Case Report

Clinico-pathological review of ciliated muconodular papillary tumour of lung

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Abstract

Pulmonary tumours with cilia formation are rare and are usually recognized as benign. Ciliated muconodular papillary tumour (CMPT) of lung is a rare entity recently described in 2002. It is characterized by the presence of papillary tumour consisting of ciliated columnar, goblet, and basal cells. Although most of its features are of a benign tumour, some histological characteristics do suggest low-grade malignancy. We describe a case of CPMT in a 36 year old male.

Keywords: Ciliated, Papillary, Tumour, Lung

Introduction

Ciliated muconodular papillary tumour (CMPT) of lung is a rare entity recently described in 2002. It is characterized by the presence of papillary tumour consisting of ciliated columnar, goblet, and basal cells.

[1] It is not yet included in WHO classification of Lung tumours. Being a rare and newly described tumour, its clinical behaviour is not yet fully appreciated. [2] We describe a case of CPMT in a 36-year-old male.

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A 36 year old male presents with chief complain of productive cough and shortness of breath for 6 months. After thorough investigations, Computerized Tomograohy (CT) scan of chest was advised which showed diffuse nodular infiltrations in right middle lobe. (Figure 1) A transbronchial biopsy of right lung was performed which showed the presence of papillary structure lined by benign ciliated respiratory epithelium with basal cells and mucous cells along with fragments of alveolar tissue. (Figure 2, 3) Immuno histochemistry shows positivity for cytokeratin (CK) 7, Carcinoembryonic antigen (CEA) and Thyroid transcription factor 1 (TTF-1). A diagnosis of ciliated muconodular papillary tumour of lung was made. Patient has been referred to the higher centre for complete analysis and treatment.



Figure-1: Computerized Tomograohy (CT) scan of chest showed diffuse nodular infiltrations in right middle lobe.

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Figure-2: Presence of papillary structure along with fragments of alveolar tissue. (H&E 10X)

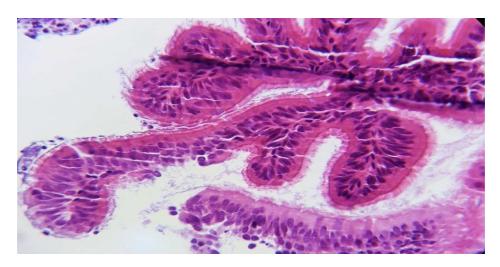


Figure-3: Papillary structure lined by benign ciliated respiratory epithelium with basal cells and mucous cells. (H&E 40X)

Discussion

CMPT of lung is a relatively new clinical entity described by Ishikawa in 2002, characterized by tubulopapillary, glandular or papillary architecture with tripartite morphology consisting of basal cells, mucinous cells, and ciliated cells involving the peripheral lung [1]. Pulmonary tumours with cilia formation are rare and are usually recognized as benign. Ciliated component has been described in the glandular component of the lung papillomas [3]. There are few case reports of presence of ciliated cells in very well differentiated adenocarcinoma [4].

CMPTs have been reported in elderly people, more common in men usually smokers, with median age of 62 years [5]. Our patient is a 36 years old man. CMPT has been reported in a teenage girl also [6].

Most of the tumors were incidentally detected. Radiologically they were interpreted as adenocarcinomas. Tumours are peripherally located and show irregular nodular appearance on CT scan ranging in size from 5 to 15 mm. Sometimes they may appear as

regular nodules or ground glass shadow [2, 5]. In our case there were diffuse nodular infiltrations in right middle and lower lobe.

Histpathologically, they are diagnosed by the presence of benign tubulopapillarytumor composed of ciliated columnar cells and goblet cells with extracellular mucin. Cells are positive for CEA, TTF-1and CK 7 and negative for CK20. Presence of basal cells can be confirmed by p63 or CK5/6. [1, 2, 5]

The malignant potential of CMPT remains unknown. Although most of its features are of a benign tumour, some histological characteristics do suggest low-grade malignancy. Presence of cilia and basal cells with benign nuclear and cytological features suggest its benign naturebut endo and peribronchial growth pattern with proliferation alongthe alveolarwalls and skip lesions, lack of encapsulationand staining for CEA are suggestive of malignancy. The immunohistochemical staining patterns for CK7, TTF-1, and CK20 are identical to pulmonary adenocarcinoma [7].

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CMPT must be differentiated from peribronchial metaplasia, glandular papillomas, well differentiated mucinous bronchoalveolar adenocarcinoma. Glandular lung papillomas if ciliated are difficult to be distinguished from CMPT. Their location is commonly central endobronchial but rarely theycan be peripheral bronchiolar types. These peripheral bronchiolar papillomas are similar to CMPT and distinguishing features between the two are unclear [8].

Peribron-chiolar metaplasia can be seen associated with various interstitial lung disorders. Adenocarcinomas can be distinguished by the presence of mucin-producing epithelium growing within the alveolar spaces and by the presence of nuclear and cytological atypia [2, 5].

CMPT cases are treated by wedge resection of the lung with wide freemargins. However, true malignant potential of this tumour is not known because of its rarity. Careful histological analysis should be done to exclude coexistent malignant tumours. More studies and data are needed for complete clinico-pathological analysis of this tumour [5].

Conclusion

Pulmonary tumours with cilia formation are rare and are usually recognized as benign. Ciliated muconodular papillary tumour (CMPT) of lung is a rare differential. True malignant potential of this tumour is not known because of its rarity and is occasionally misdiagnosed as lung adenocarcinoma.

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