CASE REPORT

Occult primary pulmonary synovial sarcoma presenting as recurrent spontaneous pneumothorax and explosive progression

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Abstract
Primary pulmonary synovial sarcoma (PPSS) is a relatively rare neoplasm with highly progressive potential. We present an extremely rare case of PPSS presenting as recurrent pneumothorax with bullous lesions. Bullectomy was performed at the local hospital. Unfortunately, the patient was initially misdiagnosed as atypical carcinoid. Although a negative resection margin was obtained during the first surgery and a remedial operation and chemotherapy followed, the patient developed severe disease progression and died soon after. This report demonstrates that PPSS can easily be misdiagnosed and should be seriously considered in the differential diagnosis of pneumothorax.

Introduction
Synovial sarcomas (SS) typically occur as primary neoplasms of the soft tissues, accounting for 8% of all soft tissue tumors in the body.1 SS occur not only in the para-articular tissues, but also can occur in the lung, head and neck, mediastinum, heart, kidney, prostate, esophagus, and vulva.2 Primary pulmonary SS (PPSS) was first described by Zeren et al. in 1995.1 It is a relatively rare neoplasm accounting for less than 0.5% of malignant lung tumors.3 We report a case of occult PPSS presenting as recurrent pneumothorax with bullous lesions.

Case report
A 37-year-old man presented three times with right-side pneumothoraces over a period of seven months. The previous two pneumothoraces were confirmed by plain chest film without any obvious pulmonary lesions and were cured by chest tube drainage. At the third occurrence, chest computer tomography (CT) scanning revealed bullous lesions located at the right upper lobe without any obvious pulmonary lesions (Fig 1a). Thoracoscopic bullectomy and pleural abrasion was performed at the local hospital. The postoperative course was uneventful. Postoperative pathologic examination showed a dense proliferation of spindle cells (Fig 2). Further immunohistochemical staining showed that the tumor was positive for vimentin, epithelial membrane antigen, CD99, Ki67, partly positive for cytokeratin (CK)8, CK19, and neuron...
specific enolase, but negative for S-100, synaptophysin, or chromogranin A. A reverse transcriptase-PCR assay identified the t(X;18) translocation with the SYT-SSX1 variant. The diagnosis was reconfirmed to be PPSS.

Bone scintigraphy and whole body CT scans did not identify any other disease.

Multidisciplinary discussion including the thoracic surgeon, medical oncologist, pathologist, and radiologist was conducted after general assessment. Considering the malignant nature of PPSS and suspicious local recurrence, we performed progressive right upper lobectomy and lymphadenectomy of levels 2, 3, 4, 7, 8, 9, 10, and 11 lymph nodes (LNs) via thoracotomy a month and a half after the first surgery. Postoperative pathological examination revealed local SS recurrence at the previous operative site. No LN metastasis was found among the 30 LNs dissected. The tumor classification was pT2aN0M0 and stage IB according to the 7th Lung Cancer Tumor Node Metastasis Staging system. The patient recovered well. Four weeks after the surgery, he received two cycles of adjuvant chemotherapy with ifosfamide and doxorubicin. Unfortunately, he developed progressive dyspnea and hemoptysis during adjuvant chemotherapy administration. Chest CT scans revealed massive recurrent lesions both in the right residual lung and pleural cavity (Fig 1c,d). He died three months after the second surgery.

Figure 1 Chest computed tomography scanning. (a) Bullous lesions located at the anterior segment of the right upper lobe before the first operation (red arrow). (b) A cord-like shadow and a nodule developed at the previous operative site one month after the first operation (red arrow). (c,d) Multiple pleural nodules and mass like lesions in the residual lung developed only two months after the second operation (red arrows).

Figure 2 Microscopic pathologic examination showing histopathological features of spindle cell sarcoma (hematoxylin and eosin x40).
Discussion

Primary pulmonary synovial sarcoma is a highly aggressive tumor. Most patients diagnosed with PPSS present with symptomatic or asymptomatic well-circumscribed nodules or masses. However, PPSS presenting with occult pneumothorax and manifesting as a cystic/bullous lesion is not well recognized. Only a few cases of PPSS with occult pneumothorax have been reported in the literature. Our patient was an extremely rare case presenting as recurrent pneumothorax with bullous lesions. However, plain chest film failed to detect the occult lesions during the previous two episodes.

Histologically, PPSS can be classified into four subtypes, including monophasic fibrous, monophasic epithelial, biphasic, and the poorly differentiated monophasic subtype. Because of its unusual histological features and often benign appearance, PPSS can easily be misdiagnosed. For those tumors with spindle cell proliferation on microscopic view, if the patients present with pneumothorax, a small set of tumors should be taken into consideration for differential diagnosis, including lymphangioleiomyomatosis, solitary fibrous tumor of the pleura, pleuropulmonary blastoma, and metastatic SS. Although cytokeratin, CD99, B-cell lymphoma 2 and epithelial membrane antigen are positive in most SS cases, these makers are unspecific. This factor makes confirmation of diagnosis difficult. The most important diagnostic tool for SS is the identification of typical translocation t(X;18)(p11.2;q11.2) by fluorescence in situ hybridization or reverse transcriptase-PCR. Usually, this translocation produces a fusion transcript of the SYT gene (exon 10) on 18q and the SSX1 gene on Xp (exon 6). Our patient was also initially misdiagnosed at the local hospital as atypical carcinoid because of its subtle pathological features. Immunohistochemical staining and cytogenetic examination finally confirmed the diagnosis of PPSS; however, because of the limitations of medication for SS and the absence of metastasis on general assessment, we performed remedial surgery for this patient. Although a negative resection margin was obtained during the first operation, he developed local recurrence quickly thereafter. Despite the remedial surgery and chemotherapy, he developed severe disease progression and died soon after.

In conclusion, this report demonstrates that PPSS can easily be misdiagnosed and should seriously be considered in the differential diagnosis of pneumothorax.

Acknowledgments

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Disclosure

No authors report any conflict of interest.

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