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Primary Pulmonary Biphasic Synovial Sarcoma Confirmed by Molecular Detection of a SYT-SSX2 Fusion Gene: Report of 1 Case

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Pulmonary sarcoma usually occurs through metastasis from other organs such as the extremities, and primary pulmonary sarcoma accounts for less than 0.5% of all pulmonary malignancies. We experienced a case of primary pulmonary biphasic synovial sarcoma which was confirmed by immunohistochemistry and molecular detection of SYT-SSX2 fusion transcripts. The patient was a 61-year-old men who presented well-defined mass in the left upper lung field on a routine chest radiograph. CT-guided lung needle aspiration biopsy was performed and it showed probable spindle cell tumor. No tumor was found at other site by PET-CT scan. Left upper lobectomy and lymph node dissection was performed. Histologic and immunophenotypic features were consistent with biphasic synovial sarcoma. Reverse transcription-polymerase chain reaction was performed using RNA extracted from frozen tumor samples for the detection of SYT-SSX fusion gene and amplified a single 331bp fragment characteristic of the SYT-SSX2 fusion gene. Chest radiographs taken after 16 months of follow-up showed left pleural effusion. Chest HRCT revealed pleura-based mass and enlarged mediastinal lymph nodes. CT-guided lung needle aspiration biopsy was perfomed. The histologic and immunohistophenotypic features showd the same patterns. The patient was scheduled for a total resection of the tumor along with adjacent chemoradiotherapy.

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A case of low grade malignant fibrous histiocytoma originating from pulmonary artery

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Malignant fibrous histiocytoma (MFH) is the most common soft tissue sarcoma of adults. MFH is thought to have originated from fibroblastic-hystiocytic or primitive mesenchymal cells and occurs most commonly in the extremities presented as painless mass followed by the retroperitoneum. We experienced a young male patient with malignant fibrous histiocytoma in the pulmonary artery presented as severe pulmonary hypertension. A 22-year old male under military service was admitted to our hospital because of syncope and dyspnea on exertion slowly progressed over three months. Chest computed tomography (CT) showed large filling defect involving main pulmonary trunk and all lobar branches bilaterally. On echocardiography, both right atrium and ventricle were dilated and the estimated pulmonary arterial systolic pressure was about 100 mmHg. But D-dimer was only 0.22. Immediately, he underwent palliative debulking surgery for the mass lesion in the pulmonary artery. At first sight, the tumor seemed to be a benign myxoma originating from endothelium of pulmonary artery but turned out to be a malignant fibrous histiocytoma which included focal increased cellularity and mild cytologic atypia. After the surgery, he has finished 6 cycles of chemotherapy consisting of ifosfamide and doxorubicin. As a result, a partial response was seen-the extent of residual intravascular tumor was decreased and both pulmonary hypertension and right ventricular hypertrophy were markedly improved. This is a rare case of MFH originating from pulmonary artery and progresses to severe pulmonary hypertension. In the case of any history extending for months, unresponsiveness to fibrinolytic agents or unexpectedly low level of D-dimer, intravascular neoplasm such as primary sarcoma of the pulmonary artery or metastatic tumor emboli should be suspected as a rare cause of pulmonary arterial occlusion.