

## Bronchopulmonary Sequestration

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### Introduction

Bronchopulmonary sequestration is a rare congenital lung malformation characterized by an abnormal segment of bronchopulmonary tissue supplied by an anomalous systemic artery. The diagnosis may be easily missed in adults, as many of the symptoms overlap with other pulmonary processes. Surgical resection provides definitive management and is usually reserved for patients with symptoms. This article presents a case of intralobar bronchopulmonary sequestration that presented during adulthood. A brief review of the clinical features, diagnostic strategies, and management options (with particular focus on intralobar sequestration) follows.

### Case Report

A 41-year-old man presented to the out-patient pulmonary clinic complaining of many years of intermittent, right-sided chest pain. His pain was present for the last 5 years, yet substantially worsened 2 months prior to the current presentation, when he presented to an outside clinic, complaining of chest pain, fevers, chills, and a productive cough. A chest radiograph revealed an opacity in his right lower lung field and he was treated with oral levofloxacin. The majority of his symptoms subsequently improved, but his chest pain persisted despite antibiotic therapy. He also described 2 additional similar episodes over the last 30 years, for which he was treated for pneumonia. The patient was generally healthy, denied any risk factors for the human immunodeficiency virus (HIV) or tuberculosis, and had no other important medical history. He was taking no medications. He admitted to intermittent cigarette smoking and occasional alcohol use. Family history was unremarkable.

Physical examination revealed a healthy-appearing, well-nourished man in no distress. Vital signs, including oxygen saturation, were all within normal limits. Chest auscultation revealed mild rales at the right base, but was otherwise unremarkable. Chest pain was not reproducible with palpation of the right chest wall. Extremities showed

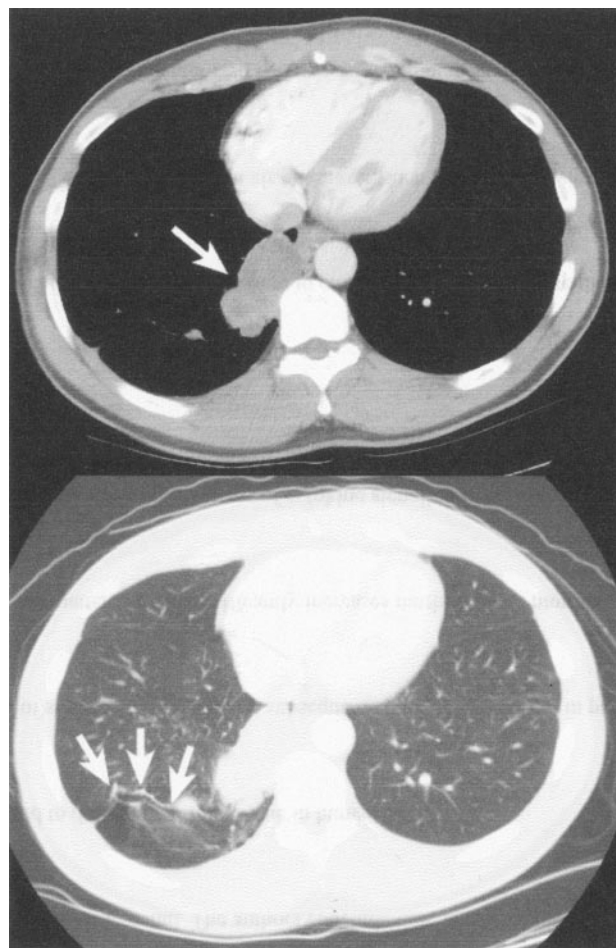


Fig. 1. Computed tomogram of a bronchopulmonary sequestration. Top panel: A multicystic mass (arrow) along the medial portion of the right lower lobe, which is a characteristic location for a sequestration. Bottom panel: Emphysematous changes in the lung parenchyma (arrows) are due to air trapping due to the sequestration.

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no evidence of clubbing. The remainder of the examination showed no abnormalities.

Laboratory results, including complete blood count, electrolytes, and liver function tests, were unremarkable. A chest radiograph brought to clinic by the patient from 2 months prior (not available) showed an ill-defined opacity behind the right heart border. Subsequent contrast-enhanced computed tomography (CT) of the chest (Fig. 1) revealed a multiloculated cystic lesion within the medial right-lower lobe, with associated right-lower-lobe emphysema and loss of vasculature. A small arterial branch from the aorta to this cystic solid mass was visualized. Given the history, imaging features characteristic of intralobar sequestration, and ongoing symptoms, the patient was referred to thoracic surgery and underwent right thoracotomy and right-lower-lobe resection. The surgeon carefully isolated and ligated the arterial branch from the aorta, and the lesion was removed without incident. Pathology of the resected specimen showed inflammatory lung parenchyma, with cystic spaces of up to 4 cm. The cysts had smooth walls and contained a viscous, brown, opaque, mucus-like material. A small artery was noted along the pleural surface. After surgery, the patient made an uneventful recovery and at a 2-month follow-up visit was free from chest pain.

### Discussion

The term "sequestration," first used in the medical literature by Pryce in 1946,<sup>1</sup> originates from the Latin word *sequestare*, which means "to separate." Bronchopulmonary sequestration is the term given to a region of lung parenchyma that is partially or completely separated from the bronchopulmonary tree of the lung proper.<sup>1</sup> Its blood supply is usually from an aberrant artery arising from the aorta or one of its branches. Sequestration is a rare congenital abnormality, representing 0.15–6% of all pulmonary malformations.<sup>2</sup> It generally is classified into 2 distinct forms: intralobar, as in our patient, and extralobar, depending on whether it has independent pleura. An intralobar sequestration is contained within the visceral pleura of the adjacent lung, while the extralobar type is contained within its own visceral pleura, separate from the involved lung.

Multiple theories of the pathogenesis of pulmonary sequestration have been proposed in the literature and have been nicely summarized by Corbett and Humphrey.<sup>2</sup> The most widely accepted hypothesis is that it results from formation of an accessory lung bud inferior to the normal lung buds during development. During embryogenesis this "accessory" lung bud develops an independent vascular supply, usually from the aorta, and remains independent from the normally developing tracheobronchial tree.<sup>2</sup>

### Clinical Features

The 2 types of sequestration are associated with very different clinical features (Table 1). Extralobar sequestration is a disease confined to neonates because of the high frequency of concomitant congenital abnormalities. In contrast, intralobar sequestration usually presents during childhood, but up to half present after the age of 20.<sup>3</sup> The vast majority of patients with intralobar sequestration are asymptomatic and carry the abnormality for years, only to be diagnosed during a routine chest radiograph for unrelated symptoms. Symptoms, when present, are often nonspecific and can include chest pain, pleuritic pain, shortness of breath, and wheezing; however, signs of recurrent infection, such as fever and productive cough, are more common. This risk of infection is counterintuitive, given the absence of a tracheobronchial connection to the sequestration. Despite this absence of a direct connection, sequestrations are not completely isolated from the native lung. Bacteria can still invade via the alveolar pores of Kohn.<sup>4</sup> Once bacteria have colonized the sequestration, frank infection can ensue because of the lack of normal bronchial drainage.

Finally, although uncommon and usually benign, the sequestration can present more dramatically with frank hemoptysis. There are, however, reports in the medical literature of life-threatening hemoptysis due to sequestration.<sup>2</sup>

The arterial supply of intralobar sequestration is a branch off of the descending thoracic aorta in up to 75% of patients.<sup>3</sup> Rarely, however, the arterial supply can come from sites other than the aorta, and have even been documented to arise from the circumflex branch of the left coronary artery.<sup>5</sup>

### Radiographic Features

Two main goals in the radiographic imaging of sequestration are: (1) to further characterize the lesion to help rule out alternative pathology, and (2) to characterize the aberrant arterial supply of the sequestration to facilitate operative management.

Plain chest radiograph is usually nonspecific, showing an ill-defined consolidation that simulates pneumonia, or shows a soft tissue mass with well- or ill-defined borders.<sup>6,7</sup> Air/fluid levels can also be seen.<sup>7</sup> Chest CT usually shows a discrete mass in the posterior- or medial-basal segment of the lower lobe, with (as in our case) or without cystic changes.<sup>7</sup> Lesions present in lobes other than the lower lobe should spur investigation of alternative diagnoses, as sequestration outside of the lower lobes accounts for only 5% of cases reported in the literature.<sup>3</sup> Cystic changes, when present, are usually multiple in nature. Emphysematous changes in the surrounding lung parenchyma

Table 1. Features That Differentiate Intralobar From Extralobar Pulmonary Sequestration

Feature	Intralobar Sequestration	Extralobar Sequestration
Relative prevalence	6:1	1:6
Age at diagnosis	Child to adult	Neonate
Distribution by sex	1:1	80% male
Location	60% left base, 40% right base	90% left
Pleura	Contained within visceral pleura	Visceral pleura independent of primary lung
Arterial supply	Usually systemic	Usually systemic
Pulmonary arterial supply	Absent	May be present
Presence of bronchi	Rare	May connect to gastrointestinal tract
Association with congenital abnormalities	Rare	Common

(Adapted from Reference 6)

are a characteristic CT feature of sequestration and are probably related to air trapping from the sequestration itself.<sup>6,7</sup>

Pre-operative identification of the aberrant artery supply is critically important in preventing operative morbidity. CT angiography, also known as spiral CT, is the radiographic method of choice for identifying the artery. Magnetic resonance imaging is not as useful.<sup>3,6,7</sup> Though intralobar sequestration has many characteristic clinical and radiologic features, many mimics have been described in the literature, particularly in adults.<sup>3</sup> Careful evaluation to exclude alternative pathology (Table 2) should ensure a correct diagnosis.

Table 2. Differential Diagnosis of Intralobar Sequestration

Infection
Pneumonia
Abscess
Tuberculosis
Bronchiectasis
Congenital malformations
Diaphragmatic hernia
Bronchogenic cyst
Pericardial cyst
Cystic adenomatoid malformation
Paravertebral mass—nonmalignant
Extramedullary hematopoiesis
Lateral meningocele
Malignancy
Lung cancer
Neurogenic tumor

(Adapted from References 3 and 6)

## Treatment

The consensus treatment for intralobar sequestration is surgical removal, but the timing of surgery is somewhat controversial. Most experts advocate that asymptomatic patients discovered to have an intralobar sequestration during screening for a concomitant illness be referred for surgery, because there are reported cases of death in adulthood due to massive hemoptysis from intralobar sequestration diagnosed in childhood.<sup>8</sup> A less common strategy of close monitoring for symptoms and referral to surgery if symptoms develop is occasionally advocated.<sup>9</sup> Complications of watchful waiting include recurrent pneumonia, hemoptysis, and chest pain. When symptoms develop, however, patients should be referred for definitive surgery.

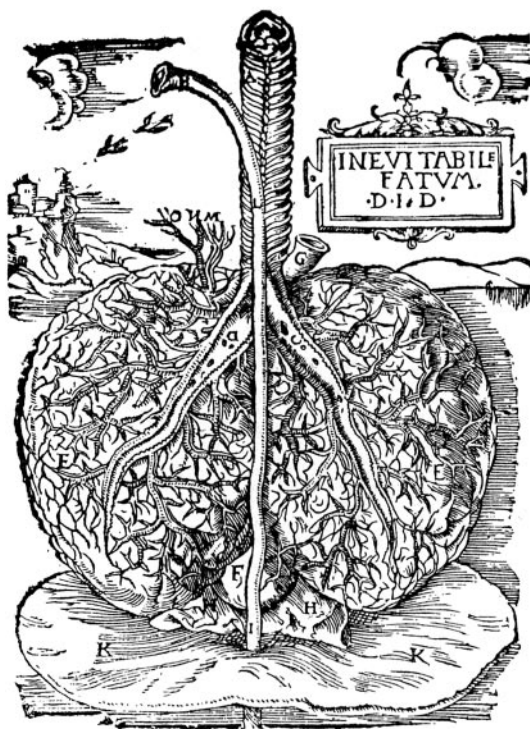
Surgery usually involves lobar resection via standard thoracotomy, although thoroscopic surgery is being used more frequently.<sup>2</sup> In cases where the risk of surgery is too great, angiographic embolization of the feeding systemic vessel can be used. This therapy has proven successful in the definitive management of intralobar sequestration and is now commonly used as an alternative treatment to surgery in children.<sup>10</sup>

This case illustrates a typical presentation of an intralobar bronchopulmonary sequestration. The constellation of recurrent pneumonia and cystic lower-lobe mass fed by an anomalous systemic arterial vessel are hallmarks of this rare disease. Patients with symptoms should be referred to surgery for definitive lobectomy. The main surgical complication can be prevented by careful preoperative investigation of the arterial supply of the sequestration.

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