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# Imaging Manifestations of Multiple Primary Pleuropulmonary Synovial Sarcoma: A Case Report

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## Abstract

Primary Pleuropulmonary Synovial Sarcoma (PPSS) is a rare subtype of synovial sarcoma. Most reported cases involve single lesions, but our case involves multiple lesions occurring in different pleurae with both CT and MRI. A 42-year-old man presented to the Fifth People's Hospital of Shanghai, Fudan University, with pain in the left upper chest without obvious cause for nearly half a year. There were no obvious abnormalities in multiple laboratory indicators of the patient. CT and MRI examinations showed multiple lesions on the left oblique fissure pleura and diaphragmatic pleura. The patient underwent surgery on January 19th, 2021 and multiple chemotherapy treatments subsequently. The postoperative pathology confirmed that it was monophasic-fibrous type synovial sarcoma. PPSS is highly malignant and rare; the correct diagnosis of radiologist will be of great importance for treatment and prognosis. We have CT and MRI images of the lesion for this patient, combined with the imaging characteristics of previously reported cases, looking forward to provide valuable information regarding the imaging manifestations of PPSS.

## Case Presentation

A 42-year-old male presented with pain in the left upper chest with no obvious cause for nearly six months. The patient's clinical laboratory indicators such as complete blood count, clinical urine tests, cardiac markers and biochemical indicators were generally normal. CT (Figures 1-4) showed lesions on the left oblique fissure pleura and diaphragmatic pleura respectively. The largest was on the left oblique fissure pleura. It was 5.5 cm × 4.4 cm in size, had non-specific form and clear boundary, exhibited isodensity mainly with some patchy hypodense areas and had a pleural connection with wide substrate. There were no signs of lymphadenectasis in the mediastinum or hilum pulmonis. On contrast-enhanced CT, the patchy hypodense areas manifested no enhancement and the remaining substantive areas showed mild enhancement. The lesions on the left diaphragmatic pleura revealed uniform mild enhancement. A small volume of pleural effusion was observed in the left thorax. MRI (Figures 5-7) demonstrated the lesion on the left oblique fissure pleura. T1WI-FS showed iso-hypointense lesions with some patchy more hypointense areas. While in T2WI-FS, it was slight hyperintensity with some patchy hypo-hyperintense areas. The patchy hypointense areas revealed no enhancement and the remaining substantive areas showed mild enhancement on contrast-enhanced T1WI-FS. A thoracoscopic lobectomy and a biopsy of the patient's lesions were performed. The hematoxylin-eosin staining (Figure 8,9) revealed that the lesions were monophasic-fibrous type synovial sarcoma. The immunohistochemistry results were as follows: CK (-/+), Vim (+), SMA (bit -/+), Desmin (bit -/+), EMA (+/-), D2-40 (+/-), Ki67 (+, hot pot 15-20%), BcL-2 (+), and S-100 (-/+). Subsequently, the patients received multiple chemotherapy treatments.

## Discussion

Synovial Sarcoma (SS) is a relatively rare malignancy representing a Soft Tissue Sarcoma (STS). SS often occurs in young adults and mostly occurs in the limbs, especially the tendons, tendon sheaths, and synovial areas [1]. Most studies suggested that synovial sarcoma did not originate from synovial tissue but from pluripotent interstitial tissue that differentiated into both the epithelial and interstitial tissue types [2]. Primary Pleuropulmonary Synovial Sarcoma (PPSS) is a rare subtype of synovial sarcoma that usually originates from the chest wall, pleura or lung and accounts for only 0.5% of all primary lung malignancies [3]. The clinical manifestations of PPSS are not specific. Typical symptoms include chest pain (24% to 80%), difficulty breathing (8% to 36%), cough (8% to 33%) and hematosis (20% to 25%). Approximately 40% of patients have no obvious symptoms, and lesions are found only on physical examination [4]. Regarding imaging manifestations, PPSS appeared iso-hypodensity with clear boundary, no bone damage or tissue invasion, pleural reactions

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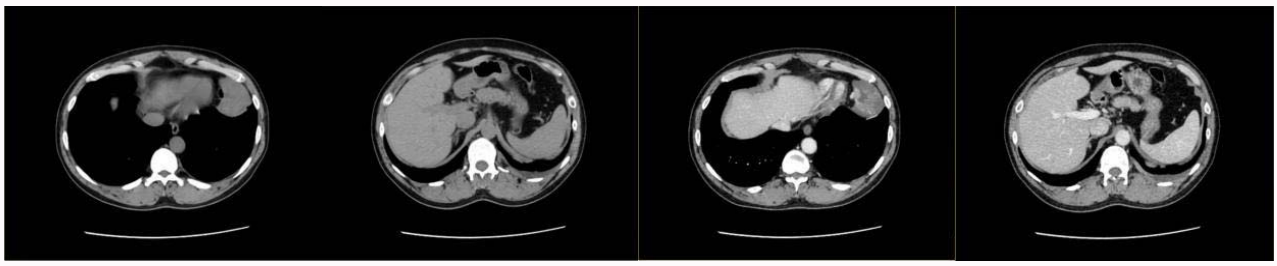
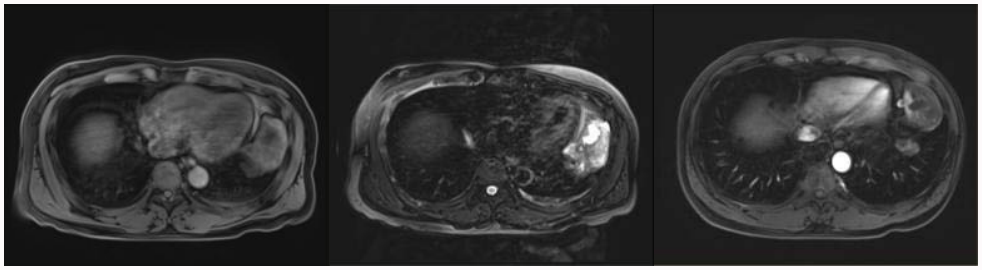


**Figures 1-4:** CT images before surgical resection of primary pleuropulmonary synovial sarcoma on both left oblique fissure pleura and diaphragmatic pleura.



**Figures 5-7:** MRI images before surgical resection of primary pleuropulmonary synovial sarcoma on the left oblique fissure pleura.

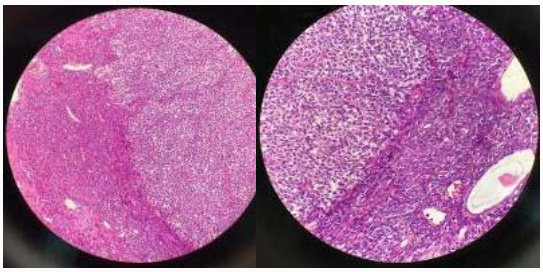


**Figure 8 and 9:** He 200x and He 400x: A large number of spindle cells arranged in bundles with unclear gaps.

and pleural effusion could be seen on CT scan [5]. Few lesions showed calcifications [6] and lymphadenopathy in the mediastinum and hilum pulmonis [4,7,8]. The cystic necrosis areas manifested no enhancement with mild enhancement in the remaining substantive areas after enhanced, thin and irregular ring-like enhancement was also a common feature [4,7,9,10]. On MRI scan, T1WI demonstrated lesions were iso-hypointensity while hyperintensity in T2WI [4,7]. Effusion of the lesion caused by bleeding or cystic necrosis was also reported [11]. On contrast-enhanced MRI, this tumor commonly appears as a well-defined heterogeneous mass with a thin and irregular peripheral enhancing rim. The internal complexity of this heterogeneous tumor is known to be due to enhanced nodular soft-tissue components combined with hypoattenuated cystic, necrotic or hemorrhagic [4,7]. In Summary, uneven density/intensity, clear boundary, lack of calcification, uneven enhancement, pleural effusion and lack of lymphadenopathy in our study were consistent with the features of previous studies while the peripheral ring-shaped enhancement was not. According to the composition and degree of differentiation of the synovial tissue determined using an optical microscope, PPSS is usually divided into 4 pathological types: Biphasic, monophasic-fibrous, monophasic-epithelial, and poorly differentiated. The biphasic and monophasic-fibrous types are common. More precisely, poorly differentiated tumors can actually develop in either the monophasic or biphasic subtype, and can be regarded as a form of tumor progression rather than a distinct subtype [12]. Immunohistochemical markers of PPSS include CK (+), EMA (+), Vim (+), BcL-2 (+), CD99 (+/-), Calponin (+/-), SMA (-), Desmin (-) [13]. Currently, some scholars have reported that large-scale tumor resection and surrounding tissue resection are the best choices for the treatment of synovial sarcoma. Chemotherapy and radiotherapy are supplementary strategies that can be used to achieve a negative tumour resection margin [14]. However, PPSS is prone to recurrence and metastasis, the five-year survival rate of it about 50% [15].

## Conclusion

In summary, PPSS is a rare soft tissue tumor derived from pluripotent interstitial tissue. Although its radiographic findings lack specificity, they still have certain characteristics. Typical images of PPSS present as well-defined boundary lesions with uneven density/intensity, uneven enhancement, often accompanied by cystic necrosis and pleural effusion, no signs of lymphadenectasis in the mediastinum or hilum pulmonis and related to the pleura closely. Pathological and immunohistochemical tests are also helpful in the diagnosis of the disease.

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