Fine needle aspiration cytology of giant chondroid hamartoma of lung with review of literature.

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Abstract: Giant pulmonary hamartoma (diameter > 9cms) is a very uncommon benign tumor with only eleven reported cases. Here we report such a rare case of giant pulmonary hamartoma measuring 12cms in diameter occupying almost whole of the left hemithorax. CT guided FNAC was done in suspicion of a malignant tumor but the cytomorphological features were of a benign cartilaginous neoplasm. The cytological findings were correlated with the radiological features and a diagnosis of chondroid hamartoma was suggested on FNAC. The diagnosis was later confirmed by histopathological examination.

Key Words: lung, giant chondroid hamartoma, fine needle aspiration cytology

I. Introduction

Pulmonary hamartoma, also known as chondroma, chondroid hamartoma, chondroid adenoma, mesenchymoma, is the most common type of benign lung tumours, composed of varying proportions of mesenchymal tissues with entrapped respiratory epithelium. They were considered as developmental abnormalities in the past but now considered as benign mesenchymal neoplasms. [1-5]

Incidence of pulmonary hamartoma is 0.25% with a two to four fold male preponderance, most occurring in 6^{th} decade of life. Most often it is incidentally discovered as solitary nodule on chest x-ray. [1,2,3] They are usually peripheral and less than 5cm in diameter. Giant pulmonary hamartoma (measuring >9cm in diametre) is very rare and only 11 cases have been published till date [6-13]. Here we present a 27 years old female with a giant chondroid hamartoma, measuring 12 cms in diametre extending from periphery to bronchus, in the left lung.

II. Case Report

A 27 year old female was admitted to our hospital with cough, fever, and left-sided chest pain of 8 months duration. She had an episode of haemoptysis. Physical examination revealed decreased breath sounds in the left side. Routine blood and biochemical parameters were within normal limits. Sputum was negative for acid-fast bacilli.

X-ray revealed a large homogeneous opacity occupying almost whole of the left hemithorax blurring the left C-P angle. CT-scan (Figure 1) confirmed a large, heterogeneous mass measuring 12 cms in diameter occupying almost entire left hemithorax with extensive nodular (popcorn) calcification. Mediastinum was displaced to right. Left bronchus was compressed. No mediastinal lymphadenopathy or pleural effusion was documented. Radiological impression suggested a malignant neoplasm.

CT-guided FNAC of the lesion was done. Smears were stained by May-Grünwald-Giemsa and Papanicolaou (Pap) stains. Smears were moderately cellular comprising of round to ovoid cells with nucleomegaly and anisonucleosis, moderate amount of pale cytoplasm in a fibromyxoid background. There were also sheets of broncholiolar epithelium interspersed with fat cells and with foci of calcified spherules (Figure 2). Based on cytology a strong suspicion of chondroid hamartoma was made.

Percutaneous transthoracic needle biopsy, the main diagnostic approach, showed predominantly cartilaginous tissue & confirmed the diagnosis (Figure 3A, 3B, 3C).

Endobronchial biopsy showed immature cartilaginous cells having acidophilic cytoplasm that resemble histiocytes along with plump fibroblast & smooth muscle fibres (Figure 3D, 3E).

The patient is on follow-up and awaiting surgery.

III. Discussions

The term "hamartoma" was coined by Albrecht ^[14] in 1904 to describe tumor-like malformations resulting from a presumptive developmental abnormality. In 1934, Goldsworthy ^[15]applied this term to benign lung tumors which were composed predominantly of a combination of fat and cartilage. Cytogenetic studies have identified chromosomal bands of recombination located at 6p21 and 14q24 positions which supports the theory that hamartomas represent mesenchymal clonal neoplasms.^[16]

Pulmonary hamartomas are the most common benign tumours of the lung. While most of the usual hamartomas measure 1-5 cm in diameter, giant hamartomas reported measure between 9 to 30cm. [1,3,6] Petheram and colleagues reported a massive hamartoma with a diameter of 30cm. [6] Most of the giant hamartomas are seen in female patients in contrast with the usual hamartomas which have a male preponderance. The age of presentation is wide -30 to 63 years, mean age being 48.9 years. Some of the patients are younger than the expected age. Most of the giant hamartomas reported are localized in the right lung. Our patient is a 27-year-old female with a giant hamartoma in the left lung with endobronchial extension.

The peripheral pulmonary hamartomas are usually asymptomatic. Occasionally, the tumors are central or giant with endobronchial extension causing hemoptysis, bronchial obstruction, coughing, wheezing, expectoration, leukocytosis and fever. Our patient also presented with cough, fever, and chest pain of 8 months duration and an attack of hemoptysis due to the giant size of the tumor with compression of bronchus and endobronchial extension.

Usually diagnosis of hamartomas are incidental. These tumors constitute about 8% of all "coin" lesions in chest radiographs^[17]. X-ray shows solitary well-defined pulmonary nodules with varying patterns of calcification like irregular popcorn, stippled, or curvilinear pattern or even a combination of all three. In CT scan specific popcorn-type calcification seen in about 30% cases is almost pathognomonic. [1-4,11]. Our case has also typical popcorn calcification.

Fine needle aspiration cytology is diagnostic of majority of chondroid hamartomas (90%) and hence widely used. [9,18] Cytological criteria for diagnosis is presence of mature cartilage, myxoid connective tissues, sheets of bronchiolar epithelium & adipose tissue. Sometimes anisonucleosis & large intranuclear inclusion may lead erroneous diagnosis of malignancy. [1,2,8]

The histology of the parenchymal lesions in both giant and usual hamartomas usually reveals a predominant chondroid differentiation (80%), with fibroblastic (12%), fatty (5%) and osseous (3%) differentiation making the rest. [1,2] Endobronchial lesions tend to have more fat. [6] Myxomatous connective tissue, smooth muscle, blood vessels and other mesenchymal elements may also be seen histologically. [1,2]

Hasen and associates observed that preoperative diagnosis from transthoracic needle aspiration biopsy could be obtained in 85% of the patients with pulmonary harmatomas.^[18]

Most tumors grow slowly (average of 3 mms per year) during follow-up. Surgical treatment is the gold standard in intraparenchymal hamartomas including enucleation, lobectomy or sleeve resection, wedge resection, segmentectomy and pneumonectomy. [18]

This case is being reported because of its rarity.

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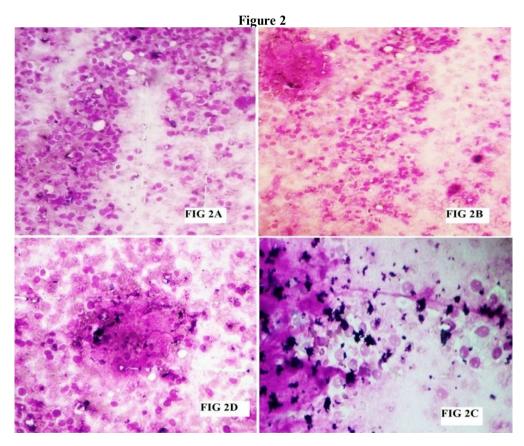
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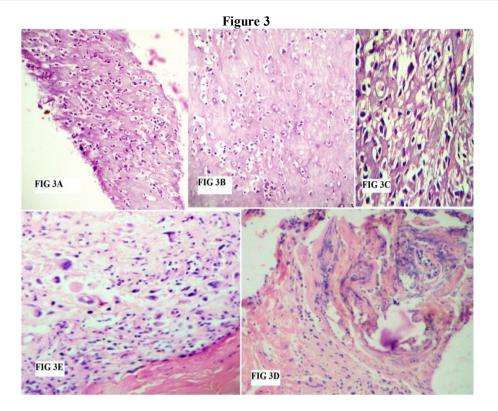
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Figures:

Figure 1 SLICE SPIRAL CISCAN





Legend to Figures:

Figure 1: CTscan- large, heterogeneous mass occupying left hemithorax with extensive nodular (popcorn) calcification.

Figure 2: FNAC smears- moderately cellular with round to ovoid cells showing nucleomegaly and anisonucleosis, moderate amount of pale cytoplasm in a fibromyxoid background with foci of calcified spherules

Figure 3: Figure 3A, 3B, 3C- Percutaneous transthoracic needle biopsy showing predominantly cartilaginous tissue; Figure 3D, 3E-Endobronchial biopsy showing immature cartilaginous cells having acidophilic cytoplasm that resemble histiocytes along with plump fibroblast & smooth muscle fibres.