

Progressive Growth of Primary Synovial Sarcoma of the Lung

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An 80-year-old male was admitted because of a giant mass in the left lower lobe of the lung on a routine chest X-ray. Chest computed tomography verified this to be a well-defined heterogeneous mass as described with no associated lymphadenopathy. FDG-PET depicted moderately marginal FDG uptake. The patient underwent a left lower lobectomy and lymphadenectomy. Grossly, the tumor measured 60 × 50 mm and was uniformly filled with a pure white, pudding-like friable substance. No lymph node metastasis was observed microscopically. Histologically, the tumor showed a dense proliferation of rounded or spindled malignant cells with a frequent mitotic activity and an increased nuclear-to-cytoplasmic ratio. The immunohistochemical staining was positive for vimentin, negative for cytokeratin, keratin-wide, EMA, CD34. A SYT-SSX2 fusion gene transcript was detected as a result of RT-PCR analysis. Because of these results, the tumor was diagnosed as a monophasic synovial sarcoma. (Ann Thorac Cardiovasc Surg 2010; 16: 194–197)

Key words: synovial sarcoma, lung, operation, SYT-SSX

Introduction

Primary lung sarcoma is rare and accounts for less than 0.5% of all malignant tumors of the lung.¹ Previous reports suggested that synovial sarcoma is the third common pathological type of soft-tissue sarcoma, next to liposarcoma and malignant fibrous histiocytoma.² Synovial sarcoma is usually classified into three major histological subtypes: monophasic (epithelial and fibrous type), biphasic, and poorly differentiated type. Although the leading treatment for a synovial sarcoma of the lung is considered to be a surgical resection, several patients with this condition have also received chemoradiotherapy pre- or postoperatively.³

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Received October 20, 2009; accepted for publication May 20, 2009
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A preoperative diagnosis of synovial sarcoma is so difficult that most diagnoses for this condition are made postoperatively, with supplementary immunohistochemistry, as we experienced in this case.³

Here we report a case of synovial sarcoma of the lung receiving curative surgical resection.

Case Report

An 80-year-old male with a history of previous smoking was admitted because of a giant mass in the left lower lobe of the lung on a routine chest X-ray (Fig. 1a). He had no complaints, and a physical examination revealed no problems in particular. Chest computed tomography (CT) verified a well-defined heterogeneous giant mass in the left lower lobe of the lung with no associated lymphadenopathy. Neither spicula nor pleural indent was observed. He had received a chest CT by chance six months before admission, but no abnormal shadow was then observed at all (Figs. 1b, 1c). Thus the giant mass grew within only six months. The FDG-PET depicted a moderately marginal FDG uptake coincident with CT findings (SUV; 6.58). A preoperative

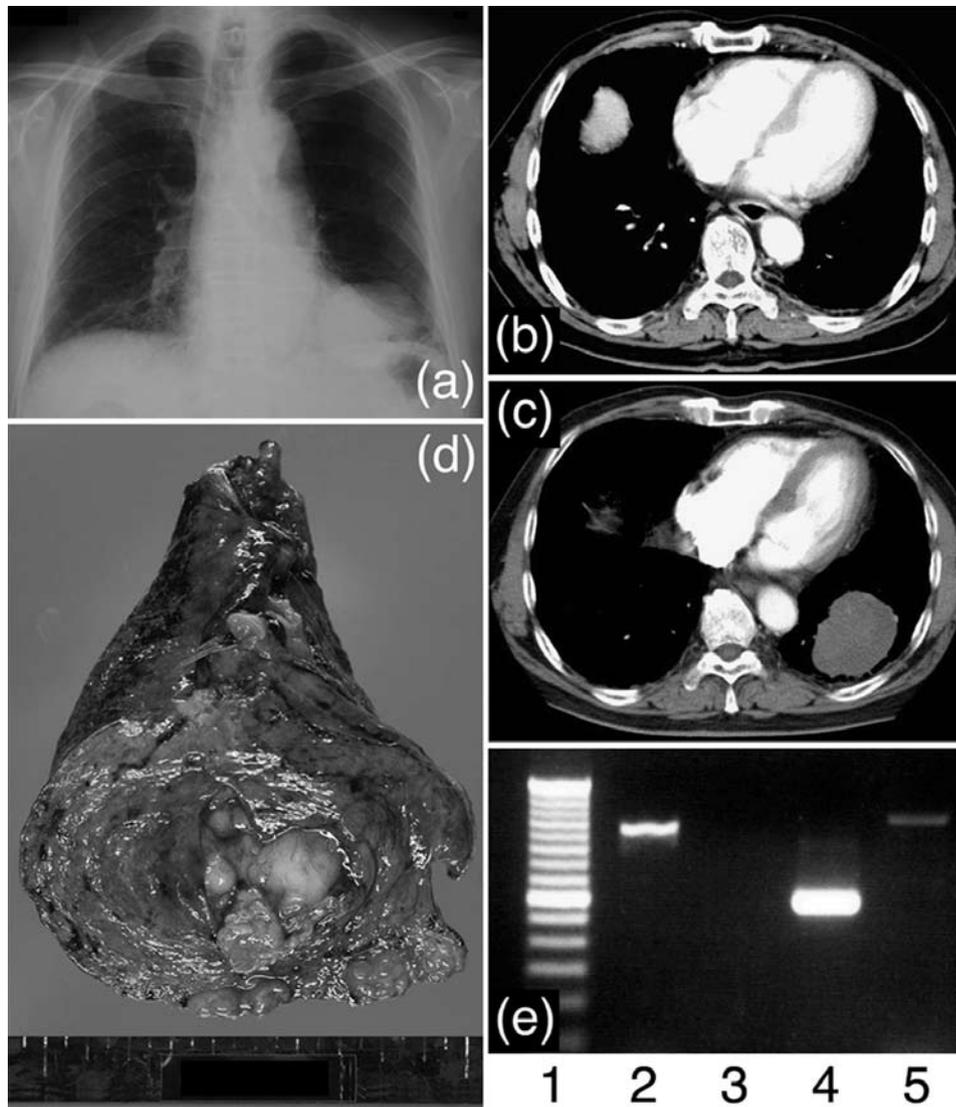


Fig. 1. (a) Chest X-ray shows a giant mass in the left lower lobe of the lung. (b) No abnormal shadow is observed on chest CT. (c) A well-defined heterogeneous giant mass in the left lower lobe of the lung without any associated lymphadenopathy. (d) The tumor measured 60 × 50 mm and was uniformly filled with a pure white, pudding-like friable substance. (e) RT-PCR of SYT-SSX fusion gene transcripts: (1) marker, (2) SYT-SSXs, (3) SYT-SSX1, (4) SYT-SSX2, (5) GAPDH.

transbronchial lung biopsy showed several malignant cells with poor differentiation, which is symptomatic of a sarcoma or a malignant solitary fibrous tumor.

He underwent a left lower lobectomy and lymphadenectomy. The tumor was entirely confined to the left lower lobe; thus no exposure to the pleura or pleural effusion was observed. Grossly, the tumor measured 60 × 50 mm and was uniformly filled with a pure white, pudding-like friable substance. There were no solid parts or areas of calcification present (Fig. 1d).

Histologically, the tumor showed a dense proliferation

of rounded or spindle malignant cells with a frequent mitotic activity (10–20 mitoses per 10 HPFs) and an increased nuclear-to-cytoplasmic ratio (Fig. 2a). Furthermore, there were extensive necrotic findings and vascular invasion. Surgical margin was free, and no lymph node metastasis was observed. The immunohistochemical staining was positive for vimentin, weakly positive for bcl-2 and CD99, and negative for cytokeratin, keratin (polyclonal), EMA, CEA, TTF-1, α -SMA, desmin, S-100, CD34, CD117, and carletinin (Figs. 2b, 2c). Also, an increased number of Ki-67 positive cells were observed (Fig. 2d). Furthermore,

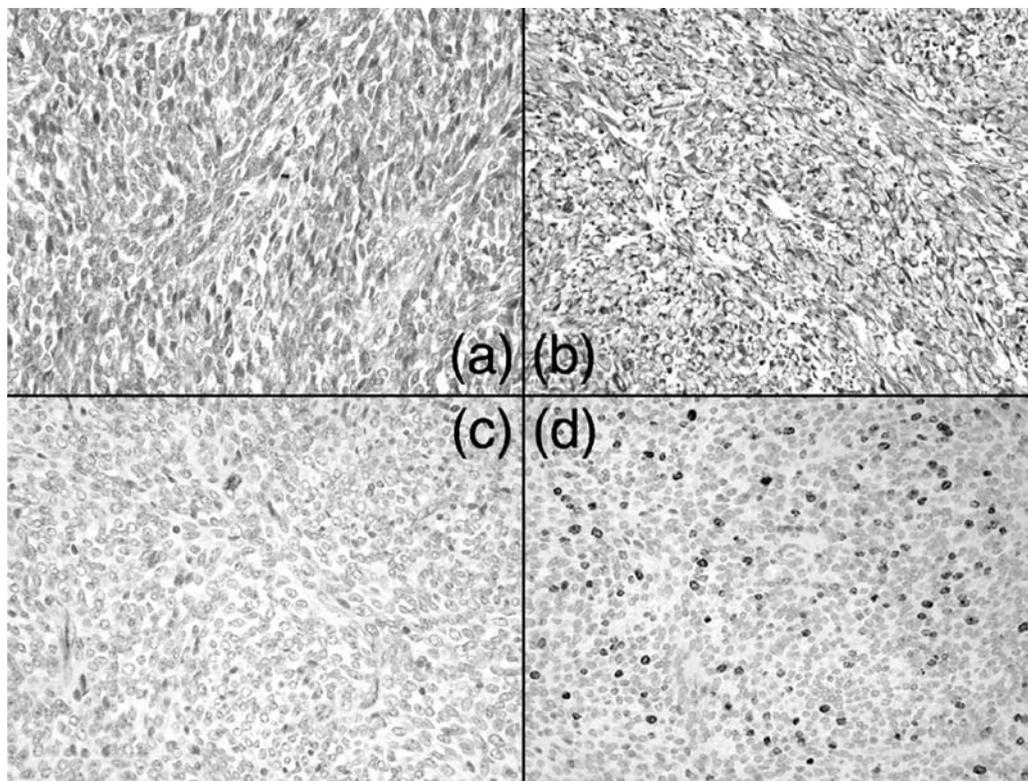


Fig. 2. Immunohistochemical staining of the tumor. (a) Tumor with dense proliferation of rounded or spindled malignant cells with a frequent mitotic activity and an increased nuclear-to-cytoplasmic ratio (H.E. $\times 100$). (b) Tumor with positive stain for vimentin. (c) Tumor with negative stain for CD34. (d) Tumor with increased number of Ki-67 positive cells.

a SYT-SSX2 fusion gene transcript was detected as the result of an RT-PCR analysis for the SYT-SSX fusion gene (Fig. 1e). Because of these results, the tumor was diagnosed as a monophasic synovial sarcoma.

The patient is still alive 11 months after operation, and no recurrence was observed.

Discussion

Primary synovial sarcoma is scarcely rare in malignant lung neoplasms. Synovial sarcoma occurs mainly in the large joints, but it has also been observed in the head and neck, mediastinum, prostate, lung, and many other organs.⁴⁾ Until now, only about 60 cases of synovial sarcoma of the lung had been reported in English literature. In general, the age distribution in patients with synovial sarcoma is higher in comparison to those with other soft tissue counterparts. It is interesting that Chan et al. suggested that elderly patients with synovial sarcoma often show a poorly differentiated histology and develop the condition at an unusual location, such as a limb.⁵⁾ Our case

had an extremely high age, and though the histological subtype was monophasic, the differentiation was relatively poor.

Recent cytogenetic studies have revealed that synovial sarcoma has a consistent chromosomal translocation $t(x;18)(p11;q11)$ and that this translocation fuses the SYT gene to either of the two homologous genes SSX1 or SSX2.⁶⁾ These fusion genes are thought to be associated with tumorigenesis, but the target gene is still unknown.

A variety of parameters have been reported to be possible predictors of the outcome of synovial sarcoma. Bergh et al. divided patients into high- and low-risk groups.⁷⁾ Overall disease-free survival of the low-risk group (patients' ages < 25 , tumor size < 5 cm, and no histological evidence of a poorly differentiated tumor) was 88%; that of the high-risk group (patients' ages ≥ 25 , tumor size ≥ 5 cm, and evidence of a poorly differentiated tumor) was only 18%. Furthermore, Trassard et al. suggested that high mitotic rate might be a poor prognostic factor.⁸⁾ Because our case satisfies at least three of these high-risk criteria (age, tumor size, high mitotic rate), a strict follow-up of this patient will therefore be necessary.

Immunohistochemically, most synovial sarcomas that test positive for vimentin, cytokeratins, and EMA have a lower immunoreactivity for S-100 and CD34. Our case was positive for vimentin, but negative for any cytokeratin subtypes. From this perspective, the present case did not appear to have a typical synovial sarcoma.

Because the best treatment for synovial sarcoma remains undefined, a complete resection with a free surgical margin seems to be one of the best options. Several reports have suggested that a local or wedge resection may create a risk of recurrence and result in a shorter survival.⁹⁾ In our case, the tumor was entirely confined to the left lower lobe and no lymph node metastasis was observed; therefore a lobectomy could be a curative operation.

In conclusion, we experienced a case of synovial sarcoma of the lung. The progression of the tumor was very rapid, but the operation seemed to be curative. Regardless, because of the high-risk nature of the patient, a long-term follow-up will thus be necessary.

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