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Incidental Finding of Bronchopulmonary Sequestration in a 64-Year-Old Female

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Background: Bronchopulmonary sequestration is a congenital abnormality of the primitive foregut. In adults, the typical age at presentation is 20-25 years.

Case Report: A 64-year-old female was referred for evaluation of an 8 × 6-cm right lower lobe cystic lesion. Her medical history was significant for recurrent right lower lobe pneumonia requiring multiple hospitalizations. Her physical examination was significant for crackles at the right lung base. Computed tomography (CT) of the chest with contrast showed cystic changes with thickened septation of the medial segment of the right lower lobe lacking distinct visceral pleura and with arterial supply from the anomalous branch of the thoracic aorta arising near the celiac trunk. Pulmonary angiography confirmed the diagnosis of intralobar pulmonary sequestration. The patient underwent celiac endovascular coil embolization of the anomalous artery to lessen the risk of hemorrhage prior to video-assisted thoracoscopic surgery (VATS) resection of the right lower lobe. She recovered well and was discharged home 1 week after VATS lobectomy. Follow-up CT of the chest 2 months later showed normal postsurgical changes related to right lower lobe lobectomy. The patient remained asymptomatic and resumed her daily activities.

Conclusion: Pulmonary sequestration can present with recurrent pneumonia in late adulthood. Physicians must review any previous imaging studies of the chest to identify the structural abnormality and be cognizant of differential diagnoses such as infected cystic bronchiectasis, bronchogenic cyst, congenital diaphragmatic hernia, or cystic adenomatoid malformation that can occur in conjunction with bronchopulmonary sequestration. Pulmonary angiogram is the gold standard to confirm the diagnosis of bronchopulmonary sequestration. Surgical resection is the standard of care.

Keywords: *Bronchopulmonary sequestration, congenital abnormalities, respiratory system abnormalities*

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INTRODUCTION

Bronchopulmonary sequestration was first described in 1946[1] and is the second most common pulmonary congenital abnormality.[2,3] The actual prevalence of bronchopulmonary sequestration is not available in the literature because of the rarity of this abnormality. The typical age at presentation in adults is 20-25 years.[3-5] Bronchopulmonary sequestration is a nonfunctioning mass of the lung parenchyma that lacks normal communication with the tracheobronchial tree and receives arterial blood supply from the systemic circulation. This abnormality originates from an abnormality of the primitive foregut during embryonic development.[6] We describe a case of bronchopulmonary sequestration incidentally diagnosed in a patient who presented with recurrent right lower lobe pneumonia.

CASE REPORT

A 64-year-old female with a medical history significant for asthma and recurrent right lower lobe pneumonia requiring repeated hospitalizations was referred for evaluation of an 8 × 6-cm right lower lobe cystic lesion. At the time of presentation, she reported right chest discomfort but said she otherwise felt well and denied fever, cough, shortness of breath, or weight loss. Physical examination was significant for crackles at the right lung base. Her pulmonary function test was unremarkable. Computed tomography (CT) of the chest with contrast showed normal lung parenchyma and cystic changes with thickened septation of the medial segment of the right lower lobe lacking distinct visceral pleura and with arterial supply from an anomalous branch of the thoracic aorta arising near the celiac trunk. No definite anomalous venous drainage was identified (Figure 1). This finding was consistent with intralobar pulmonary sequestration and similar to a CT of the chest from 3 years prior to presentation (Figure 2). The patient had deferred surgical treatment at that time but now opted for surgical intervention for relief from her recurrent pneumonic episodes. She underwent celiac endovascular coil embolization of the anomalous artery to lower the risk of hemorrhage prior to video-assisted thoracoscopic surgery (VATS) right lower lobe

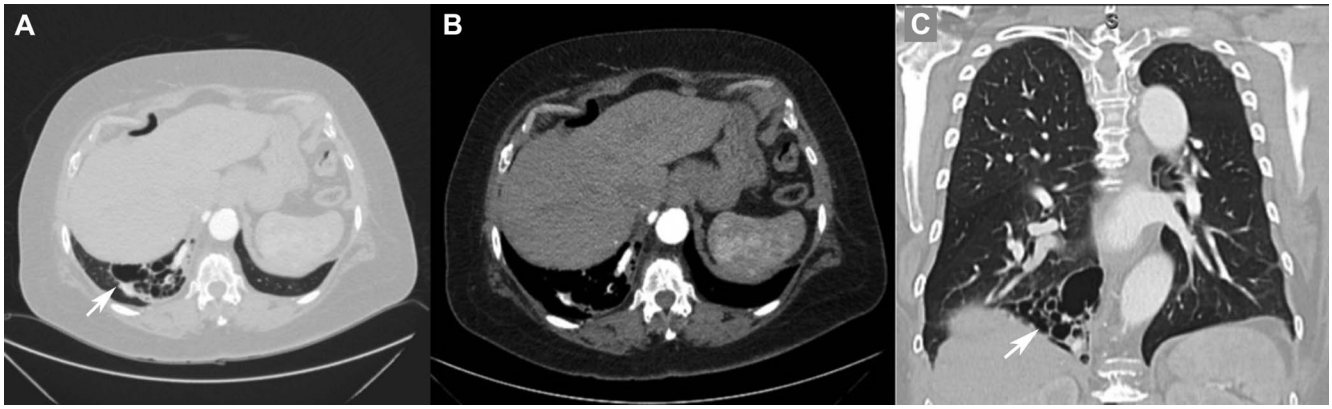

Figure 1. Axial (A and B) and coronal (C) computed tomography views of the chest with contrast show cystic changes with thickened septation of the medial segment of the right lower lobe lacking distinct visceral pleura with arterial supply from an anomalous branch of the thoracic aorta arising near the celiac trunk. No definite anomalous venous drainage was identified.

lobectomy. A 4-cm access incision was made in the right fourth intercostal space. Extensive thin adhesions visualized along the diaphragm adhering to the right lower lobe were subsequently divided using a LigaSure device (Medtronic). The lung was reflected superiorly, and an aberrant artery was seen originating from below the diaphragm into the right lower lobe. The vessel was circumferentially dissected. A stapler was used to clamp across the right lower lobe bronchus. Test inflation confirmed patency of the right middle lobe and right upper lobe, and the stapler was fired. The right lower lobe was retrieved using the Echelon Flex GST (Ethicon US, LLC), an anchor bag endoscopic retrieval system, via the access incision.

Gross specimen examination confirmed the CT finding of cystic changes and septation of the medial segment of the right lower lobe lacking distinct visceral pleura with arterial supply from an anomalous branch of the thoracic aorta and revealed the absence of tracheobronchial communication to the nonfunctional segment in the medial aspect of the right lower lung (Figure 3). Histologically, a large systemic feeder vessel with atherosclerotic changes was also identified (Figure 4A), and a sharply circumscribed area with dilated remnants of bronchi containing inspissated mucus, extensive interstitial fibrosis, epithelial metaplasia, and thick-walled vessels was seen (Figures 4B-4C), confirming the clinical impression of intralobar pulmonary sequestration.

The patient had no complications after VATS lobectomy and was discharged home 1 week after the procedure. Follow-up CT of the chest 2 months later showed normal postsurgical changes related to right lower lobe lobectomy. She continued to be asymptomatic and resumed her daily activities.

DISCUSSION

Bronchopulmonary sequestration is categorized as extralobar pulmonary sequestration or intralobar pulmonary sequestration. Extralobar pulmonary sequestration is located outside the normal lung and has its own visceral pleura. Thus, extralobar pulmonary sequestration is less prone to infectious complications but is associated with other congenital anomalies usually discovered during routine prenatal ultrasound. Bronchopulmonary sequestration presents with recurrent pneumonias in up to 70% of patients.[3] Up to 25% of patients are asymptomatic, and the sequestration is incidentally found on routine chest imaging studies.[3] The Table presents the characteristic features of intralobar and extralobar pulmonary sequestration.[4,5] Differential diagnoses include infected cystic bronchiectasis, bronchogenic cyst, congenital diaphragmatic hernia, or cystic adenomatoid malformation that can occur in conjunction with bronchopulmonary sequestration.[7] Congen-

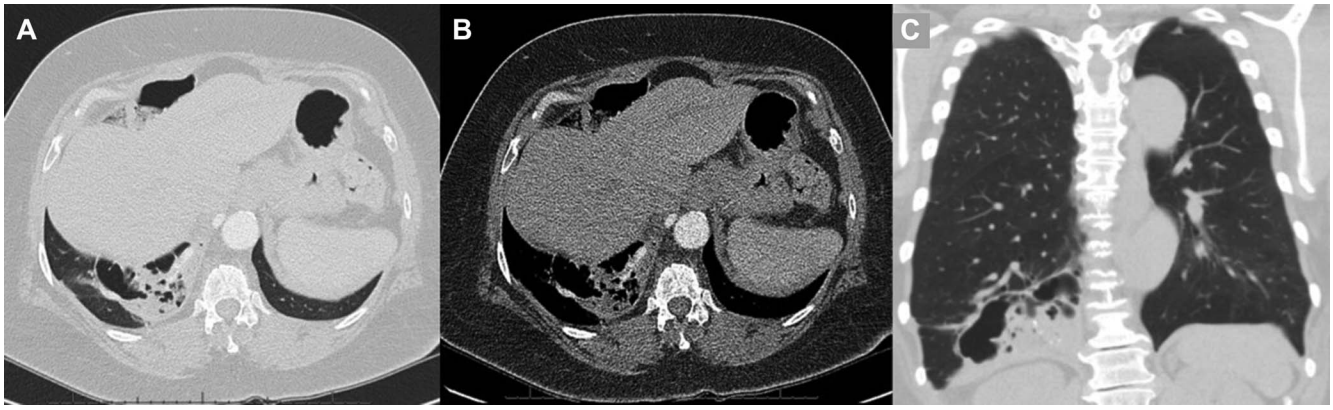

Figure 2. Axial (A and B) and coronal (C) computed tomography views of the chest without contrast from 3 years before presentation show cystic structures with intracystic fluid.

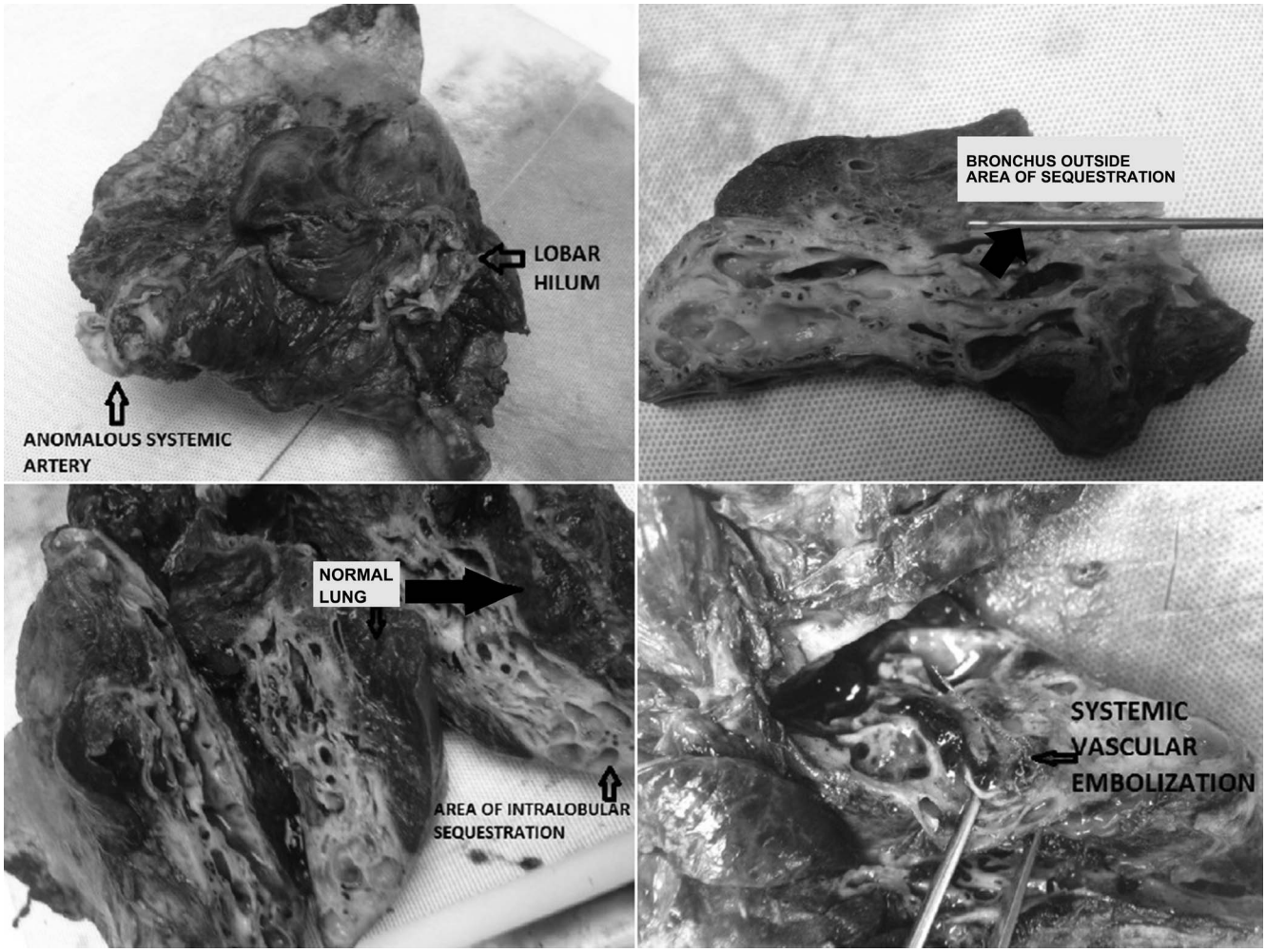

Figure 3. Gross images of the lobectomy specimen show a circumscribed area of intralobar pulmonary sequestration with common visceral pleural investment and normal lung and separate systemic arterial supply. (To see this image in color, visit https://education.ochsner.org/publishing-services/toc/tunsupon-17-0005-fig3.)

ital pulmonary malformation should be suspected in patients with an incidentally found cyst or recurrent pneumonia. Pulmonary angiography plays an important role in establishing the diagnosis and identifying the vascular structures that may require embolization prior to surgical resection to avoid inadvertent vascular injury during surgery. Pulmonary angiography demonstrating a portion of the lung parenchyma that lacks normal communication with the tracheobronchial tree and that receives arterial blood supply from the systemic circulation is the gold standard for diagnosis of bronchopulmonary sequestration. While treatment for intralobar pulmonary sequestration is lobectomy, treatment for extralobar

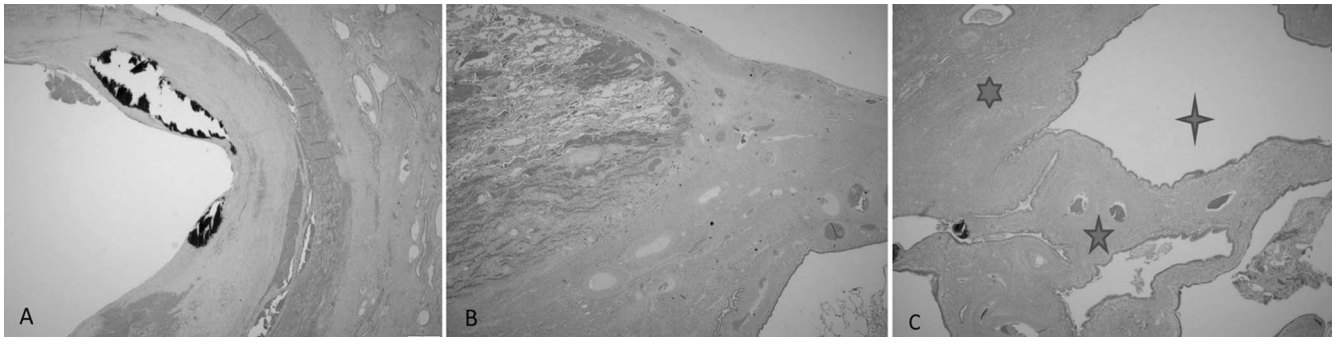

Figure 4. A: Systemic arterial feeder with atherosclerotic changes. B: Sharp junction of normal lung with intralobar pulmonary sequestration shows common visceral pleural investment. C: Intralobar pulmonary sequestration shows prominent vessels (5-point star), dilated bronchi (4-point star), and fibrotic stroma (6-point star) (hematoxylin and eosin stain, 2×). (To see this image in color, visit https://education.ochsner.org/publishing-services/toc/tunsupon-17-0005-fig4.)

Table. Characteristic Features of Intralobar and Extralobar Pulmonary Sequestration

	Intralobar Sequestration (ILS)	Extralobar Sequestration (ELS)
Sex	Equally affects male and female	Male to female ratio of 4:1
Clinical onset	Late adolescence or early adulthood Recurrent pneumonia (most common) Hemoptysis Hemothorax	Early in infancy before 6 months Respiratory distress Feeding difficulties
Common location	Left posterior basal segment	Left posterior costodiaphragmatic sulcus
Pleural involvement	Shares visceral pleura with normal lobes	Has own visceral pleura
Radiographic presentation	Homogenous or heterogenous opacity Cystic lesions filled with fluid and air Cavitation Emphysematous change surrounding the lesion	Homogenous opacity with distinct border Emphysematous change surrounding the lesion
Arterial blood supply (applies to both ILS and ELS)	Descending thoracic aorta (most common) Upper abdominal aorta Celiac trunk Splenic arteries Intercostal arteries Subclavian arteries Internal thoracic or coronary arteries	
Venous drainage	Pulmonary veins to left atrium	Systematic veins Intercostal veins Azygos-hemiazygos vein Superior or inferior vena cava
Associated congenital anomalies	Uncommon	Common Congenital diaphragmatic hernia Diaphragmatic eventration Diaphragmatic paralysis Polyhydramnios Fetal hydrops

pulmonary sequestration is simple excision (sequestrectomy) with ligation of the anomalous systemic artery.

CONCLUSION

Pulmonary sequestration can present with recurrent pneumonia in late adulthood. Physicians must review any previous imaging studies of the chest to identify the structural abnormality and should be cognizant of differential diagnoses such as infected cystic bronchiectasis, bronchogenic cyst, congenital diaphragmatic hernia, or cystic adenomatoid malformation that can occur in conjunction with bronchopulmonary sequestration. Surgical resection is the standard of care for pulmonary sequestration.

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REFERENCES

1. Pryce DM. Lower accessory pulmonary artery with intralobar sequestration of lung: a report of seven cases. *J Pathol Bacteriol*. 1946 Jul;58(3):457-467.
2. Biyyam DR, Chapman T, Ferguson MR, Deutsch G, Dighe MK. Congenital lung abnormalities: embryologic features, prenatal diagnosis, and postnatal radiologic-pathologic correlation. *Radiographics*. 2010 Oct;30(6):1721-1738. doi:10.1148/rg.306105508.
3. Ha D, Yadav R, Mazzone PJ. Cystic lung disease: systematic, stepwise diagnosis. *Cleve Clin J Med*. 2015 Feb;82(2):115-127. doi: 10.3949/ccjm.82a.14020.
4. Abbey P, Das CJ, Pangtey GS, Seith A, Dutta R, Kumar A. Imaging in bronchopulmonary sequestration. *J Med Imaging Radiat Oncol*. 2009 Feb;53(1):22-31. doi: 10.1111/j.1754-9485.2009.02033.x.
5. Tsolakis CC, Kollias VD, Panayotopoulos PP. Pulmonary sequestration. Experience with eight consecutive cases. *Scand Cardiovasc J*. 1997;31(4):229-232.
6. Correia-Pinto J, Gonzaga S, Huang Y, Rottier R. Congenital lung lesions—underlying molecular mechanisms. *Semin Pediatr Surg*. 2010 Aug;19(3):171-179. doi: 10.1053/j.sempedsurg.2010.03.003.
7. Conran RM, Stocker JT. Extralobar sequestration with frequently associated congenital cystic adenomatoid malformation, type 2: report of 50 cases. *Pediatr Dev Pathol*. 1999 Sep-Oct;2(5):454-463.

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