# Bronchopulmonary Sequestration Presenting as Recurrent Pneumonia

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## ABSTRACT

Bronchopulmonary sequestration is a rare congenital malformation of the lower respiratory tract. Its presentation is varied, and it rarely presents in adulthood. We report the case of a 31-year-old woman who was admitted with recurrent pneumonia. She had been treated for pneumonia multiple times within the last 3 years. On subsequent workup, she was found to have intra-lobar bronchopulmonary sequestration in the left lower lobe of the lung, which was diagnosed on computerized tomography (CT) of the chest. The chest CT revealed an anomalous blood vessel from the abdominal aorta ascending to the left lower lobe of the lung and supplying an abnormal part of the left lung, which was the key to the diagnosis. Bronchopulmonary sequestration was found to be the cause of her recurrent pneumonia. She subsequently underwent resection of the left, lower lobe of the lung. We recommend that bronchopulmonary sequestration be included in the differential diagnosis of recurrent pneumonia in relatively healthy patients.


## INTRODUCTION

Bronchopulmonary sequestration (BPS) is an extremely rare congenital malformation of the lower respiratory tract that usually manifests in infants and adolescents.[1] It is rare to see it in adults, and its presentation is varied. We report a young adult female who presented with recurrent pneumonia and was eventually diagnosed with BPS.

## CASE REPORT

A 31-year-old relatively healthy Asian American woman presented with complaints of purulent productive cough, hemoptysis, and subjective fever for 2 weeks. She denied any chest pain, shortness of breath, weight loss, night sweats, or loss of appetite. She also denied exposure to any illness, contact with animals, or any recent travel outside the United States. Her past medical history was significant for 2 episodes of pneumonia in the past 3 years and a chronic cough with unknown etiology for the same duration. She had been admitted twice to a different medical institution for pneumonia, where she had been treated with antibiotics and had responded well. She brought her old records with her, which included a chest radiograph that showed left lower lobe lung infiltrate. All of her other tests, including blood cultures, had been negative. A computed tomography (CT) of the chest had not been performed, because her illness was thought to have been a simple case of community-acquired pneumonia. She worked as a waitress at a local bar and had a history of 15-pack years of cigarette smoking. On physical examination she appeared anxious, but her vital signs were within normal limits. Chest examination revealed crackles in the left lower lobe of the lung. Examination of other systems (including oral cavity, cardiovascular, abdominal, and neurological) all were unremarkable.

Laboratory investigation revealed leucocytosis, with a white blood cell count of 14.0 x 10*3/uL and a neutrophil count of 92%. Her purified protein derivative (PPD) test and human immunodeficiency virus (HIV) test were negative. Chest radiograph showed left lower lobe consolidation. Sputum examination showed growth of normal flora, and sputum for acid-fast bacilli was negative. An echocardiogram was negative without any associated abnormality. Because this was her third episode of pneumonia, a CT scan of the chest was ordered, which showed dense consolidation in left lower lobe with areas of cavity, cysts, and air fluid level. The CT scan also revealed an anomalous blood vessel from the abdominal aorta ascending to the left lower lobe of the lung and supplying an abnormal part of the left lung (Figures 1–5). The patient's presentation, based on history, physical findings, and chest radiography, was consistent with bronchopulmonary sequestration. She eventually was referred to cardiothoracic surgery and underwent a left lower lobe lung resection.

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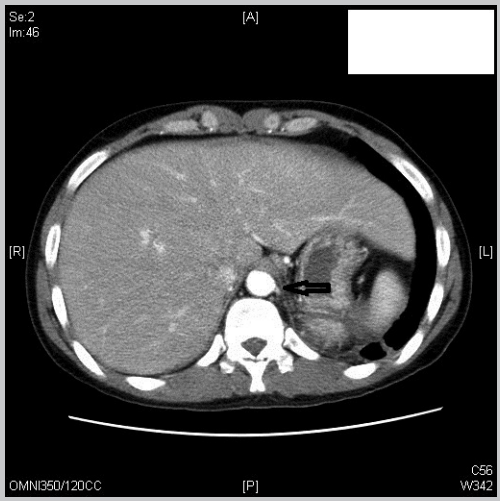
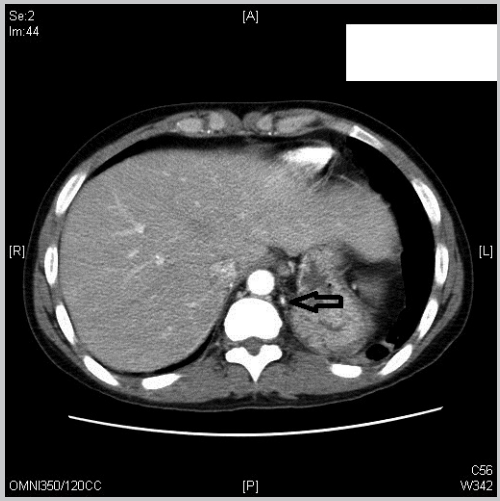



**Figure 1.** Computed tomography scan of chest and abdomen showing a small artery arising from abdominal aorta.



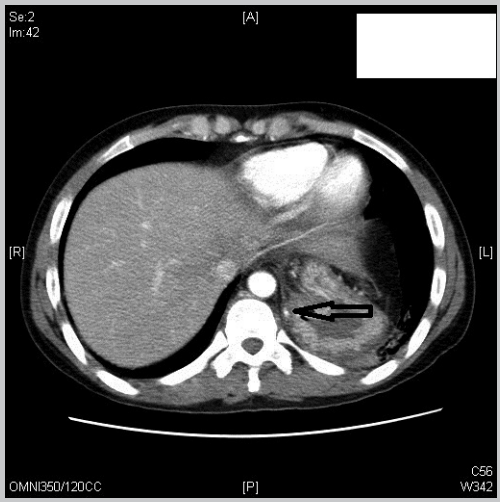




**Figure 2A (top) and B.** Computed tomography scans of chest and abdomen showing the artery travelling upward toward the thorax.

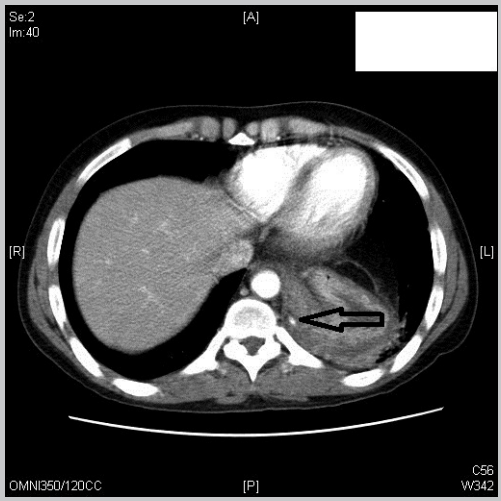



**Figure 3.** Computed tomography scan of chest and abdomen showing that the artery now can be traced into the thorax.

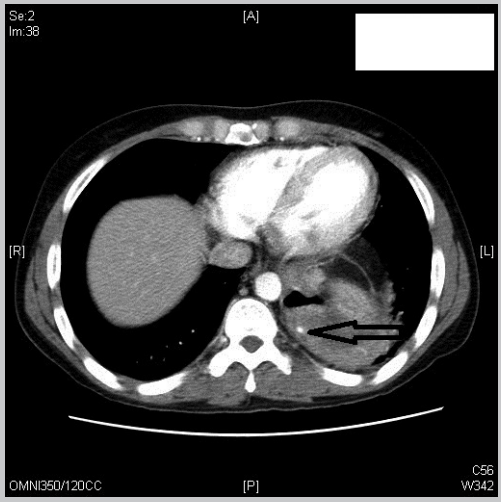



**Figure 4.** Computed tomography scan of chest and abdomen showing the artery now in the lung parenchyma.

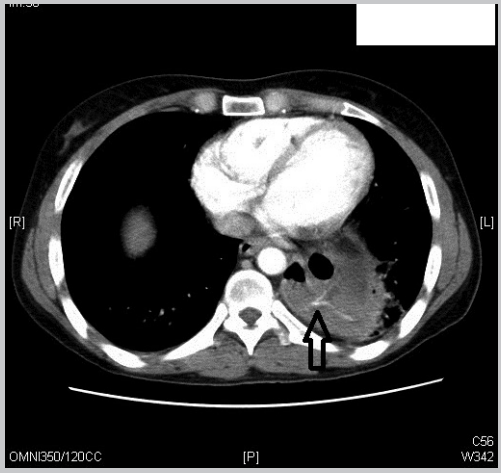



**Figure 5.** Computed tomography scan of chest and abdomen showing that the artery bifurcates within the bronchopulmonary sequestration.

Histopathological examination of the tissue from the lung resection included gross and microscopic examination. Gross examination revealed "left lower lobe sequestration" consisting of a hemorrhagic and fibrinous left lower lobe (315 g; 15.0 x 10.9 x 4.5 cm). The inferior portion of the specimen (7.0 x 4.5 x 2.3 cm) was an area of previous disruption with a silk-sutured vessel at the edge (0.8 cm x 0.6 cm). The hilar area had several stapled resection margins, including a bronchial resection margin (0.8 cm in length x 3.1 cm in greatest diameter), a curved lung resection margin (4.2 cm in greatest dimension), and 4 vessel branches ranging from 0.3 cm to 1.2 cm in length and from 1.6 cm to 2.1 cm in greatest dimension. The pleural surface was fibrinous and hemorrhagic, and the serial section of the lobe from the superior to inferior surface revealed an ill-defined tan-firm hemorrhagic cystic lesion with cavity formation of the bronchus that was partially filled with a brown mucus plug. The rest of the lung was spongy maroon-red with tan speculations. Microscopic examination showed acute and chronic inflammation with cystic dilatation consistent with intralobar sequestration. No fungi or acid-fast bacilli were identified (confirmed by special Gomori methenamine silver and acid fast bacilli stains).

## DISCUSSION

Bronchopulmonary sequestration was first described by Pryce in 1946 in a report of 7 cases.[2] BPS is defined as a nonfunctioning mass of lung tissue that lacks normal communication with the tracheobronchial tree and receives its arterial blood supply from the systemic circulation.[3] It is an extremely rare disorder: BPS accounts for only 0.16% to 6.4% of all pulmonary congenital malformation.[1] The differential diagnosis may include bronchial atresia, cystic adenomatoid malformation, intrapulmonary bronchogenic cyst, and arteriovenous fistula.[4] These conditions are differentiated through the finding of an anomalous systemic arterial supply in bronchopulmonary sequestration.

Depending on its location, BPS is subdivided into intralobar and extralobar sequestration.[5] Extralobar sequestration is located outside the normal lung and has its own visceral pleura, whereas intralobar sequestration is located within the normal lung parenchyma and shares the viscera pleura of the parent lobe of the lung. Intralobar sequestration has normal pulmonary venous return, while extralobar sequestration is associated with aberrant pulmonary venous drainage. Intralobar sequestration is more common than extralobar sequestration, and the majority of intralobar sequestrations are likely acquired lesions.[6-8] Approximately 75% of bronchopulmonary sequestrations are intralobar.[5-8] Extralobar sequestration may be an incidental finding on prenatal ultrasound done during the second to third trimesters. It usually presents in infancy as respiratory distress syndrome or, less commonly, as pneumonia.[9] Intralobar sequestration usually presents in early childhood and adolescence with recurrent respiratory infections. The blood usually is supplied by an aberrant artery arising from the descending thoracic aorta (70%) or abdominal aorta (20%).[5] The defense mechanism is impaired in the abnormal lung tissue, making it prone to recurrent infection, chronic inflammation, cystic changes, and fibrosis. The standard treatment is resection of the segment or lobe that contains the sequestered tissue; the prognosis is favorable.[7,8] Our patient had intralobar sequestration and subsequently underwent a left lower lobe lung resection.

## CONCLUSION

In the adult population, BPS can present as recurrent pneumonia and should be included in the differential diagnosis of recurrent pneumonia in relatively healthy patients.[10] Physicians should be aware of this rare congenital condition that can present in adults with symptoms of common diseases.

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