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Review Article

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PLEUROPULMONARY SYNOVIAL SARCOMA

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ABSTRACT

Pleuropulmonary synovial sarcoma (PPSS) is an extremely rare malignant tumour, which is increasingly recognized as a subtype of sarcoma with a distinctive chromosomal translocation specific to synovial sarcoma. It is often presented like any thoracic tumour with symptoms such as chest pain or cough, breathlessness. Here we report a case of PPSS in a 19-year-old boy presenting with cough, shortness of breath and chest pain. And who were found upon histologic examination of the resection specimen to have cystic primary pleuropulmonary synovial sarcoma.

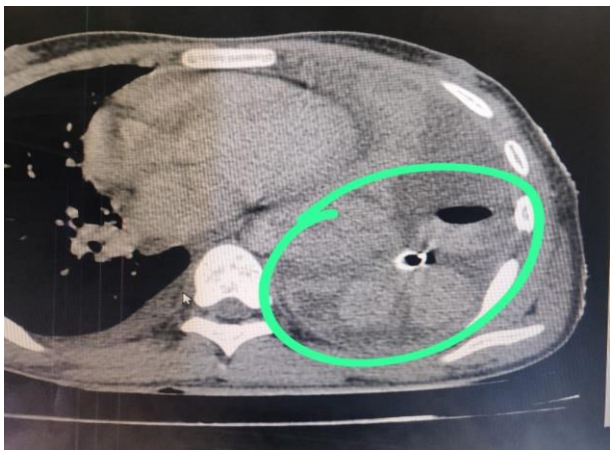
KEYWORDS: Synovial sarcoma, Neoplasm.

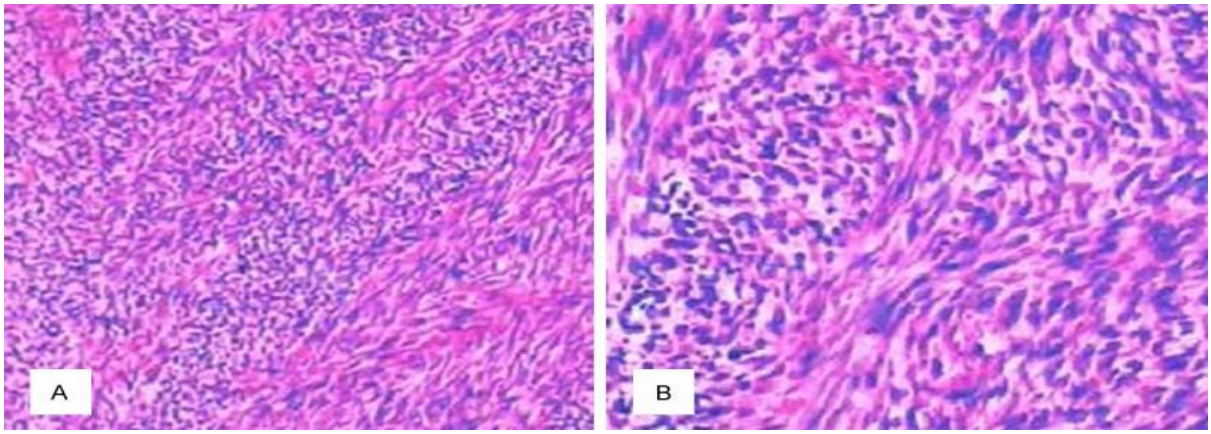
INTRODUCTION

Synovial sarcoma is a malignant mesenchymal neoplasm, which most commonly occurs near the joints of the extremities. However, primary pleuropulmonary synovial sarcoma (PPSS) is an extremely rare tumour. We present a case of PPSS confirmed by Histopathology.

MATERIALS AND METHODS

A previously healthy, 19-year-old boy was admitted to our hospital with a cough, shortness of breath and chest pain for one month. Chest computed tomography (CT) showed a large heterogeneous mass, 7.1 cm×6.4 cm×9.4 cm, in the left hemithorax with solid and cystic components. There was a wide area of severe adhesion to the adjacent structures. The adjacent lower lobe of the right lung was compressed and consolidated, and there was mild ipsilateral pleural effusion. No lymph node involvement or distant metastatic disease was noted. On gross examination, the tumour contained a variety of necrotic, cystic and solid components with 7 cm in greatest diameter. Pathological examination was done in the outside lab, revealed proliferation of oval to spindle-shaped tumour cells with variation in morphology in fascicular and herringbone patterns, Immunohistochemical staining results demonstrated that the tumour cells were positive for vimentin, keratin and CD99, focally positive for EMA, but negative for CD34, S-100, smooth muscle actin and desmin. Pathologic diagnosis was synovial sarcoma with extensive necrosis.





DISCUSSION

Synovial sarcoma is a type of spindle cell tumour that mainly affects the extremities in adolescents and young adults and accounts for 2.5%~10% of all soft-tissue sarcomas.[1] However, PPSS is an extremely rare malignant tumour, which is increasingly recognized as a subtype of sarcoma because of the recent identification of a distinctive chromosomal translocation specific to synovial sarcoma.[2] PPSS demonstrates more aggressive clinical behaviour than soft-tissue synovial sarcoma.[3] It can originate from the lung parenchyma, the pleura, the mediastinum or the chest wall. PPSS chiefly affects young and middle-aged adults.

On chest radiographs, PPSS typically appears as a sharply marginated mass with uniform opacity, based either in the pleura or in the lung, and often accompanied by an ipsilateral pleural effusion.[2] The most common CT findings of PPSS have been described to be a heterogeneously enhancing soft-tissue mass with a well-defined margin and ipsilateral pleural effusion, but without lymphadenopathy.[2,4,5] Although calcification is a common finding in soft-tissue synovial sarcoma at para-articular sites and can be seen in 30% of lesions, PPSS usually lack tumour calcifications.[2,5] Magnetic resonance imaging provides superior demonstration of nodular soft tissue and multilocular fluid-filled internal components of PPSS and often allows more accurate delineation and localization of the tumour and is helpful for determining the presence and extent of tumour invasion and for tissue characterization.[6] PPSS usually demonstrates internal heterogeneity, with predominantly intermediate signal intensity (isointense to the signal in the chest wall musculature) on T1- and T2-weighted images. After the administration of a gadolinium-based contrast material, T1-weighted images may show areas of dramatic heterogeneous enhancement that correspond to lobules of viable tumour. The radiologic manifestations of PPSS overlap with those of many other lesions of the lung and pleura. Differential diagnosis includes malignant mesothelioma, metastatic tumour, primary lung cancer, solitary fibrous tumour and other rare primary mesenchymal sarcomas. The final diagnosis depends on the histologic and immunohistochemical staining results.

Synovial sarcoma is a mesenchymal spindle-cell tumour characterised by variable epithelial differentiation and the specific chromosomal translocation t(X; 18) (p11.2; q11.2) that results in fusion of the *SYT* gene on chromosome 18 with the *SSX1* or *SSX2* gene on chromosome X. It encompasses two histologic subtypes, monophasic and biphasic, with the monophasic variant being the more common. The presence of poorly differentiated tumour cells within lesions of either subtype is considered indicative of a poorer prognosis.

Although there is no gold standard of treatment for PPSS, a multidisciplinary approach, including surgical resection, chemotherapy, and radiotherapy has been suggested. Radical resection is the mainstay of treatment. Neoadjuvant chemotherapy can be beneficial prior to radical resection since it can cause reduction in tumour volume and potentially treat micrometastasis. In available case series and reports, the 5-year survival rate for synovial sarcoma is variable, depending on the patient's age, the tumour size, high grade, neurovascular invasion, *SYT-SSX1* variant, and its resectability. An age greater than 20 years at diagnosis and the trend in size (≥ 5 cm) were associated with a significantly worse prognosis. In our case both prognostic factors (> 20 years of age, size of tumour > 5 cm) suggests a relatively poor prognosis for the patient. Thus, the patient was offered neoadjuvant chemotherapy followed by complete resection and long-term follow-up is being carried out.

CONCLUSION

Because the morphologic features of primary and metastatic synovial sarcomas are similar, clinical and radiologic evaluation is essential to exclude the presence of a primary tumour outside the thorax before a diagnosis of PPSS can be confirmed. Despite being extremely rare, PPSS should be considered on the differentiation of a primary lung or pleura mass, especially in young and middle-aged adult patients. Current treatment consists of surgical resection followed by chemotherapy, radiation therapy, or both.

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