Case Report Primary pulmonary synovial sarcoma: a case report

Junfeng Liu^{1*}, Zhaoming Nie^{1*}, Jianyao Lv², Yingrong Lai¹

Departments of ¹Pathology, ²Medical Imaging, The First Affiliated Hospital of Sun Yat-sen University, Guangzhou, People's Republic of China. *Equal contributors.

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Abstract: Primary pulmonary synovial sarcoma (PPSS) is a relatively rare mesenchymal tumor accounting for 0.5% of all primary lung malignancy with a highly progressive potential. We describe a case of a 52-year-old man whose chest CT-imaging revealed a heterogeneous mass (8.5 × 7.7 × 6.5 cm), with multiple nearly-circular nodules in bilateral pulmonary tissues. EBUS-TBNA and pathological H&E and immunohistochemical examination were performed subsequently, and sarcoma was suspected. Then molecular detection was used and PPSS was confirmed by the detection of the expression of SYT gene (positive) and EWSR1 gene (negative) in situ hybridization. This report shows that EBUS-TBNA combined with in situ hybridization could serve as a helpful diagnostic tool.

Keywords: Primary pulmonary synovial sarcoma, diagnosis, SYT gene, EWSR1 gene

Introduction

Synovial sarcoma commonly occurs in periarticular tissue, but it has also been reported in other sites [1]. synovial sarcoma, especially primary synovial sarcoma, rarely occurs in the lungs. primary pulmonary synovial sarcoma (PPSS) accounts for < 0.5% of all lung cancers, while metastatic synovial sarcoma in other tissues is far more common than PPSS [2, 3]. As a very malignant tumor, the 5-year overall survival of PPSS is significantly worse than soft tissue sarcomas of the extremities (35% vs 71%) [4]. The treatment of PPSS is diverse and nonstandard on account of its rarity. However, complete surgical excision reportedly gives the best chance of a cure [5]. This case study describes a patient presenting with PPSS and who was suffering from bilateral pulmonary multiple change and is alive with a good prognosis after holistic pulmonary chemotherapy.

Case report

A 52-year-old man presented with the left-side chest pain, polypnea, shortness of breath, and increasing pain when coughing, pyrexia to 39 degrees (this symptom resolved after treatment) for the previous 2 months. His pain slowly increased, was insidious in its onset, and was a dull ache. However, no other symptoms were apparent, including pyrexia, rigor, cough, hemoptysis, or obvious weight loss. The mental and sleep states of the patient were normal.

The physical health of patient was good, and he had no coronary pulmonary tuberculosis, heart disease, hepatitis, hypertension or diabetes. However, he had a long history of smoking and alcohol intake. a general physical examination revealed there was no cervical lymphadenopathy or clubbing. There was a lot of effusion in the left thoracic cavity.

A contrast-enhanced computed tomography (CT) scan showed a $8.5 \times 7.7 \times 6.5$ cm soft tissue density in the apical segment of the left upper lobe (**Figure 1**). The mass was abutting the hilus of the lung. Multiple nearly-circular nodules were apparent in bilateral pulmonary tissues, and the biggest soft tissue density was almost $2.0 \times 2.0 \times 2.0$ cm. An endobronchial ultrasound-guided transbronchial needle aspiration (EBUS-TBNA) from the lung mass indicated that a small round cell neoplasm was infiltrating into the lung parenchyma.

In addition, routine immunohistochemical analyses were performed from formalin-fixed, paraffin-embedded specimens. The panel of antibodies mainly included CK, CD99, CD56, Syn, TTF-1, Napsin a and Ki-67. all the antibodies were products of DAKO, and the staining was performed with the DAKO envision kit. The sec-

Primary pulmonary synovial sarcoma



Figure 1. CT-scan of the patient. The arrows point to the sentinel lesion areas in segments of the left upper lobe.

tions were developed with 3,3'-diaminobenzidine tetrahydrochloride and counterstained with hematoxylin. Immunohistochemical staining showed that a few of the tumor cells were positive for CK, CD56 and Syn, and these cells were strongly positive for CD99. The Ki-67 proliferation index was about 20%. The tumor cells had no expression of TTF-1, napsin a (Figure 2). Meanwhile, in situ hybridization assays (Abbott laboratories, Chicago) detected the expression of the SYT and EWSR1 genes. The cut-off values were estimated by the red and green separation signals (> 15% is positive, and \leq 15% is negative) in 100 cancer cells. The results showed the tumor cells were positive for SYT and negative for EWSR1 (Figure 3).

The patient had not undergone a lobectomy of the lungs. six cycles of adjuvant chemotherapy with ifosfamide and doxorubicin were given after the diagnosis. with post-adjuvant chemotherapy, the patient was kept on a regular follow-up.

Discussion

Synovial sarcoma is a mesenchymal tumor, accounting for 5-10% of all soft tissue sarcomas [2]. It occurs most commonly in the soft tissues of the extremities of adolescents and young adults, especially near large joints, and also in the parts of the head, neck, eyelids, mediastinum, as well as in the temporoman-



Figure 3. In situ hybridization results (SYT and EWSR1 genes) of the EBUS-TBNA tissues and lymph nodes.

munohistochemical investigations to exclude alternative primary tumors and metastatic sarcoma [7]. At present, PPSS is known to consist of four subtypes, in-

dibular joint, infratemporal fossa, and pulmonary valve [6]. The lung is an extremely unusual anatomical site for synovial sarcoma, and it has rarely been reported there. Primary pulmonary sarcoma is also quite rare and accounts for only < 0.5% of all primary lung malignancies.

The clinical manifestations of patients are mainly chest pain, dyspnea, cough, or hemoptysis [7]. The CT finding of PPSS is typically a wellcluding monophasic fibrous (spindle), monophasic epithelial, biphasic, and poorly differentiated, with the monophasic subtype being the most common [3]. The poorly differentiated subtype is the most complicated, consisting of spindle, polymorphic, and round cells. It is hard to differentiate the poorly differentiated subtype of synovial cell sarcoma from the others through cell morphology, so immunohistochemistry is essential [9]. In immunohistochemistry, PPSS is often found positive for cytokeratin, EMA, bcl-2, and vimentin, and negative for S-100, desmin, smooth muscle actin, and vascular tumor markers [10]. Synovial sarcoma is characterized by a reciprocal chromosomal translocation (X;18)(p11.2;q11.2) which results from the fusion of the SYT gene on chromosome 18 [11]. Current treatment for PPSS primarily consists of surgical resection, radiation therapy, and chemotherapy. The use of adjuvant chemotherapy for soft tissue sarcoma is controversial. The prognosis for PPSS is poor, and there is an overall five-year survival rate of 50%.

In our case, a physical examination and CT were initially used to examine the patient's clinical symptoms. A big soft tissue density and multiple small soft tissue densities were separately found in the left lung lobe and in the bilateral lung lobe. Lung cancer and synovial sarcoma were considered and excluded subsequently by immunohistochemical results. The result of molecular detection showed these tumor cells were positive for SYT, and negative for EWSR1, a gene expressed in Ewing's sarcoma. Hence, the patient was diagnosed with PPSS (poorly differentiated subtype). Because there were multiple lesions which were distributed in the bilateral pulmonary lobe, the patient was treated with chemotherapy, without any surgical resections. The prognosis of the patient was satisfied in 6 months.

Conclusions

In conclusion, we demonstrate a rare case of poorly differentiated primary pulmonary synovial sarcoma. The use of EBUS-TBNA and in situ hybridization in the diagnosis of PPSS is a minimally invasive and accurate technique, rather than having to resort to more invasive surgical biopsy and immunohistochemical analyses.

Disclosure of conflict of interest

None.

Address correspondence to: Yingrong Lai, Department of Pathology, The First Affiliated Hospital of Sun Yat-sen University, 58 Zhongshan 2nd Road, Guangzhou 510080, People's Republic of China. E-mail: laiyr@126.com

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