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MUCOEPIDERMOID CARCINOMA OF THE TRACHEOBRONCHIAL TREE

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Abstract

Primary salivary type lung cancers are extremely rare intrathoracic malignancies. Mucoepidermoid tumor is one of the salivary gland tumors that originates from submucosal glands of the tracheobronchial tree. These are very slow-growing low-grade malignant tumors. Surgery is the mainstay of treatment and rarely requires adjuvant therapy. In this case report we describe a 65-year-old woman who presented with a solitary cough yet on further investigation was found to have a mucoepidermoid tumor originating from the hilum of the left lung.

Introduction

Mucoepidermoid carcinoma (MEC) is derived from the submucosal glands of the tracheobronchial tree and bears structural homology with exocrine salivary glands. It is a rare-occurring tumor comprising only 0.1% to 0.2% of primary lung malignancies.[1] Although considered an indolent tumor, local invasion and lymph node metastases may occur. Computed tomography (CT) usually reveals a solitary nodule or an endobronchial mass with or without postobstructive pneumonia or atelectasis.[2] Imaging by ^{18}fluorodeoxyglucose (18F-FDG) positron emission tomography (PET)-CT usually demonstrates increased uptake in lung malignancies.[3] Complete surgical resection is the treatment of choice and is associated with excellent prognosis. Herein, we report our experience with a patient with an endobronchial lesion that was diagnosed as mucoepidermoid carcinoma, with cough being the solitary presenting symptom.

Case Report

The patient is a 65-year-old woman with a longstanding history of smoking who presented with solitary cough. She was evaluated by her primary care physician and found to have a left lung mass. Multiple tissue samples taken during bronchoscopy showed only suspicious cells with no concrete evidence of malignancy. An endobronchial ultrasound-directed biopsy was then performed but failed to identify any malignant cells. We decided to proceed with an ^{18}F-FDG PET scan, which revealed a 5.8 cm mass at the hilum with a markedly elevated standardized uptake value of 9 (Figure 1). The mediastinum and other areas in the body did not show any evidence of involvement. A split perfusion study showed that 91% of perfusion was to the right lung and only 9% was to the left lung. The decision was made to perform a mediastinoscopy followed by a left thoracotomy if lymph node biopsies were negative.

In the operating room, a small transverse incision was made in the suprasternal notch, and the video mediastinoscopy was inserted into the pretracheal plane. Careful dissection was performed down to the level of the carina. Subcarinal nodes were biopsied, and pathology showed these to be negative. No lymph nodes were seen in the left paratracheal region. The incision was closed, and a left thoracotomy was then performed. After adhesions between the lung and the chest wall were taken down, the centrally located mass was easily felt along with the

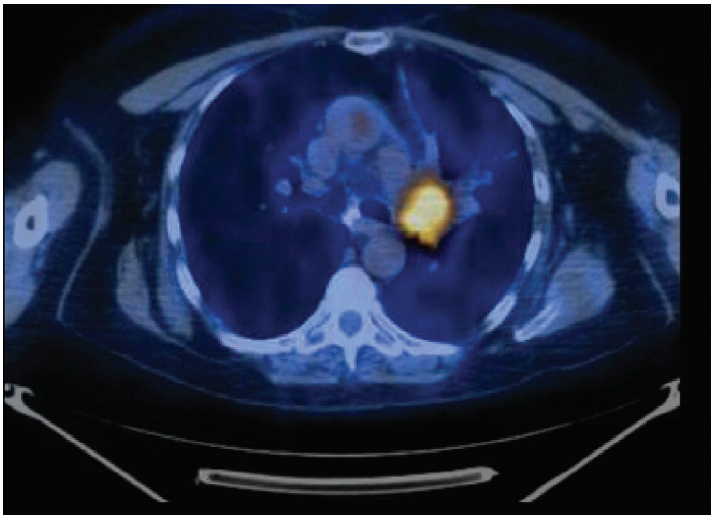

Figure 1. Positron emission tomography–computed tomography image revealing a 5.8 cm mass at the hilum of the left lung.

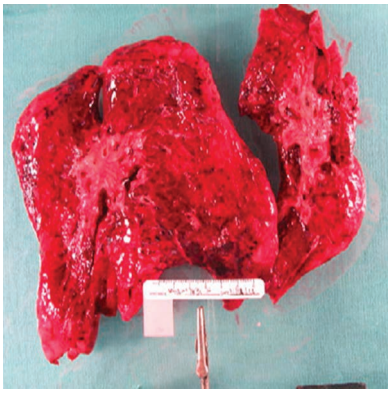

Figure 2. A cut surface of the mucoepidermoid carcinoma showing gray-white-tan with a glistening mucoid texture.

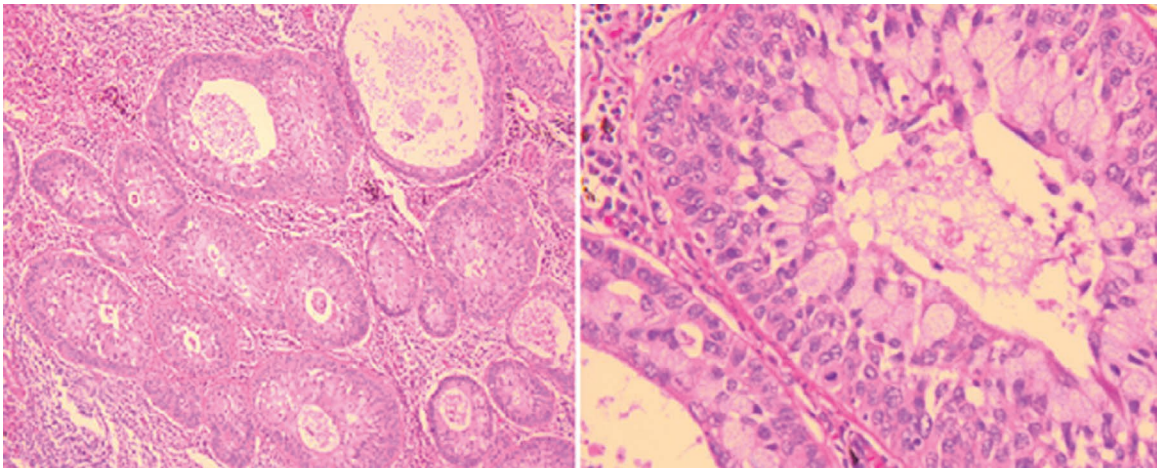

Figure 3. Histologically, the left lung mass shows a mucoepidermoid carcinoma characterized by infiltrating nests of tumor cells with peripheral palisading of squamoid cells and mucin-containing goblet cells in the center. Rare, small, mucin-containing cysts are seen. No obvious keratinization is identified. The tumor invades the bronchus, peribronchial tissue, cartilage, and lung parenchyma.

hilum, which had a hard, woody feel (Figure 2). The pleural reflection was incised, and the mass appeared to be growing into the aortopulmonary window. It was shaved off the aorta by removing the adventitia. Extensive lymph node sampling did not reveal any regional spread. It was not possible to encircle the pulmonary veins within the chest, therefore the pericardium was opened. The superior and inferior pulmonary veins were encircled within the pericardium and transected with a Covidien vascular stapler. Similarly, the main trunk of the left pulmonary artery was transected. The bronchial resection margin was negative by frozen section. Finally, the left main stem bronchus was transected with a stapler. Her postoperative course was uneventful and did not require any adjuvant therapy.

As part of our assessment, we performed a follow-up imaging study that included a CT scan 6 months following the surgical resection, and there has been no evidence of any mediastinal mass. The patient continues to do well 19 months following the surgery.

Discussion

Mucoepidermoid carcinoma is an uncommon lesion accounting for less than 1% of primary malignant bronchial tumors. Although generally indolent, local invasion and lymph node metastases may occur. The tumor generally affects patients age 30 years and older. Patients usually present with cough, hemoptysis, wheezing, and recurrent pneumonia due to bronchial obstruction, but 9% to 28% of cases may be asymptomatic.[2,4] Histologically, mucoepidermoid carcinomas are composed of varying mixtures of mucus-secreting, columnar and goblet cells. They are classified as high-grade or low-grade based on histologic appearance. The histopathological grading also reflects the prognosis of these tumors (Figure 3).[2]

PET-CT is emerging as a helpful tool for the evaluation of bronchopulmonary tumors. However, experience in mucoepidermoid carcinomas is limited, with 12 cases reported from six studies.[3] The range of SUVmax values on ^{18}F-FDG PET-CT scan varies from zero to 6.2 for low-grade mucoepidermoid carcinomas and from 2.86 to 23.4 for high-grade mucoepidermoid carcinomas. Bronchial carcinoids constitute a common differential diagnosis for MECs, both by structural radiology and by ^{18}F-FDG PET-CT. In doubtful cases, such as those with an inconclusive preoperative biopsy, a ^{68}Gallium 1,4,7,10Tetraazacyclododecane-N^{I},N^{II},N^{III},N^{IIII},- tetra acetic acid (D) - Phel1-Tyr^{3}-octreotide (^{68}Ga-DOTATOC) PET-CT scan can be performed, results of which are usually positive for typical bronchial carcinoids and negative for MECs.[5]

Surgical resection is the mainstay of treatment. Complete tumor removal with nodal dissection and preservation of functional parenchyma is the goal of the therapy.[6] Sleeve lobectomy is commonly done and occasionally requires pneumonectomy in more extensive disease. Adjuvant radiotherapy is required in cases of unresectable or incompletely resectable tumors.[6] Adjuvant chemotherapy is not necessary, however in a few case reports tyrosine-kinase inhibitor Gefitinib has shown good response in patients with MEC having EGFR gene mutations.[7] Overall survival for primary salivary gland-type lung cancer after surgical resection is excellent with 5 year and 10 year survival of 97.6% and 86.7%, respectively. Molina et al. reported excellent survival in surgically resected mucoepidermoid tumors (87% at both 5 and 10 years) and poor survival in surgically resected adenoid cystic carcinomas (57% and 45% at 5 and 10 years, respectively).[8]

Conclusion

Mucoepidermoid carcinoma of the bronchus is a rare primary lung malignancy. It usually presents with symptoms of airway obstruction and recurrent pneumonia. Familiarity with the entity by the clinician, radiologist, and pathologist may allow a preoperative diagnosis of mucoepidermoid carcinoma to be reached. Surgical resection is the mainstay of treatment. Metastasis to regional lymph nodes is rare, and the prognosis is excellent.

Conflict of Interest Disclosure: The authors have completed and submitted the *Methodist DeBakey Cardiovascular Journal* Conflict of Interest Statement and none were reported.

Keywords: bronchus, mucoepidermoid carcinoma, bronchoscopy, sleeve resection

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