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Primary Epithelial Myoepithelial Lung Carcinoma

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Primary epithelial-myoepithelial carcinoma (EMC) of the lung is an extremely rare neoplasm that originates from submucosal bronchial glands and has been found in the salivary glands, breast tissue, and sweat glands. However, only a few cases in the respiratory tract have been identified. In the literature, most pulmonary EMCs have been reported to have developed endobronchially although a few EMC cases have been presented as intraparenchymatous tumors. We have identified a case of primary EMC that developed in the peripheral lung parenchyma.

Key words: 1. Lung neoplasms
2. Salivary gland neoplasms

CASE REPORT

A 51-year-old female was admitted to the hospital with an abnormal chest radiographic finding on a periodic medical inspection. The patient was a non-smoker, and her past medical history was unremarkable. She did not have any respiratory symptoms, such as cough, hemoptysis, dyspnea, fever, or pneumonia. There were no palpable adenopathies, and cardiopulmonary auscultation and laboratory study results, including an electrocardiogram and blood analysis, were within reference limits. A chest radiograph showed a mass 3.2×2.8 cm in size in the left peripheral lung field. A chest computed tomography (CT) scan revealed a well-defined mass of 3×2.9 cm that bordered the visceral pleura at the lingular segment of the left upper lobe (LUL) (Fig. 1A, B). A percutaneous needle biopsy was performed, and the diagnosis indicated that the mass was a salivary gland tumor. Positron emission tomography (PET)-CT revealed a LUL mass with a maximal standardized uptake value of 3.2 and uptake, which indicated distant metastasis. The patient was treated by a lobectomy of the left upper lung with lymph node dissection via video-assisted thoracic surgery. Grossly, the excised section of the left upper lung showed a well-circumscribed, gray-white to yellow tumor mass, measuring 3.3×2.4×1.9 cm (Fig. 2). A microscopic examination revealed that the cut section of the tumor was composed of two types of cells: tubular and glandular structures filled with eosinophilic material, mixed with solid spindle and polygonal cell areas (H&E, ×200). The inner tubular layer showed epithelial cell characteristics, whereas the outer layer exhibited myoepithelial cell characteristics. A few mitotic figures were also seen (H&E, ×400) (Fig. 3A, B). Immunochemical staining for epithelial membrane antigen was positive in the inner tubular layers and epithelial components (EMA, ×200). Immunostaining for actin was positive in the outer tubular layer and solid areas (Actin, ×200). Immunostaining for S-100 showed a focal positive reaction in the outer tubular layer and solid areas (S-100, ×200) (Fig. 3C–E). The postoperative tumor-node-metastasis (TNM) stage was T2aN0M0, and the pathologic stage was Ib. The patient did not receive any adjuvant treatment and was followed up for

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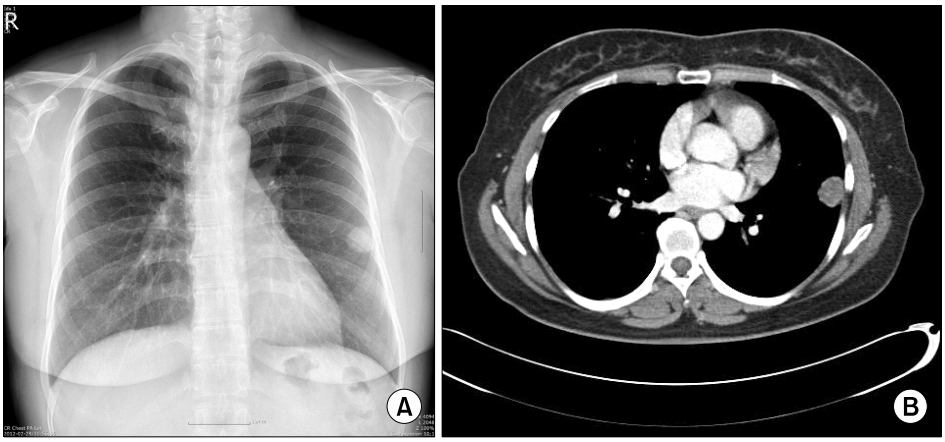

Fig. 1. (A) Preoperative chest radiography and (B) computed tomography reveal a well-defined mass of 3×2.9 cm that borders the visceral pleura at the left upper lobe lingular segment.

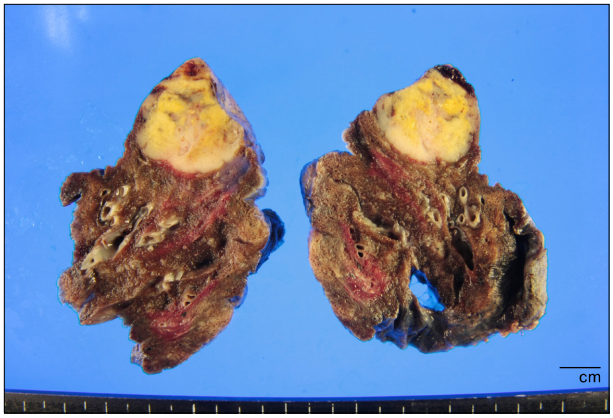

Fig. 2. Cross section of the left upper lung, including the tumor mass, a peripherally located well-circumscribed, gray-white to yellow tumor mass, measuring 3.3×2.4×1.9 cm.

16 months without any local recurrence or distant metastasis.

DISCUSSION

Epithelial-myoepithelial tumors are rare neoplasms that originate from salivary glands and comprise approximately 1% of all primary tumors. Epithelial-myoepithelial carcinoma (EMC) was first described in the salivary glands in 1972, and the World Health Organization has classified this carcinoma as a malignant tumor composed of variable proportions of the two cell types, which typically form duct-like structures [1]. The submucosal tracheobronchial glands, which are the pulmonary equivalent of the minor salivary gland system of the head, neck, and breast, and of most other tumors that develop, originate from these glands. However, EMC is rarely found in the respiratory tract. A review of the literature reveals that the incidence of primary lung EMC does not exceed thirty cases worldwide, and most of these cases have been presented as polypoid endobronchial masses [2]. In Korea, Cho et al. [3] were the first to report a case of primary endobronchial epithelial-myoepithelial carcinoma. Munoz et al. [4] was the first to describe EMC that developed in the lung parenchyma without any connection to other visceral pleura or the bronchial tree. In our case, the tumor demonstrated peripheral parenchymal development without a bronchial connection and was also characterized by visceral pleura invasion. An epithelial-myoepithelial carcinoma diagnosis is based on pathological characteristics, including tubular differentiation with a mixture of epithelial and myoepithelial cells. The tubules are lined by epithelial cells next to the lumina and myoepithelial cells beneath the epithelial cells. An immunohistochemical analysis indicated that the inner epithelial cells were positive for cytokeratin and epithelial membrane antigen. In contrast, the outer myoepithelial cells were positive for S100 and muscle-specific actin. An EMC differential diagnosis can be broad and depends on the relative predominance of the myoepithelial cells or the biphasic cell pattern. A differential diagnosis of a predominant biphasic pattern also includes pulmonary pleomorphic adenoma. In addition, myoepithelioma, myoepithelial carcinoma, and sugar tumors can be differentiated from EMC if myoepithelial cells are predominant. Epithelial-myoepithelial carcinoma symptoms vary; endobronchial lesions, cough, hemoptysis, dyspnea, fe-

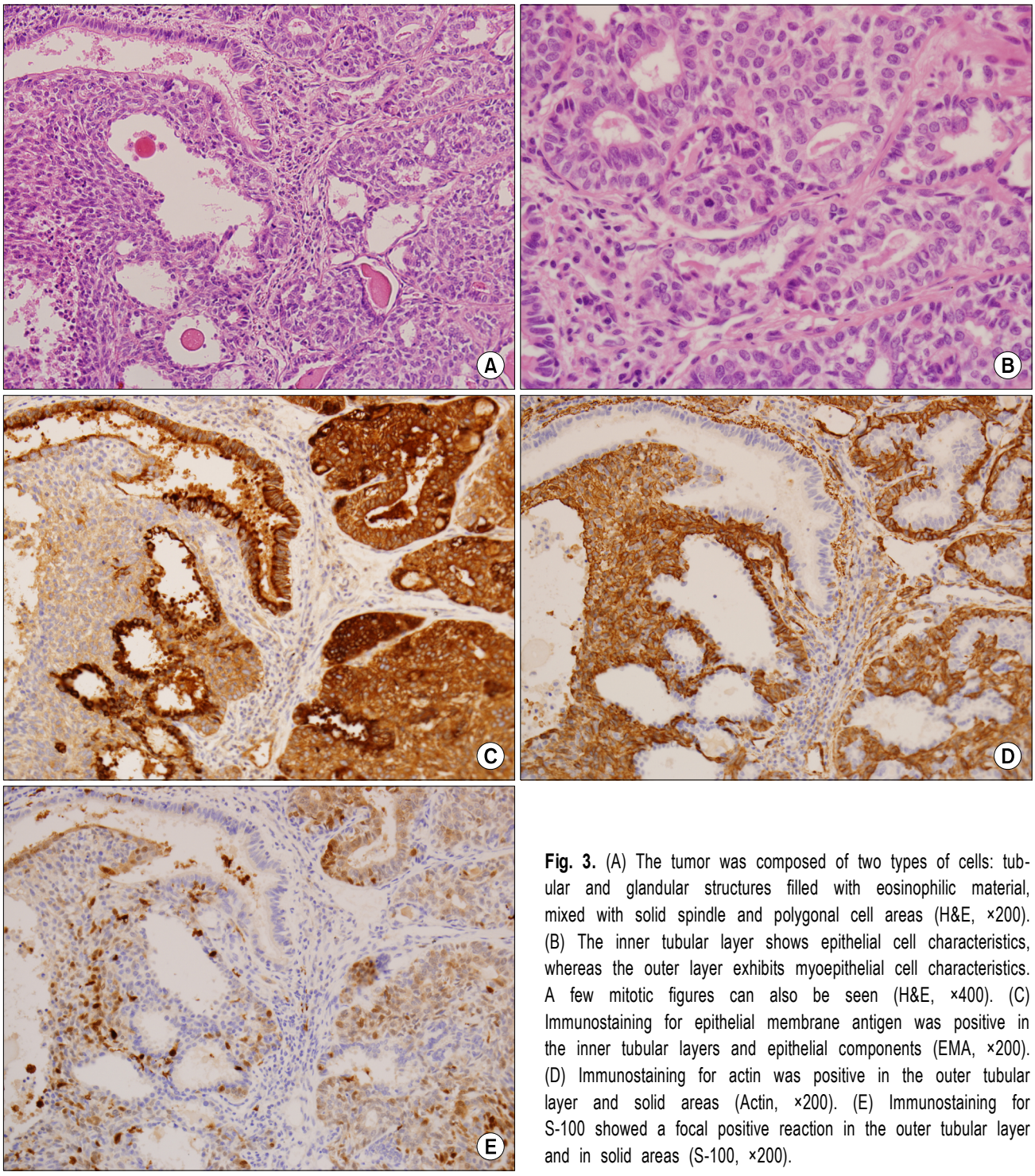

Fig. 3. (A) The tumor was composed of two types of cells: tubular and glandular structures filled with eosinophilic material, mixed with solid spindle and polygonal cell areas (H&E, ×200). (B) The inner tubular layer shows epithelial cell characteristics, whereas the outer layer exhibits myoepithelial cell characteristics. A few mitotic figures can also be seen (H&E, ×400). (C) Immunostaining for epithelial membrane antigen was positive in the inner tubular layers and epithelial components (EMA, ×200). (D) Immunostaining for actin was positive in the outer tubular layer and solid areas (Actin, ×200). (E) Immunostaining for S-100 showed a focal positive reaction in the outer tubular layer and in solid areas (S-100, ×200).

ver, and recurrent pneumonia can occur, while thoracic pain may occur in EMC that develops peripherally [5,6]. In this case, unlike previous reports, the tumor was identified before symptoms developed in the patient due to the location. A percutaneous needle biopsy was performed preoperatively, and the pathological findings suggested a salivary gland-like tumor. Based on these results, we decided to perform a left upper lobe lobectomy with lymph node dissection. The patho-

logical tests determined that the mass exhibited immunoreactive EMC diagnostic criteria. There was no lymphatic metastasis or bronchial involvement. Epithelial-myoepithelial carcinoma is regarded as a low-grade malignant neoplasm. A majority of patients are treated without local recurrence or metastases, although some authors have described cases of peribronchial lymph node metastases that occurred in a series of five cases [7]. Therefore, complete resection of EMC is currently regarded as the preferred treatment, and adjuvant chemotherapy or radiotherapy is not necessary for treatment.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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