


CASE REPORT

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# A surgical case of ciliated muconodular papillary tumor of the lung

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## Abstract

Ciliated muconodular papillary tumor (CMPT) is a rare type of tumor with both benign and malignant characteristics. Herein, we report the surgical case of a 65-year-old man with CMPT. Chest computed tomography revealed a solitary cavitory lesion with a maximum diameter of 11-mm in S<sup>10</sup><sub>b</sub> of the right lower lung. A thoracoscopic lung wedge resection was subsequently performed. On microscopic examination, the tumor was composed of highly columnar cells with tubular-to-papillary and cystic growth patterns. On immunostaining, it was positive for the epidermal growth factor receptor. The tumor was diagnosed as a CMPT, exhibiting no recurrence after two years of follow-up.

**Keywords** Ciliated muconodular papillary tumor, Lung nodule, EGFR

## Introduction

Ciliated muconodular papillary tumor (CMPT) is a rare lung tumor that was first reported in 2002. This tumor involves ciliated, goblet, and basal cell proliferation with mucin secretion [1]. On computed tomography (CT), CMPT commonly presents as ground-glass opaque peripheral nodules, while cavitory formation is rarely observed. This study presents a surgical case of CMPT in a 65-year-old man with a cavitory lesion in the right lower lung field identified on chest CT. The diagnostic pitfalls and considerations are further discussed.

## Case

A 65-year-old man had been referred to our hospital for evaluation 4 years prior, after the detection of a cavitory

lesion on chest CT during routine health screening. He had a history of hyperlipidemia and impaired glucose tolerance. Laboratory examination revealed elevated triglyceride and LDL-cholesterol levels. The serum levels of the tumor markers carcinoembryonic antigen (CEA), pro-gastrin-releasing peptide, and squamous cell carcinoma antigen were within the normal limits. CT revealed a solitary cavitory lesion with a maximum diameter of 11 mm in S<sup>10</sup><sub>b</sub> of the right lower lung (Fig. 1). <sup>18</sup>F-fluorodeoxyglucose (FDG) positron emission tomography (PET) revealed no abnormal FDG accumulation in the tumor or lymph nodes. Thoracoscopic lung wedge resection was subsequently performed under one-lung anesthesia to verify the presence of a primary malignancy (Fig. 2). The resected specimen showed an 11-mm cavity, and intraoperative analysis of the frozen section revealed only inflammatory changes; therefore, additional lobectomy was not performed. On microscopic examination, the tumor was composed of tall columnar cells, exhibiting tubular-to-papillary and cystic growth patterns (Fig. 3a, b). Lymphocyte infiltration was further observed in the interstitium. On immunohistochemical staining, basal cells were positive for p40, and alveolar epithelial cells were positive for thyroid transcription factor 1 (TTF-1)

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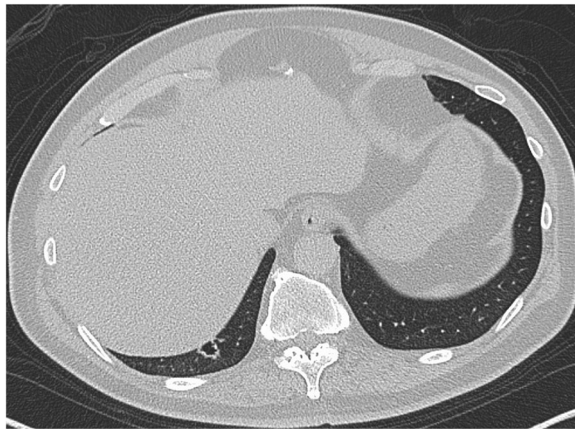
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**Fig. 1** Chest computed tomography (CT) showing a cavitary lesion in S10b of the right lower lobe

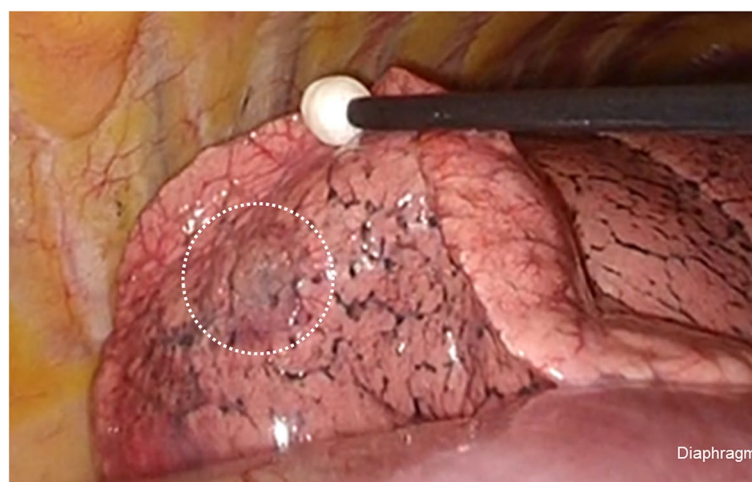
(Fig. 4a, b). Tall columnar cells strongly expressed epidermal growth factor receptor (EGFR) and v-raf murine sarcoma viral oncogene homolog B1 V600E (BRAF V600E) and anaplastic lymphoma kinase (ALK) were not expressed (Fig. 4c–e). The final pathological diagnosis was CMPT. The postoperative course was uneventful, with no recurrence 2 years later.

## Discussion

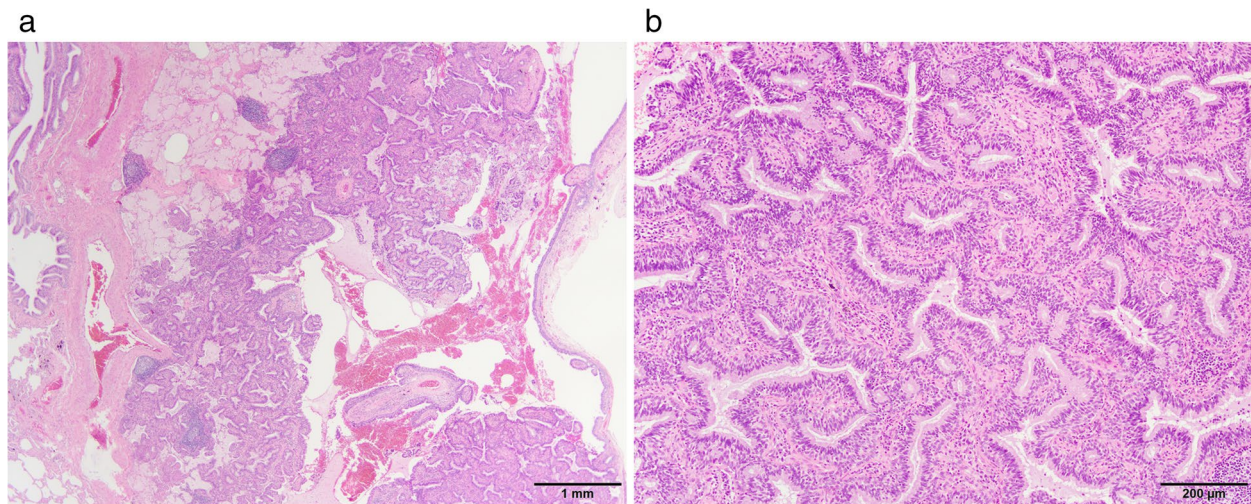
CMPT (which was first reported in 2002) is a rare tumor that develops in the peripheral lung. This lesion exhibits papillary growth during alveolar replacement, and ciliated and goblet cells are accompanied by mucus production [1]. CMPT was classified as an adenoma in the 5th edition of the WHO classification [2]; however, CMPT has been described as having both benign and malignant

features. The presence of cilia, basal cells, and a low Ki-67 index are all indicators of a benign tumor. Conversely, findings, suggestive of a malignant tumor include a ruptured alveolar structure, central fibrosis, proliferation along the alveolar walls and skip lesions, lack of encapsulation, micropapillary patterns, and positive staining for CEA. The immunohistochemical findings of this tumor are similar to those of lung adenocarcinoma. CMPT is typically positive for CEA, TTF-1, and cytokeratin (CK) 7, but negative for CK20 [3, 4]. Although the malignant potential of CMPT remains unknown, some studies have associated it with various gene alterations. Recently, EGFR, ALK, AKT1, KRAS, BRAF, and HRAS gene mutations were all detected via gene analyses among patients with CMPT [5–7]. These findings are suggestive of a neoplastic change. Thus, their malignant potential requires further investigation.

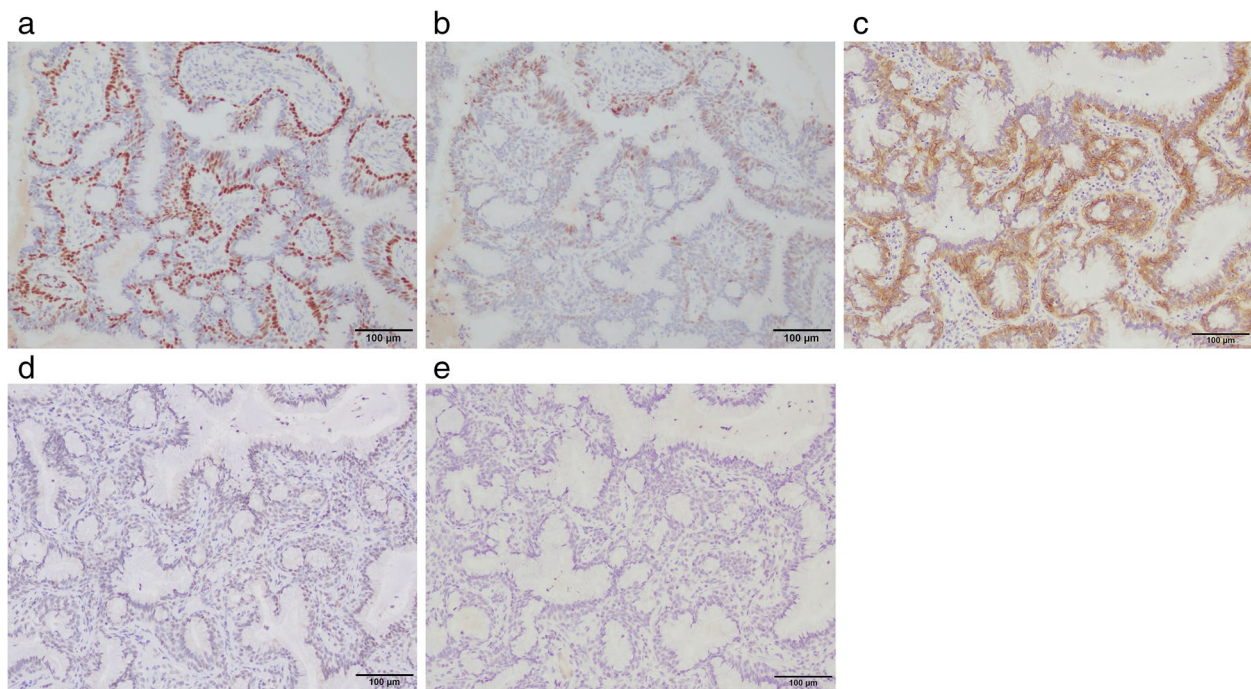
CMPT is frequently detected incidentally as a small, peripheral, ground-glass opacity or nodule on CT. In a previous study examining 38 cases of CMPTs (17 men and 21 women), the mean age at identification was 68 years (range 50–84 years), and the average tumor diameter was 1.1 cm (range 0.4–4.5 cm) [8]. Most CMPT lesions developed in the peripheral lesions of the lung with approximately 79.4–81.5% occurring in the lower lobe [8, 9]. CT findings included nodular shadows in 28 patients (73.7%) and hollow shadows and ground-glass-like shadows in the remaining 10 (26.3%). In another study, the CT findings of 16 patients with CMPTs, including eight (50.0%) with solid nodules, seven (43.8%) with high-density ground-glass nodules (GGNs), and one (6.3%) with pure GGNs, were examined. CMPTs grew very slowly at 0.49 mm/year in maximal diameter.



**Fig. 2** Intraoperative findings revealed the nodule (white dotted circle) in the dorsobasal segment of the right lung. The photograph was extracted from the digital video data



**Fig. 3** Hematoxylin and eosin staining showed that the tumor was composed of tall columnar cells exhibiting a tubular-to-papillary and cystic growth pattern. **A** Low-power field. **B** High-power field



**Fig. 4** Immunohistochemistry. **a** Basal cells were positive for p40. **b** Alveolar epithelial cells were positive for TTF-1. **c** Tall columnar cells strongly expressed EGFR. Conversely, **d** BRAF V600E and **e** ALK expression was not observed

Moreover, 10 out of 16 lesions (62.5%) were found near the pleura [9]. Based on this, it may be difficult to distinguish CMPT from lung cancer, when pleural changes are present.

FDG-PET findings of 15 patients with surgically resected CMPTs showed moderate FDG uptake in one patient, with the maximum standardized uptake values

(SUVmax) of 3.67. The remaining 14 patients showed mild FDG uptake, with the SUVmax ranging from 0.57 to 1.35. According to the relationship between FDG accumulation and pathological lymphocyte infiltration, cases with mild FDG accumulation exhibit minimal lymphocyte infiltration and large amounts of mucin.

Meanwhile, cases with moderate FDG accumulation exhibit significant lymphatic infiltration [10].

Since CMPT often presents as a small nodule in the peripheral lesion of the lung, it is difficult to diagnose on bronchoscopy. As such, surgical resection is often performed to confirm the diagnosis. In 38 CMPT cases, partial resection was performed in 27 (71.1%), while segmentectomy was performed in one, and lobectomy was performed in 10 [8]. When sufficient excision margins are obtained in the excision range, a partial excision is considered curative [11]. In this case, the maximum tumor diameter was 11 mm, and malignant findings were not observed in the intraoperative analysis of the frozen sections. Therefore, a sufficient resection margin was secured, and the operation was successfully completed via partial resection.

The prognosis for CMPT is good, and no recurrence or metastasis was reported during the mean follow-up period of 33.8 months [8]. Since the pathological findings of CMPT exhibit both benign and malignant characteristics, careful follow-up is necessary. In conclusion, this study reports a rare case of CMPT, which was managed by thorascopic lung wedge resection, with no recurrence detected in 2 years of follow-up.

#### Abbreviations

CMPT	Ciliated muconodular papillary tumor
CT	Computed tomography
CEA	Carcinoembryonic antigen
FDG	18 F-fluorodeoxyglucose
PET	Positron emission tomography
EGFR	Epidermal growth factor receptor
ALK	Anaplastic lymphoma kinase
CK	Cytokeratin
AKT1	Serine/threonine kinase 1
KRAS	v-Ki-ras2 Kirsten rat sarcoma viral oncogene homolog
BRAF	v-raf murine sarcoma viral oncogene homolog B1
SUVmax	Maximum standardized uptake values

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Not applicable.

#### Authors' contributions

Hiroaki Shidei, Hiroe Aoshima, and Masato Kanzaki conceived of this study design. Hiromi Onizuka, Yoji Nagashima, and Yukio Nakatani participated in providing the pathological findings. All authors performed this study and collated this manuscript as a group effort. Hiroaki Shidei wrote the first draft of the manuscript. Hiroe Aoshima assisted Hiroaki Shidei in preparing the manuscript. All authors read and approved the final manuscript.

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#### Availability of data and materials

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#### Declarations

#### Ethics approval and consent to participate

Not applicable.

#### Consent for publication

Informed consent was obtained from the patient for the publication of this case report and the accompanying images.

#### Competing interests

The authors declare that they have no competing interests.

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