, he has been treated with Chinese medications for recurrent bronchitis. He denied a history of tuberculosis.

The physical examination revealed a BP of 110/65 mm Hg; pulse rate, 80 beats/min; and respiration rate, 18 breaths/min. Cachexia was present, but the patient was in no acute distress. The jugular venous distention was 7 cm, without hepatojugular reflux. There were diminished breath sounds bilaterally and crackles in the left lower lung field. The cardiac examination was notable for a normal S1 and S2, and a grade II/VI systolic murmur at the apex. There were no gallops, opening snap, or diastolic murmurs. His extremities were without clubbing, cyanosis, or edema.

The patient’s chest radiograph and thoracic CT scan are shown in Figures 1–3.

What is the diagnosis?
**Figure 1.** Chest radiograph shows left lower lobe infiltrate, bilaterally downward displaced hila, and an enlarged cardiac silhouette.

**Figure 2.** Thoracic CT scan demonstrates right and left atria enlargement, volume loss and chronic scarring medially in the left lower lobe, as well as small collateral vessels seen throughout the density in the left lower lobe (arrow).

**Figure 3.** Thoracic CT scan shows multiple enhancing collateral vessels in the aortopulmonary window (arrow).
Diagnosis: Intralobar pulmonary sequestration with abnormal systemic arterial supply of both congenital and acquired origins.

DISCUSSION

A chest radiograph (Fig 1) revealed a chronic left lower zone infiltrate. A thoracic CT scan (Fig 2) demonstrated left lower lobe atelectasis and fibrosis, mild bronchiectatic changes in the lingula, emphysema in the lower lung fields, and pulmonary artery enlargement. The CT scan (Fig 3) also showed numerous vessels in the aortopulmonary window, which are actually collateral vessels arising from the aortic arch seen on the sagittal reformatted image (Fig 4). An echocardiogram demonstrated mild left ventricular hypertrophy, left atrial enlargement, mitral regurgitation, right atrial and ventricular dilatation, tricuspid regurgitation, and normal systolic function. A cardiac catheterization was then performed; an aberrant artery arising from the left circumflex artery was noted to lead to the left lower lobe (Fig 5).

This represents an intralobar sequestration supplied by a large aberrant circumflex artery with multiple collateral vessels arising from the aortic arch to the abnormal pulmonary parenchyma in the left lower lobe. The aberrant circumflex artery is likely a congenital anomaly, given its large caliber and its rarity. The collaterals originating from the aortic arch most likely demonstrate an acquired phenomenon, secondary to chronic inflammation, which was secondary to a chronic inflammatory process.

A pulmonary sequestration is a portion of pulmonary tissue that does not have communication with the tracheobronchial system through a normal bronchus and is supplied by anomalous systemic arteries.1-3 Sade et al3 proposed a sequestration spectrum that they defined as a collection of anomalies of the lung parenchyma and of its vascular supply, often with associated foregut anomalies. The spectrum includes sequestered pulmonary tissue within or outside of the visceral pleura of the lung; the arterial supply may be from a systemic or pulmonary artery or both; the venous drainage may be to a systemic or pulmonary vein or both; communication with the GI tract may occur; and the diaphragm may not be normal. Within this spectrum lies sequestered lung with an anomalous vascular supply and, at the other end, abnormal pulmonary tissue with a normal vascular supply (ie, bronchopulmonary cyst). 3,4 Pulmonary sequestrations are anatomically divided into two categories, intralobar and extralobar. An intralobar sequestration lies within the pleural cavity of the normal lung. An extralobar sequestration has its own visceral supply.2,3

Intralobar pulmonary sequestration has an equal gender distribution. The intralobar sequestration is usually diagnosed after the second decade of life. It occurs in the left lung 60% of the time, most commonly in the posterior basal segment of the lower lobe. The caliber of the anomalous artery in the intralobar sequestration is usually large, and venous drainage generally occurs through the pulmonary vein.3,5 It is infrequently associated with other congenital anomalies (11.9%).6 Intralobar sequestration does not, initially, communicate with the tracheobronchial system. However, mucous secretion with in the intralobar sequestration may cause cystic swelling, leading to atelectasis of the normal surrounding lung tissue. Superimposed infection can develop, extending to the sequestered area, causing capsular erosion and airway communication.4

Extralobar pulmonary sequestration affects male patients more frequently, in 80% of cases. It is usually diagnosed in infancy or childhood. Extralobar sequestration occurs in the left lower hemithorax.
nary segments do not have normal communication resulting anomalous, nonfunctioning lung tissue is the base of the fetal lung do not regress. The normal differentiation at the site of the GI and respiratory questration as a congenital anomaly suggests that genital vs an acquired anomaly. The theory of sequestration is frequently associated with other congenital anomalies 58% of the time.7

Frequently, sequestration is discovered when a patient develops symptoms that are usually secondary to infection or associated cardiac disease. Pulmonary signs and symptoms include recurrent pneumonia, abscesses, hemoptysis, recurrent purulent sputum, chest pain, dyspnea, fever, fatigue, bruist, and, rarely, clubbing. Cardiovascular symptoms result from left-to-right and left-to-left shunting. Fifteen percent of the patients remain asymptomatic, and sequestration is discovered incidentally on the chest radiograph.1,5,7

The vascular supply to a pulmonary sequestration is generally via a single artery; however, multiple arteries supply 15% of sequestrations. The vascular supply originates from the abdominal aorta (73%), thoracic aorta (18%), and, rarely, from the ascending aorta, aortic arch, subclavian, innominate, and celiac arteries.2 In our review of the literature, there are four reported cases of a coronary artery supplying the sequestered lung.8–11 The left circumflex artery was the single arterial supply to the intralobar left lower lobe sequestration in three of the cases.8,9,11 There was dual arterial supply by the right coronary artery and left circumflex artery to the posterior basal segment of the left lower sequestered lobe in the fourth case.10

An ongoing debate exists in the literature regarding the etiology of intralobar sequestrations, a congenital vs an acquired anomaly. The theory of sequestration as a congenital anomaly suggests that differentiation at the site of the GI and respiratory tracts is disrupted. Systemic arterial connections to the base of the fetal lung do not regress. The normal merging of the main pulmonary artery into a primitive arterial plexus does not occur. Therefore, the resulting anomalous, nonfunctioning lung tissue is supplied by a systemic artery. Sequestrated pulmonary segments do not have normal communication with the tracheobronchial tree.2 The arguments in favor of the acquired (pseudosequestration) theory suggest that bronchial obstruction by aspiration or inflammation leads to the transformation of normal lung into intralobar sequestration. Infection leading to intralobar sequestration differs from typical lower lobe pneumonia that resolves with little or no sequelae, in that there is a persistently occluded bronchus to the involved segment and a partial or complete occlusion of pulmonary artery to the infected region.2,6 Extensive granulation tissue, pleural adhesions, and increased metabolic requirements of hypertrophied bronchial muscle, chronic inflammation, and lymphoid tissue stimulate neovascularization from the high- pressured systemic circulation, perhaps via angiogenesis factors.2,12 This series of reactions can occur from a single or recurrent episodes of pneumonia.6 A study by Stocker and Malczak6 of 11 randomly selected pediatric autopsies revealed the presence of one or more pulmonary ligament arteries off of the aorta in 90% of cases, demonstrating that vessels are available to potentially supply abnormal areas of the lung. Others have shown extensive systemic pulmonary anastomoses at the precapillary level in lungs with bronchiectatic changes.12 These studies indicate that the systemic arteries to visceral pleura are available for incorporation in sequestered pulmonary tissue after the occurrence of bronchial obstruction and pneumonia.

On plain chest radiograph, pulmonary sequestration can appear as a homogenous, well-circumscribed round, oval, or triangular infiltrate. It can also appear as a solid, poorly defined opacity, or as a cystic lesion.2 Bronchography reveals normal bronchi branching down and around the sequestrated area, but not directly to this portion of the lung.2 Aortograms are performed in order to define the systemic blood supply and venous drainage.2 On CT, various parenchymal abnormalities have been observed, including cysts, soft tissue masses, emphysema surrounding the cysts or masses, and hypervascularity of a region of lung.5

Since it is difficult to conceive how a large single artery from the coronary system could be stimulated by chronic inflammation, this aberrant artery is most likely congenital in origin. However, the multiple small vessels arising from the aorta are likely secondary to chronic inflammation, which led to angiogenesis and neovascularization. Therefore, this patient has both congenital and acquired intralobar sequestration.

The majority of this patient’s symptoms resolved spontaneously. His dyspnea was most likely multifactorial, including sequestration, cardiac disease, and high altitude. Therefore, no further diagnostic studies or interventions were performed. The patient returned to China.

References

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