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Case Report OMICS International

Primary Pulmonary Synovial Sarcoma: One Case Report

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Abstract

Primary pulmonary synovial sarcoma (PPSS) is a rare disease. Diagnosis is made postoperatively following resection of the tumor. We present an extremely rare case of PPSS in a 28-year-old, smoker-man whose chest imaging revealed a giant mass (5 × 6 × 7 cm), with dullness and decrease in breath sound in the left side of the chest. A fiber optic bronchoscopy was performed and there was necrosis, hemorrhage, and left lower bronchial obstruction. Subsequently diagnosis was confirmed by immunohistochemistry and the detection of a SYT-SSX fusion gene. We believe that a diagnostic approach for lung sarcoma would provide helpful information to clinicians.

Keywords: Synovial sarcoma; Pulmonary; Sarcoma

Introduction

Primary pulmonary synovial sarcoma is a very uncommon but aggressive primary lung tumor. It was first described by Zeren et al. [1,2]. It is a rare neoplasm accounting for less than 0.5% of malignant lung tumor [3]. It can easily be misdiagnosed because of its unusual histological features. Its diagnosis depends on immune histochemical staining and RT-PCR analysis. Here, we review one case of it in our hospital.

Case Report

A 28-year-old man presented with hemoptysis for 10 days. Physical examination shows dullness and decrease in breath sound in the left side of the chest. There was no enlargement of lymph nodes. Chest computed tomography (CT) revealed a well-defined giant mass in the left lower lobe. Brain CT or bone scans show no metastasis. The smoking history is 18 pack years. He underwent a resection of hemangioma in the left foot 3 years ago in our hospital. Cytological analysis of sputum was negative. A fiber optic bronchoscopy was performed and there was necrosis, hemorrhage, and left lower bronchial obstruction. Biopsies were obtained, which shows there were sheets of spindle cells with plump nuclei, suggesting spindle cell carcinoma. Finally, a left lower lobectomy with radical lymph nodes dissection was performed, adjacent pleura was infiltrated by the tumor during the operation. The soft tumor measured 5 × 6 × 7 cm. It was well circumscribed and compressed with the adjacent tissues, hemorrhaging. At high power magnification, the tumor was characterized by a dense proliferation of atypical short spindle cells with indistinct cytoplasm and short fusiform or small rounded nuclei. An epithelioid appearance of the neoplastic cells was also evident, but immune histochemical staining was positive for EMA(+), Vim(+), BcL-2(+), CD5/6(+), CD99(+), CK7(focal,+), Desmin(few,+), Ki-67(+, 40%), negative for PCK(-), CK(-), CD34(-), SMA(-), TTF-1(-) (Figures 1-6).

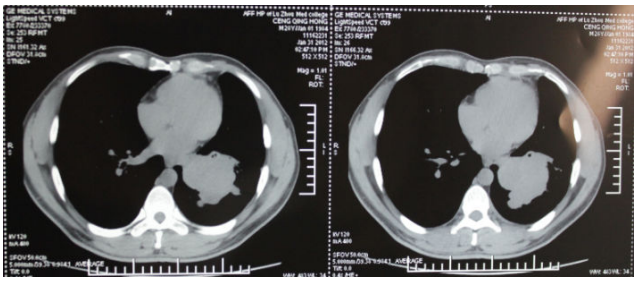

Figure 1: CT scanning: a 4 × 5 cm, well-defined giant mass in the left lower lobe.

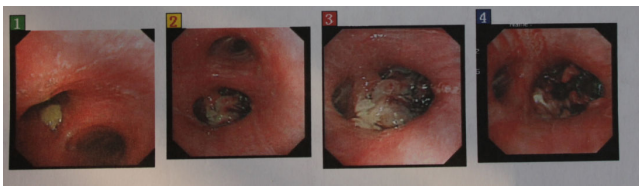

Figure 2: Necrosis, hemorrhage, and endobronchial obstruction under bronchoscopy; Soft tumor measured 5 × 6 × 7 cm.

These findings were compatible with a monophasic synovial sarcoma.

Discussion

Synovial sarcomas usually occur in the soft tissues of the extremities of adolescents and middle-aged patients, in the vicinity of large joints. Rare variants may occur in non-soft tissue locations such as the larynx, heart, pericardium, pleura, lung, mediastinum, and peritoneal cavity [4].

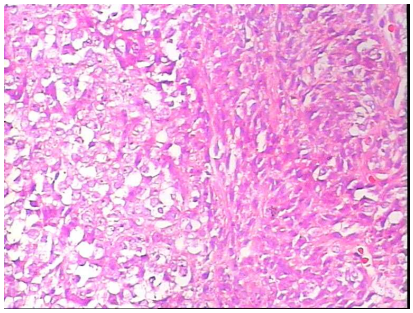

Figure 3: HE staining; packed fascicular proliferations of spindle cells with eosinophilic cytoplasm, (×200).

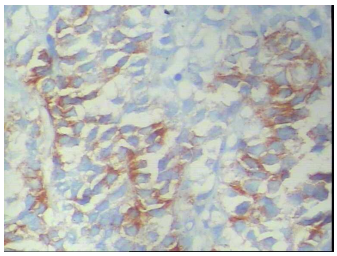

Figure 4: Tumor cells are positive for EMA by immunohistochemical staining (×400).

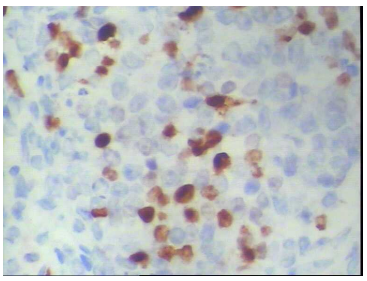

Figure 5: Tumor cells are positive for Ki67 by immunohistochemical staining (×400).

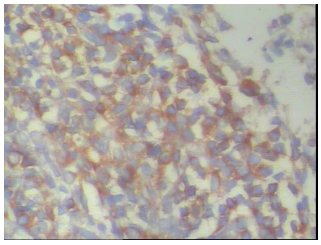

Figure 6: Tumor cells are positive for Vim by immunohistochemical staining (×400).

The diagnosis of primary synovial sarcoma of the lung should base on clinical manifestations, pathological features, immunohistochemical staining, chromosomal translocation and gene fusion. Cytogenetic studies are helpful, as adequate tissue biopsy is required for confirmatory diagnosis based on morphology, special staining and chromosomal studies, which shows the chromosomal translocation t(x;18) (p11.2; q11.2) by fluorescence in situ hybridization or reverse transcriptase- PCR [5]. The presence of the translocation t(X:18) on fluorescent In situ hybridization is also confirmatory of synovial sarcoma [6-8]. The chromosomal translocation t(X;18) (p11.2; q11.2) has been found in about 90% of synovial sarcoma [9]. This translocation involves the SYT gene on chromosome 18 and one of the several highly homologous genes (SSX1, SSX2, and SSX4) on the X chromosome. About two-thirds of cases have a SYT/SSX1 fusion and one-third has SYT/SSX2 fusion, the fusion between SYT/SSX4 is rare. SYT/SSX1 gene is associated with biphasic subtype while SYT/SSX2 with monophasic subtypes.

There are four subtypes of primary synovial sarcoma of the lung: monophasic fibrous (spindle), monophasic epithelial, biphasic and poorly differentiated, Monophasic is most common.

The differential diagnosis includes the fibrous pleural tumor, sarcomatoid subtype of malignant pleural mesothelioma, spindle cell carcinoma, and malignant peripheral nerve sheath tumor [10].

Complete surgical resection is strongly suggested [11].The surgical principle is resection of the tumor with dissection of adjacent tissue planes. Adjuvant radiation therapy is used in patients with tumors >5 cm in size. Chemotherapy in patients with primary disease has been controversial.

The 5-year survival rate is more than 50%. The prognostic factors including gender (male) [12] age (>25 years), size (>5 cm), margin, mitotic activity (>10/10 HPF), bone or neurovascular invasion, histological subtype (monophasic), p53 over expression, Ki67 proliferative index, SYT-SSX fusion type, incomplete resection. For this case, the left lower lobe was completely resected and no lymph node metastasis was observed. So the prognosis is good.

In summary, although synovial sarcoma tends to be aggressive and its progression is very rapid, it can be cured through surgery. It is necessary to have a follow-up for this case.

Conflict of Interests

The authors declare that they have no conflict of interests.

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