Primary biphasic synovial sarcoma presenting as a lung mass

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Abstract

Synovial sarcoma was defined by the World Health Organization (WHO) in 2002 as a type of mesenchymal tissue cell tumour that exhibits epithelial differentiation and represents the third most common soft-tissue sarcoma in adults, accounting for approximately 10% of soft-tissue sarcomas. To date, there are only a few reports of primary pulmonary synovial sarcoma. This report describes a case of a 12 cm primary pulmonary giant synovial sarcoma, diagnosed in a 20-year-old patient admitted in our Department of Surgery with a six-month history of cough productive of bloody sputum.

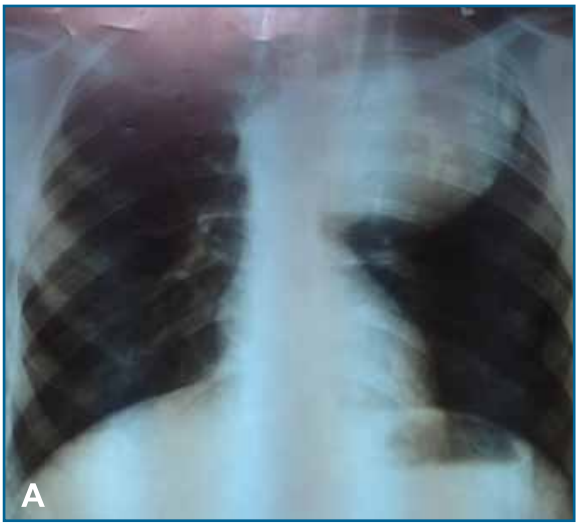
Introduction

Most lung tumours are malignant and tend to be carcinomas. Primary pleuropulmonary synovial sarcoma is one of the rarest pulmonary soft-tissue malignancies. It presents great diagnostic challenges in areas where there are no well-established pathology and radiology facilities, especially if there are no usual histological patterns. This is a disease that commonly affects the deep soft tissues of the extremities and young adults. Molecular testing for the pathognomonic t(x;18) chromosomal translocation has enabled diagnostic confirmation in almost all cases. In t(x;18)-negative cases, diagnosis must rely on histological and immunophenotypic features. We present a case of primary pulmonary synovial sarcoma in a young patient presenting with cough.

Case summary

Our patient was a 20-year-old man who presented with occasional cough that had been present for one year; the cough was productive of a small amount of blood-tinged sputum. The cough was later accompanied by dull, aching left posterior chest pain. The patient also claimed he had lost weight, but this was unquantified. Otherwise he had no shortness of breath, night sweats, or loss of appetite. He was treated with anti-tuburculosis (TB) drugs, initiated based on chest X-ray and completed six months' treatment to no avail. The patient claimed that he had no contact history with a known TB patient with chronic cough. Physical examination revealed nothing except relatively decreased air entry on the left upper lung field. He was investigated and laboratory investigations (such as complete blood count and blood chemistry) were within the normal limits. Serum tumour markers such as alpha-feto protein and human chorionic gonadotropin (hCG) were also determined and they were found to be normal.

A postero-anterior chest X-ray showed a densely radio-opaque shadow on the left upper chest, occupying almost one third of the hemi-thorax. There were no other apparent parenchymal changes (Figure 1(a)). Chest computed tomography (CT) scan revealed a heterogeneously enhancing left upper lobe rounded mass with no areas of calcification, measuring about 12 cm in its



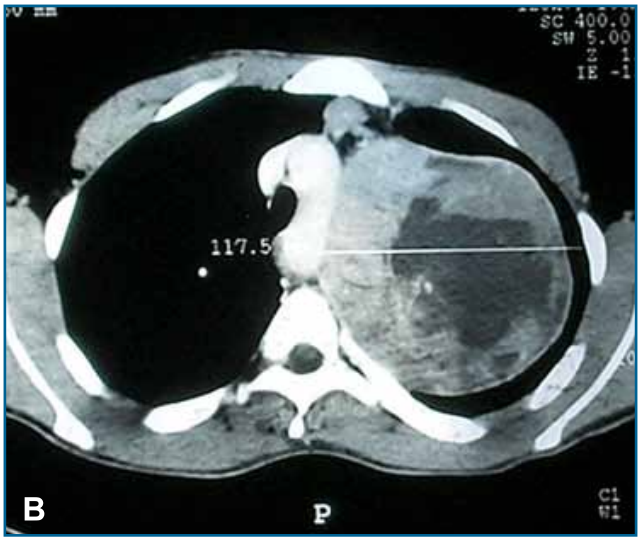


Figure 1: Chest X-ray and CT scan of the chest: (a) a large densely radio-opaque shadow in the left upper lobe area; (b) heterogeneously enhancing mass with central hypodensity which indicates necrosis.

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greatest dimension.

With the radiologic and clinical suspicion of primary lung teratoma, the patient was prepared for surgery. The approach was through a standard posterolateral thoracotomy. There was minimal adhesion between the chest wall and the lung, which was released without much difficulty. The tumour occupied almost the whole of the upper lobe and it had some inflammatory adhesion with the mediastinal pleura. This was released with some difficulty and moderate bleeding. Subsequently the vessels, followed by the upper bronchus, were divided and an upper lobectomy was carried out and a specimen sent for histopathology. The histology showed features consistent with biphasic synovial sarcoma (Figure 2). The patient was counselled regarding subsequent adjuvant treatment but he was lost from follow-up.

Discussion

Synovial sarcoma was defined by the World Health Organization (WHO) in 2002 as a type of mesenchymal tissue cell tumour that exhibits epithelial differentiation[1] and represents the third most common soft-tissue sarcoma in adults

Synovial sarcoma accounts for 5–10% of all soft-tissue sarcomas, occurring mainly near the big joints of the extremities; however it can occur in many sites in the body, including the lungs and mediastinum, and peritoneal and retroperitoneal areas. Synovial sarcoma of the lung is one of the rarest sarcomas that primarily originates from the lung tissue; as a result of this it is overlooked as a differential diagnosis. It is a highly aggressive tumour with a slight male predilection and is not related to cigarette smoking.[2] It is prevalent in individuals between the ages of 15 and 40 years,[3] commonly presenting with chest pain. The term 'synovial' sarcoma was given because of the synovial differentiation of the tumour that is believed to originate from multipotential mesenchymal cells. The diagnosis of primary pulmonary synovial sarcoma requires clinical, radiological, pathological, and immunohistochemical investigations to exclude alternative primary tumours and metastatic sarcoma.

The tumours are of four types: (a) monophasic fibrous (spindle), composed of homogeneous spindle cells with pale-staining nuclei arranged in fascicles and sheets, embedded in a variable background of myxoid to densely collagenous elements; (b) monophasic epithelial, composed of relatively uniform spindle cells with elongated nuclei, slightly basophilic cytoplasm and indistinct cell borders with tumour cells densely packed and little intervening stroma; (c) biphasic, composed of both spindle and epithelial cells; and (d) the poorly differentiated form, monophasic subtype being the most common.[4] Calcification in these tumours occurring in the periarticular region is said to be relatively common but is seldom found in thoracic lesions.[5] The monophasic type is difficult to diagnose, because it has a uniform spindle cell pattern; thus it may be confused with other malignant spindle cell neoplasms, such as fibrosarcoma, hemangiopericytoma, leiomyosarcoma, and spindle cell carcinoma or carcinosarcoma. Differentiation of these tumours requires immunohistochemistry of different membrane proteins such as cytokeratin, epithelial membrane antigens, CD99, and calponin which are positive in synovial sarcoma. Diagnosis of the biphasic type is not difficult because it contains both epithelial and spindle cell components.[2] However, care should be taken not to confuse the epithelial component with entrapped epithelium of alveoli or bronchioles; this was not the case in our patient.

Synovial sarcoma is characterised by translocation t(x: 8)(p11; q110). PCR studies for this translocation can help differentiate between the types of synovial sarcoma in the lung but despite its high sensitivity, molecular testing is not required if the diagnosis of synovial sarcoma is certain or probable on the basis of clinical, histological, and immunohistochemical evaluations.[6,7]

Two-thirds of primary pulmonary synovial sarcomas are centrally located and present with post-obstructive pneumonia, atelectasis, and haemoptysis. Peripheral tumours are less common and usually asymptomatic, but may infiltrate adjacent pleura, thoracic wall, and mediastinum, or metastasize to hilar or mediastinal lymph nodes, adrenal, brain, and spinal cord.[8] Our patient presented with a centrally located tumour with mild haemoptysis and chest pain, and the mediastinal pleura was involved and had to be excised with the tumour. With regard to imaging tools, a chest X-ray is usually the first and least accurate examination performed in all thoracic pathologies; 78% of these tumours have well-delineated circular opacity on posterior–anterior chest X-rays, with no sign of calcification.[9] A CT scan may reveal space-occupying lesions in the thoracic cavity with no specific radiological pattern compared with other mediastinal stromal tumours, including necrotic, haemorrhagic, or cystic components on section with areas of soft tissue density. There is no propensity for sidedness.[10] Pneumothorax may be evident in some patients. The real site of origin (lung, pleura or mediastinum) is often unclear, but acute or recurrent haemothorax and a rim of ground-glass opacity surrounding the mass has been reported in a case of pulmonary synovial sarcoma.[11]

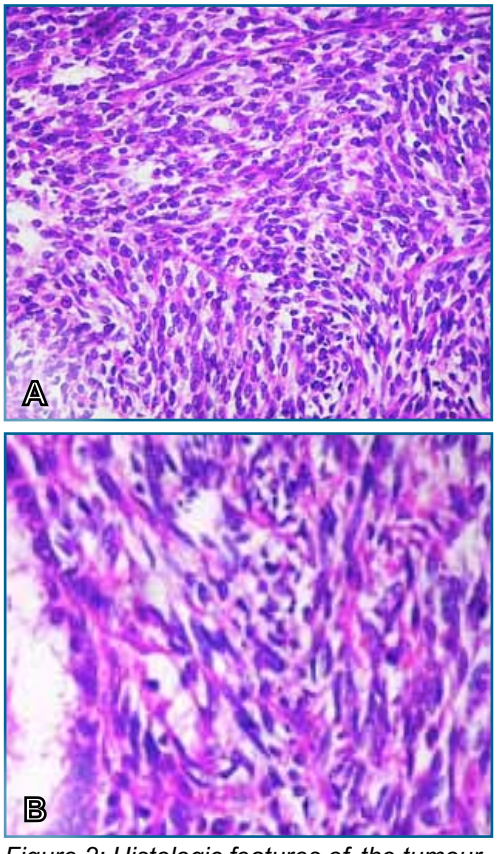

Figure 2: Histologic features of the tumour. (a) The dominant spindle cell components of the tumour consisted of intervening fascicles of densly packed elongated cells. (b) The smaller epithelial areas show glandular differentiation.

Owing to its rarity and the paucity of data

regarding its natural history, there are no guidelines for optimal treatment. Therefore, current treatment includes complete surgical resection (lobectomy or pneumonectomy) followed by adjuvant radiotherapy or chemotherapy.[12] Cure rates are related to how radical the resection was, even then recurrence has been seen.[13] Hence long-term follow up is mandatory. Prognosis is related to the disease stage and is usually poor. In available case series and reports, the five-year survival ranges between 36 and 76%.[8] A full- body scan is part of the management of these patients to rule out the presence of other primary sites. In our patient extensive clinical examination (done after histologic confirmation of the disease) did not reveal any other site of mass or dissemination of the tumour.

In conclusion, primary pulmonary synovial sarcoma is an extremely rare neoplasm. Clinical and imaging investigation is necessary to exclude alternative primary sources, while a definitive diagnosis requires detailed immunohistochemical staining. Surgical excision with clear margins and possibly adjuvant chemo-radiotherapy is the currently accepted treatment. In addition, due to the high risk of recurrences, long-term follow up is needed.

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Author Declaration

Competing interests: none.

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