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SHORT COMMUNICATION

Intralobular bronchopulmonary sequestrations associated with bronchogenic cysts ☆

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Summary We present three cases of intralobar bronchopulmonary sequestrations with associated congenital bronchogenic cysts. As congenital abnormalities tend to be found together, these cases question the notion that intralobar sequestrations only occur secondary to chronic inflammation or infection, and suggest they can be congenital lesions.
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Introduction

Bronchopulmonary sequestrations are regions of non-functioning pulmonary tissue that have inadequate communication with the tracheobronchial tree.[1] Sequestrations were first described by Pryce in 1946 and further characterized by Sade in 1974.[2,3] Two forms of sequestrations exist, extralobar and intralobar, with 75% being of the intralobar variety.[3] These anomalies have different anatomic findings and presentations and, as such, are considered to have different etiologies. Extralobar sequestrations (ELS) are universally accepted to be congenital anomalies arising from the primitive foregut and are frequently associated with other foregut malformations. ELS and bronchogenic cysts are the two most common foregut anomalies and, therefore, are the most frequent associated congenital thoracic malformations.

Unlike ELS, intralobar sequestrations (ILS) present later in life, do not have their own investing visceral pleura, are located within the normal lung architecture and are thought to arise from chronic inflammation or recurrent infections. We present

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three cases of ILS associated with a bronchogenic cyst implying that ILS may be congenital in nature.

Case presentations

Case 1: A 47-year-old woman presented with multiple episodes of right lower lobe pneumonia since early adulthood. She was a non-smoker and denied known tuberculosis exposure. She had no risk factors for HIV infection or aspiration. Chest radiographs (CXRs) revealed a right lower lobe opacity, which had been present on prior images obtained during previous episodes of pneumonia. Computed tomography (CT) showed a right posterior basal infiltrate and 2 cm mediastinal cyst (Fig. 1). The CT also revealed an aberrant artery, branching from the thoracic aorta, directly feeding the posterior basal segment. Bronchoscopy was unremarkable. She was diagnosed with an ILS and underwent surgical resection. Pathologic sections of the right lower lobe revealed marked interstitial fibrosis with intrabronchial and intraalveolar abscess formation. In addition to confirming the ILS, pathologic examination incidentally discovered an attached cystic space lined by a tan, smooth wall that was determined to be a bronchogenic cyst. The patient's post-operative course was uncomplicated.

Case 2: A 44-year-old, asymptomatic, non-smoking woman was referred for a mass incidentally noted on CXR. The CXR showed a 3 cm, well-circumscribed mass in the right lower lobe. CT revealed consolidation of the medial basal segment and 2.7 cm cystic lesion with mediastinal involvement (Fig. 2). An aberrant branch of the aorta supplying this segment was identified. Bronchoscopy was unremarkable. She was diagnosed with an ILS and underwent right lower lobe resection. Pathologic review of the sections of lung revealed a

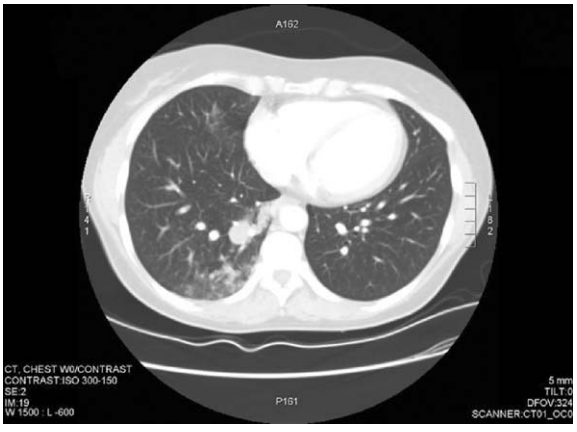

Figure 1 Case 1—CT (lung windows)—right posterior basal infiltrate and 2-cm mediastinal cyst.

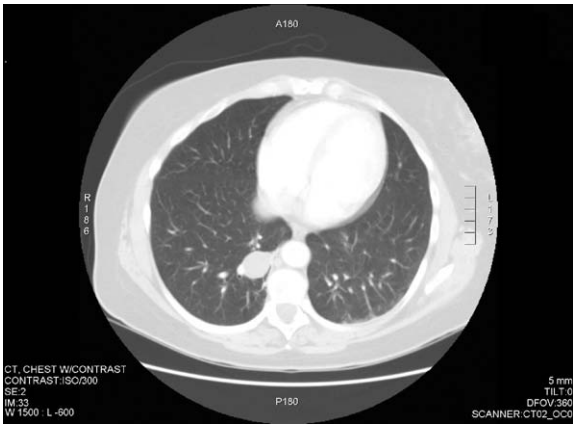

Figure 2 Case 2—CT (lung windows)—2.7 cm mediastinal cyst.

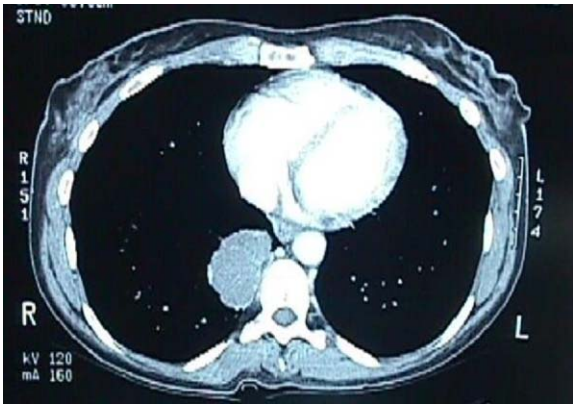

Figure 3 Case 3—CT (mediastinal windows)—dense consolidation of right medial basal segment.

4.0 cm abscess with purulent material consistent with an ILS. The cyst, which involved the mediastinum, was not removed. The patient tolerated the surgery without complication and had no subsequent complications.

Case 3: A 39-year-old, asymptomatic woman was referred for PPD-conversion. Screening CXR revealed a right lower lobe infiltrate and sputum was negative for acid-fast bacilli. Subsequent CT and MRI both showed a 3.2 cm dense consolidation of the right medial basal segment (Fig. 3). Angiography identified an aberrant artery branching from the aorta, supplying a consolidated right lower lobe. The CT and MRI also revealed a 4.5×2 cm mass in the superior mediastinum. Bronchoscopy was unremarkable. She was diagnosed with an ILS with underwent surgical resection. Histologic examination revealed a cyst with respiratory type epithelium consistent with an intralobular sequestration. The superior mediastinal mass was subsequently removed to exclude malignancy. Histology revealed benign respiratory epithelial-lined cyst

consistent with a bronchial cyst. She tolerated both procedures without post-operative complications.

Discussion

Bronchogenic cysts are congenital anomalies arising from the ventral foregut during the first 16 weeks of development and arise from abnormal budding from the tracheobrochial tree.[4] Foregut cysts account for approximately 49% of all cysts in the mediastinum and commonly present with other congenital malformations.[5] While usually asymptomatic, bronchogenic cysts may be associated with recurrent pneumonias or airway compression and can clinically resemble bronchiectasis.[6]

Bronchopulmonary sequestrations are regions of non-functioning pulmonary tissue that have inadequate communication with the tracheobronchial tree.[1] Of the two forms of sequestration, ELS are often associated with other congenital lesions, including diaphragmatic hernias and bronchogenic cysts. However, congenital anomalies are thought to be rarely associated with ILS. This led to the theory that these are acquired lesions, resulting from chronic inflammation or recurrent infections. It is believed that chronic inflammation leads to the interruption of the connection to the bronchial tree and obliteration of the normal pulmonary artery feeding that area.[7] In these three presented cases, an ILS was associated with a congenital bronchogenic cyst. Two were not associated with prior infections, suggesting that one anomaly was not the result of the other. In all three cases, no evidence of airway compression, mass or stenosis was found on bronchoscopy. The associated cyst implies that ILS is a congenital malformation and not an acquired lesion resulting from recurrent infections. We cannot exclude the possibility that the patients had previous asymptomatic childhood infections whose consequences were only presenting in adulthood. However, the recent discovery of intralobular sequestration in newborns who had not had chronic infections or inflammation suggests that ILS are not always the result of infection.[8]

Aberrant systemic arteries supply sequestrations, and demonstration of this vessel confirms the diagnosis and guides surgical resection. While angiography is still considered the gold standard, newer, less-invasive imaging techniques are showing promise; including CT angiogram, Doppler ultrasound and magnetic resonance angiography.[8–11] More recently, diagnoses of sequestrations have been made by prenatal ultrasound.[12] Treatment of choice remains surgical resection to prevent future infections and hemoptysis.[13,14]

Although not common, the discovery of ILS with bronchogenic cysts argues that these are congenital, foregut malformations and not acquired lesions. CT angiography is an invaluable diagnostic tool that can both demonstrate the aberrant arterial supply and identify other anomalous lesions.

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