

Solitary Fibrous Tumor of the Pleura as a Cause of Type II Respiratory Failure

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ABSTRACT

Solitary fibrous tumor of the pleura is a rare type of tumor originating from the mesenchyma of the pleura. It is traditionally a benign lesion. However, in some cases malignant features have been observed. The majority of solitary fibrous tumors of the pleura are noticed by accident on chest X-ray, while the main symptoms include cough, thoracic pain and dyspnea. When growing within the thoracic cavity, these tumors exert pressure on vital adjacent tissues and large vessels. In addition, these tumors can be accompanied with paraneoplastic syndromes that are completely resolved after tumor resection. Respiratory failure is a rare complication of this tumors, which is reported in a handful of cases. Herein, we report a rare case of a benign solitary fibrous tumors of the pleura in a 75-year-old woman complicated with type II respiratory failure.

Keywords: solitary fibrous tumor, respiratory failure, pleural tumor.

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Article received on the 13th of January 2021 and accepted for publication on the 18th of January 2021

INTRODUCTION

Solitary fibrous tumor of the pleura (SFTP) is an infrequent, slow-growing primary pleural neoplasm accounting for <5% of pleural tumors. It originates from the submesothelial mesenchymal layer and arising usually from visceral pleura. Typically, it is a benign lesion. However, some of resected SFTP have malignant features (1). Definitive malignant criteria have not been identified. Some SFTPs recur after complete excision and their clinical behavior is unpredictable (2).

Diagnosis of SFTPs based on radiology before surgical resection is difficult. Moreover, it is not feasible to distinguish the malignant from benign tumors by radiographic features (3). The diagnosis of SFTP is established by typical histological findings of short spindle malignant cells mixed with collagen, growing in a 'patternless' or in a manner like hemangiopericytoma in combination with compatible immunohistochemical staining (positive for CD 34, CD 99, bcl-2) (4).

Solitary fibrous tumors of the pleura have traditionally a subclinical course, with the majority of patients being asymptomatic at the time of diagnosis as an incidental routine chest radiograph finding. Occasionally, non-specific symptoms such as cough, dyspnea, chest pain, weight loss and hemoptysis can occur, especially with large lesions (5). Some patients present with paraneoplastic syndromes such as refractory hypoglycaemia, clubbing and pulmonary hypertrophic osteoarthropathy, probable due to ectopic secretion of hormone-like factors (6).

Complete resection of the tumor remains the gold standard of treatment. For malignant SFTP, therapeutic approach with radiotherapy or conventional chemotherapy showed limited efficacy and the investigation on targeted therapy is under progress (7).

Herein, we report a case of SFTP in a 75-year-old woman who was diagnosed with a SFTP as a cause of respiratory failure. □

CASE REPORT

A 75-year-old woman, non-smoker, with a body mass index (BMI) 26 kg/m² and a history of arterial hypertension and venous insufficiency was admitted to our Pulmonology De-

partment with vomiting, fatigue, dyspnea at rest and dry cough over the last week.

Physical examination revealed a patient with decreased breath sounds on auscultation at middle and lower lung field of the right lung and without abnormal signs from abdominal/neurological assessment. Blood pressure was 165/100 mm Hg, heart rate 85 beats per minute, oxygen saturation 93% with Venturi mask delivering a fraction of inspired oxygen (FiO₂) 31% and body temperature 36°C on admission. Electrocardiography showed no abnormal findings. Arterial blood gas analysis revealed pO₂ 60 mm Hg, pCO₂ 59 mm Hg, pH 7.52 and HCO₃⁻ 48.2 mmol/L on FiO₂ 31%. Chest X-ray showed a large mass lesion in the right lung that shifted the mediastinum to the opposite site (Figure 1).

Laboratory findings included hemoglobin (Hb) 14.8 g/dL (normal 12-15 g/dL), white blood cells (WBC) 12.89 x 10³/μL (normal 4-11 x 10³/μL), neutrophils 11.92 x 10³/μL (normal 2-8 x 10³/μL), lymphocytes 0.48 x 10³/μL (normal 1.1-4 x 10³/μL), platelets (PLTS) 346 x 10³/μL (normal 150-400 x 10³/μL) and C-reactive protein (CRP) 15 mg/L (normal <6 mg/L). Urinalysis was normal.

The other blood biochemistry parameters, including TSH, were normal, with the exception of decreased potassium 2.5 mEq/L (normal



FIGURE 1. Chest X-ray showing a large mass lesion in the right lung that shifts the mediastinum to the opposite site



FIGURE 2. Chest computed tomography (CT) showing a large lesion at the anterior and apical-posterior segment of the right upper lobe, extended from the right hilum to the lung periphery, small pleural effusion and atelectasis of the affected lung segments



FIGURE 3. Complete obstruction of the apical segmental bronchus and about 80% obstruction of the posterior segmental bronchus of the right upper lobe observed during bronchoscopy

3.5-5.3 mEq/L) and decreased albumin 3.1 g/dL (normal 3.5-5.1 g/dL).

The patient underwent abdominal ultrasound, in which imaged liver, gall bladder, spleen and pancreas had a normal appearance. Com-

puted tomography (CT) of the chest, abdomen and brain were also performed. Chest CT revealed a large space occupying lesion at the anterior and apical-posterior segment of the right upper lobe, extended from the right hilum to the lung periphery, small pleural effusion and atelectasis of the affected lung segments (Figure 2). Abdomen CT revealed a slightly enlarged left adrenal gland and brain CT had no abnormal findings.

The patient underwent bronchoscopy. During endoscopic procedure, complete obstruction of the apical segmental bronchus and about 80% obstruction of the posterior segmental bronchus of the right upper lobe with mucosal invasion were observed (Figure 3). Bronchial biopsies and washings were obtained from the affected segments. Cytological and microbiological examination of bronchial washings was negative. Histological examination of bronchial biopsies revealed no malignancy.

CT-guided fine needle aspiration biopsy was conducted. Histological examination of the biopsy established the diagnosis of a SFTP. Malignant features were not observed. The patient denied undergoing surgical resection of the tumor and was discharged with oxygen therapy. □

DISCUSSION

Solitary fibrous tumor of the pleura was described for the first time by Lietaud in 1767 (8). To date, less than 2000 cases have been reported globally (9). Utilization of immunohistochemical techniques and antibodies to mesenchymal elements and the use of electron microscope allowed the recognition of SFTP (10). Common features on chest X-ray include a solitary opacification of round or oval shape and with a size that varies, often in continuation with the chest wall (11). Chest CT is the test of choice because it is a more accurate imaging tool, allowing an assessment of size, morphology, anatomic location and relations with other tissues. The majority of patients present with an oval or elliptical homogeneous SFTP with a median diameter of 6 cm, most located in the chest wall, intrapulmonary fissure, diaphragm and mediastinum (12). 18F-FDG/PET-CT has been used as a potentially useful imaging modality for differentiating malignant from benign SFTPs in few single cases or small retrospective studies (13), while in a retrospective study by Tazeler *et al.*

18F-FDG/PET-CT had a limited role in identifying malignant SFTP (14).

Respiratory failure is one of the rarest clinical manifestations of SFTP. This neoplasm has been described as a cause of respiratory insufficiency in a case series by Abe *et al.*, with one case requiring mechanical ventilation (2). The majority of patients have no symptoms and the most common reported symptoms include chest pain, cough and dyspnea (10, 15-17). Respiratory failure is usually a result of compression of adjacent structures and local enlargement of the tumor leading to restrictive ventilatory defect (7). Pleural tumors are space occupation extrapulmonary diseases that can result in type II respiratory failure (18). In our case, type II respiratory failure was attributed to SFTP based on lack of abnormal findings on brain CT and neurological examination, absence of obesity, history of airways disease and medication causing reduced breathing effort, and absence of thyroid hormones dysfunction.

Other uncommon clinical presentations of SFTP include hypoglycemia due to secretion of insulin-like growth factor II, known as Doege Potter syndrome (19-21), hypokalemia (3), hypertrophic osteoarthropathy, also called as Pierre-Marie-Bamberger syndrome, characterized by clubbing of tissue of the fingers (7), gynecomastia, irregular menstruation and flushing (22-23), due to increase of human beta chorionic gonadotropin-releasing factor, cerebellar degeneration (24), hemoptysis due to compression of lung parenchyma and hoarseness due to compression of laryngeal nerve. In addition, superior vena cava syndrome due to compression of the mediastinum (25) and loss of appetite/weight loss and fatigue due to compression of the liver (26) have been described. Soli-

tary fibrous tumor of the pleura has also been associated with complications such as autoimmune hemolytic anemia (27) and hemothorax due to spontaneous intra-tumor bleeding (28).

The diagnosis of SFTP is rarely confirmed before surgical excision and histological examination of the mass. In some cases, preoperative diagnosis is feasible with large-bore cutting needle biopsies (11). Complete resection of the primary lesion and local recurrent tumors is the preferred treatment for SFTP. Resection usually leads to decompression of lung tissue and full re-expansion and to treatment of the related paraneoplastic syndromes (29). Thoracotomy is often used for large tumors, while video assisted thoracoscopic surgery (VATS) is mostly performed for resection of small tumors [30]. The prognosis of SFTP is generally good. However, due to local recurrence, long-term follow-up is required (7). □

CONCLUSION

This is a rare case of SFTP causing respiratory failure type II. These tumors usually have a subclinical behavior and clinical presentation depends on the tumor size. Respiratory failure is an uncommon manifestation of SFTP. Total surgical resection can resolve symptoms, complications and paraneoplastic syndromes associated with this entity. □

Conflicts of interest: none declared.

Financial support: none declared.

Written consent: Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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