Case Report

Cytomorphology of Primary Adenoid Cystic Carcinoma of Lung: An Exceedingly Rare Case

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ABSTRACT

Primary adenoid cystic carcinoma (ACC) of lung is a rare tumor, and probably accounts for 0.04-0.2% of all primary pulmonary tumors. It was formerly referred to as bronchial adenoma implying a benign glandular neoplasm. However, it is now considered to be a low-grade bronchial carcinoma. Pulmonary ACC usually arise in the proximal tracheobronchial tree, and are regarded as a slowly growing tumor.

A 20 year female was admitted with complain of cough without expectoration, weight loss, fever and anorexia since 20 days. In Ultrasound Sonography (USG) approx. 10x7 cm in size of well defined mixed echogenic lesion was noted at left upper zone. USG guided Fine Needle Aspiration Cytology was done. Cytopathology was suggesting of primary adenoid cystic carcinoma of lung. Fine needle aspiration cytology is simple method and useful in pre-operative diagnosis. However, diagnosis of adenoid cystic carcinoma of lung is difficult by cytologically. So, it is confirmed by histolopathological study.

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Introduction

Adenoid cystic carcinoma (ACC) occurs most commonly in the salivary glands and less commonly, at other sites such as the breast, skin, uterine cervix, upper aero digestive tract, and lung. [1]Primary ACC of lung is a rare tumor, and probably accounts for 0.04-0.2% of all primary pulmonary tumors. [2] Primary lung ACC tends to occur in the 4th and 6th decades of life and there is no sex predominance. It is very rarely seen below 30 years. [1] The development of the disease has no association with cigarette smoking or other carcinogens. [3]

The tumors are often asymptomatic, but can also be present with dyspnea, cough, fever and hemoptysis.[1] It was formerly referred to as bronchial adenoma implying a benign glandular neoplasm. However, it is now considered to be a low-grade bronchial carcinoma.[4] Pulmonary ACC usually arise in the proximal tracheobronchial tree, and are regarded as a slowly growing tumors.[5]

Case Report

A 20 year old female was admitted with complain of dyspnea on exertion, cough without expectoration, and chest pain since 20 day, also having weight loss, fever and anorexia but no history of hemoptysis. No history of any addiction or old pulmonary tuberculosis. Other systems were normal on physical examination. Laboratory findings were normal.

Than USG is done and approx. 9x8 cm in size of well defined mixed echogenic lesion was noted at left upper zone and chest X ray (postero-anterior view) revealed homogeneous soft tissue opacity in left upper and mid zone (figure 1). Thorax Computerized tomography (CT) was performed and finding were showing a well defined large lobulated heterogeneously enhancing soft tissue density mass lesion of size 104x 84x 88 mm is noted in left upper lobe (Figure 2).

USG guided FNAC was done with 22 G needle and 10 cc syringe. Blood mixed aspirate came out. Slides were prepared by crush method. Slides were stained by May-Grunwald-Giemsa, haematoxylin & eosin stains. On microscopy- Smears show moderate cellularity and contain poorly cohesive clusters and complex sheets of epithelial cells. Cells show monomorphic, bland finely granular hyperchromatic nuclei, smooth nuclear border and scanty cytoplasm. Epithelial cells embedded within eosinophilic homogenous hyaline globules with even distribution. (fig.3-4) Cells also dispersed individually on blood mixed background. Overall findings are suggestive of primary pulmonary salivary gland type of tumor most likely adenoid cystic carcinoma of lung. However that is

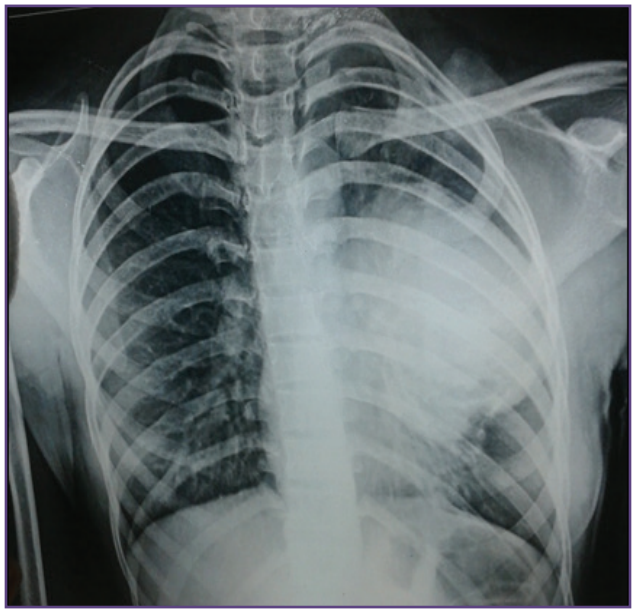

Fig. 1: (Chest X Ray PA view) Homogenous Opacity In Left Upper Zone

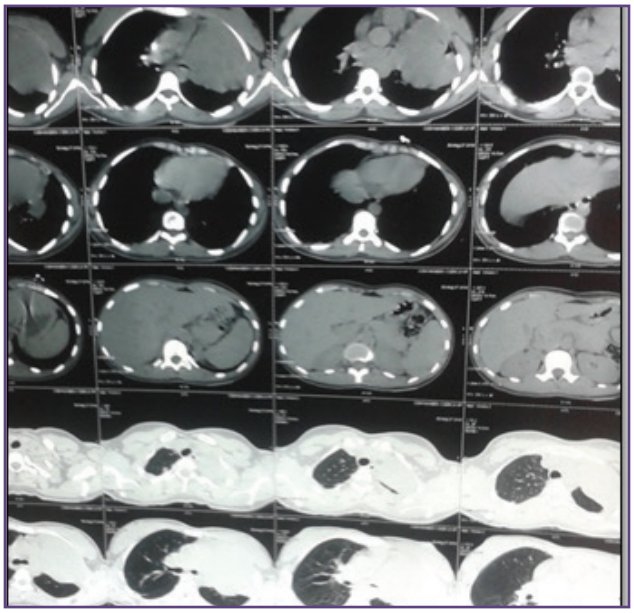

Fig. 2: (CT scan) Heterogeneously Enhancing Mass In Left Upper Lobe.

also confirmed by true cut biopsy taken from upper zone of left lung with histopathological examination.

Grossly the tissue was 0.5x0.2 cm sized and grayish white in color. Histopathological section showed proliferation of neoplastic cells arranged in cribriform pattern forming cystic spaces containing eosinophilic hyaline material. Cell shows round, mildly atypical nuclei and scanty to moderate amount of cytoplasm (Figure 5). Overall findings are in favors of salivary gland type tumor – adenoid cystic carcinoma of lung.

On immunohistochemistry study, tumor cells were found to be C-kit (CD117) positive, MIB1 positive (2-5%). (fig.6).

Discussion

ACC, also known as cylindroma in the past, is a variant of adenocarcinoma with distinct histopathologic and clinical features. [1] ACC is a rare, less than one percent, distinctive salivary gland-type malignant epithelial neoplasm that arises infrequently as a primary tumor in the lung.

ACC of the lung typically arises in the trachea and large airways. There are some reports that it may arise as peripheral lung tumour[5].In our case also tumour was located in the upper lobe of the lung.

In an earlier published report [6] primary adenoid cystic carcinoma of lung presenting as peripherally located lung mass in 33 year old male, and in second case[7] the age of patient was 29 year old female. It is very rarely seen below 30 years.[2] In our case the patient's age was 20 year, which is an uncommon age for this tumor to present with. So it is an exceedingly rare case presentation.

Cytologically tumor shows monomorphic epithelial cells embedded with hyaline globules. Tumor cells show

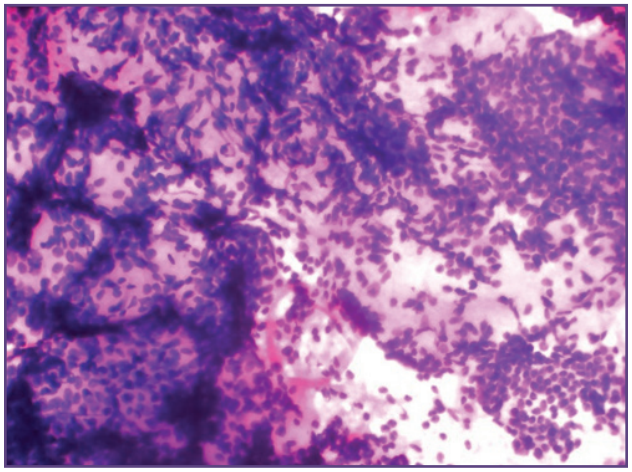

Fig. 3: Prominent Hyaline Globule and monomorphic with hyperchromatic Tumor cells (H & E, 10x)

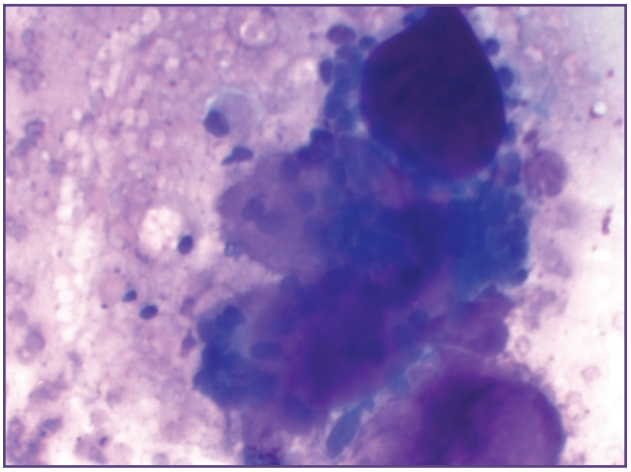

Fig. 4: Prominent Hyaline Globule (MGG stain 40x)

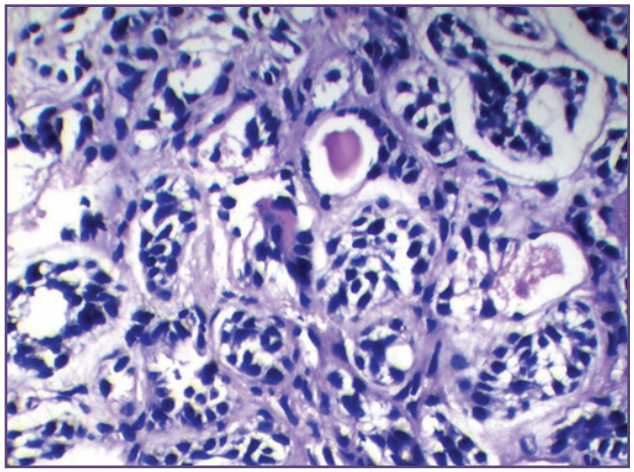

Fig. 5: Tissue section: Cribriform pattern of growth of ACC (H & E, 10 X)

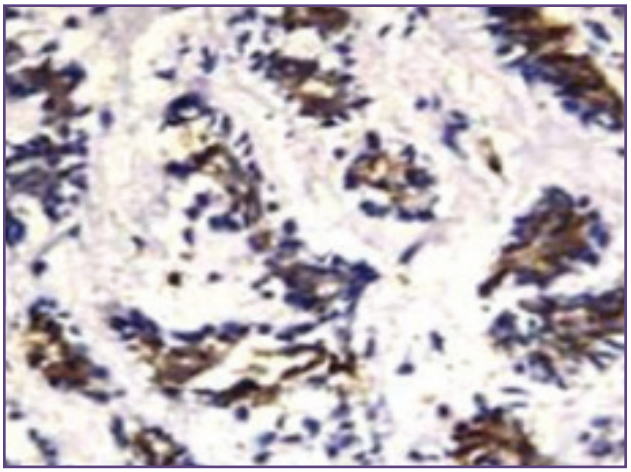

Fig. 6: Tissue section: Positivity for C-KIT In Tumor Cells

hyperchromatic nuclei, high nuclear/cytoplasmic ratio and scanty cytoplasm and at places nuclear molding and naked nuclei. So overall findings are characteristic and specific diagnostic criteria help to differentiate other tumor /lesions and it can diagnosed easily by cytologically. The cytopathological differential diagnosis of pulmonary ACCs include reserve cell hyperplasia, carcinoid tumor, small cell carcinoma, well differentiated adenocarcinoma etc. [8] In reserve cell hyperplasia cells are compactly arranged in sheets with characteristic, attached, columnar cells. cribriform and tubular structures are also not seen. In carcinoid tumor, absence of mucoid globules within glandular structure differentiates it from ACCs. In small cell carcinoma, small cell show typical nuclear molding and nuclear streaking. The presence of spherical hyaline globules and basement membrane material surrounded by neoplastic cells favors a diagnosis of ACCs over small cell carcinoma. The well differentiated Adenocarcinoma can be ruled out by glandular arrangement of cells, clusters of uniform cells with cystic spaces.

Histologically the tumor has 3 major growth patterns namely cribriform, tubular and solid .The most commonly found subtype is the cribriform pattern [9] .In our case the classical cribriform pattern was evident on histopathological examination. The tumor cells containing dense basophilic nuclei inconspicuous nucleoli and cell borders are indistinct. Prognosis depends on the histological pattern (tubular having the best prognosis and solid having the worst) and clinical stage of the tumour. [9] C-kit expression is commonly detected by IHC in adenoid cystic carcinoma. [10] In our case C-kit (CD117) positive (fig 8) MIB1 positive (2-5%), TTF1 negative.

Treatment for this tumor is not well defined. Complete excision of the tumor followed by adjuvant radiotherapy is by far the consensus treatment. [11] In one study from India[12], oral imatinib, tyrosine kinase inhibitor, showed good response in an inoperable case of primary adenoid cystic carcinoma of lung. In our case the patient is shift to higher centre for further treatment .Now patient is on tab imatinib, and having good prognosis.

Conclusion

This study highlights the utility of Radiological guided FNAC in the diagnosis of primary pulmonary ACC, mainly in peripheral tumors .Radiological guided aspiration cytology of lung lesions is easy to perform, rapid and useful in pre-operative diagnosis. Primary pulmonary salivary gland type neoplasms are difficult to diagnose on cytomorphology. However adenoid cystic carcinoma of lung has a characteristic cytomorphology which is helpful in diagnosis or to rule out other tumor/lesions of lung. But still final confirmation of diagnosis should be done by histopathology examinations. IHC study can be also useful for confirmation of diagnosis. So ACC should be considered in the differential diagnosis of a primary lung tumor, even when present in the peripheral lung, although it is rare.

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Competing Interests

None declared

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