CASE REPORT

Pulmonary mucinous cystadenocarcinoma: an extremely rare tumor presenting as a cystic lesion of the lung

Teruo Iwasaki, MD · Kunimitsu Kawahara, MD · Teruaki Nagano, MD · Katsuhiro Nakagawa, MD

Abstract
Pulmonary mucinous cystadenocarcinoma (PMC) is a rare tumor characterized by mucin production. It is similar to tumors of the same name arising in the ovaries and pancreas. Here we describe the 20th case of PMC reported in the English literature. The patient was a 75-year-old woman with a 3-day history of bloody sputum. Chest radiography and computed tomography revealed a cavitary mass 5 cm in diameter in the posterior segment of the right lung. 18F-fluorodeoxyglucose positron emission tomography demonstrated intense uptake in the wall of the lesion. Right lower lobectomy was performed, and the pathology examination revealed this tumor to be a PMC. The preoperative serum CA 19-9 level was 162.3 U/ml (cutoff 37 U/ml) and decreased to 22.8 U/ml after resection. No mutation of epidermal growth factor receptor or K-ras gene was detected. Thoracic surgeons should bear in mind this rare tumor for the differential diagnosis of a pulmonary cystic lesion.

Key words
Pulmonary mucinous cystadenocarcinoma · 18F-Fluorodeoxyglucose positron emission tomography · Prognosis · Surgery

Introduction
Pulmonary mucinous cystadenocarcinoma (PMC), so designated by Devaney et al. in 1989, is a cystic adenocarcinoma with copious mucin production; it may resemble tumors of the same name arising in the ovaries, breast, and pancreas. Preoperative diagnosis is difficult, and its prognosis is controversial. Thoracic surgeons should keep in mind this rare tumor for the differential diagnosis of a cavitary or cystic lesion of the lung.

Case
A 75-year-old nonsmoking woman with a 3-day history of bloody sputum was admitted for evaluation. On admission, no abnormalities were found on physical examination. Serum carcinoembryonic antigen (CEA) and cytokeratin-19 fragment levels were within normal limits, but the serum CA 19-9 level was 162.3 U/ml (cutoff 37 U/ml). She had mild obstructive ventilatory impairment. Microbiological and cytological examinations of the sputum showed nothing of particular significance. A chest radiograph revealed a cavitary mass 5 cm in diameter overlapping the hepatic shadow (Fig. 1A). Chest computed tomography (CT) disclosed a thick-walled cavitary mass with an irregular margin in the posterior segment of the right lung (Fig. 1B). 18F-fluorodeoxyglucose positron emission tomography (FDG-PET) and fused positron-emission tomography (PET/CT) images demonstrated intense uptake (maximum standardized uptake value 6.7) in the wall of the lesion (Fig. 1C). No other abnormal uptake of FDG was detected. The mass
was diagnosed as adenocarcinoma by bronchoscopic biopsy.

Right posterolateral thoracotomy through the fifth intercostal space and a right lower lobectomy with mediastinal lymph node dissection were performed. The specimen was a fibrous-walled cyst $4.8 \times 3.8$ cm in diameter containing mucus and blood located in the subpleural area (Fig. 2A). Solid areas were found in the periphery of the cyst. Microscopically, the cyst consisted of a fibrous capsule and abundant extracellular mucin; the capsule and multiple septa were lined by tall columnar mucinous tumor cells with basally oriented nuclei (Fig. 2B). Periodic acid-Schiff (PAS) and alcin blue stains revealed strong positivity for mucin. The solid areas exhibited features of invasive adenocarcinoma (Fig. 2C). We diagnosed this tumor as PMC (pathological T2N0M0, stage IB). Immunohistochemically, CEA and CA 19-9 were focally expressed in tumor cells, and the mitotic index determined using anti-Ki-67 antibody was 10%. No mutation of the epidermal growth factor receptor (EGFR) gene in exons 19–21 or K-ras gene was detected.

The postoperative course was uneventful. The patient has now been followed for 14 months and has shown no signs of recurrence. The serum CA 19-9 level decreased to 22.8 U/ml.

Discussion

The PMC is considered part of a spectrum of mucinous cystic neoplasms, including mucinous cystadenoma and mucinous cystic tumor of borderline malignancy. PMC differs from these other entities in that the lining epithelium exhibits frankly malignant cytologic features or that solid invasive areas are present.

We screened the English literature based on the PubMed database and found that 19 cases of PMC had been reported so far, although some controversy exists over the number of PMC cases except that of borderline-malignant cases. The characteristics of PMC cases including ours are summarized as follows. There were 11 men and 9 women who ranged in age from 29 to 75 years (median 64 years). Twelve (63%) patients

![Fig. 1 A Chest radiograph showing a thick-walled cavitary mass (arrowheads) in the right lower lung field overlapping the hepatic shadow. B Chest computed tomography (CT) showing a thick-walled cavitary mass with an irregular margin in the posterior segment. C Combined $^{18}$F-fluorodeoxyglucose positron emission tomography and CT images demonstrating abnormally intense uptake (arrows) in the wall of a right pulmonary lesion](image)