CLINICAL VIGNETTE

A Subcarinal Mass

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Case Presentation

A 28-year-old female without any previous past medical history presented to pulmonary with complaints of shortness of breath with exertion. She reported no difficulty when walking at a slow pace, but dyspnea occurred as her exertional pace increased. Palpitations coincided with the dyspnea. She also reported occasional wheezing, mainly at night and with exertion, as well as a dry cough that occurred intermittently, mostly with exertion. She denied dysphagia, odynophagia, chest pain, hemoptysis, fever, chills, weight loss, recent travel, or putrid sputum production. She was a lifelong nonsmoker. She owned a cat, and was a college student. Her vital signs were unremarkable. Her physical exam was normal, revealing no stridor, wheezing, or abnormal lymphadenopathy. Pulmonary function studies were unremarkable, showing no obstructive or restrictive lung disease, and a normal diffusion capacity for carbon monoxide. A computer tomography of the chest was performed. It revealed a well-defined low-attenuation lesion in the subcarinal region measuring 5.6 x 6.1 x 5.7cm, with mild compression of the bronchus intermedius, left atrium, and right pulmonary artery. There was no lymphadenopathy, pleural effusion, or consolidation. Given the location and imaging features, the lesion was suspected to be a bronchogenic cyst.

Discussion

Introduction: Bronchogenic cysts are congenital lesions that arise from anomalous budding of the ventral foregut during development.[1] Bronchopulmonary cysts represent part of the spectrum of bronchopulmonary foregut malformations, and are associated with other congenital pulmonary malformations such as congenital lobar emphysema, and pulmonary sequestration.[1] Bronchogenic cysts occur most commonly within the mediastinum, but can occur at any point throughout the tracheobronchial tree. When they form early, they are located near the trachea and esophagus, or close to the carina and main bronchi. When they occur later, during bronchial budding and branching, they grow within the lung.[2,3] The later the development, the more peripheral it is in the lung.

Epidemiology: The prevalence of bronchogenic cysts is difficult to determine because of their different forms and natures. Cooke et al estimated prevalence of 0.04% to 0.06%.[4] Schenck et al showed that 10% of cases were recognized at birth, 14% were discovered in the first year of life, and 57% were found over the age of 15y/o.[5] It is difficult to ascertain what percentage of undiagnosed or diagnosed, untreated cysts remain asymptomatic during a lifetime.

Presentation: Although some bronchogenic cysts are asymptomatic, and found incidentally after imaging, most cysts are symptomatic. Symptoms are frequently nonspecific, prompting testing, which then reveals the lesion. Affected individuals typically present during the second decade of life with symptoms such as recurrent coughing, wheezing, pneumonia, chest pain, and chest pressure. Symptoms are likely due to compression or irritation of adjacent structures.[6] Bronchogenic cysts may also present with symptoms from cyst communication with aerodigestive structures, leading to bleeding, or localized infections.[1] It is uncommon for the cyst to have a patent connection with the airway, but when present, it may promote infection of the cyst by allowing bacterial entry.[7] The cyst can also rupture into the trachea, the pericardial cavity, or the pleural cavity.[8] Pneumothorax is not an uncommon complication, and is usually accompanied by pleuritic pain.[8,9] Severe hemoptysis is a rare complication.[8] Newborns with rapidly enlarging central cysts can develop respiratory distress, cyanosis, and feeding problems.[10]

Diagnosis: Diagnosis of bronchogenic cysts is by imaging. Chest radiographs and computed tomography (CT) scans are the most valuable diagnostic studies. Bronchogenic cysts appear as spherical or oval masses with smooth outlines, and are usually unilocular and noncalcified.[8,11] CT is beneficial in documenting the size and shape of the cyst and determining its position relative to other structures. The differential diagnosis for bronchogenic cysts in the lung parenchyma includes lung abscess, hydatidosis, fungal disease, tuberculosis, infected bullae, vascular malformations, and malignancy.[8]

Treatment: Despite the effectiveness of noninvasive diagnostic studies, a definitive diagnosis is not always possible preoperatively.[7] The management of bronchogenic cysts therefore consists of complete surgical excision by partial or total lobectomy. Complete resection is recommended in all cases of symptomatic and asymptomatic suspected bronchogenic cysts given their unclear diagnosis, infectious complications, erosive complications, likelihood of eventual development of symptoms and the potential for serious illness, including harboring

malignancy.[12] The pathologic hallmark of bronchogenic cysts is the presence of ciliated pseudostratified columnar epithelium, cartilage, and smooth muscle within the cyst wall.[6] However, as in all congenital cystic anomalies, malignant degeneration may occur with this disorder,[13] with some excised cysts showing the presence of squamous metaplasia and raising the possibility of stepwise carcinogenesis in these lesions.[6] Bronchogenic cysts also have a high recurrence rate if incompletely excised.[8] The prognosis after complete excision is excellent in all patients.[8,14]

Case Outcome

As imaging findings were most consistent with bronchogenic cyst, the patient was referred to thoracic surgery for complete excision of the mass. She underwent robot-assisted resection and evacuation of mediastinal cyst. Post-operative course was uncomplicated and she was discharged home on post-operative day 3. Pathological diagnosis revealed bronchogenic cyst lining. She recovered well and her symptoms of dyspnea, palpitations, wheezing, and cough resolved post-operatively.

Figures

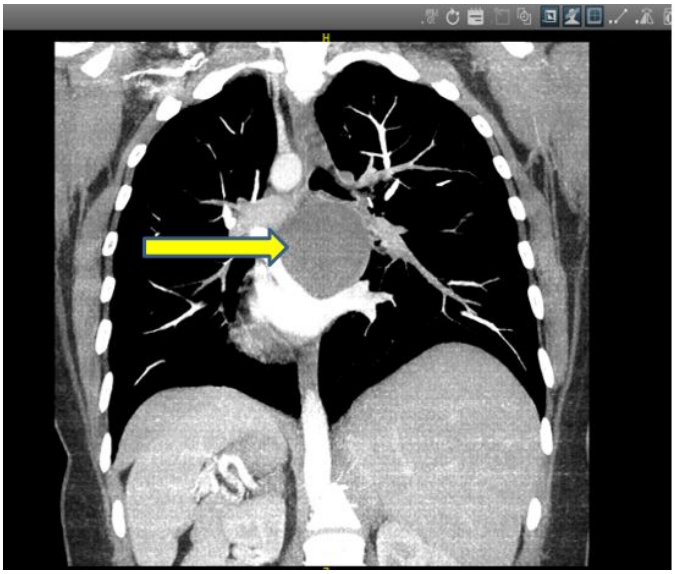

Figure 1: CT scan of chest showing 5.6 x 6.1 x 5.7cm subcarinal mass (yellow arrow) with mass effect on the bronchus intermedius, left atrium, and right pulmonary artery.

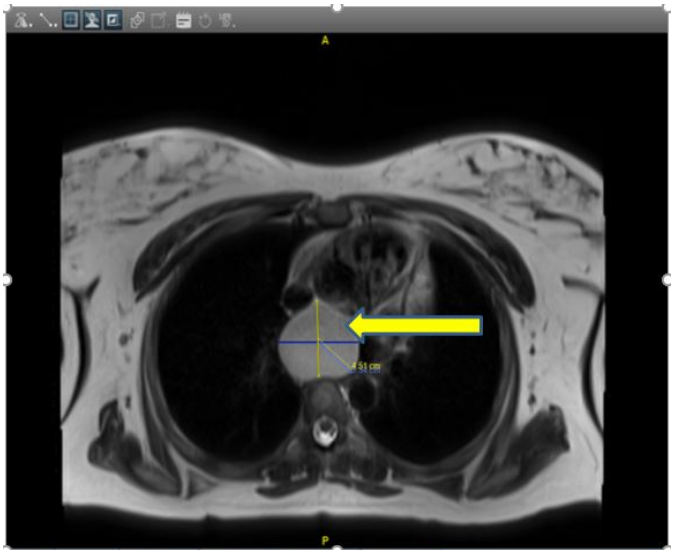

Figure 2: MRI chest showing homogenous T2 hyperintense cystic lesion (yellow arrow) without internal enhancement or associated solid or nodular components.

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