CLINICAL VIGNETTE

Dynamic Airway Obstruction Secondary to Rare Endobronchial Lesion

Eric J. Kwoh, MD, Casey Y. Kaneshiro, MD and Jaime Betancourt, MD

Introduction

Benign lung tumors account for less than 1% of all lung tumors. Of these, pulmonary hamartomas are the most common with an incidence between 0.025% and 0.32%.[1] The majority of pulmonary hamartomas are located within the lung parenchyma, while endobronchial hamartomas account for only 5-10% of such lesions.[2,3] Pulmonary hamartomas are usually discovered incidentally since the vast majority of patients are asymptomatic.[4] We report a unique case of a symptomatic pulmonary endobronchial hamartoma.

Case Description

A 76-year-old male with a 50 pack-year smoking history and moderate chronic obstructive pulmonary disease (COPD) presented to our pulmonary clinic for continued management of his COPD. He reported progressive dyspnea on exertion, an increase in his chronic productive cough, and worsening exercise tolerance over the preceding 2 years. During this period, he experienced multiple acute exacerbations of his COPD requiring glucocorticoids and antibiotics. He was also hospitalized twice in the preceding year for *Klebsiella pneumoniae* pneumonia. Chest radiograph demonstrated several small faint nodular opacities in the right upper lung field that appeared stable over the past 3 years and possibly represented scar from sequela of prior infectious/inflammatory processes. Transthoracic echocardiogram was without significant abnormalities. Computed tomography (CT) scan of the chest and pulmonary function tests were ordered to further evaluate the cause of his recurrent symptoms and progressive dyspnea.

Diagnostic Tests

Chest CT identified a polypoid lesion in the right mainstem bronchus arising from the anterior wall at the level of the right upper lobe origin (Figure 1). There were significant inflammatory changes with pleural-parenchymal scarring seen in the right upper lobe. Pulmonary function testing demonstrated a moderate obstructive ventilatory defect with evidence of air trapping.

Flexible fiberoptic bronchoscopy discovered a nodular endobronchial lesion affixed to the anterior wall of the right secondary carina with significant dynamic obstruction of the right upper lobe (Figure 2). During exhalation, the lesion nearly completely obstructed the right upper lobe.

Treatment

The patient underwent a follow-up interventional flexible bronchoscopy and the lesion was successfully snared and removed using argon plasma coagulation (APC), cryotherapy, and manual debulking. At the conclusion of the procedure, the right upper lobe takeoff was clearly patent and without distal obstruction (Figure 3). Pathological review of the lesion was consistent with a pulmonary hamartoma.

Discussion

Pulmonary hamartomas are rare benign tumors usually discovered incidentally as they are often asymptomatic. They arise from primitive bronchial mesenchymal tissue which can differentiate into various components. They are usually histologically heterogeneous composed of cartilage, bone, fat, and muscle tissues.[2,3,5]

Pulmonary hamartomas typically exhibit slow growth over many years. Consequentially, they are most commonly identified between the ages of 50 and 70 years.[2] Men are several times more likely than women to be affected.[2,6] In less than 10 percent of cases, pulmonary hamartomas can present as endobronchial lesions, which can lead to symptoms related to endobronchial obstruction including cough, dyspnea, and recurrent infection. Hemoptysis is rare.[2,3,7] Recurrent pneumonia secondary to bronchial obstruction can irreversibly damage the affected areas of the lung causing bronchiectasis and scarring.[8]

The differential diagnosis for endobronchial lesions is broad and includes both benign and malignant etiologies. Benign endobronchial lesions are typically caused by abnormal tissue growth in the form of hamartoma, leiomyoma, papilloma, or granulation tissue.[7] Malignant endobronchial lesions are either primary endoluminal carcinomas or metastatic carcinomas from other sites such as breast carcinoma, renal cell carcinoma, colon carcinoma, melanoma, and sarcoma.[7]

Chest radiography is non-specific for the diagnosis of pulmonary hamartomas but can demonstrate soft tissue attenuation or a well-circumscribed mass.[9] On chest CT, pulmonary parenchymal hamartomas appear as rounded soft tissue masses that frequently exhibit a popcorn-like calcification (30%) and intralesional fat (60%).[9] Presence of fat in a well-circumscribed solitary pulmonary nodule which does not demonstrate significant growth is highly indicative of a pulmonary hamartoma.[10]

In cases where chronic obstruction has occurred due to an endobronchial location, inflammatory changes and scarring secondary to recurrent pneumonias can be seen in the affected area on both chest radiography and chest CT as demonstrated in our patient.

While chest CT aids in diagnosis, biopsies are necessary to differentiate a hamartoma from other causes of endobronchial lesions. Bronchoscopy is preferred for tissue biopsy. Bronchoscopically, endobronchial hamartomas are smooth, fleshy, pedunculated masses that are tan to pink in color and are often polypoid in nature. Histologically, the mesenchymal components are usually varied with a predominance of adipose tissue.[2,3,5] In our patient, histology of the specimen demonstrated a combination of mixed mesenchymal components composed mostly of cartilage with focal areas of calcification, adipose tissue, and myxoid connective tissue. The mixture of these benign mesenchymal tissues with the added presence of fat tissue established the diagnosis of hamartoma rather than chondroma.

Once the diagnosis of a symptomatic endobronchial hamartoma is established, complete removal is recommended. In the absence of symptoms, complete removal of endobronchial hamartomas should be considered as they may eventually cause bleeding and obstruction of the tracheobronchial tree.[3] Historically, endobronchial hamartomas were treated by lung resection.[11] However, given the advancements in endobronchial techniques, bronchoscopic intervention is now considered the preferred modality because it is less invasive. Additionally, definitive surgical management is not necessary since malignant degeneration of hamartomas is extremely rare and recurrence rates are low.[2,3] Rigid bronchoscopy with either laser, electrocautery, or cryotherapy is considered first-line treatment; however, a flexible bronchoscope can be used in select cases, as in our case. [12,13]

Conclusion

Our patient reported significant improvement in his respiratory symptoms in follow-up visits following his procedure. In the 9 months since the procedure, he has not experienced any further COPD exacerbations or recurrent pneumonias. His chronic productive cough is largely resolved, and he can now walk from the parking lot to clinic without taking any rest breaks. Given our patient's significant clinical improvement following bronchoscopic intervention, it is highly likely that his endobronchial hamartoma with dynamic airway obstruction largely accounted for his worsening respiratory symptoms and recurrent pneumonias over the past 2 years.

This case demonstrates the clinical importance of classifying and managing pulmonary hamartomas based on their location. While pulmonary hamartomas are benign tumors, an endobronchial location can result in significant morbidity due to tracheobronchial tree obstruction. Suspicion for a possible obstructive endobronchial lesion should be elevated in a patient with unexplained recurrent lung infections and exacerbations of respiratory symptoms even if they carry the diagnosis of COPD. Expedient identification and removal of symptomatic endobronchial hamartomas is crucial and long-term outcomes are often favorable with a low rate of recurrence.

Figures

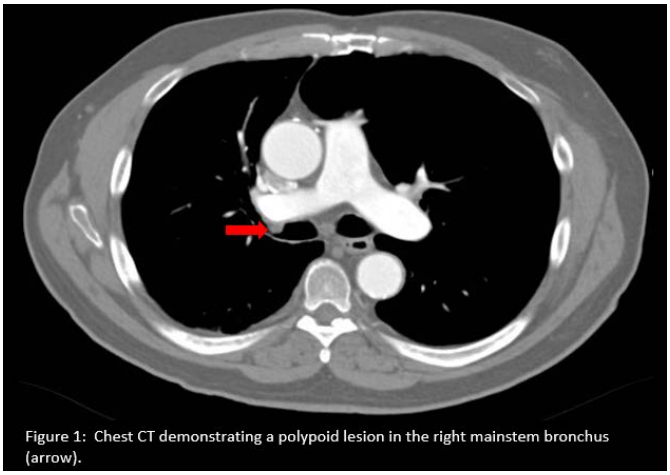

Figure 1: Chest CT demonstrating a polypoid lesion in the right mainstem bronchus (arrow).

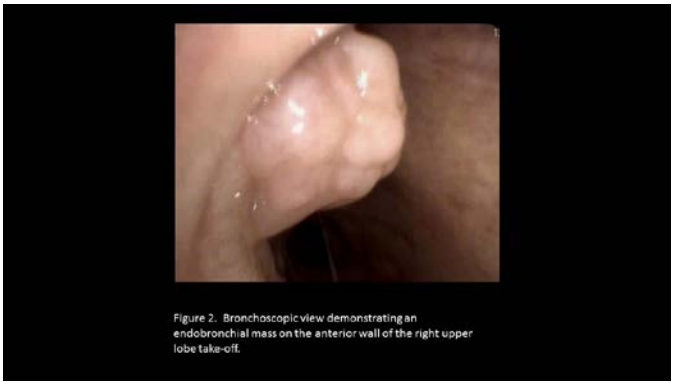

Figure 2. Bronchoscopic view demonstrating an endobronchial mass on the anterior wall of the right upper lobe take-off.

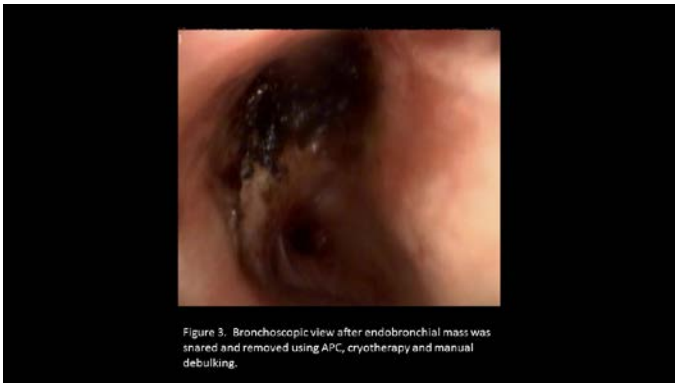

Figure 3. Bronchoscopic view after endobronchial mass was snared and removed using APC, cryotherapy and manual debulking.

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