**A young man with primary pulmonary myxoma**

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Key clinical message

Primary pulmonary myxoma is a rare tumor and usually diagnosed incidentally in clinically asymptomatic patient as a well-defined lung mass without metastasize. It should also be considered that diagnostic methods such as bronchoscopy or transthoracic aspiration may be non-diagnostic. So tumor resection is essential for diagnosis and treatment.

Introduction

Myxoma is a morphologically benign tumor that frequently occurs in the left atrium.[1]Primary pulmonary myxoma is a rare tumor and usually found as an incidental well-demarcated pulmonary mass without symptoms. Myxomas are usually found as a single mass, growing from the primary lesion and do not produce distant metastases. The pathological characteristic of these tumors is the presence of elongated and stellate cells in a myxoid stroma. Since it is difficult to diagnose these tumors based on a small biopsy samples or cytology, tumor resection is necessary for diagnosis and treatment. [2] Given the few reports on primary pulmonary myxoma, we present a case of this tumor in a young adult man.

Case History/examination

A 29-year-old male patient without any underlying disease presented to the emergency department of our hospital with productive cough and pleuritic chest pain in left hemithorax with radiation to the left shoulder since 2 months ago. He also complained of occasional hemoptysis and orthopnea for 4 months. The patient had a non-significant weight loss and mentioned occasional cigarette smoking.

On presentation vital signs and laboratory investigations were unremarkable.

CT scan demonstrated a large oval-shaped homogeneous mass sized 50\*80 mm with well demarcated margin in left lower lobe extending from the left hilum to the periphery as a pleural base density and increased pleural thickness with surrounding reticular opacity (Figure 1).

*Figure 1:* CT scan showed large homogeneous oval-shaped, well-demarcated mass in left lower lobe

Differential diagnosis, investigation and treatment

The differential diagnosis of a lung mass in a 29-year-old man includes a broad range of infectious, neoplastic, inflammatory, and congenital conditions. Therefore, different diagnostic methods should be used to diagnose the patient.

Initially, flexible fiberoptic bronchoscopy was performed for the patient which revealed a LLL bronchus fish mouth stenosis and LB6 complete obstruction due to external compression and mucosal erythema in LC2. BAL was taken from LLL and EBB was performed in LC2 (figure 2). BAL pathological examination reported mostly degenerated cells and few alveolar macrophages in mucoid back ground. It was negative for malignancy.

Diagnostic transthoracic aspiration was performed under ultrasound guidance. About 10 cc of very thick and mucinous fluid was drawn and sent for culture and Gram stain smear and pathologic examination.

Fluid pathological examination revealed several mesothelial cells in hemorrhagic background with negative microbiologic result.

Ultimately, the patient became a candidate for surgical resection. So surgical resection of left pulmonary mass was scheduled and performed by thoracic surgeon. There was a large lung mass with adhesion to the surrounding tissues in the posterior segment of left lower lobe (figure 3). The cut surface of the mass showed large amounts of fragmented gelatinous contents (figure 4). The frozen section specimen was reported to be consistent with hamartoma.

Gross pathological examination of resected tumor revealed a cream brownish mucoid gelatinous circumscribed tissue and a piece of cream whitish gelatinous mass. Under high microscopic magnification benign hypocellular neoplasm composed of delicate spindle shape and stellate cells in abundant myxoid stroma with no mitotic figures lined by collagenized capsule was seen. The findings were compatible with myxoma. Lymph nodes had non-specific pathologic changes (figure 5).

Immunohistochemical analysis revealed positive staining for vimentin (+), Ki67 index of less than 1% positive and negativity for AE1/AE3, S100, Myogenin, Calretinin, GFAP, TTF1 and P63 which was compatible with pulmonary myxoma.

Conclusion and Results

As mentioned, the final diagnosis was primary pulmonary myxoma. The patient did not have any complication after surgery and was finally discharged with stable respiratory symptoms.

Figure 2: Bronchoscopic view with stenosis at left lower lobe bronchus

Figure 3: Surgical view of the tumor

Figure 4: Fragmented gelatinous contents within the tumor

Figure 5: Pathologic examination of resected tumor revealed hypocellular neoplasm composed of delicate spindle shape and stellate cells in abundant myxoid stroma with no mitotic figures.

Discussion

We present a young adult male patient with pulmonary myxoma with a left lower lobe mass, in whom initial evaluations at bronchoscopy and transthoracic aspiration were not diagnostic. Interestingly, the patient's frozen sample was reported hamartoma. Finally, a definitive diagnosis of pulmonary myxoma was made based on pathological and immunohistochemical analysis of the resected mass.

As noted in the patient, initial evaluations with transthoracic aspiration was non-diagnostic due to the low cellularity of the tumor.

Although the median age of patients with myxoma is reported to be 69 years (2), our patient was a young adult man.

Barkley and Cardozo reported the earliest case of pleuro- pulmonary myxoma in 1957.[3] Myxomas are benign mesenchymal tumors that can be found in multiple locations. The most common type of myxoma, which is mostly common in adults, is primary cardiac myxoma, accounting for 30-50% of all cases of cardiac tumors.[4] As previously mentioned, primary pulmonary myxoma is rare and usually diagnosed as an incidental well-defined lung mass in clinically asymptomatic patient.[2] Myxomas usually do not metastasize , but occasionally metastasis to brain , bone and soft tissues are seen. On the other hand, local recurrence may be diagnosed as emboli or sarcomatous origin. Therefore, long-term follow-up is recommended in these patients.[4] In addition to pleuropulmonary myxomas, there are case reports describing myxomas arising in the pulmonary artery and mimicking pulmonary embolism and subacute cor pulmonale (5,6). There are also some reports of trachea and endobronchial myxoma with symptoms resembling asthma and repeated pneumonia (4). Differential diagnoses for pulmonary myxoma include: myxoid liposarcoma, myxoid chondrosarcoma, myxoid malignant fibrous histiocytoma, pulmonary hamartoma, angiomyolipoma and pulmonary lipoma. [3]

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