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CASE REPORT

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Ciliated muconodular papillary tumor of the lung with cavitary change: A case report with 11-year preoperative follow-up

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Abstract

Ciliated muconodular papillary tumor (CMPT) is a rare benign lung tumor characterized by ciliated columnar cells, mucous cells, and basal cells. Herein, we report a case of CMPT with 11 years of preoperative follow-up, depicting the natural course of the tumor and changes in computed tomography (CT) findings. A 39-year-old man had a 5-mm solid pulmonary nodule in the right lower lobe that had slowly grown and transformed into a thin-walled cavitary lesion. Right lower lobe lobectomy was performed and the tumor was confirmed to be a CMPT. Although it is difficult to diagnose CMPT with CT findings alone, CMPT should be considered as a possible diagnosis when a slowly growing nodule undergoes cavitary changes.

KEYWORDS

computed tomography, follow-up studies, lung neoplasms

INTRODUCTION

Ciliated muconodular papillary tumor (CMPT) is a rare benign tumor, with only 65 cases reported to date in English literature[1] since the first case was reported by Ishikawa in 2002.[2] Histologically, the tumor consists of ciliated columnar, mucous, and basal cells, with a predominant papillary structure.[3] The tumor appears as a solitary small peripheral ground-glass nodule or solid nodule on computed tomography (CT) imaging. Although the tumor is known to be a slow-growing benign tumor,[4] most of the reported cases have described only postoperative follow-up and recurrence. To date, long-term follow-up studies that show the natural course of the tumor have rarely been reported. Herein, we report a rare case of CMPT showing cavitary change with an 11-year follow-up period.

CASE REPORT

A 39-year-old man with epigastric pain was referred to our hospital. Abdominal CT revealed an incidental finding of a solid pulmonary nodule with a maximum diameter of 5 mm in the right lower lobe (RLL). Because the nodule was small, a specific disease could not be identified (Figure 1a). The patient underwent regular image follow-up and the nodule showed a slight increase in size during the follow-up period. On the CT image at 8 years of follow-up, the nodule showed a new central cavitary change and the maximum diameter of the nodule increased to 7 mm (Figure 1b). Three years later, the size of the nodule further increased to 10 mm, and the cavitary wall became slightly thinner on chest CT (Figure 1c). No fluorodeoxyglucose (FDG) uptake was observed on FDG positron emission tomography (PET)/CT. The nodule slowly grew, with a 40% increase in the longest diameter over 8 years and a doubling of the longest diameter over 11 years. Video-assisted wedge resection was performed for diagnosis. Initially, the tumor was misdiagnosed as mucinous adenocarcinoma in intraoperative frozen section analysis due to the proliferation of mucin-secreting tumor cells with an abundant mucin pool, therefore RLL lobectomy was performed. The final diagnosis was confirmed as CMPT in the postoperative analysis. The tumor showed abundant mucous lakes (Figure 2a) and a mixture of ciliated columnar and basal cells with papillary structures interspersed between the mucous lakes (Figure 2b). The

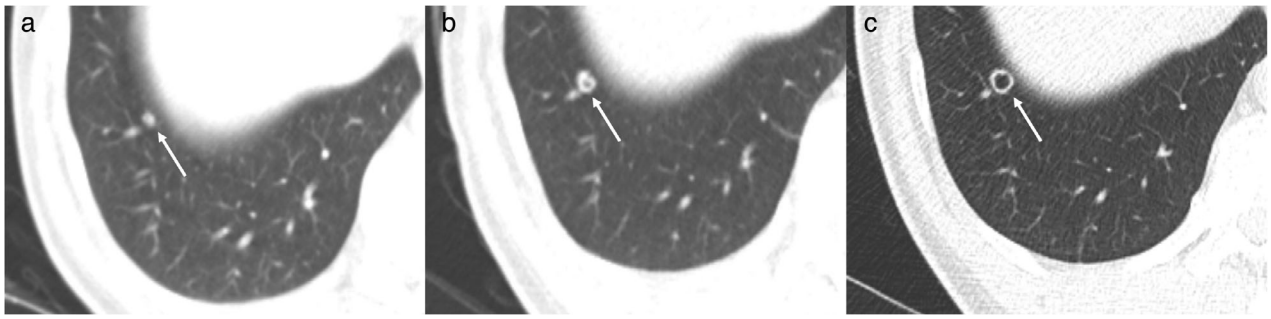

FIGURE 1 Initial computed tomography (CT) image showing (a) 5-mm size oval shape nodule in right lower lobe. (b) Chest CT image taken after 8 years depicting that the maximum diameter of the nodule increased to 7 mm. (c) Chest CT image taken after 11 years depicting that the maximum diameter of the nodule further increased to 10 mm. The internal cavity became larger and the cavitary wall became thinner

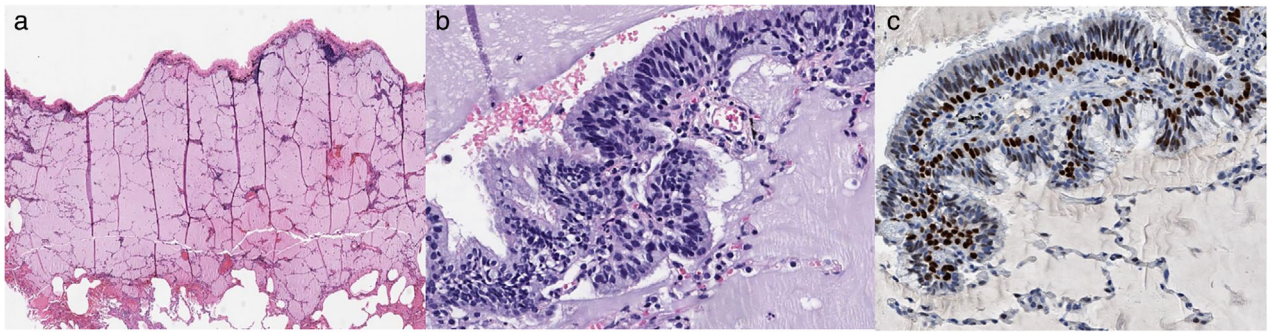

FIGURE 2 Histopathological findings of the case. (a) The tumor showing abundant mucous lakes (hematoxylin & eosin [H&E] ×25). (b) The tumor consists of ciliated columnar cells with papillary structure surrounded by extracellular mucin in high-power examination (H&E ×400). (c) Basal cells were stained with p40. There was no definite nuclear atypia or mitosis

TABLE 1 Summary of CT findings in previous reports and present case

		CT finding				
Author	**Age/sex**	**Lesion characteristics**	**Cavity**	**Size (mm)**	**Location**	**Treatment**
Wang et al.[1]	64/F	Solid nodule	−	12	RLL	N/A
Kamata et al.[3]	56–78 (median age: 62)/7 men and 3 women	Solid or part-solid nodule	+	10 (on average)	RUL ($n = 1$), RLL ($n = 5$), LLL ($n = 4$)	Partial resection ($n = 9$) Lobectomy ($n = 1$)
Onishi et al.[4]	70.7 (mean age)/ 10 men and 6 women	Ground glass nodule ($n = 8$) Solid nodule ($n = 8$)	−	9.1 (on average)	RML ($n = 1$), RLL ($n = 8$), LUL ($n = 2$), LLL ($n = 5$)	N/A
Chu et al.[7]	56/M	Solid nodule	−	11	LUL	Partial resection
Chuang et al.[8]	68/M	Ground glass nodule	−	7	RLL	Partial resection
Hata et al.[9]	76/F	Solid nodule	−	7	LUL	Lobectomy
Kon et al.[14]	80/M	Solid nodule	+	7	LLL	Partial resection
	67/M	Solid nodule	−	10	RLL	Partial resection
	66/M	Solid nodule	+	13	RLL	Partial resection
	73/F	Solid nodule	+	9	LUL	Partial resection
	70/F	Solid nodule	−	8	RLL	Partial resection
Matsushima et al.[15]	60/M	Ground glass nodule	−	4	LLL	Partial resection
Murakami et al.[16]	70/F	Solid nodule	+	2	RLL	Partial resection
Present	50/M	Nodule	+	5	RLL	Partial resection

Abbreviations: RLL, right lower lobe; RUL, right upper lobe; LLL, left lower lobe; RML, right middle lobe; LUL, left upper lobe; N/A, not available.

basal cells were stained with p40 (Figure 2c). No nuclear atypia or mitotic figures were observed. No tumor recurrence was noted during the 15-month postoperative follow-up period.

DISCUSSION

CMPT is a rare benign tumor and newly classified in the 2021 World Health Organization classification of thoracic tumors.[5] The tumor is usually observed in middle-aged and elderly patients with an average age of 70.1 years.[1] The tumor occurs in both males and females, with a very slight male predominance (M:F ratio = 1.10:1). Most of the described cases have originated in Asia. Approximately half of the patients had a history of smoking. Patients are usually asymptomatic and the tumor is usually found incidentally.[1]

Pathologically, the tumor exhibits trilineage differentiation with ciliated columnar cells, mucous cells, and basal cells and shows papillary growth.[3] Mucus is observed in the periphery or center of the tumor.[3] Occasionally, the tumor has been misdiagnosed as a mucinous adenocarcinoma on intraoperative frozen section analysis. Shirsat et al. reported that only 16.7% of cases were diagnosed as CMPT during intraoperative frozen studies.[6] It is challenging to distinguish CMPT from mucinous adenocarcinomas in frozen slides because of the abundant mucous lakes and the difficulty in recognizing ciliated columnar cells in the small floating cells.[6,7] However, CMPT does not represent nuclear atypia or mitotic activity, as against mucinous adenocarcinoma.[8,9]

A distinctive molecular feature of the tumor is a single driver mutation. BRAF driver mutation is the most common and other alterations involve EGFR, KRAS, ALK, and AKT1. This molecular profile is similar to that of bronchiolar adenoma and supports a nosological relationship between bronchiolar adenoma and CMPT.[5,10–12]

Radiologically, CMPTs usually appear as peripheral lung nodules, most of which are located in the lower lobes. The average size of CMPTs reported in the literature is 11.8 mm, ranging from 4 to 45 mm.[1] CMPTs can vary from ground-glass nodules to solid nodules on CT images (Table 1). CMPTs presenting as ground-glass nodules contain more mucin than those presenting as solid nodules.[4] Internal cavities have been reported in a few cases, and mucous lakes are likely to be found as cavities.[13–16] CMPTs show no or moderate increase in FDG uptake on PET/CT.[17]

CMPT is a slow-growing tumor, but little is known about its growth rate. In the present case, the maximum diameter increased from 5 to 10 mm in 11 years, with a growth rate of 0.45 mm/year. This is similar to a previously reported rate of 0.49 mm/year.[4] This rate is slower than the growth rate of hamartomas, which is 3.2 mm/year,[18] and this reflects the benign nature of CMPT.

To the best of our knowledge, there has been only one case of long-term preoperative follow-up that showed the natural course of the tumor. In the case of a patient with 12 years of follow-up reported in 2020, the tumor initially appeared as a ground-glass nodule, then increased in density to become a solid nodule and a central cavity eventually developed.[15] Similarly, in this case, a central cavitary change was observed in the solid nodule during the follow-up period. In the present case, the tumor had abundant mucous lakes, and as the tumor grew, the mucous lakes formed the central cavity.[14]

Cavitation can appear in either benign or malignant tumors. Although smooth and thin cavitary walls (<5 mm in thickness) are characteristic of benign lesions,[19] it is difficult to accurately evaluate the thickness of the cavitary wall in small nodules <10 mm. Moreover, Shi et al. described cavitation as a risk factor for malignancy in tumors <10 mm,[20] therefore malignancy cannot be completely ruled out in cases of cavitary changes in small nodules, and the diagnosis of CMPT is challenging if based on CT imaging alone. This long-term follow-up case shows that CMPT can be considered as a differential diagnosis when a slowly growing nodule undergoes cavitary change.

In conclusion, CMPT is a rare benign tumor with a slow growth rate that can exhibit cavitary change. Radiologically, it is difficult to diagnose CMPT. However, it is important to keep CMPT in mind when there is a slow-growing nodule with cavitary change on chest CT imaging.

CONFLICT OF INTEREST

The authors declare no potential conflicts of interest.

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