

**Case  
Report**

# Malignant Solitary Fibrous Tumor of Pleura Accompanied with First Symptoms of Chest Pain and Hemoptysis: A Case Report

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**Solitary fibrous tumor of the pleura (SFTP) is a rare tumor especially presents malignant features. Such symptoms of hemoptysis and dyspnea were rarely seen and take 5% and 4% respectively in malignant SFTP. A 26-year-old Chinese man, presenting with hemoptysis in the emergency room, was hospitalized because of dyspnea. The X-ray examination revealed a tumor in the right chest cavity. The patient refused treatment, and the tumor grew rapidly, which complicated the symptoms of the patient. En-bloc excision of tumor plus the involved lung was performed. There was at least a 5000-ml mixture of blood and tumor tissue in the right chest cavity because of continuous bleeding, leading to a tumor capsule split. Histopathology and Immunohistochemistry identified the tumor as malignant SFTP, but CD34 was negative. In this case, the tumor grew rapidly and aggressively in two months, indicating that close follow-up and active treatment are needed.**

**Keywords:** solitary fibrous tumor, hemoptysis, rapid growth

## Introduction

Solitary fibrous tumor (SFT) is a rare tumor, especially when presenting invasively and malignantly, even though it usually shows a benign clinical course. We report a rare case of tumor that most likely derived from pleura and showed malignant behavior in a short time, accompanied by symptoms of chest pain, hemoptysis and dyspnea.

## Case Report

A 26-year-old man presented with symptoms of mild, continuous chest pain on the right side, accompanied with

a small mass hemoptysis, bloody sputum (<100 ml/24h) for nearly three days. The color of blood was dark red, and the quantity was 5–15 ml, according to the report of the patient. Then the patient had dyspnea and rushed to the emergency room at night. The patient had a history of right chest pain, 2 months ago. During that time, he was hospitalized in the department of respiratory of our hospital. He had symptom of right chest pain aggravated by deep inspiration. No other systematic disorders were found nor were there any relevant family history of the disease. The chest X-ray, taken during the first hospitalization, revealed a round, well-distributed and high-density mass, occupying the right lower lung field. Only part of the mass's edge was clear, and its size was 5.4 cm × 6.5 cm (**Fig. 1**). Initial diagnosis was “pulmonary sequestration associated with cystic degeneration”. The department of thoracic surgery recommended surgical treatment after consultation; unfortunately, the patient rejected the treatment program and requested a discharge.

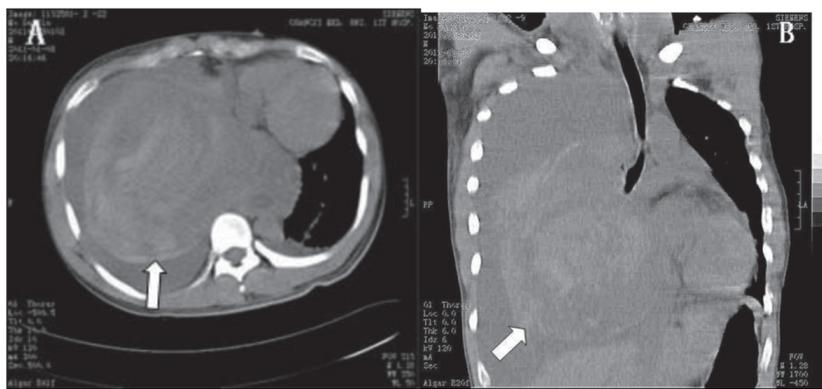
Under admission, we measured his body temperature, which was 39.5°C; pulse, 133/min; breathing rate, 28/min; and blood pressure, 113/78 mmHg. The bulbar conjunctivas were not stained yellow, and no edemas of superior

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**Fig.1** X-ray films of the chest performed in November 8th 2010 showed right chest cavity tumor, 5.4 cm × 6.5 cm (arrow).



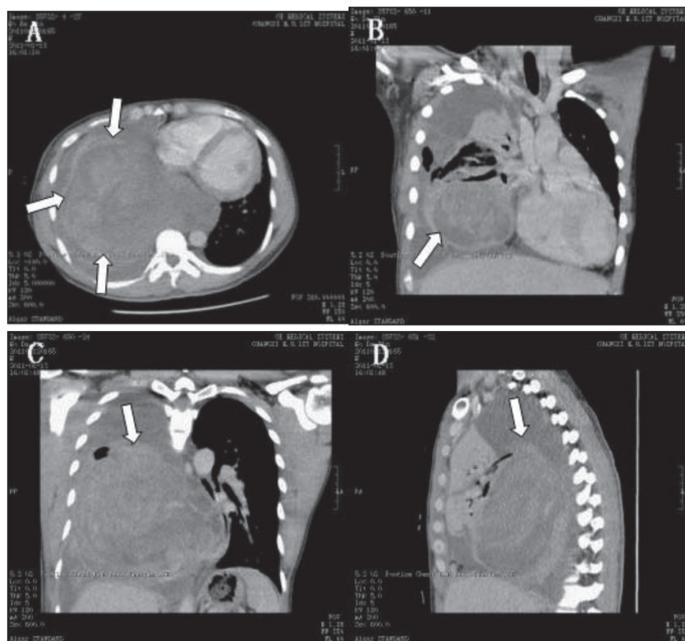
**Fig. 2** Chest CT scans performed in January 8th 2011 showed chest tumor (emergency room), **A** (arrow) and **B** (arrow) revealed the tumor in horizontal and coronal plane respectively.

palpebras were found, but the palpebral conjunctiva showed a bit of pallor, indicating anemia. No superficial lymph nodes were palpable. The right chest wall was heaved, and the intercostal space was widened, compared to the left side in the chest inspection, and the trachea was positioned to the left. Chest auscultation revealed no respiratory sounds on the right side, and there were considerable coarse crackles detected in the median and outer zone of the left lung field. There were no abnormalities of the abdomen, no neurological abnormalities, and no leg edema was observed. The WBC in the blood chemistry results was abnormal at  $24.79 \times 10^9/L$ , indicating severe infection and inflammation. A chest computed tomography (CT) revealed an enormous, soft tissue occupation between the right lower lung and posterior mediastinum, measuring 18.2 cm × 17.1 cm × 14.8 cm, and the computerized tomography number was 22–87 Hu. There was a large effusion in the right thoracic cavity, leading to pulmonary atelectasis, and small amounts of effusion in the left thoracic cavity (**Fig. 2**). We performed chest drainage and hemorrhagic effusion and subsequently drained nearly 800 ml. The effusion was reduced to 600 ml after 24 hours. We also gave the patient hemostatic drugs and antibiotics for prophylaxis. After the treatment, annihilation was relieved, breathing rate fell to 22 breaths/min, WBC  $20.54 \times 10^9/L$ . Contrast-enhanced CT images revealed a huge, heterogeneous mass at the inferior lobe of the right lung, enhancement scanning showed a heterogeneous substance and an indistinct edge, with a size, equal to that in the CT done in the emergency room. The trachea, heart, and mediastinum were with apparent compression and were positioned to the left. We detected large effusions in right thoracic cavity, as well as small

amounts of effusion in the left thoracic cavity (**Fig. 3**). These demonstrations led us to suspect “malignant tumor or pulmonary sarcoma.”

By the consent of the patient, he underwent thoracotomy through an incision of the sixth intercostal space on the right posterior-lateral. The findings showed a split tumor cyst split; a bloody, fluid-filled right chest cavity and mixed, cracked tumor tissue together, which was tofu-like. The thickness of the tumor capsule wall was about 1 cm. The tumor had already invaded the inferior lobe of the right lung and part of the diaphragm. Most of the remained cyst wall bed were broad-based and adhered to parietal pleura and contained significant feeding vessels that were continuous bleeding. The tumor was resected with adequate margins, included inferior lobe of the right lung and part of diaphragm, except those margins we could not resect because of the tight connection to the pleura. The volume of bleeding and cracked tumor tissue during the operation was nearly 5000 ml.

Postoperative histopathological examination showed that the tumor cells were a spindle-shaped, interlacing arrangement. Mitoses, with local necrosis, were easy to see. The mitotic activity was 15 mitotic figures per 10 high-power fields. The lymph nodes had merely showed reactive hyperplasia (**Fig. 4**). They stained positive for Vimentin, CD99, and Bcl-2 by immunohistochemistry and negative for CD34, SMA, S-100, CK, CK19, Calretinin, MC, CK5/6, CD117 and Dog-1. The final diagnosis was malignant solitary fibrous tumor of the pleura (SFTP), according to the pathologic criteria of England et al.,<sup>1)</sup> showing that the inferior lobe of the right lung and part of visceral pleura tissues had been invaded. Preoperative symptoms disappeared after surgery in our patient, and the

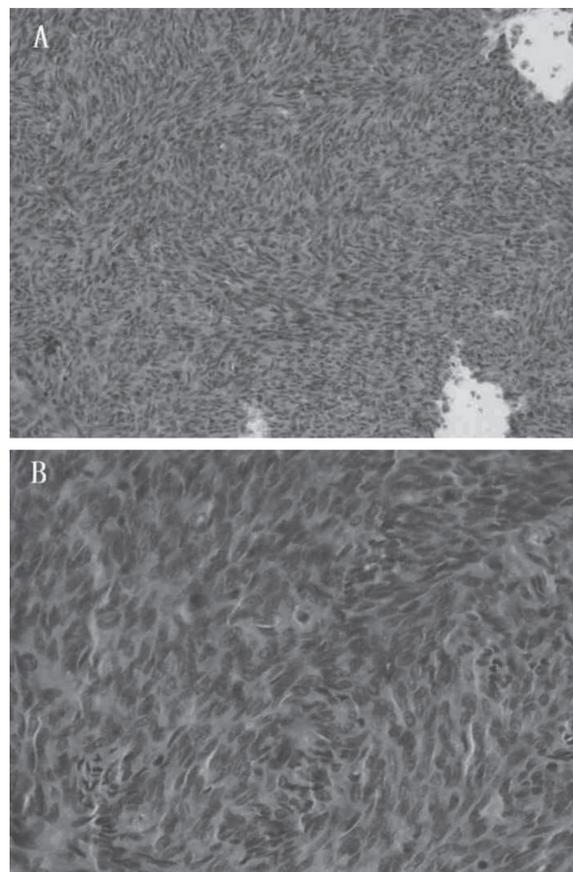


**Fig. 3** The contrast-enhanced chest CT scans performed in January 13th 2011 showed right chest tumor. **A, B, C** and **D** (arrows) revealed the size of tumor was  $18.2 \times 17.1 \times 14.8$  cm, there was heterogeneous enhancement in the mass, accompanied large effusion in right chest cavity, and mediastinum moved to the left because of occupying disease (**B**, arrow).

had no pulmonary edema. However, SFTs may recur after an incomplete resection,<sup>2)</sup> thus we recommended radiotherapy for eliminating residual margins in parietal pleura, but our patient refused treatment, so he was discharged after healing of the incision. The patient did not have a relapse at the 2-month follow-up.

### Discussion

SFT is a rare, spindle cell neoplasm, derived from mesenchymal cells; it was first discussed by Wagner in 1870, and Klember and Rabin described it in pathologic ways in 1931 and proposed the classification of primary pleural tumors into localized and diffuse mesotheliomas.<sup>3)</sup> Even though SFT was described like that, many reports have indicated that SFT is derived from mesenchymal cells that are outer ectopygma and more than mesothelium derived.<sup>4)</sup> These kinds of cells exist generally in human connective tissue; therefore, except for the morbidity associated with tumor in the pleura, clinical symptoms are predominantly based on the site of occurrence and tumor size. The tumor can originate from many locations, such as peritoneum, mediastinum, fossa orbitalis, liver and lung tissues, and even in thymus etc.<sup>5,6)</sup> One of



**Fig. 4.** Histopathological examinations of the tumors showed tumor cells were spindle-shaped, with seeing mitotic easily, and accompanied with local necrosis, which indicated the evidence of malignancy. (HE stain: A  $\times 100$  B  $\times 200$ )

them is a solitary fibrous tumor of the pleura (SFTP), originating from mesenchymal cells in the submesothelial tissue. These are immunohisto-chemically negative for cytokeratines and positive for CD34, bcl-2 and CD99.<sup>7)</sup>

Benign SFTs are more common to see; most of them develop slowly and have a better prognosis.<sup>1)</sup> No symptoms occur until after the tumor grows into a large mass, leading to local compression. Symptoms of SFT are intrathoracic, and include chest pain, cough and dyspnea—mainly, and a minority of patients could present with carcinoid syndrome related to hypoglycemia. Partial pleura tumors mostly lead to chest pain, for cough and dyspnea are mostly caused by giant-sized tumors, which compress the trachea and main bronchus. Our patient's dyspnea was caused by a large pleural effusion in the right thoracic cavity, leading to the shift of the trachea and mediastinum. One of his first symptoms was hemoptysis, and this finding was confirmed during the operation; we

found the tumor and its cyst invading the inferior lobe of the right lung tissues and bronchia. Fortunately, the invasion did not involve arteries or veins of bronchus. This can explain the small quantity of hemoptysis, which may be caused by the rupture of blood capillaries or necrosis of tumor, and it was tunica mucosa bronchiorum alone that was invaded by the tumor. His breathing rate was 28 breaths/min, combined with infection (WBC  $24.79 \times 10^9/L$ ) and high fever ( $39.5^\circ C$ ), and he had a continuous bleeding intrathoracic cavity. The series of nonspecific symptoms and lack of diagnostic standards are a barrier in the diagnosis of SFT.

There are super impositions between benign and malignant SFTs in histologic evidence, for instance, heman-giopericytoma, leiomyoma, nodular fasciitis, inflammatory myofibroblastic tumor, fibromatosis and benign peripheral nerve sheath tumor, etc.<sup>8)</sup> SFT is very difficult to detect because of its lack of typical imaging characteristics. In X-rays, it shows an opaque, solitary mass, commonly seen; ultrasonic examinations demonstrate a well defined tumor of a solid homogeneous echo pattern; while CT provides a homogeneous, capsule-surrounding mass. When the tumor is small, its homogeneous scanning time is prolonged during the artery to venous phase, and the degree of enhancement increased gradually; sometimes it will present with multiple, small non-enhanced areas.<sup>9)</sup> During 2 months between initial discoveries to emergency hospitalization, our patient's tumor increased, apparently. CT scans demonstrated that the tumor had an unsmoothed edge and heterogeneous density enhancement; the manifestation might be due to necrosis, hemorrhage, or cystic changes; all of these were coincident with the CT diagnosis of SFT. Moreover, SFT is a tumor with a full blood supply, so there are obvious enhancements at the cell density area and vascular rich area in imaging, whereas, they are not obvious at the cell sparse zone and vascular deficient zone.

Microscopically benign SFT is described as circular or spindle-shaped cells, admixed with thick or thin collagen bands, and prominent branching vasculatures. Nuclear chromatin is distributed homogeneously, and mitoses are rare to see, generally less than 2 mitoses per 10 hp fields.<sup>8,10)</sup> The malignant ones, apart from benign characteristics, also have nuclear atypia, and there are 4 mitoses per 10 hp fields at least, and accompany necrosis, furthermore.<sup>11)</sup> Compared to benign tumors, malignant SFT always has multi-cell lesions, moderate to severe cell atypia, and obvious tumor necrosis, edge of infiltration, as well as high frequency of mitoses.<sup>12)</sup> Our case's histopathological re-

port described tumor cells that were a spindle-shaped, interlacing arrangement. Mitosis were easy to see (15 mitoses per 10 hp fields), and with local necrosis. These corresponded with the diagnosis of pathologic criteria of malignant SFT.<sup>1)</sup>

Immunohistochemistry has a momentous significance in the diagnosis of SFT. The immunoreactivities of vimentin, Bcl-2, CD 99 and CD34 are important because they can distinguish SFT from other tumors. Anders et al.<sup>13)</sup> stated CD99 was usually positive, and others like CK, S-100, desmin and SMA were negative. Our patient's results were consistent with the above statement, except that CD34 was negative. A positive CD34 result is suggestive of an epithelial origin, and the tumor is considered to arise from differentiated mesenchymal cells in the sub-pleural connective tissue.<sup>14)</sup> However, immunohistochemical expression of CD34 tends to be reduced in malignant SFTs,<sup>15)</sup> just like ours. In some cases, it was reported that c-kit, CAM 5.2, factor XIIIa, HMB-45, AE-1, SMA, CD31 and Fli-1 were immunohistochemical negative, while there was the emergence of high-expression in Ki-67.<sup>16,17)</sup> No tumor expressed epithelial differentiation, found immunohistochemically or ultrastructurally, and the attached criteria of diagnosis<sup>1)</sup> indicated that our patient suffered from SFTP.

Although it is generally believed that SFTs are relative chemoresistance, the most effective drugs seem to be anthracyclines and ifosfamide, followed by gemcitabine and dacarbazine, which are commonly used in soft tissue sarcomas.<sup>18)</sup> The targeted drug imatinib mesilate seems to play a role in SFT expressing the wild type platelet-derived growth factor receptor- $\beta$ , and its in vitro and in vivo experiments of inhibitory activity in chemo- and radioreistant malignant SFT have been reported.<sup>19,20)</sup> Radiotherapy may have some benefit, but if applicable, it was recommended to be used in combination with chemotherapy.<sup>21)</sup>

## Conclusion

It is reported that malignant SFT coupled with hemoptysis and dyspnea as symptoms were 5% and 4% respectively.<sup>1)</sup> It is uncommon for patients with SFT to have both of these symptoms. The tumor had grown rapidly from the first discovery to the preoperative period, in this case. Symptoms were chest pain, haemoptysis and dyspnea. The tumor body was split due to continuous bleeding in the capsule and resulted in 5000 ml of bleeding, at least. The tumor had also invaded the inferior lobe of the right

lung, part of the pleura and diaphragm. SFTs of malignant transformation and larger ones certainly have recurrence and poor prognosis.<sup>22)</sup> Accordingly, recommendations of radiotherapy, as well as long term follow-up, especially for an unresectable residual capsule wall attached to parietal pleura, are particularly important.

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