

Pulmonary Vascular Anomaly

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Case Presentation

A 32-year-old man presented with a two-year history of chronic cough and episodes of hemoptysis. He had been previously managed as having pulmonary tuberculosis. A chest CT was performed at another institution, which revealed a vascular malformation. CT angiography of the pulmonary arteries and aorta (**Fig.**) was advised and subsequently performed at our center.

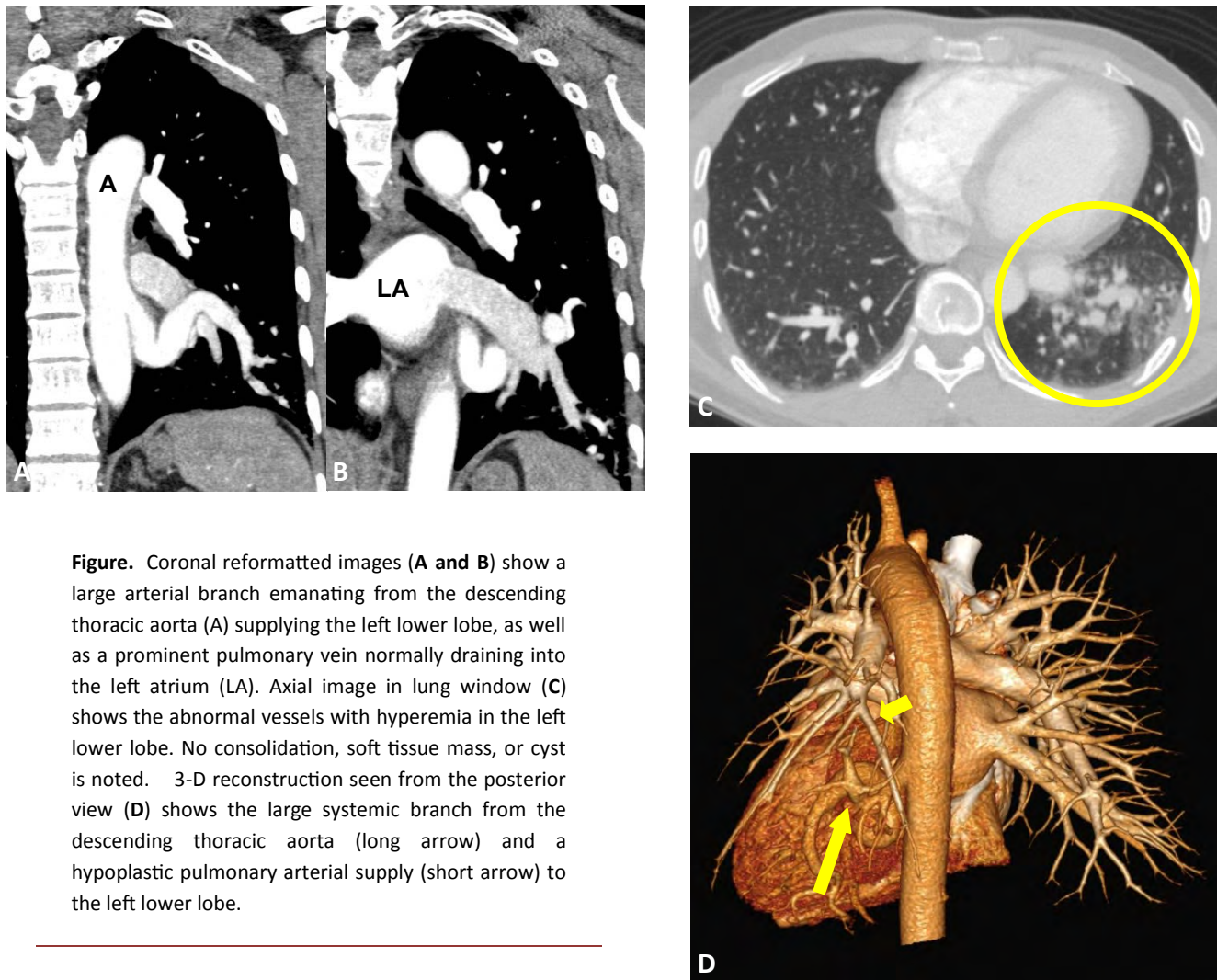


Figure. Coronal reformatted images (**A and B**) show a large arterial branch emanating from the descending thoracic aorta (**A**) supplying the left lower lobe, as well as a prominent pulmonary vein normally draining into the left atrium (**LA**). Axial image in lung window (**C**) shows the abnormal vessels with hyperemia in the left lower lobe. No consolidation, soft tissue mass, or cyst is noted. 3-D reconstruction seen from the posterior view (**D**) shows the large systemic branch from the descending thoracic aorta (long arrow) and a hypoplastic pulmonary arterial supply (short arrow) to the left lower lobe.

Key Clinical Finding

Chronic cough with hemoptysis

Key Imaging Findings

Pulmonary vascular anomaly

Systemic arterial supply to the left lower lobe with prominent pulmonary venous drainage

Differential Diagnoses

Pulmonary arteriovenous malformation

Pulmonary varix

Pulmonary sequestration

Pulmonary pseudosequestration

Discussion

Congenital bronchopulmonary foregut anomalies may involve the lung parenchyma, airways, and/or vascular arterial supply and venous drainage.^{1,2} These are included in a spectrum, which ranges from abnormal lung parenchyma with normal vasculature to abnormal vasculature with normal lung parenchyma. In between are lesions with mixed parenchymal and vascular abnormalities.^{3,4,5}

Although chest radiographs play a role in the incidental detection and initial imaging evaluation for such lung lesions,⁵ CT is very useful in confirming the presence of a lesion, determining its extent, defining associated abnormalities,¹ and as pre-operative evaluation for surgical cases.⁵ 3-D and multiplanar reformations can be particularly helpful in delineating abnormalities of the bronchi and arterial and venous vasculature.¹

Pulmonary Arteriovenous Malformation.

Pulmonary arteriovenous malformations (AVMs) usually present with dyspnea, hemoptysis, cyanosis, or clubbing; they may also be asymptomatic and found incidentally. These lesions are caused by abnormal communication between the pulmonary arteries and veins and occur most frequently in the lower lobes.^{1,2,5} CT may demonstrate more complex appearances, such

as a plexiform mass of dilated vascular channels or with dilated, tortuous direct communication between an anomalous feeding artery and draining vein (nidus). AVMs may be solitary or multiple. Multiple lesions are often associated with hereditary hemorrhagic telangiectasia (HHT), also known as Osler-Weber-Rendu syndrome.^{1,5} An AVM is excluded in our patient because of the presence of a systemic arterial supply to the left lower lobe and the lack of a definite direct communication between the pulmonary artery and vein.

Pulmonary Varix.

Pulmonary varix refers to an enlargement of a segment of a pulmonary vein without an enlarged feeding artery or nidus. This is typically seen near the left atrium with contiguity with the pulmonary vein. Varices may be congenital or acquired. Patients are usually asymptomatic and generally not treated, but may also present with hemoptysis; hence, surgery may be required.⁵ Although the left inferior pulmonary vein is prominent in our patient, the presence of a large systemic arterial supply makes this diagnosis unlikely.

Pulmonary Sequestration.

Pulmonary sequestration is characterized by dysplastic, nonfunctioning lung parenchyma that does not communicate with the tracheobronchial tree and has an anomalous systemic arterial supply,^{1,2,3,4,5} usually from the thoracic or abdominal aorta; arterial supply from the celiac, splenic, intercostal, subclavian, or even coronary arteries is less common.^{3,4} A sequestration may appear as a persistent opacity or mass. It may be associated with congenital pulmonary airway malformation (CPAM), in which case air may be present within the lesion. Lesions may also contain air when infected. The most common location is within the left lower lobe.^{1,2,4} Sequestrations can be intralobar (within visceral pleura and venous drainage via the inferior pulmonary vein) or extralobar (separate pleural covering with venous drainage usually via systemic veins, typically the azygous vein and less commonly via the portal, left subclavian, or internal mammary veins).^{1,2,3,4,5} The imaging findings in our patient are very similar to a pulmonary sequestration, except that the involved lung only shows hyperemia with a normal tracheobronchial tree.

Pulmonary Pseudosequestration.

Pseudosequestration is within the sequestration spectrum and is characterized by lung parenchyma that is perfused by a systemic artery but maintains a normal tracheobronchial tree. The pulmonary artery is often rudimentary or hypoplastic with poor arborization.⁶ Although the cause of the systemic arterial supply is unknown, it is thought that persistence of an embryonic connection between the aorta and the pulmonary parenchyma leads to the anomaly.⁷

According to Yamanaka and colleagues,⁸ patients may range from 0 to 68 years of age and are predominantly male. Pseudosequestration is often left-sided and supplied by a branch of the descending thoracic aorta. The pulmonary veins drain normally into the left atrium. Most patients are asymptomatic; when symptomatic, hemoptysis, exertional dyspnea, and congestive heart failure from left heart overload are the most common presentations.⁸ A cardiac (continuous or systolic) murmur is the most common clinical manifestation in children.^{7,8}

Based upon the above-mentioned characteristics, our patient has pulmonary pseudosequestration in the left lower lobe.

Diagnosis

Pulmonary pseudosequestration

Summary

With the advances in CT having multi-detector technology, the evaluation and diagnosis of congenital bronchopulmonary foregut malformations is greatly enhanced. Thorough and careful assessment of the airway, lung parenchyma, esophagus, arteries, and veins should be systematically analyzed in order to arrive at the correct diagnosis. The treatment plan is dependent in the proper identification of the pulmonary and systemic vessels.

In our 32-year-old patient who presented with a two-year history of chronic cough and hemoptysis, CT findings in the left lower lobe of a predominantly systemic arterial supply from the descending thoracic aorta, a small pulmonary arterial supply, a prominent left inferior pulmonary vein draining into the left atrium, and a normal tracheobronchial tree leads to the diagnosis of pulmonary pseudosequestration.

References

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