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A 39-year-old man, a smoker without significant medical history, presented with vague chest pain, cough, and hemoptysis. His symptoms had started 2 weeks earlier. Radiography showed an area of decreased lucency (ground glass appearance) in the right lower lobe. CT scan confirms the presence of a triangular area of inhomogeneous parenchymatous increased attenuation mainly of ground glass with inlying bronchocoeles and tree-in-bud appearance compatible with an intrapulmonary sequestration. The arterial supply is derived from the lower thoracic aorta. The venous drainage is to the left atrium.

The hemoptysis is a result of a supratherapeutic International Normalized Ratio (INR) on Sintrom intake for atrial fibrillation (AF). Treatment of sequestration consisted of a thoracoscopic lobectomy.

Comment

Pulmonary sequestration is a relatively rare anomaly comprising 0.15-6.4% of all congenital pulmonary malformations. An area of dysplastic and non-functioning lung parenchyma is present without a normal connection to the tracheobronchial tree and the pulmonary arteries. It receives its vascular supply from a systemic artery. It is anatomically classified as intralobar or extralobar.

Intralobar pulmonary sequestration is located within the visceral pleura and is surrounded by normal lung, whereas extralobar pulmonary sequestration is separated from the lung by a pleural envelope. The venous drainage of intralobar pulmonary sequestration is generally through the pulmonary veins, whereas the extralobar variant usually has a systemic venous return. Intralobar sequestration, the most common form of pulmonary sequestration, is most commonly localized in the lower lobes.

Patients may present with an incidental pulmonary lesion on imaging and otherwise remain asymptomatic. More commonly however, they may manifest varying types of pulmonary conditions such as pleural effusions or recurrent pneumonia. A few articles have reported more severe complications such as aspergillosis and even fatal hemoptysis. Although the etiology of the hemoptysis is uncertain, it is thought to be related to high-pressure blood flow in the sequestrated lung from the anomalous systemic arteries.

Imaging studies are important in the diagnostic process with the modality dependent upon patient age and also in the differential diagnosis. Chest radiography may reveal a homogenous lesion in the lung base (left side > right side) in uncomplicated intralobar sequestration, although complicated sequestration may present as a cystic lesion with air-fluid levels. In the adult, chest CT scan is most helpful, and will provide information about the vascular anatomy, crucial for operative treatment. Previously, catheter angiography was performed to diagnose sequestration and demonstrate its blood supply. However, presently helical CT angiography offers a less invasive way to demonstrate the abnormalities. Visualization of the anomalous arteries and veins is important to make the correct diagnosis of pulmonary sequestration and differentiating it from other lung abnormalities. The capability of CT angiography to simultaneously image the arterial supply, venous drainage, and parenchymal changes in a single examination makes it the imaging modality of choice.

The main treatment for pulmonary sequestration should be surgical resection with a ligation of an aberrant artery because of recurrent pulmonary infection and the unfavorable cardiac consequences caused by the existing aorto-pulmonary shunt.

Reference