Pulmonary Hamartoma

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Abstract
Pulmonary hamartoma are common benign tumor of lung. These are composed of variable components of cartilage, fat, smooth muscles and respiratory epithelium. Most patients are asymptomatic with few present with pulmonary symptoms. The neoplasm is less common in young with surgical excision being the treatment of choice. Histopathology revealed the true pathological nature of the tumor.

Key words: Lung, benign mass, pulmonary hamartoma, adipose tissue, chondroid tissue.

Introduction:
Hamartomas are the benign tumors that can occur in the lungs, heart, breast and other organs. Greek word hamartia is the origin for this term which means erroneous or faulty. Pulmonary hamartoma is the lesion first described in 1904 by a German pathologist cells Eugen Albrecht.¹ These are benign neoplasm composed of varying amount of at least two mesenchymal elements along with entrapped respiratory epithelium. The exact etiology is unknown.² These are slow growing well defined neoplasms detected incidentally on radiology in most of the patients; at the periphery of lung. However endo-bronchial lesions present with respiratory obstructions.³ Our patient is a 40 years old female who presented with lung mass, peripheral in position with clinical history of back pain. She underwent Video assisted thoracic surgery (VATS) surgery with excision of mass with wedge resection of segment four. Histopathology proved that the mass was pulmonary Hamartoma.

Case history
A 40 years old female presented to the thoracic outpatient department with history of pain at the upper back for 3 months. On examination her vitals were stable. All the laboratory investigations were within normal limits. CT scan of the chest showed a lung mass of 5x5 cm with possible origin from the fissure of the lungs likely benign in nature with a differential diagnosis of tumor of fibrous origin. She underwent video assisted thoracic surgery (VATS) surgery with excision of the mass. On histopathological examination there were islands of mature chondroid tissue with fibromyxoid stroma. Multiple foci of mature adipose tissue was also noted. There are slit like respiratory epithelium trapped in between these mesenchymal elements.
Discussion:
Pulmonary hamartomas are common benign tumours of the lung with an incidence of between 0.025-0.32% as shown in different autopsy studies. These are slowly growing neoplasms with male predominance. These are commonly seen in fifth to sixth decade of life. They mainly have two types of clinical presentations. First type is seen as peripheral lung lesion (90%) and next type is the endobronchial lesion (10%). Clinical features range from asymptomatic to obstructive features and their consequences. Symptoms like cough, dyspnea and chest pain are seen due to recurrent pneumonia, bronchiectasis, collapse, pneumonitis and even atelectasis. Cytogenetically there is abnormal karyotype and recombination between chromosomal bands 6p21 and 14q24, thus supporting that hamartoma of lung is a true neoplasm. Radiologically they present as solitary coin lesion. Areas of calcifications presenting as pop corn calcification has be described. The lesions present as smooth, fleshy polypoidal to sessile, tan to pink mass in bronchoscopy. FNA during bronchoscopy can be done. Though the aspirate may be scanty due to the density of the lesion. The fine needle aspirate material is characterized by fibromyxoid-stroma, cartilage, bronchial cells, adipose tissue and rarely bone marrow. Bronchial cells with reactive atypia may be a source of false-positive result. In such cases, immunocytochemical stain for S-100 protein can help to highlight the chondroid and fibromyxoid stroma. In gross histology, these are nodular, well circumscribed lesion with irregular lobules of cartilage along with foci of calcification. Myxoid areas are noted in the periphery of the lobules. Hamartoma without cartilaginous component resembles fibroadenoma of breast. On cut section there is bulging with irregular nodular border. Adipose tissues are also prominent in some specimen. The tumor size ranges from 1-5 cm. Histopathologically, these tumors are composed of predominantly chondroid or chondromyxoid tissue mixed with variable proportion of other mesenchymal components like fat, connective tissue, smooth muscles, bone, respiratory epithelium. The mesenchymal components of endobronchial hamartoma are highly varied. Parenchymal tumours tend to have more epithelial clefts than endobronchial tumors. Endobronchial tumours tend to have more fat. It has been found that sometimes there is associated peculiar change in pulmonary hamartoma in which placental villous like formations are noted named as placental transmogrification. Immunohistochemically, some of the spindle cells of the lesion have features of myoepithelial cells, positivity for actin and S-100 protein. There is also expression of estrogen receptor, progesterone receptor and Androgen Receptors in males mostly seen in myoepithelial cells.

Multiple pulmonary chondromatous hamartomas have been noted as manifestations of either the Carney triad or Cowden syndrome. The former is predominantly seen in young women and with concurrent appearance of gastrointestinal stromal tumours, pulmonary hamartoma and extra-adrenal paraganglioma. Patients with Cowden disease often display multiple hamartomas, manifesting as mucocutaneous lesions, multiple benign tumours of internal organs and increased risk for several forms of cancer, including breast and digestive tract malignancies. Treatment of choice is in the form of conservative surgery, lung sparing or bronchoplastic surgery. Regarding the principles of the surgery, it is believed that normal lung tissue should be preserved as much as possible. Furthermore, due to lack of the malignancy after operation it was presumed that the wedge resection was safe enough. However, a potential trend of recurrence has to be kept in mind so it must be emphasized that the patients with Pulmonary hamartoma should be submitted to a complete evaluation and regular follow-up.

Conclusion:
We have presented a case of pulmonary hamartoma in a female, at a younger age group with a peculiar presentation of back pain. These are common benign neoplasms with slow annual growth however, sometimes a rapid enlargement especially in endobronchial mass may present with severe symptoms. Recurrences and sarcomatous transformations though rare should be kept in mind. Identification of these lesions are important because these are surgically resectable and have a good prognosis.
References


