Case report

Sarcomatoid carcinoma (SC) is a rare primary malignant tumor in which both carcinomatous and sarcomatous elements occur. It can occur in many different organs and anatomical locations, such as the skin, thyroid gland, bone, urinary tract, breast, pancreas, liver and other areas. Of them, pulmonary sarcomatoid carcinoma (PSC) is a rare malignant cancer composed of sarcoma and sarcoma-like tumors with spindle or giant cell features. Here a case of a 75-yearold Chinese man with a six-month history of cough and hemoptysis is reported. Chest X-ray showed a tumor shadow in the left lung field. Chest computed tomography (CT) scan showed a lobulated mass in his left hilum and even the left pulmonary artery. Pleomorphic interstitial cells were found by bronchoscopic brushing. To establish a definitive diagnosis for PSC, a left pneumonectomy was performed. The pathological stage was IIB (pT2N1M0) based on the tumor node metastasis (TNM) staging system. The tumor's pathology, histology, immunohistochemistry and treatment methods are discussed.

Key words: lung neoplasms; carcinoma; diagnosis; treatment.

Pulmonary sarcomatoid carcinoma: a case report

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Introduction

Sarcomatoid carcinoma (SC) is a rare primary malignant tumor in which both carcinomatous and sarcomatous elements occur. It can occur in many different organs and anatomical locations, such as the skin, thyroid gland, bone, urinary tract, breast, pancreas, liver and other areas [1–6]. Of them, pulmonary sarcomatoid carcinoma (PSC) is particularly uncommon, representing less than 1% of all lung tumors [7]. We herein report a PSC case to discuss the clinical manifestations, diagnosis, treatment, and prognosis of this condition.

Case report

On March 10, 2011, a 75-year-old man was admitted to our department because of coughing and hemoptysis for six months. He also had a 45-year history of cigarette smoking (20 cigarettes per day) and a significant medical history of hypertension. His chest X-rays showed a tumor shadow presenting in the left lung field (arrow in Fig. 1). Chest computed tomography (CT) scan revealed a lobulated mass occupying the left hilum and even stretching mediastinal structures including the left pulmonary artery (arrow in Fig. 2). Bronchoscopic examination demonstrated significant obstruction involving the superior segmental bronchus (BVI) due to pressure by abnormal vessels. Pleomorphic interstitial cells were further observed by bronchoscopic brushing.

However, for definitive diagnosis of this disease, a left pneumonectomy and lymph node dissection were performed on March 18, 2011. The results showed a $6.5 \times 6.5 \times 6$ cm³ lobulated mass in the left hilum. The tumor was ovoid in shape, poorly circumscribed, yellow-tan in color, heavy, hemorrhagic, and necrotic. Histologically, the tumor was composed of compact cells arranged in cords. The tumor cells were pleomorphic, irregular, ovoid, or spindle-like in shape, and some of them had an indistinct cell boundary. Importantly, severe necrosis was found inside (Fig. 3). Immunohistochemically, the tumor cells were positive for epithelial membrane antigen (EMA) (Fig. 4A), creatine kinase (CK) (Fig. 4B), and vimentin (Fig. 4C). They were negative for thyroid transcription factor-1 (TTF-1) (Fig. 4D), smooth muscle actin (SMA) (Fig. 4E), and cytokeratin 5/6 (Fig. 4F). Based on the new revision of the tumor node metastasis (TNM) staging system, this pulmonary carcinoma was classified as stage IIB (pT2N1MO).

Discussion

Pulmonary sarcomatoid carcinomas (PSCs) are poorly differentiated non-small cell lung carcinomas (NSCLCs) containing a sarcoma-like element (malignant spindle or giant cells) or sarcomatous component (neoplastic bone, cartilage, or striated muscle) [8]. In the World Health Organization (WHO) clas-

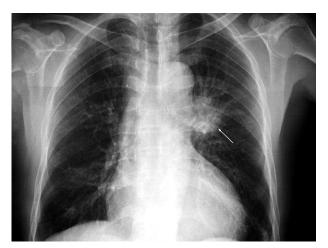
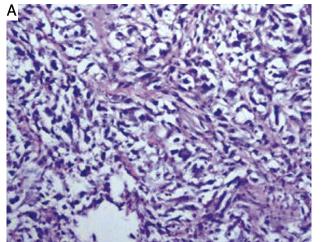


Fig. 1. Tumor shadow in the left lung



Fig. 2. Lobulated mass in the left hilum and even the left pulmonary artery



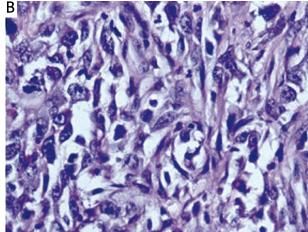


Fig. 3. Histological analysis of the resected tumor specimen A) Tumor cells were pleomorphic, irregular, ovoid, or spindle-like in shape, and some of them had an indistinct cell boundary. The tumor was composed of compact cells arranged in cords (HE, magnification 200×). B) A higher magnification of the sarcomatoid component (HE, magnification 400×)

sification, spindle cell carcinoma is classified as a variant of squamous cell carcinoma and giant cell carcinoma is classified as a variant of large cell carcinoma [9]. However, Fishback *et al.* designated SC as pleomorphic (spindle/giant cell) carcinomas of the lung because both spindle and giant cell components are often found in the same tumors [10].

PSC patients are predominantly male and smokers, with a male-to-female ratio of 4.4: 1 and average age at presentation of 51.4 years in the published series [11]. The patient in our case also had a long history of cigarette smoking. Besides, it is believed by some authors that radiotherapy for the treatment of a patient's other cancers may also induce a sarcomatoid change in lung tissue, which can accelerate the subsequent development of SC [12].

Pulmonary sarcomatoid carcinoma presents as either a central or peripheral lesion, most often in the right upper lobes. It grows by invading the bronchial tree, the pulmonary parenchyma, and the adjacent anatomical structures (mediastinum and chest wall) in the form of widely necrotic and hemorrhagic, round to bosselated large masses [13]. In our case, the lobulated mass occupied the left hilum and even

stretched to mediastinal structures, such as the left pulmonary artery. Significant BVI obstruction was also observed. The tumor was ovoid in shape, poorly circumscribed, yellow-tan in color, heavy, hemorrhagic, and necrotic.

The clinical signs and symptoms may be related to tumor localization. Symptoms such as chest pain, accumulation of excessive fluid in the pleural cavity, and difficulty breathing are shared by most pulmonary carcinomas. This makes it hard to distinguish them from each other. In endobronchial tumors, coughing and blood-tinged sputum usually occur, but peripheral tumors are asymptomatic [14]. However, no paraneoplastic syndrome was reported to accompany PSC, although such symptoms occurred in about 15–20% of small cell lung carcinomas (SCLCs) and 5–8% of NSCLCs [15]. PSCs metastasize via lymph and blood vessel routes to the same anatomical sites as conventional NSCLCs. Therefore, in general, PSC is classified using the same TNM criteria as other NSCLCs.

Although the final diagnosis of PSC depends on the close scrutiny of histopathology, its immunohistochemical analysis may help to better highlight the different cell components

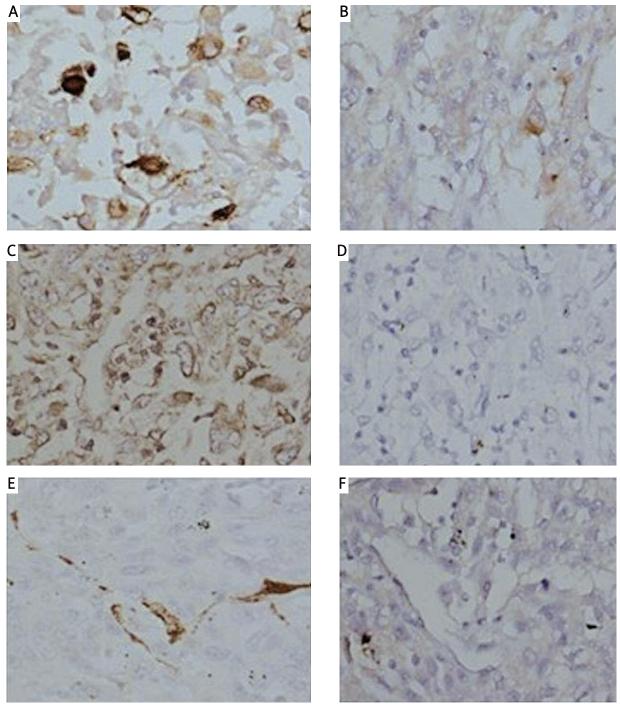


Fig. 4. Immunohistochemical staining of the resected tumor specimen. There was positive expression of: A) EMA, B) CK, and C) vimentin, but negative expression of D) TTF-1, E) SMA, and F) cytokeratin 5/6

occurring in these tumors [16]. The epithelial component is stained with antibodies to cytokeratins, EMA, and carcinoembryonic antigen (CEA), whereas the opposite holds true for the sarcomatous or sarcomatoid component, which is instead consistently immunoreactive for vimentin. In our case, we found that the PSC specimen was positive for EMA, CK, and vimentin, but negative for TTF-1, SMA, and cytokeratin 5/6. This result suggested that our case had sarcomatoid components and epithelial phenotype, which was in accordance with previous reports [17–19].

Pulmonary sarcomatoid carcinoma is an aggressive cancer and the resultant tumors are frequently symptomatic, locally advanced, and have high rates of recurrence. Recently, although radical surgery is still reported as the best option for PSC patients [20], many non-surgical treatments are recommended for patients with clinically advanced sarcomatoid cancer, such as chemotherapy. Chemotherapy is often performed to treat patients according to the results of trials that enrolled patients with lung squamous cell carcinoma or adenocarcinoma. Italiano *et al.* [21] reported the short-

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term outcomes of using doxorubicin and ifosfamide in the treatment of metastatic SC. Half of their patients with metastatic SC experienced an initial response to chemotherapy with doxorubicin and ifosfamide. However, all patients ultimately died of disease despite continued treatment [22]. Therefore, due to the limited treatment, the five-year survival rate for patients with PSC was only 24.5%, relatively worse than for patients with other forms of NSCLC (46.3%).

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