Bronchopulmonary sequestration in a 60 year old man

Lena Naffaa[1*] , Jay Tank[2], Sara Ali[2], Cesar Ong[3]

1. Department of Radiology, Akron Children's Hospital, Akron, Ohio, USA

2. Department of Radiology, Harlem Hospital, New York, New York, USA

3. Department of Radiology, University of Minnesota Children's Hospital, Columbia, Missouri, USA

* **Correspondence:** Lena Naffaa, 1242 Emerald Creek Drive, Broadview Heights, OH, 44147, USA (*lnaffaa@chmca.org*)

Radiology Case. 2014 Oct; 8(10):32-39 :: DOI: 10.3941/jrcr.v8i10.2235

ABSTRACT

We report a case of bronchopulmonary sequestration (BPS) in a 60 year old man with recurrent cough. After failed antibiotic therapy for presumed left lower lobe (LLL) pneumonia seen on chest radiographs, bronchoscopy was performed revealing cryptogenic organizing pneumonia. Further work-up with thoracic imaging demonstrates a feeding artery from the thoracic aorta to the LLL consolidation indicating the presence of BPS. A brief review of the clinical and radiological features and management options of BPS are listed, with particular emphasis on the various imaging modalities and techniques in the diagnosis and pre-surgical planning of intralobar sequestration.

CASE REPORT

CASE REPORT

A 60 year old man presented to the emergency room (ER) with a chief complaint of a recurrent cough which had been recently worsening. His past medical history was significant for pneumonia. A PPD test was performed, which was negative. His HIV test was negative. He was not a smoker or alcohol abuser. He took multivitamins at home, denied taking any other medications. In the ER, the patient was afebrile and in no respiratory distress. Physical exam was remarkable for diminished breath sounds in the left lower lung field and wheezing in the upper lung fields but without rales or rhonchi. Based on the physical exam and past medical history, he was not at an increased risk for pulmonary embolism. Laboratory tests including complete blood count, basic metabolite profile, urinalysis and troponins were negative.

Imaging Findings

PA and lateral chest radiographs in the ER (Figure 1) demonstrated a heterogeneous opacity in the left lower lobe which was initially thought to represent pneumonia. Further evaluation with contrast enhanced CT scan of the chest was obtained on a 64 multi-slice Siemens CT scanner with the following technique: axial images, 641 mA, 100 kV, 5 mm slice thickness, 100 mL Omnipaque (Figure 2) and Sagittal and coronal reconstructed images, 641 mA, 100 kV, 5 mm slice thickness, 100 mL Omnipaque (Figures 3 and 4). On the axial images (Figure 2), a circumscribed triangular-shaped opacity was seen in the posterobasal segment of LLL having a heterogeneous appearance with internal small foci of diminished enhancement. A feeding artery was seen on the axial CT images originating from the descending thoracic aorta and coursing towards the LLL opacity. These findings were better appreciated and confirmed on the sagittal and coronal reformats (Figures 3 and 4) and on the maximum intensity projection (MIP) and volume rendering technique (VRT) (Figures 5 and 6), indicating the presence of sequestered lung parenchyma. As it was confined to the posterobasal segment of the LLL and sharing the pleural covering of the left lung, it was felt to represent an intralobar type of BPS. Figure 7 shows a magnetic resonance image (MRI) of the lower lobes demonstrating the feeding vessel coursing from the aorta directly into the consolidation.

Management/Follow-up

In light of the patient's age and atypical presentation, the patient was admitted for further evaluation by the pulmonary team. Pulmonary function tests were performed revealing no obstructive or restrictive pattern. Bronchoscopy yielded a pathologic diagnosis of cryptogenic organizing pneumonia. The patient was treated with a course of antibiotics and steroids. The patient was followed up routinely in the clinic, and the cough resolved after a few months. Given the patient's age, rapid resolution of symptoms, and lack of malignant cells on bronchoscopy, the pulmonary team decided on a conservative management.

DISCUSSION

BPS is a congenital defect where a portion of the lung does not have a connection with the bronchial tree or pulmonary arteries and instead receives its blood supply from an abnormal branch originating from the aorta [1,2,8]. There are two subtypes, intralobar and extralobar, which are differentiated based on their pleural lining and venous drainage. The intralobar sequestration shares the visceral pleural lining of the lung where it is located and drains into the pulmonary veins [1,2,8]. The extralobar type has its own pleural covering and drains into the systemic veins [1,2,8]. BPS is thought to represent 0.15-6.4% of all congenital pulmonary malformations [1]. The intralobar variant is more common than the extralobar variant, roughly in a 3:1 ratio [1]. The sequestered lung is not functional and is dysplastic, which commonly leads to chronic infection [3-7]. The treatment is surgical resection in the younger patients, while older or asymptomatic patients may be treated with conservative management, including antibiotics and corticosteroids. A summary of the main features of pulmonary sequestration is listed in Table 1.

The diagnosis of intralobar BPS is usually considered in childhood when a patient presents with recurrent lower respiratory tract infections as well as a persistent opacity in the lungs, especially in the lower lobes, more common on the left side [3-7]. Rare cases of life-threatening hemoptysis due to BPS have been reported [9]. For the less common extralobar type, the diagnosis is typically thought of in neonates and early infancy with opacity found in the lower lobes [1,2] or less commonly in sub-diaphragmatic location, around 10% [10].

Echo Doppler ultrasound plays an important role in the prenatal diagnosis and follow-up of BPS as it can identify the origin of the aberrant vessel feeding the lung mass which is usually echogenic [11,12]. MRI and nuclear scintigraphy may also be used. MRI can demonstrate signal voids of the aberrant feeding vessel on T2-weighted imaging. Magnetic resonance angiography (MRA) and Nuclear angiography can demonstrate the systemic blood supply to the sequestered lung. Pneumonia has virtually similar findings as a sequestered lung, and is difficult sometimes to discern off imaging alone. The nuclear scintigraphy scan would show increased blood flow in the infected portion of the lung. A neoplasm would have different findings, with increased Doppler flow on ultrasound, lymphadenopathy and mass effect on CT and MRI, and increased 18-fluorodeoxyglucose (FDG) uptake on positron emission tomography (PET) scan. Pulmonary embolism is usually not visible on chest radiographs, but a wedge-shaped opacity, called the "Hampton's hump" is sometimes seen when pulmonary infarction has occurred as a consequence to pulmonary embolism. A ventilation perfusion mismatch on V/Q scan or a filling defect in the pulmonary artery on CTA are the two mainstays of diagnosis on imaging. MRI and ultrasound are not usually used in diagnosing pulmonary embolism. A summary of differential diagnosis and findings on the various imaging modalities is listed in table 2.

The role of imaging has two main goals: further characterization of the lesion to exclude alternative pathologies that can mimic BPS and to characterize the aberrant arterial supply to facilitate surgery. The findings on chest radiographs are non-specific, ranging from a consolidation that mimics pneumonia such as in our case, to a soft tissue mass with well or ill-defined borders [8,13]. Occasionally, air-fluid levels can be seen [13]. CT scan with intravenous contrast and preferably CT angiography (CTA) is the method of choice for identifying the arterial supply. Multi-planar reformatting of CT images in the sagittal and coronal planes, focused reformatting to the area of abnormality, MIP and VRT are essential techniques in identifying the origin and course of the aberrant feeding vessel to the sequestered lung. Preoperative identification of the aberrant arterial supply is critically important in preventing operative morbidity. MRI is not as useful [8,13]. The CT findings range from a discrete mass in the posterior-basal or medial-basal segment of a lower lobe, with (as in our case) or without heterogeneous appearance/cystic changes [8,13]. The aberrant arterial supply on cross sectional imaging is the hallmark for the diagnosis of sequestered lung. Up to 75% of intralobar BPS receives their arterial supply from the descending thoracic aorta [13]. Rarely, arterial supply can come from sites other than the aorta such as the circumflex branch of the left coronary artery [14]. It is worth mentioning that lung cancer can be associated with BPS [15]. This possibility was felt unlikely in our patient as no malignant cells were found on pathologic examination of the bronchoscopy specimen.

The approach to treatment depends upon whether the patient has respiratory distress, recurrent infections, or is asymptomatic. In symptomatic patients, BPS is treated by surgical excision which is usually curative and has minimal morbidity [16,17]. In our patient, due to the advanced age at the time of initial diagnosis and due to rapid symptoms ' resolution following conservative treatment, no surgery was contemplated.

Two other alternative treatment approaches should be noted in symptomatic patients. One consists of occluding the aberrant arterial supply by endovascular approach; however this carries the risk of recurrent infection in the retained unaerated lung parenchyma [18]. The other option, which is widely used in the pediatric population is resection via minimal-access procedures such as video-assisted thorascopic surgery (VATS) [19].

TEACHING POINT

In patients with chronic or recurrent pulmonary consolidation, the diagnosis of BPS needs to be considered as a possibility, regardless of the patient's age. On imaging, an abnormal branch most commonly originating from the descending aorta and supplying the sequestration is the hallmark of this entity. CTA with multiplanar reformatting, MIP and VRT techniques are essential for the diagnosis and pre-surgical planning of BPS.

REFERENCES

1. Dahnert W. Radiology Review Manual. New York: Lippincott 2003;471-473.

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;30:1721-1738. PMID: 21071385

3. Hertzenberg C, Daon E, Kramer J. Intralobar pulmonary sequestration in adults: three case reports. J Thorac Dis 2012 October; 4(5): 516-519. PMID: 23050118

4. Sugio K, Kaneko S, Yokoyama H, Ishida T, Sugimachi K, Hasuo K. Pulmonary sequestration in older child and in adults. Int Surg 1992 Apr-Jun;77(2):102-7. PMID: 1644534

5. Gowrinath K, Kamath GS, Rajagopal KV, Kudva R. Intralobar pulmonary sequestration with aspergilloma in an adult. Respiratory Medicine CME. 2010 Volume 3, Issue 3, Pages 142-145) No PMID found

6. Cooke CR. Bronchopulmonary Sequestration. Resp Care 2006;51:661-64. PMID: 16723043

7. Somja J, De Leval L, Boniver J, Radermecker MA. Intrapulmonary lung sequestration diagnosed in an adult. Rev Med Liege 2011;66:7-12. PMID: 21374954

8. Zylac CF, Eyler WR, Spizarny DL, Stone CH. Developmental lung anomalies in the adult:radiologic-pathologic correlation. Radiographics 2002;22 Spec No:S25-S43. PMID: 12376599

9. Corbett JH, Humphrey GME. Pulmonary sequestration. Paediatr Respir Rev 2004;5(1):59-68. PMID: 15222956

10. Laje P, Martinez-Ferro M, Grisoni E, Dudgeon D. Intraabdominal sequestration. A case series and review of the literature. J Pediatr Surg 2006;41:1309. PMID: 16818069

11. Becmeur F, Horta-Geraud P, Donato L, Sauvage P. Pulmonary sequestrations: prenatal ultrasound diagnosis, treatment, and outcome. J Pediatr Surg 1998;33:492-6. PMID: 9537564

12. Nicolini U, Cerri V, Groli C, Poblete A, Mauro F. A new approach to prenatal treatment of extralobar pulmonary sequestration. Prenat Diagn 2000;20:758-60. PMID: 11015708

13. Petersen G, Martin U, Singhal A, Criner GJ. Intralobar sequestration in the middle-aged and elderly adult: recognition and radiographic evaluation. J Thorac Cardiovasc Surg 2003;126(6):2086-2090. PMID: 14688733

14. Silverman ME, White CS, Ziskind AA. Pulmonary sequestration receiving arterial supply from the left circumflex coronary artery. Chest 1994;106(3):948-949. PMID: 8082386

15. Okamoto T, Masuya D, Nakashima T, Ishikawa S, Yamamoto Y, Huang CL, Yokomise H. Successful treatment for lung cancer associated with pulmonary sequestration . Ann Thorac Surg 2005;80:2344-6. PMID: 16305905

16. Samuel M, Burge DM. Management of antenatally diagnosed pulmonary sequestration associated with congenital cystic adenomatoid malformation. Thorax 1999; 54:701. PMID: 10413723

17. Haller JA Jr, Golladay ES, Pickard LR, Tepas JJ, Shorter NA, Shermata DW. Surgical management of lung bud anomalies: lobar emphysema, bronchogenic cyst, cystic adenomatoid malformation, and intralobar pulmonary sequestration. Ann Thorac Surg 1979; 28:33. PMID: 454042

18. Marine LM, Valdes FE, Mertens RM, Bergoeing MR, Kramer A. Endovascular treatment of symptomatic pulmonary sequestration. Ann Vasc surg 2011;25:696. e11-5. PMID: 21724108

19. Gonzalez D, Garcia J, Fieira E, Paradela M. Video-assisted thoracoscopic lobectomy in the treatment of intralobar pulmonary sequestration. Interact Cardiovasc Thorac Surg 2011;12;77-9. PMID: 20972207

FIGURES

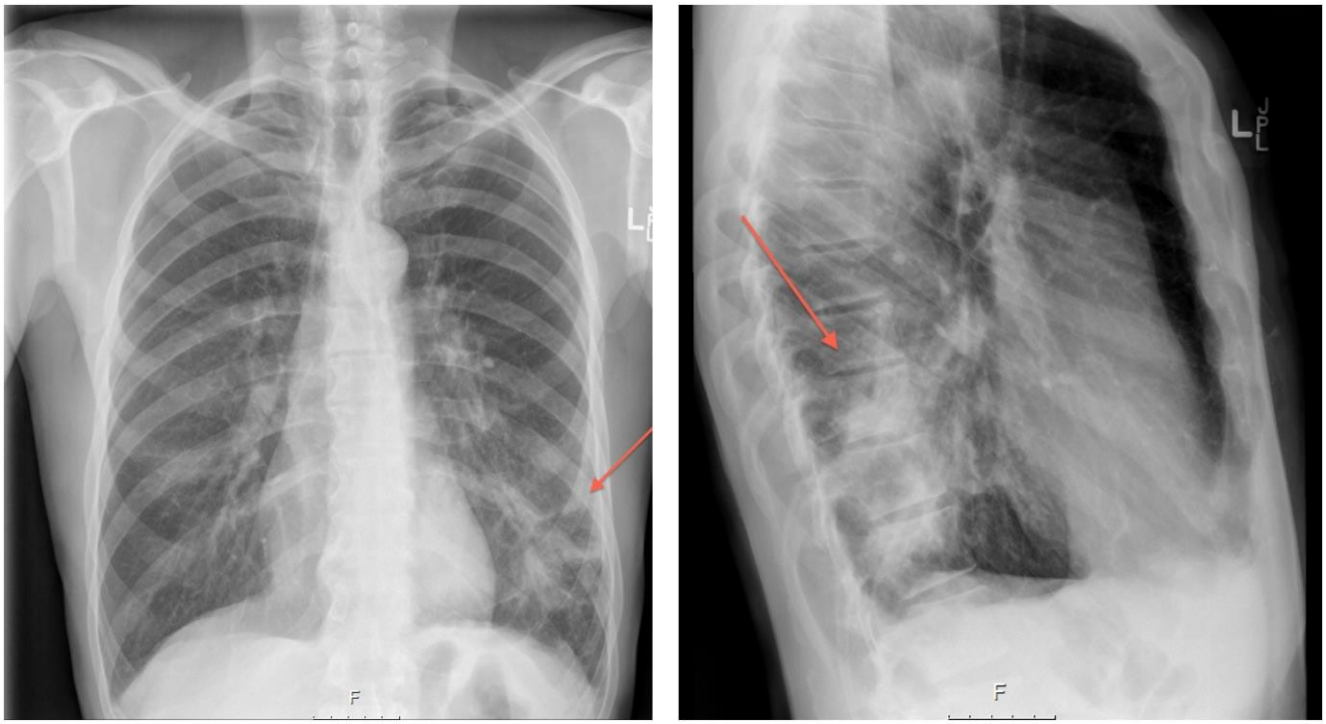


Figure 1: 60 year old male with left lower lobe pulmonary sequestration.
Findings: PA and lateral radiographs of the chest demonstrate a heterogeneous opacity (arrows) in the posterobasal segment of the left lower lobe which may represent pneumonia.

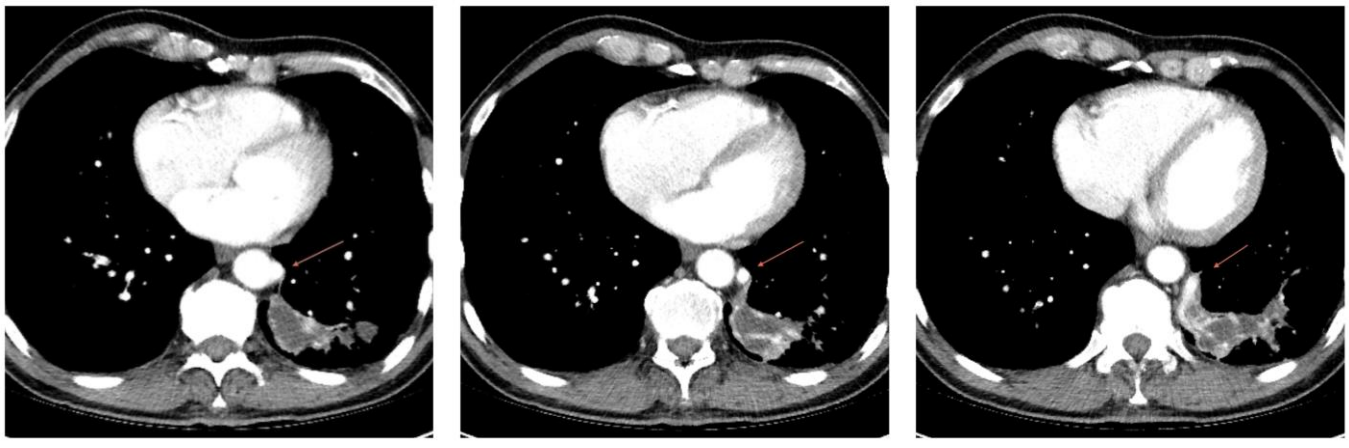


Figure 2: 60 year old male with left lower lobe pulmonary sequestration.
Findings: Axial contrast enhanced CT of the chest demonstrates a heterogeneous mass in the posterobasal segment of left lower lobe and abnormal branch originating from the descending thoracic aorta (arrows) supplying the left lower lobe mass indicating the presence of a pulmonary sequestration.
Technique: Axial CT, 641 mA, 100 kV, 5mm slice thickness, 100 mL Omnipaque.

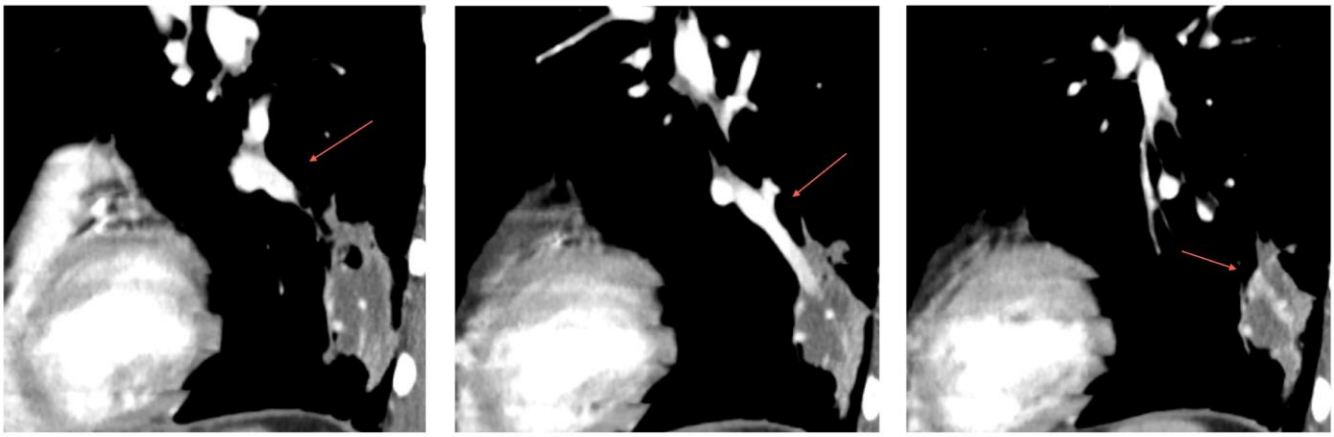


Figure 3: 60 year old male with left lower lobe pulmonary sequestration.
Findings: Sagittal reconstructed contrast-enhanced sequential CT images of the chest demonstrate the abnormal branch originating from the descending thoracic aorta (arrows) coursing to the sequestered portion of the lung in the left lower lobe.
Technique: Sagittal reconstructed CT, 641 mA, 100 kV, 5mm slice thickness, 100 mL Omnipaque.

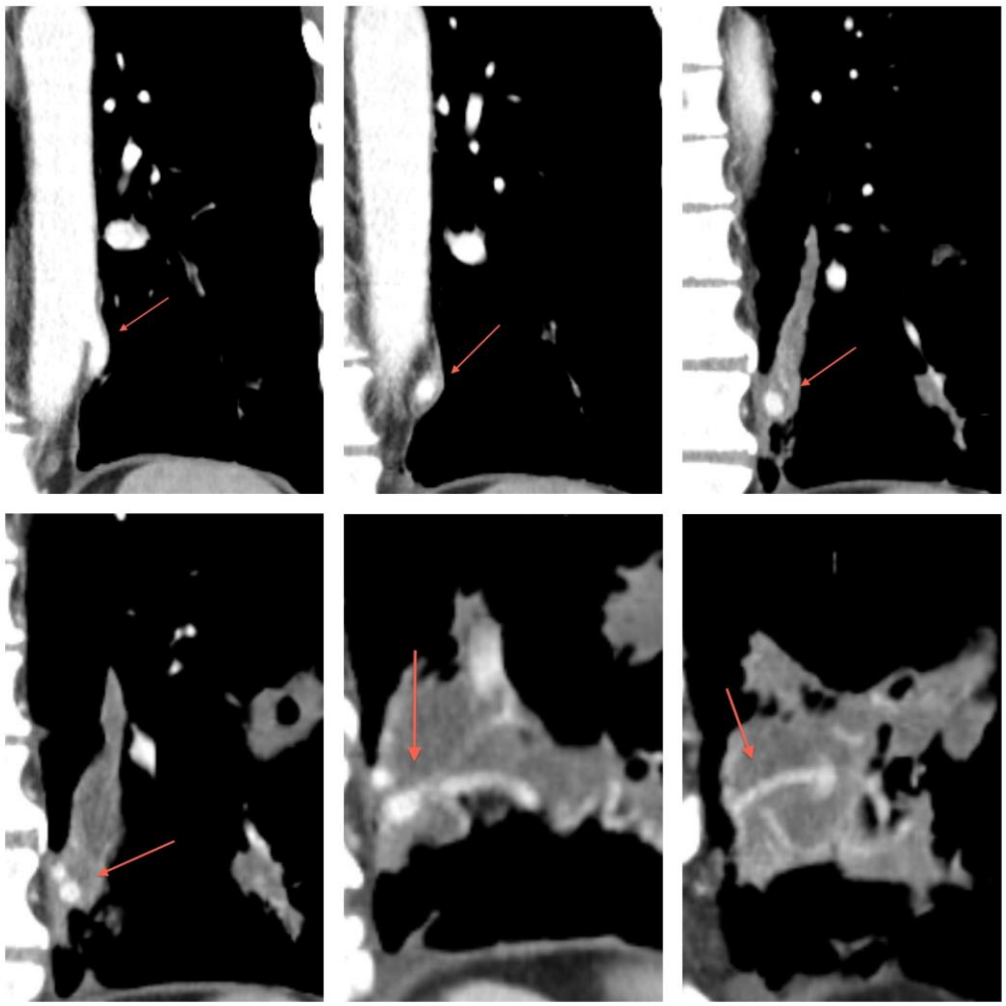


Figure 4: 60 year old male with left lower lobe pulmonary sequestration.
Findings: Coronal reconstructed contrast-enhanced sequential CT images focused on the left lower lung field demonstrate an abnormal branch (arrows) off the descending thoracic aorta supplying the left lower lobe lung sequestration.
Technique: Coronal reconstructed CT, 641 mA, 100 kV, 5mm slice thickness, 100 mL Omnipaque.

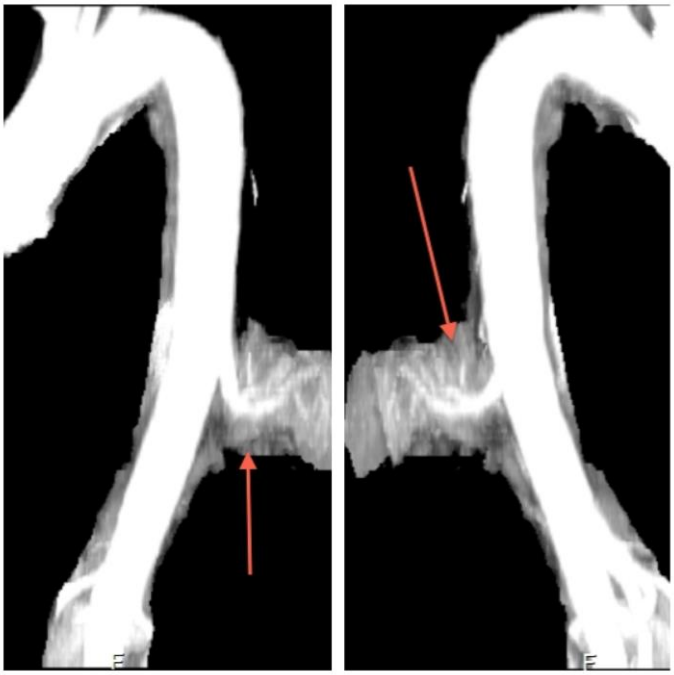
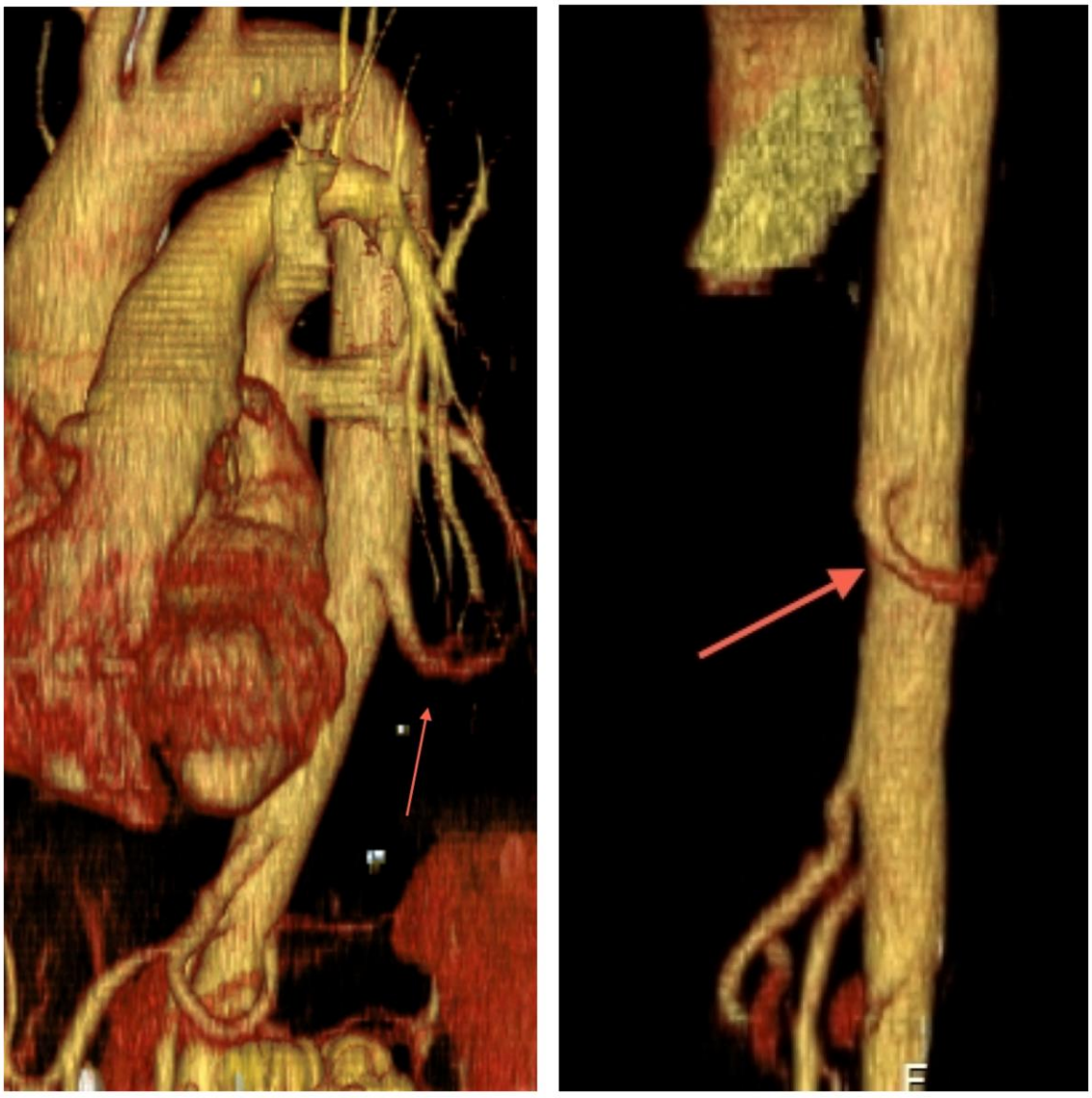


Figure 5 (left): 60 year old male with left lower lobe pulmonary sequestration.
Findings: MIP reconstructed images, left anterior oblique (a) and posterior oblique (b), clearly show the abnormal vessel (arrows) arising from the thoracic aorta.
Technique: Maximum intensity projection (MIP)

Figure 6 (bottom): 60 year old male with left lower lobe pulmonary sequestration.
Findings: VRT reconstructed images show nicely the orientation and location of the abnormal vessel (arrows) arising from the descending thoracic aorta and supplying the left lower lung.
Technique: Volume rendering technique (VRT)



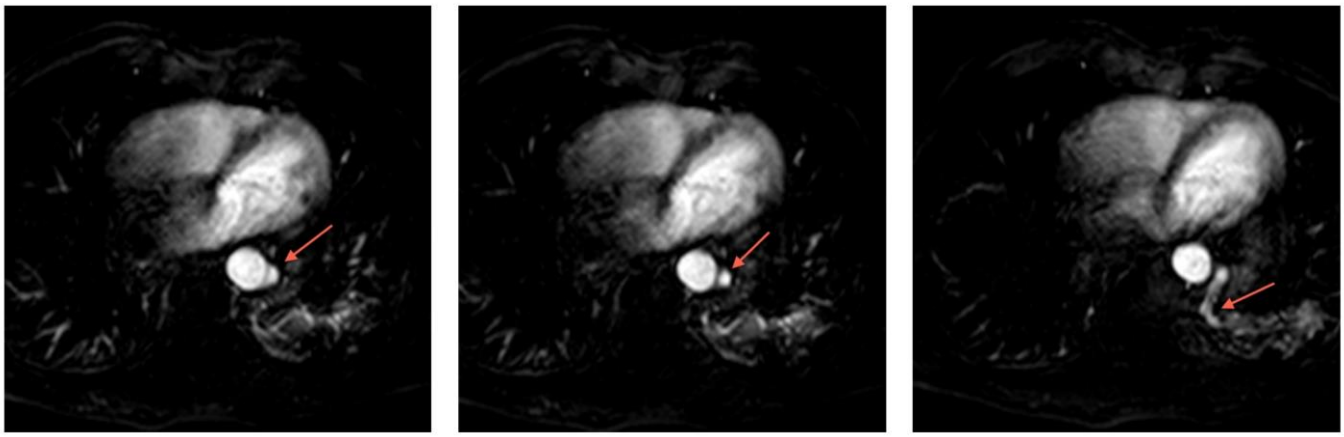


Figure 7: 60 year old male with left lower lobe pulmonary sequestration.
Findings: T1-post contrast axial MRI images demonstrate the abnormal vasculature (arrows) supplying the left lower lobe sequestration.
Technique: MRI axial T1 THRIVE sequence 20 seconds post-contrast, TR 3.91, TE 1.84, FOV 375 mm, SENSE-XL-Torso coil, Magnevist 20 mL.

Etiology	Congenital
Incidence	0.15-6.4% of all congenital pulmonary malformations.
Gender Ratio	1:1 male to female ratio
Age Predilection	Extralobar is more common in the neonatal period; Intralobar is more common in childhood.
Risk Factors	None
Treatment	Surgical resection in younger patients, conservative management in older or asymptomatic patients
Prognosis	Very good
Findings on imaging	Rounded mass in lung parenchyma with aberrant vessel from aorta providing blood supply.

Table 1: Summary table for pulmonary sequestration

	Pulmonary Sequestration	**Pneumonia**	**Tumor**	**Pulmonary Embolism**
X-ray	Dense well-defined mass in the lung parenchyma.	Dense mass with possible air bronchograms.	Variable; air-fluid levels, irregular borders, invasion.	Wedge-shaped opacity, "Hampton's hump" in the lung periphery.
Ultrasound	Echogenic homogenous mass with feeding artery seen on Color Doppler imaging originating from the aorta.	Hyperechoic mass in lung parenchyma with surrounding vasculature.	Increased vascularity on Color Doppler, irregular mass.	n/a
CT/CTA	Well-defined mass with possible cystic changes and surround emphysematous changes in the lung parenchyma. Aberrant vessel from aorta to the sequestered mass (CTA).	Dense consolidation with air-bronchograms, pleural effusion.	Ill-defined mass with heterogeneous enhancement with lymphadenopathy and mass effect.	Filling defect in one or many pulmonary arteries.
MRI/MRA	Flow voids to demonstrate aberrant artery and venous drainage (MRA).	T2 hyperintense edema surrounding consolidation.	Heterogeneously enhancing lesion with irregular borders and possible invasion.	Filling defect in one or many pulmonary arteries.
Nuclear scintigraphy	Nuclear angiography demonstrates the systemic arterial blood supply to the sequestered lung.	V/Q scan may show decreased blood flow in necrotic or infarcted lung.	Increased 18-FDG uptake in highly metabolic cancerous lesions.	Mismatch between ventilation and perfusion.

Table 2: Differential diagnosis table for pulmonary sequestration

ABBREVIATIONS

BPS = Bronchopulmonary sequestration
CT = Computed tomography
CTA = Computed tomography angiogram
ER = Emergency room
FDG = Fluoro deoxyglucose
LLL = Left lower lobe
MIP = Maximum intensity projection
MRA = Magnetic resonance angiography
MRI = Magnetic resonance imaging
PET = Positon emission tomography
V/Q = Ventilation-perfusion
VATS = Video assisted thorascopic surgery
VRT = Volume rendering technique

KEYWORDS

Intralobar; extralobar; sequestration; pulmonary; pneumonia

ACKNOWLEDGEMENTS

Kari Brugeman RT and Ramon Nunez