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Radiology Corner Case #8

Intralobar Bronchopulmonary Sequestration

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Introduction

Intralobar bronchopulmonary sequestration is a relatively uncommon pulmonary disorder that is characterized by non-functioning lung tissue that lacks normal connection with the tracheobronchial tree and has a systemic arterial blood supply. Computed tomography and magnetic resonance imaging can provide the necessary information required for diagnosis and pre-operative planning. The following case report reviews the typical clinical presentation and imaging findings in a young adult with intralobar bronchopulmonary sequestration.

History

An otherwise healthy 18-year-old man presented for a routine physical examination, which included frontal (Figure 1A) and lateral (Figure 1B) chest radiographs. The patient denied any respiratory or cardiovascular complaints or any history of cardiopulmonary or gastrointestinal disease. The patient was afebrile and had an unremarkable physical exam. Contrast-enhanced thoracic CT (Figures 1C and 1D) was ordered for further evaluation.

Imaging Findings

Chest radiographs (Figure 1A, frontal; and Figure 1B, lateral) revealed a large 12 cm (superior-inferior) x 5 cm (right-left) x 5 cm (anterior-posterior) right lower lobe soft tissue mass. The mass had a smooth contour and was not associated with calcification or air-fluid levels. CT examination (Figure 1C) confirmed the features identified on chest radiographs and the mass was noted to have a homogeneous fluid density, with a smooth contour and clearly defined margins with adjacent lung parenchyma.

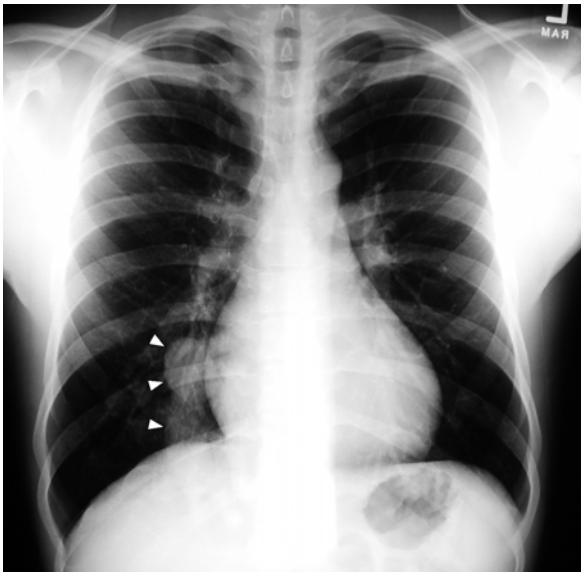

Figure 1A

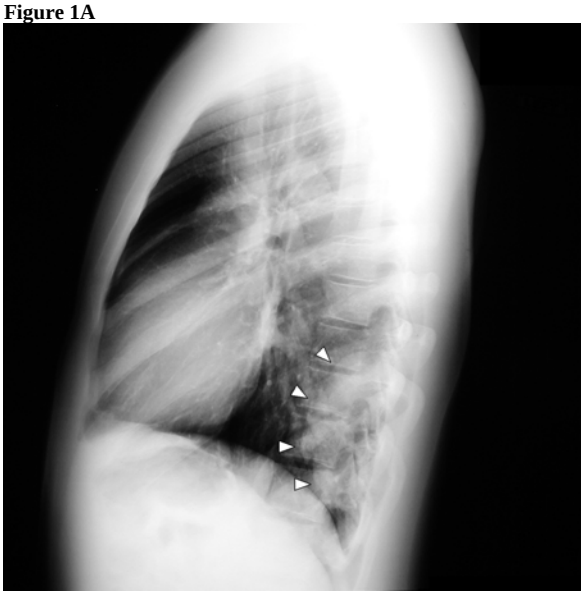

Figure 1B

Figure 1A and 1B. On the frontal (A), a large soft tissue mass (arrowheads) is noted in the medial right lower lung field. On the lateral view (B), the mass (arrowheads) is noted to be within the right lower lobe.

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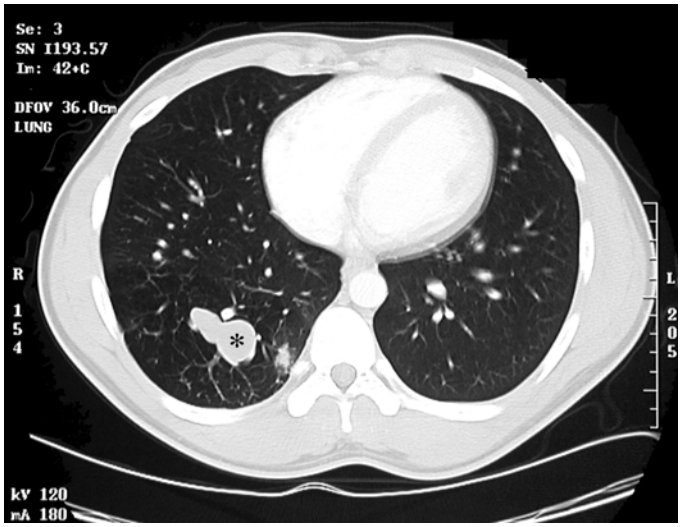

Figure 1C

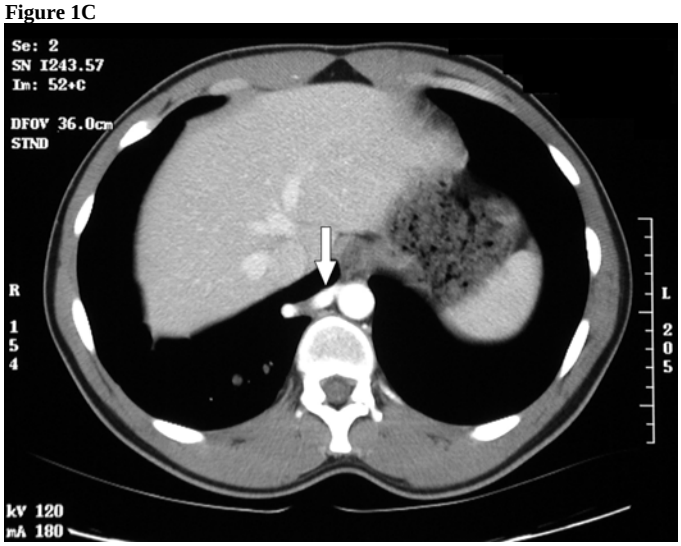

Figure 1D

Figure 1C and 1D. Contrast-enhanced CT (C, lung windows) demonstrates a well-circumscribed right lower lobe mass (*) with a lateral finger-like projection. The mass is surrounded by normal lung parenchyma. On more caudal images (D, soft tissue windows), a feeding vessel (arrow) from the aorta is identified but no anomalous pulmonary veins were noted, suggesting the diagnosis of intralobar bronchopulmonary sequestration.

Lower in the thorax, an anomalous artery was noted emanating rightward from the aorta at the thoracoabdominal junction (Figure 1D). Of note, no definite anomalous pulmonary venous drainage was identified. The presumptive diagnosis was intralobar bronchopulmonary sequestration.

Magnetic resonance angiography (MRA) was ordered to better delineate the arterial and venous anatomy for pre-operative planning. On pre-contrast T1-weighted spoiled gradient echo images (Figure 1E), the right lower lobe mass was noted to be homogeneously hyperintense. The margins of the mass, moreover, were well-circumscribed with smooth and "finger-like" radiating projections, especially along its inferior margin. On T2-weighted fast spin echo (not shown) the mass was of intermediate signal intensity. Multi-phase coronal breath-hold Gadolinium (Gd)-enhanced three dimensional (3D) MRA was performed using a 0.2 mmol/kg dose of Gd-chelate contrast agent administered intravenously at a rate of 2mL/sec and followed by a 20 mL saline flush injected at the same rate. On Gd-enhanced 3D MRA (Figure 1F and 1G), a solitary anomalous artery was identified arising from the lateral aspect of the juxta-diaphragmatic abdominal aorta and traversing superolaterally, through the medial aspect of the right hemidiaphragm, into the right lower lobe. Normal venous drainage of the mass into the inferior pulmonary vein was also identified. There was no evidence of a separate anomalous draining vein to the inferior vena cava, azygous vein or hemiazygos vein. The parenchymal mass itself was not noted to enhance following contrast administration. The features of a systemic arterial supply and normal pulmonary venous drainage were consistent with intralobar sequestration.

Based on concerns for potential recurrent pulmonary infections, a right lower lobe resection was performed. At thoracotomy, the large anomalous artery originating from the lateral wall of the abdominal aorta was identified and ligated. The inferior pulmonary vein was also identified and transected. A right lower lobectomy was performed successfully without complication. Pathologic examination of the resected right lower lobe confirmed the presence of a large mucous plug and the diagnosis of intralobar sequestration, which was also noted histologically to be associated with elements of acute and chronic inflammation with fibrosis.

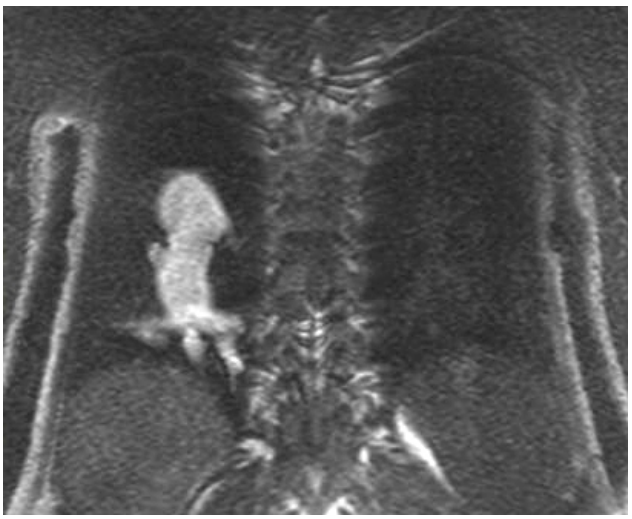

Figure 1E

Figure 1E. Coronal T1-weighted spoiled gradient echo image demonstrates a large irregular but smooth contoured mass in the right lower thorax. The mass has high signal intensity and finger like projections which are consistent with mucous plugs, a common feature in bronchopulmonary sequestration.

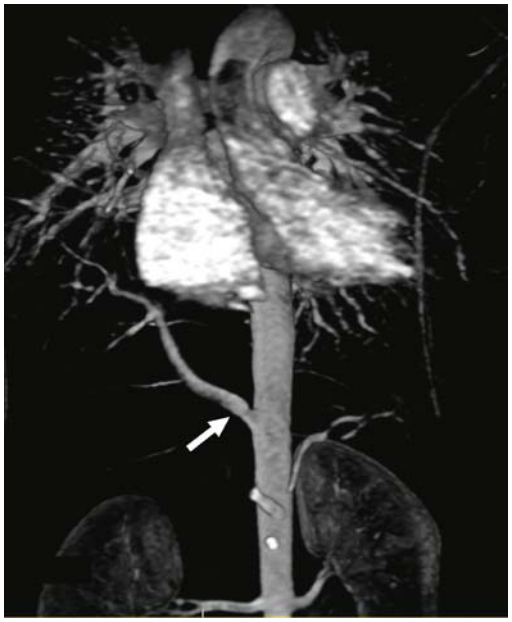

Figure 1F

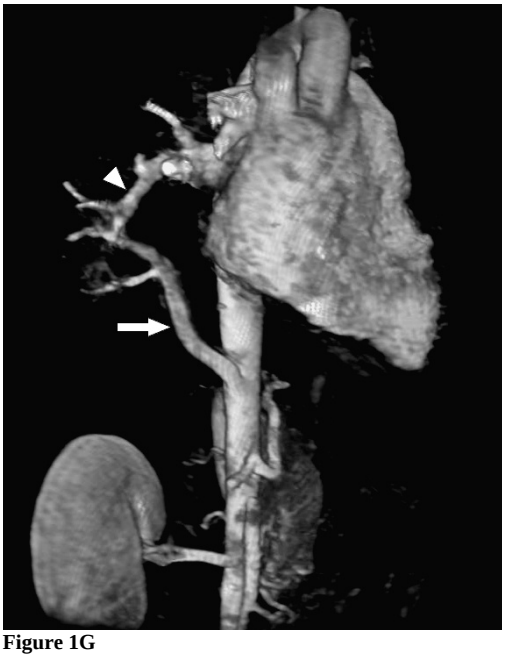

Figure 1G

Figure 1F and 1G. Coronal (F) and oblique (G) volume rendered reconstructions of a Gd-enhanced 3D MRA demonstrate a large feeding artery (arrow) arising from the thoracoabdominal aorta and normal pulmonary venous drainage of the mass via the inferior pulmonary vein (G, arrowhead).

Discussion

Bronchopulmonary sequestration is a pulmonary anomaly of non-functioning bronchopulmonary tissue that lacks a normal bronchial connection and is characterized by a systemic arterial blood supply.[1-6] Bronchopulmonary sequestrations are categorized as intralobar and extralobar. Approximately 75% of bronchopulmonary sequestrations are intralobar.[7] Intralobar sequestrations typically have normal pulmonary venous return and share the visceral pleura of their parent lobe. Extralobar sequestration, on the other hand, is contained within its own distinct visceral pleura and associated with aberrant systemic pulmonary venous drainage. The distinctive features for the two varieties of sequestration are summarized in Table 1.[4, 7-11] Rarely, both have been reported to co-exist.[7, 12]

Intralobar sequestrations are almost always located in the lower lobes (98%), and slightly more commonly (60%) on the left side.[8] Upper lobe or bilateral involvement is rare. Patients with an intralobar sequestration typically present before the age of 20 years with histories of recurrent pulmonary infections.[6, 8] Occasionally, as in our case, patients may be asymptomatic. Most intralobar sequestrations are acquired lesions and their association with congenital anomalies is uncommon.[8, 9]

Table 1. Pulmonary Sequestration[1-10, 18, 20, 21]

Characteristic	Intralobar	Extralobar
Pleura Investment	Shares visceral pleura of parent lobe	Separate visceral pleura
Location	Posterior basal segments (Approx. 60% on left)	Above, below or within diaphragm (Approx. 90% on left)
Gender Incidence	Equal	Greater in males (4:1)
Arterial Supply	Systemic	Systemic
Venous Drainage	Pulmonary venous	Systemic venous (IVC, azygos, hemiazygos, portal vein)
Foregut Communication	Very rare	More common
Associated Anomalies	Uncommon	Common
Presentation	Usually found in early adulthood and is associated with a history of hemoptysis, pulmonary infection, chronic cough, chest pain, or asthma. Asymptomatic mass is less common (15%)	Approximately 60 % found during first 6 months of life due to respiratory or feeding problems. Often found in conjunction with another anomaly. Only about 10% are asymptomatic.
Pathogenesis Theory	Controversial. Thought to be usually acquired.	Congenital
Histological Features	Extensive fibrosis and chronic inflammation, may have cysts	Normal lung parenchyma with dilated structures
Radiographic Features	Homogeneous consolidation with irregular margins or uniformly dense mass with smooth or lobulated contours.	Single well defined, homogeneous, triangular shaped opacity in the lower thorax. Mass may present else where in the thoracic cavity.

Extralobar sequestration is thought to be primarily a congenital anomaly and is thus often associated with other anomalies such as diaphragmatic hernia, complex congenital heart disease, adenomatoid malformation type II, and gastrointestinal tract anomalies, which probably accounts for its presentation during infancy or early childhood.[9, 11, 13] Contained within its own pleura, extralobar sequestration has also been called accessory lung.[11, 13]

The typical radiographic appearance of a bronchopulmonary sequestration is that of a soft-tissue or cystic mass in the lower lung in association with a systemic arterial feeder.[4, 6, 9, 10, 14, 15] As in our case, the mass is well circumscribed and has a homogeneous appearance on CT and

MRI. Furthermore, the blind bronchi within the sequestered lung may become distended with trapped mucous secreted by bronchial glands. This can result in mucous plugs and mucocele formation,[14] as seen in our patient (Figure 1E). On occasion, air may enter a sequestration via the pores of Kohn or through small parenchymal or bronchial leaks as a complication of an infection.[3, 6] An important key to the diagnosis of bronchopulmonary sequestration as in our case was the finding of an anomalous systemic arterial supply (Figures 1D, 1F and 1G). In most instances (90%), the systemic arterial supply is from thoracic aorta (69%) or abdominal aorta (21%).[7]

Aside from the differences in their clinical presentation (i.e. age, sex, symptoms), bronchopulmonary sequestrations can be distinguished by their location, venous return and associated features. Table 1 details some of the more common differences. In our case, the age of the patient, lack of associated congenital lesions as well as the radiological findings of a normal pulmonary venous drainage and a more central pulmonary location (not abutting the hemidiaphragm) strongly favored the diagnosis of intralobar sequestration.

The radiologic differential diagnosis of pulmonary sequestration is extensive, but the main considerations include bronchial atresia, cystic adenomatoid malformation, lobar emphysema, intrapulmonary bronchogenic cyst, bronchiectasis, pneumonia, abscess, arteriovenous fistula or malformation, and systemic arterial supply to nonsequestered lung.[4, 6, 9] Identifying anomalous systemic arterial supply to the pulmonary abnormality helps limit the differential diagnosis, as does attention to the parenchymal abnormalities.[14-16]

Classically, the diagnosis of pulmonary sequestration hinges on the identification of an anomalous arterial feeding vessel from the aorta.[6, 15, 17] Although CT and CTA[5, 16] continue to be the primary means for diagnosing bronchopulmonary sequestrations, the use of MR and Gd-enhanced MRA can serve as a suitable alternative, especially in patients in which the use of iodinated contrast agents is contraindicated such as those with known allergy to iodinated contrast agents or renal insufficiency.[15, 17] MRI has the inherent clinical benefits (i.e. no ionizing radiation exposure and no requirement for the use of iodinated contrast media with its attendant risks for anaphylaxis and nephrotoxicity). But more importantly, MRI can acquire images in oblique planes, which may be critical for the determination of lower thoracic versus upper abdominal origin of the feeding vessel(s) from the aorta—an important pre-operative determination. MRI also provides the additional opportunity to perform repeated imaging and/or oblique imaging without the concerns related to ionizing radiation exposure that would be encountered with repeated CT evaluation.

Intralobar pulmonary sequestration should be considered in any adolescent or young adult with repeated lower lobe infections, and/or persistent lower lobe abnormalities on chest radiograph. The identification of arterial and venous connections is of major importance, not only for the diagnosis, but also for pre-operative planning to prevent intra-operative hemorrhage that may result from damage to an unidentified anomalous vessel. As shown in our patient, the use of Gd-enhanced 3D MRA can obviate the need for conventional angiography.

References

1. Pryce DM. Lower accessory pulmonary artery with intralobar sequestration of lung: Report of 7 cases. J Pathol Bacteriol 1946; 58:457-67.
2. Sade RM, Clouse M. The Spectrum of Pulmonary Sequestration. Ann Thorac Surg 1974; 18:644-58.
3. Felson B. Chest Roentgenology. Philadelphia: WB Saunders Company, 1973.
4. Felker RE, Tonkin IL. Imaging of pulmonary sequestration. AJR Am J Roentgenol 1990; 154:241-249.
5. Ko SF, Ng SH, Lee TY, Wan YL, Liang CD, Lin JW, Chen WJ, Hsieh MJ. Noninvasive Imaging of Bronchopulmonary Sequestration. AJR Am J Roentgenol 2000; 175:1005-12.
6. Fraser RS, Muller NL, Colman N, Pare PD. Fraser and Pare's Diagnosis of Diseases of the Chest, 4th ed, Vol I. Philadelphia: WB Saunders Company, 1999.
7. Savic B, Birtel FJ, Tholen W, Funke HD, Knoche R. Lung sequestration: Report of seven cases and review of 540 published cases. Thorax 1979; 34:96-101.
8. Frazier AA, Christenson ML, Stocker JT, Templeton PA. Intralobar sequestration: Radiologic-pathologic correlation. Radiographics 1997; 17:725-45.
9. Panicek DM, Heitzman ER, Randall PA, Groskin SA, Chew FS, Lane EJ, Markarian B. The continuum of pulmonary developmental anomalies. Radiographics 1987; 7:747-59.
10. Rosado-de-Christenson ML, Frazier AA, Stocker JT, Templeton PA. Extralobar sequestration: Radiologic-pathologic correlation. Radiographics 1993; 13:425-41.
11. Stocker JT. Sequestrations of the lung. Semin Diagn Pathol 1986; 3:106-121.
12. Kergin FG. Congenital cystic disease of lung associated with anomalous arteries. J Thoracic Surg 1952; 23:55.
13. Stocker JT, Kagan-Hallet K. Extralobar pulmonary sequestration: Analysis of 15 cases. AJCP 1979;72(6):917-925.
14. Naidich DP, Rummancik WM, Lefleur RS, Estioko MR, Brown SM. Intralobar pulmonary sequestration: MR evaluation. J Comput Assist Tomogr 1987; 11:531-3.
15. Au VWK, Chan JKF, Chan FL. Case Report: Pulmonary sequestration diagnosed by contrast enhanced three-dimensional MR angiography. Br J Radiol 1999; 72:709-11.
16. Ikezoe J, Murayama S, Godwin JD, Done SL, Verschakelen JA. Bronchopulmonary Sequestration: CT Assessment. Radiology 1990; 176:375-9.
17. Naidich DP, Rumancik WM, Ettenger NA, Feiner HD, Harnanz-Schulman M, Spatz EM, Todar ST, Genieser NB. Congenital Anomalies of the Lungs in Adults: MR Diagnosis. AJR Am J Roentgenol 1988; 151:13-9.