Radiology Case Reports 18 (2023) 4145–4148

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Intralobar pulmonary sequestration in 2 extremes of ages: A Case report ☆,☆☆

Krishnaraj Aryal, MBBS[a,*], *Pradeep Raj Regmi, MD*[a], *Gauri Adhikari, MBBS*[b], *Ruchi Karki, MBBS*[c], *Prajwal Dhakal, MD*[a]

[a] *Department of Radiology, Institute of Medicine, Tribhuvan University Teaching Hospital, Kathmandu, Nepal*
[b] *Department of Medicine, Nepalese Army Student of Health Science- College of Medicine, Kathmandu, Nepal*
[c] *Department of Medicine, KIST Medical College and Teaching Hospital, Imadol, Lalitpur, Nepal*

ARTICLE INFO

Article history:
Received 14 June 2023
Revised 23 July 2023
Accepted 19 August 2023

Keywords:
Child
Elderly
Imaging
Pulmonary
Sequestration
Congenital

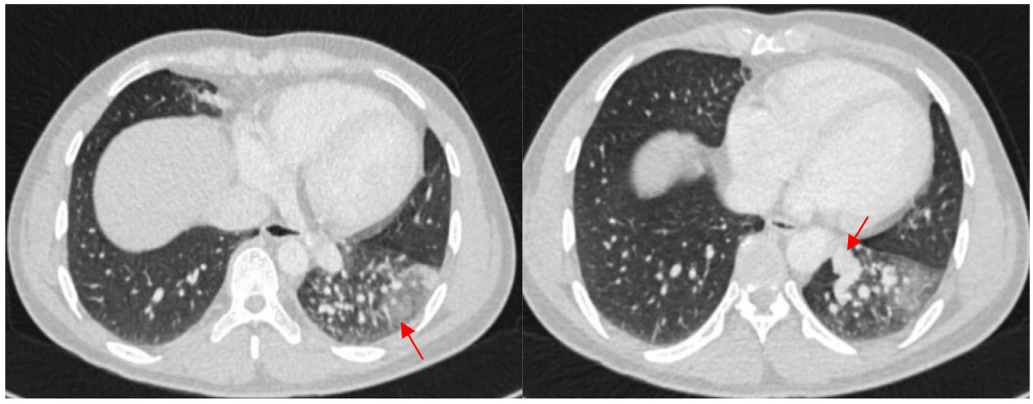
ABSTRACT

Pulmonary sequestration is a relatively rare phenomenon characterized by nonfunctional lung tissue supplied by one or more systemic arteries without direct connection to the tracheobronchial tree. Intra-lobar pulmonary sequestration comprises 75% of the total pulmonary sequestrations. Most patients with pulmonary sequestrations are often diagnosed with a childhood chest infection, so pulmonary sequestration is considered a childhood disease. However, few cases are found in adults and the elderly, with or without symptoms, and imaging findings on computed tomography (CT) or magnetic resonance imaging (MRI) are variable due to infection and inflammation. Failure to diagnose and treat this condition may lead to recurrent pneumonia and fatal hemoptysis. In this case report, we present cases of pulmonary sequestration at extremes of ages, one at 12 and the other at 65.

Introduction

Pulmonary sequestration (PS) is a rare congenital malformation [1] characterized by dysplastic and nonfunctional pulmonary tissue with 2 distinct features: First, it has no direct communication with the tracheobronchial tree. Second, it derives its blood supply from direct systemic circulation and not pulmonary circulation [2]. Therefore, identifying systemic arterial supply is important in diagnosing pulmonary sequestration. The definite diagnosis of PS is made by histopathologic examination after surgery [3]; however, PS can be confirmed by contrast-enhanced CT or MRI with or without angiography noninvasively by identifying a systemic arterial supply [4]. PS encompasses up to 0.15%-6% of broncho-pulmonary malformations and is classified as intralobar sequestration (ILS) and extra-lobar sequestration (ELS) based upon its relation to normal lung tissue. ILS is located within the visceral pleura

☆ Acknowledgments: The authors would like to express their sincere gratitude to the patients and their guardians for providing consent and additional information to write this report. We also thank all the clinical staff caring for the patients.
☆☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.
* Corresponding author.
E-mail address: aryal09kribin@gmail.com (K. Aryal).
https://doi.org/10.1016/j.radcr.2023.08.089



1 2

Fig. 1 & 2 – Axial HRCT images show ground glass opacities in the superior segment of the left lower lobe (marked by red arrow) with 1 tortuous vessel in the involved segment (red arrow).

and is surrounded by normal lung tissue, while extra lobar sequestration has a separate pleural covering [5]. Both sequestration types have a distinct blood vessel from the aorta or its branches. Venous drainage of ILS is usually via the pulmonary veins, while venous drainage of ELS is generally via systemic drainage.

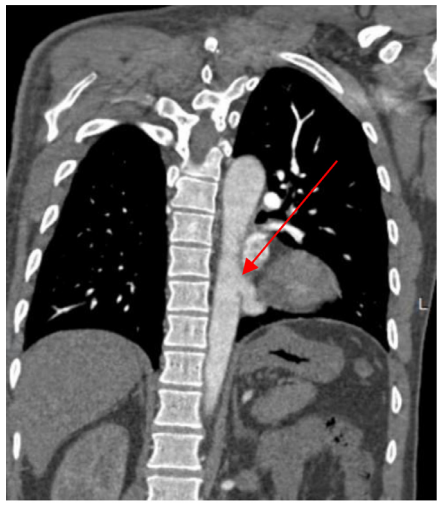
ILS accounts for 75% of pulmonary sequestrations [3]. Half of the patients with ILS are diagnosed before the age of 20 with symptoms of recurrent pulmonary infection or cardiac disease, whereas a small portion of patients are asymptomatic and diagnosed incidentally. It is rarely diagnosed after 40 years of age, and only a few cases report the initial diagnosis of this condition in patients over 50 years of age [6–8].

PS typically appears as a homogenous or heterogenous solid mass or consolidation on imaging. Less commonly, it appears as multiple cystic masses with air-fluid levels, cavitary lesions, or a single cystic mass [9]. Because of the inconsistent appearance of PS in imaging findings, identification of an aberrant systemic vascular supply to the pulmonary tissue is thus crucial in the diagnosis of PS, as the lesion has a separate blood supply than other pulmonary tissue, which is typically supplied by the pulmonary arteries [10].

Our case report presents a case of pulmonary sequestration in 2 extremes of ages, a 12-year-old young boy and a 65-year-old elderly male, based upon the contrast-enhanced multidetector computed tomography (CE-MDCT) image findings.

Case 1

A 12-year-old boy presented with a history of recurrent chest infections, cough, and chest pain. Laboratory examination showed marked leukocytosis and chest X-ray showed infiltration of the left lower lobe. CT was done because of his recurrent episodes, and it revealed pulmonary sequestration in the left lower lobe with an aberrant artery arising from the descending thoracic aorta supplying the superior segment of the left lower lobe. Ground glass opacities and patchy areas of consolidation were noted on the sequestrated segment (Figs. 1 & 2, 3). The boy was managed conservatively with antibiotics.

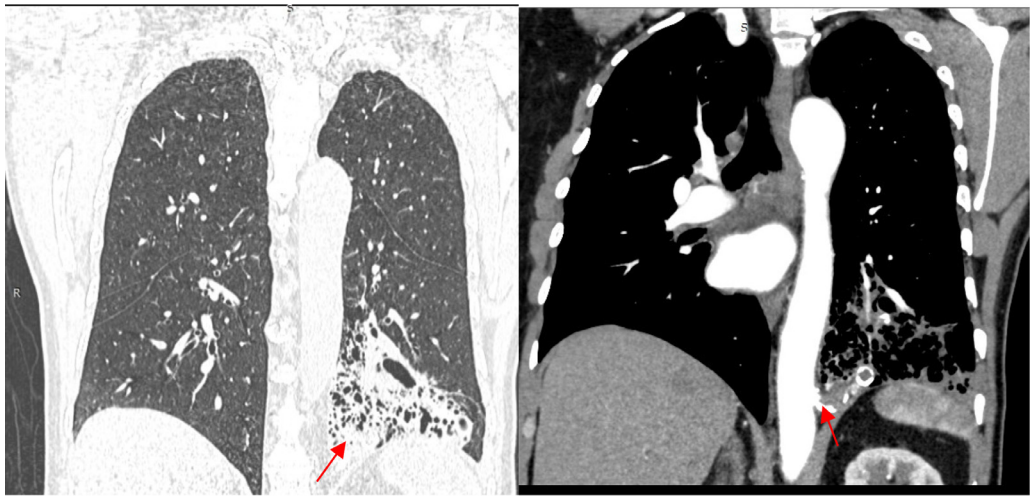


3

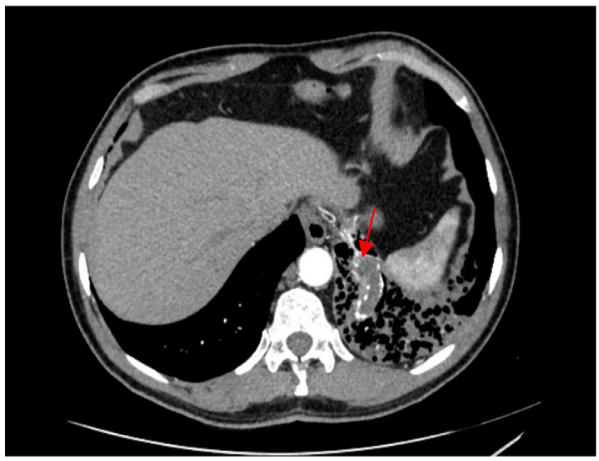
Fig. 3 – Coronal contrast-enhanced CT chest shows the aberrant artery arising from the descending thoracic aorta (shown by red arrow).

Case 2

A 65-year-old man with a history of recurrent pneumonia was referred to our department for imaging. He had a history of long-standing chronic cough with sputum. At the presentation, he complained of hemoptysis with a tinge of blood on his sputum. A chest X-ray revealed mild infiltration at the left lung base. CE-MDCT revealed an approximately 10*9*9 cm sized heterogenous lesion showing tubular and cystic air spaces scattered throughout and attenuating areas within the posteromedial aspect of the left lower lung. The lesion was supplied by a branch from the descending thoracic aorta arising from the D11-D12 level, and it was draining into the left inferior pulmonary vein (Fig. 4, Figs. 5 & 6). There was a chronic complete occlusive thrombus of the arterial supply of seques-



4 5



6

Fig. 4 – Coronal HRCT chest shows consolidation within the basal segments of the left lower lobe.
Fig. 5 & 6 - Coronal and axial contrast-enhanced CT chest show intralobar pulmonary sequestration supplied from the descending thoracic aorta with thrombus within the aberrant artery.

trated lung with sequelae of infarction in the form of tubular and cystic air-fluid filled spaces in the sequestrated lung. A bronchoscopy was also performed, which revealed bilateral tracheobronchitis. The case was managed conservatively and is kept on follow-up.

Discussion

ILS is a rare congenital anomaly [11]. Although it is a benign condition, it can be accompanied by serious complications such as recurrent respiratory infections, massive hemoptysis, and heart failure. Most cases are diagnosed in childhood; however, only a few cases are reported in adulthood and the elderly, and they can be asymptomatic otherwise. They commonly present with chronic cough, fever, pneumonia, and chest pain. Fatal complications include massive hemoptysis and superadded fungal infections like aspergillosis [12,13]. Therefore, the treatment for symptomatic patients with PS has always been surgical resection [14]. Surgical resection should be considered even in asymptomatic patients with PS [14].

Most ILSs are in the left lung's medial and posterior basal segments. Usually, they occur in the lower lobes [2]. Bilateral involvement is uncommon. The lesions of ILS may be solid, cystic, hemorrhagic, or even contain mucus. Cystic or emphysematous elements may be present, and adjacent atelectasis often exists [11].

Radiologically, PS typically appears on CT as a solid mass or consolidation with homogenous or heterogenous enhancement. Less frequently, features of PS include a collection of small cystic lesions, a sizeable cavitary lesion, or a well-defined cystic mass. Cystic lesions are often multi-cystic and

may have an air-fluid level caused by fistulous bronchial communication [4]. The lesions appear hypervascular because of the abundant systemic circulation. Super-added infection may lead to consolidation of the adjacent segments, and a chronic inflammatory process may induce localized reactive neovascularization. Emphysematous changes or hyperlucency at the margin of the lesion are characteristic findings of PS resulting from collateral air drift and air trapping due to the absence of a normal bronchial connection [3,15]. Associated congenital anomalies are rare with pulmonary sequestration [16].

As the radiological findings of PS are variable, it is vital to identify the aberrant systemic circulation that supplies the pulmonary tissue. The systemic artery supplying the PS usually arises from the lower thoracic aorta. However, it can also arise from the upper abdominal aorta and rarely from other systemic arteries such as the internal mammary artery, celiac artery, splenic artery, or coronary artery [4,11]. The vascular supply is from the descending thoracic artery in both cases.

Diagnosing a case of pulmonary sequestration is challenging as it may present variable symptoms, and imaging is often indeterminate. The most important aspect of diagnosing pulmonary sequestration is recognizing systemic arterial supply. Since the definitive step in the diagnosis of sequestration is the demonstration of the systemic arterial supply, for a long time, the diagnosis was made by conventional angiography [16]. More recently, all imaging techniques capable of showing the artery have been implicated in evaluating sequestration. MR imaging and MR angiography can also be used together to diagnose pulmonary sequestration in a single noninvasive examination and without radiation dose, however, with high scan time. However, MR cannot accurately assess lung parenchyma and the airways, and it might not be cost-effective for all in our part of the world [17]. Other noninvasive techniques for the evaluation of sequestration, such as scintigraphy, are only rarely necessary. In the cases described in this report. CE-MDCT successfully delineated the origin and course of the anomalous systemic artery. Also, venous drainage into the pulmonary veins was identified in our case.

Conclusion

Pulmonary sequestration, whether intralobar or extra-lobar, is rare, accounting for less than 6% of all congenital lung abnormalities. In this report, we presented 2 cases: one with a history of recurrent chest infections and the other with a history of hemoptysis. We assessed the pulmonary parenchyma and abnormal systemic vessels using a single imaging technique and successfully diagnosed the cases.

Imaging studies are crucial in diagnosing and managing patients with pulmonary sequestration. This is particularly important due to the potential to cause recurrent infections and life-threatening hemoptysis.

Patient consent

The authors claim that there is no personal information in this report that might be used to identify the patients. Written, informed consent was obtained from one of the patients, and written consent was obtained from the parent of the other patient.

REFERENCES

[1] Polaczek M, Baranska I, Szolkowska M, Zych J, Rudzinski P, Szopinski J, et al. Clinical presentation and characteristics of 25 adult cases of pulmonary sequestration. J Thorac Dis 2017;9(3):762–7.

[2] Savic B, Birtel FJ, Tholen W, Funke HD, Knoche R. Lung sequestration: report of seven cases and review of 540 published cases. Thorax 1979;34(1):96–101.

[3] Frazier AA, Rosado de Christenson ML, Stocker JT, Templeton PA. Intralobar sequestration: radiologic-pathologic correlation. Radiogr Rev Publ Radiol Soc N Am Inc 1997;17(3):725–45.

[4] Bolca N, Topal U, Bayram S. Bronchopulmonary sequestration: radiologic findings. Eur J Radiol 2004;52(2):185–91.

[5] Makhija Z, Moir CR, Allen MS, Cassivi SD, Deschamps C, Nichols FC, et al. Surgical management of congenital cystic lung malformations in older patients. Ann Thorac Surg 2011;91(5):1568–73 discussion 1573.

[6] Nicolette LA, Kosloske AM, Bartow SA, Murphy S. Intralobar pulmonary sequestration: a clinical and pathological spectrum. J Pediatr Surg 1993;28(6):802–5.

[7] Alsumrain M, Ryu JH. Pulmonary sequestration in adults: a retrospective review of resected and unresected cases. BMC Pulm Med 2018;18(1):97.

[8] 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):2086–90.

[9] Walker CM, Wu CC, Gilman MD, Godwin JD, Shepard JAO, Abbott GF. The imaging spectrum of bronchopulmonary sequestration. Curr Probl Diagn Radiol 2014;43(3):100–14.

[10] Amitai M, Konen E, Rozenman J, Gerniak A. Preoperative evaluation of pulmonary sequestration by helical CT angiography. AJR Am J Roentgenol 1996;167(4):1069–70.

[11] Qays Intralobar pulmonary sequestration in elderly woman: a rare case report with emphasis on imaging findings. NCBI; 2022.

[12] Rubin EM, Garcia H, Horowitz MD, Guerra JJ. Fatal massive hemoptysis secondary to intralobar sequestration. Chest 1994;106(3):954–5.

[13] Morikawa H, Tanaka T, Hamaji M, Ueno Y. A case of aspergillosis associated with intralobar pulmonary sequestration. Asian Cardiovasc Thorac Ann 2011;19(1):66–8.

[14] Liu C, Pu Q, Ma L, Mei J, Xiao Z, Liao H, et al. Video-assisted thoracic surgery for pulmonary sequestration compared with posterolateral thoracotomy. J Thorac Cardiovasc Surg 2013;146(3):557–61.

[15] Al-Timimy QAH, Al-Shamseei HF. Intralobar pulmonary sequestration in elderly woman: a rare case report with emphasis on imaging findings. Radiol Case Rep 2016;11(3):144–7.

[16] Abbey P, Das CJ, Pangtey GS, Seith A, Dutta R, Kumar A. Imaging in bronchopulmonary sequestration. J Med Imaging Radiat Oncol 2009;53(1):22–31.

[17] Kauczor HU, Knopp MV, Branscheid D, Semmler W. [Pulmonary sequestration: use of MR-angiography]. Aktuelle Radiol 1993;3(2):120–2.