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CASE REPORT

A surgical case of ciliated muconodular papillary tumor

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Abstract

Ciliated muconodular papillary tumor (CMPT) is a rare type of pulmonary tumor with mucoid features. Only a few cases confirmed by surgery have been reported worldwide. We report a case of CMPT following sublobectomy, with no recurrence detected in 10 months of follow-up. The accumulation of similar cases is essential for pathologists and surgeons to improve their understanding of such tumors.

Introduction

Ciliated muconodular papillary tumor (CMPT) is a rare type of pulmonary tumor with mucoid features. Only a few cases confirmed by surgery have been reported worldwide,[1–3] with more cases reported in East Asia than in Europe or America.[4] Because of the lack of typical clinical manifestations and image changes, it is difficult to differentiate benignity from malignancy and CMPT can easily be misdiagnosed as adenocarcinoma.[5] Herein we report a case of CMPT following sublobectomy, with no recurrence found during 10 months of follow-up.

Case report

A 67-year-old woman was initially detected with a nodule on the left upper lobe without any symptoms. No significant image changes were identified within three months of follow-up. She was admitted to our hospital for further evalutation on 2 January 2018. Contrast-enhanced computed tomography (CT) indicated an irregular solid nodule with a maximum diameter of 1.2 cm in the lingual segment of the left upper pulmonary lobe (Fig 1). No enlarged hilar and mediastinal lymph nodes were identified. The serum levels of tumor markers, including carcinoembryonic antigen, neuron-specific enolase, cytokeratin-19 fragment, squamous cell carcinoma antigen, and pro-gastrin releasing peptide were normal. No abnormalities were found in head magnetic resonance imaging, abdominal CT, or bone scan. We recommended positron emission tomography-CT (PET-CT) examination to the patient, but she refused because of the high cost. A multidisciplinary team performed comprehensive preoperative evaluation and highly suspected that the nodule was peripheral lung cancer. We performed thoracoscopic left pulmonary-lingual segmentectomy. Grossly, the tumor was solid, grey-white, with a

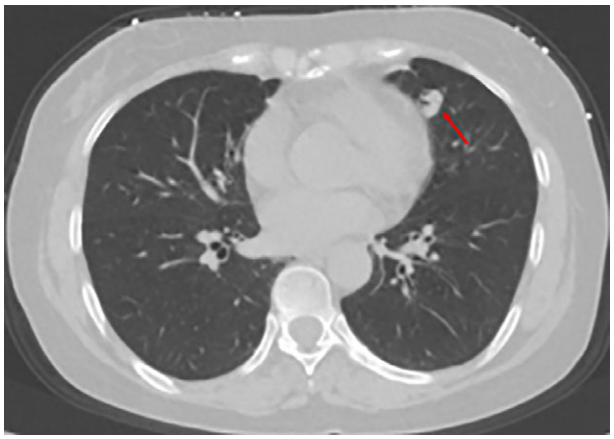

Figure 1 Contrast-enhanced computed tomography indicated an irregular solid nodule with a maximum diameter of 1.2 cm in the lingual segment of the left upper pulmonary lobe.

soft texture and clear boundaries. An intraoperative frozen section indicated a benign lesion therefore we did not perform hilar and mediastinal lymph node dissection.

Postoperative pathological examination showed that the tumor cell consisted of proliferated epithelium with adenoid and papillary structures, including ciliated columnar cells, mucous cells, and basal cells. Ciliated columnar cells were coated on the surface of adenoid or papillary structures. Basal cells were located in the outer layer. The alveolar cavity around the lesion was filled with mucus. Tumor cells showed no atypia, mitosis or necrosis (Fig 2). The immunohistochemical profile of the ciliated columnar cells and mucous cells showed that CK 7 was positive; TTF-1 was weakly positive; and CK20, CD56, NCAM1, MUC5AC, Napsin A and P63 were negative.

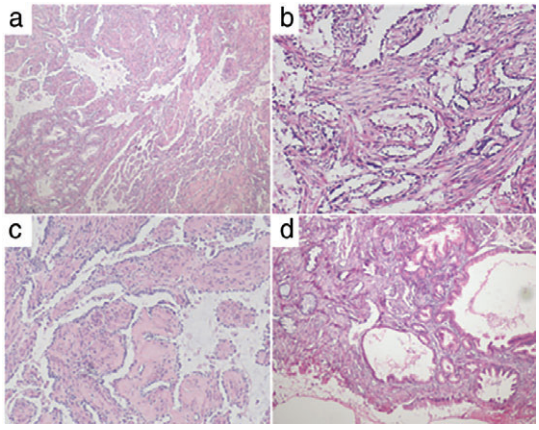

Figure 2 (**a**) Tumors exhibited papillary and glandular components. (**b**) A large number of ciliated cells is observed. (**c**) The tumor cells consisted of ciliated columnar cells, basal cells, and scattered mucous cells. (**d**) The mucous glands consisted of mucous cells.

CMPT showed a low Ki-67 proliferating index (Fig 3). Finally, the case was pathologically diagnosed as CMPT.

The patient recovered unevenly after surgery, and no recurrence was found within 10 months of follow-up.

Discussion

In 2002, Ishikawa *et al.* first reported a case of CMPT, which was characterized by tripartite cellular components with a papillary-predominant structure including ciliated columnar cells, mucinous cells, and basal cells.[6] This tumor is extremely rare, and has not been included in the World Health Organization classification of lung tumors.

CMPT does not present any symptoms and is usually detected during rountine physical examination. It can also be accompanied by some common and non-specific respiratory symptoms. CT findings of CMPT patients are mostly atypical solitary peripulmonary nodules. For pulmonary nodules with a diameter of the solid component of > 8 mm, PET-CT is helpful in differentiating benignity from malignancy.[7] However, high cost restricts the wide clinical use of PET-CT. Furthermore, it is also challenging to perform a non-operative biopsy, including bronchoscopy and percutaneous puncture. Therefore, CMPT is commonly misdiagnosed as lung cancer by surgeons and radiologists before surgery. A multidisciplinary team, including thoracic surgeons, radiologists, oncologists, and pulmonologists, should be organized to comprehensively identify the optimal diagnostic and therapeutic strategy.

The key to a diagnosis of CMPT is to identify: (i) ciliated columnar cells and basal cells; (b) papillary tumors accompanied with mucus production; and (c) tumor cells without atypia, nuclear division, or necrosis. Usually, the immunohistochemical profile of the ciliated columnar cells and mucous cells shows positive CK7 and negative CK20. Some studies have reported focally weak TTF-1 expression, while others have indicated no TTF-1 expression in CMPT. In this patient, the basal cells expressed P63 and P40; the ciliated columnar cells partly expressed MUC5AC, while mucous cells did not; and they all had a low Ki-67 proliferating index in general.[8–11]

No consensus has been reached on the benignity or malignancy of CMPT. Some scholars believe that CMPT may be a precancerous lesion or a low-grade malignant tumor of mucinous adenocarcinoma, because of the histological and molecular biological characteristics related to malignancy.[2,5,9,12] However, most research considers CMPT as a benign lesion, given that the pulmonary tumors with cilia are commonly benign.[1,7]

Local resection is the mainstream treatment for CMPT patients. However, because of its clinical rarity and the lack of uniform diagnostic criteria, pathologists continue to try and identify predicitive factors. If CMPT is misdiagnosed

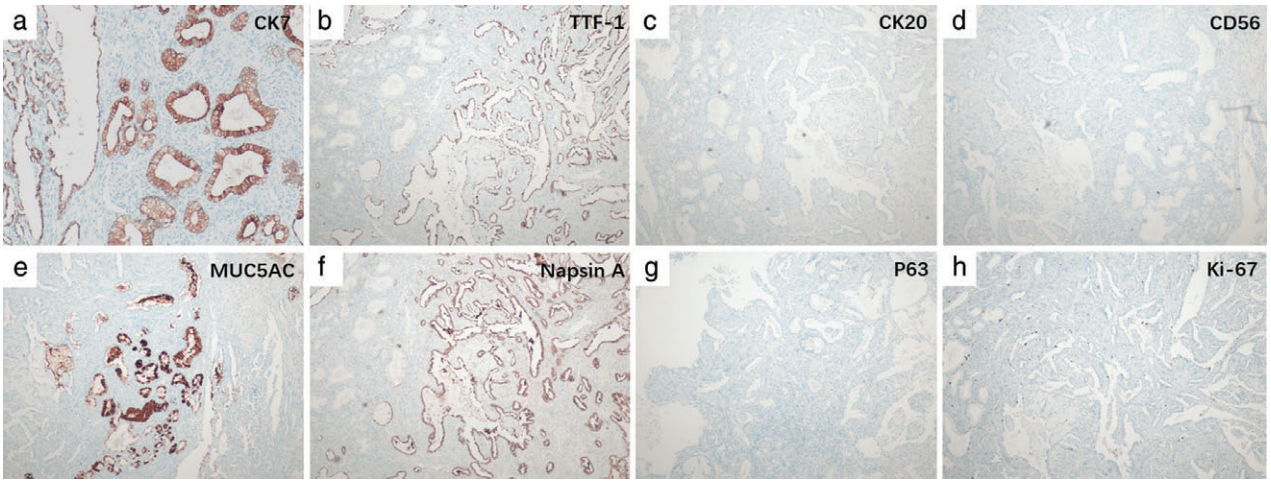

Figure 3 The immunohistochemical profile of the ciliated columnar cells and mucous cells showed that CK7 (**a**) was positive; TTF-1 (**b**) was weakly positive; and CK20 (**c**), CD56 (**d**), MUC5AC (**e**), Napsin A (**f**) and P63 (**g**) were negative. The ciliated muconodular papillary tumor showed a low Ki-67 proliferating index (**h**).

as lung adenocarcinoma in intraoperative frozen sections or malignancy cannot be excluded, lobectomy and mediastinal lymph node dissection are considered.[1,5,8] In this case, a malignant tumor was highly suspected before surgery. Lung segmental resection can achieve radical tumor resection and avoid unnecessary surgery.

In recent years, segmental resection has been widely performed for early-stage lung cancer. When a pulmonary nodule is highly suspected as malignant before surgery and meets segmental resection indications, segmental resection can be considered to achieve complete resection. The decision over whether to dissect mediastinal lymph nodes is made according to intraoperative frozen sections.

In summary, CMPT is a rare type of lung cancer. It usually presents in CT examination as an accidental lung nodule. According to the current evidence, CMPT tends to be benign, but more case data is required to determine long-term biological behavior. The accumulation of similar cases is essential for pathologists and surgeons to improve their understanding of such tumors.

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Disclosure

Authors report no conflict of interest.

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