Primary mediastinal synovial sarcoma with transdiaphragmatic extension presenting as a pericardial effusion

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ABSTRACT

Synovial sarcoma is a distinctive soft tissue neoplasm, most commonly seen in the extremities of young adults. Mediastinal synovial sarcoma is a well-documented entity; however, in many cases, the differentiation between this and other spindle cell tumours may be difficult, especially in monophasic tumours. Unlike most pleuropulmonary synovial sarcomas which are well circumscribed, mediastinal tumours are often infiltrative and resection may not be adequate, leading to a high rate of recurrence. We present a 49-year-old man with a primary pericardial synovial sarcoma, with transdiaphragmatic intra-abdominal extension, which clinically, radiologically and grossly mimicked a tuberculous pericarditis.

Keywords: mediastinal synovial sarcoma, pericardial effusion, pericardial synovial sarcoma, pericarditis, synovial sarcoma

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INTRODUCTION

Synovial sarcoma is a distinctive soft tissue neoplasm, most commonly seen in the extremities of young adults, but also well described at other sites, including intrathoracic locations. Although intrathoracic synovial sarcoma is a well described entity, most cases have been described as well-circumscribed pleural-based tumours. Pericardial involvement is very rare, and there have been no reports of transdiaphragmatic extension of this tumour. We describe a 49-year-old man who initially presented with a pericardial effusion which rapidly progressed, extending transdiaphragmatically into the abdomen, and was inoperable at the time of diagnosis. Pericardial effusions are most commonly due to infectious processes, either viral or bacterial, and the rarity of neoplasm at these sites may result in less aggressive workup, with a delay in diagnosis which may prove fatal.

CASE REPORT

A 49-year-old man presented with dyspnoea and chest pain. Initial evaluation showed a water bottle-shaped heart. Echocardiography confirmed the diagnosis of pericardial effusion. The patient was admitted for pericardiocentesis, and 400 ml of haemorrhagic fluid was drained. Cytological examination at that time did not show any atypical cells. Due to financial constraints, he was discharged, with a plan for further imaging studies on an outpatient basis. However, the patient returned only two months later, with worsening symptoms. Computed tomography showed a 13 cm × 11 cm intrapericardial heterogeneous lesion related to the left ventricle (Fig. 1), with pockets of necrosis and an

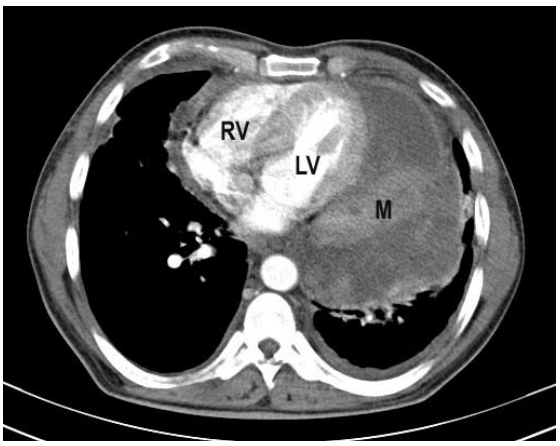

Fig. 1 Contrast-enhanced axial CT image shows a large heterogeneously-enhancing intrapericardial mass lesion (M) with areas of necrosis, and is seen in relation to the left ventricle (LV).

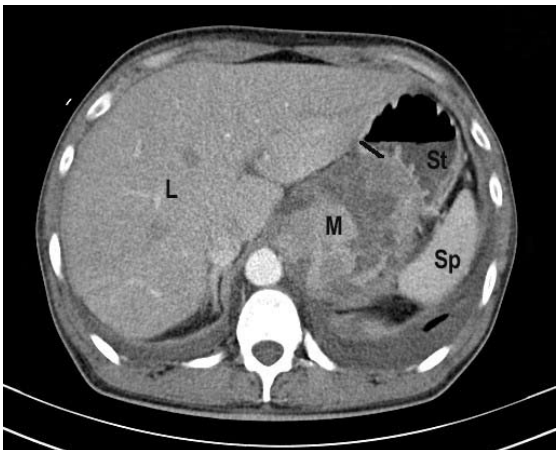

Fig. 2 Contrast-enhanced axial CT image shows intra-abdominal extension of the mass (M) through the oesophageal hiatus, between the stomach (St), liver (L) and spleen (Sp), with anterior displacement of the stomach.

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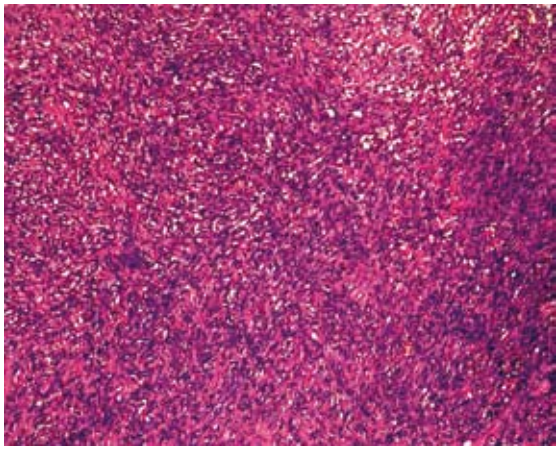

Fig. 3 Photomicrograph shows a spindle cell tumour with intervening thin-walled vascular channels (Haematoxylin & eosin, × 100).

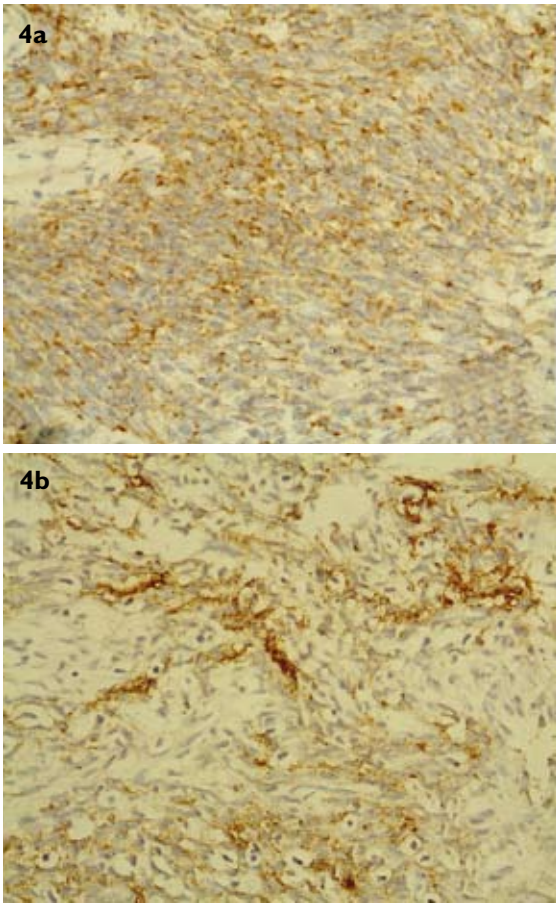

Fig. 4 Tumour cells are positive for (a) EMA and (b) Bcl-2 (Avidin-Biotin complex method, × 100).

associated loculated pericardial effusion with pericardial thickening. There was no infiltration of the myocardium. The mass extended inferiorly into the abdomen along the gastro-oesophageal junction, between the stomach and liver (Fig. 2). Small discrete omental nodes were present. There was mediastinal lymphadenopathy and mild bilateral pleural effusion. The mass did not infiltrate the pleura.

An open biopsy was performed, and at the time of surgery, the pericardium was 8–10 mm thick with minimal effusion. The heart was trapped down by a thick peel, with large amounts of exudative cheesy material in the pericardium. A frozen section was done, which showed a tumour composed of spindle cells and intervening blood vessels with a haemangiopericytoma-like pattern (Fig. 3). A preliminary diagnosis of spindle cell tumour, probably malignant, was made. Resection was not possible as the tumour extensively infiltrated the mediastinum and extended into the abdominal cavity. Immunohistochemistry was performed on representative paraffin sections (Avidin-Biotin Peroxidase method), using monoclonal mouse anti-human antibodies (DakoCytomation, Denmark). The tumour cells were positive for epithelial membrane antigen (EMA) and Bcl-2 (Fig. 4), and focally positive for cytokeratin AE1/AE3. Smooth muscle actin (SMA), S-100, CD34 and calretinin were negative. The immunoprofile ruled out other spindle cell tumours of the thorax, and a diagnosis of synovial sarcoma was made.

Following histopathological diagnosis, further examination and imaging ruled out a primary tumour in the extremities. In view of financial restraints, the patient declined palliative chemotherapy and opted for supportive care.

DISCUSSION

Synovial sarcoma is a well characterised and distinctive entity, comprising up to 5%–10% of all soft tissue sarcomas. Initially described as a biphasic tumour with both epithelial and spindle cell components and thought to arise from synovial cells,[1] it was later expanded to include monophasic tumours[2] and poorly-differentiated variants with a histogenesis from pluripotent mesenchymal stem cells showing partial epithelial differentiation. These tumours are most common in young adults, occurring in the second to fourth decades of life. Though the vast majority of cases arise in the extremities, several other sites have been documented, including the head and neck, abdominal wall, genitourinary tract, chest wall/intrathoracic, and other much rarer sites.

Begeuret et al described these tumours as being mostly large well-circumscribed masses,[3] and most of the cases studied involved only the pleura, although four tumours extended to the mediastinum and four were exclusively mediastinal. Only one of the 40 cases involved the pericardium. None of the tumours extended across the diaphragm. Essary et al, in their study of primary

pleuropulmonary synovial sarcomas, concluded that the clinical behaviour of intrathoracic synovial sarcomas was more aggressive, attributed to late presentation and inadequate surgical margins at these sites.[4]

The case described here is that of a synovial sarcoma located predominantly in the pericardium, and extending across the diaphragm into the abdominal cavity. To our knowledge, there are no other reports in the literature of a mediastinal synovial sarcoma with transdiaphragmatic extension. A primary in the abdomen extending into the thorax is also a theoretical possibility; however, since the bulk of disease was in the pericardium, it is presumed that the tumour arose from that site. Radiological studies have offered a differential diagnosis of tuberculosis and a sarcoma involving the pericardium. At surgery, the tumour was poorly delineated, and the gross picture was, in fact, not inconsistent with a tuberculous aetiology, with cheesy material within a thickened pericardium. A preliminary diagnosis was rendered at frozen section, but immunohistochemical analysis was required to rule out other spindle cell tumours before rendering a diagnosis of monophasic synovial sarcoma. The cytogenetic hallmark of synovial sarcoma is the translocation (X; 18) (p11; q11), which is present in over 90% of cases. In the absence of molecular tests for diagnosis, the use of immunohistochemistry is essential to differentiate between spindle cell tumours of the thorax.

A wide variety of clinical and histological features have been reported to be of prognostic significance. Young age (< 15 years), tumour size (< 5 cm), distal extremity location and low tumour stage have been shown to have a more favourable outcome. There are two histological subtypes of synovial sarcoma with special clinical significance. The extensively-calcified type appears to have a more indolent course and better prognosis, while the poorly differentiated tumours have a poor outcome.[5,6]

Extensive surgery with good margins is the therapy of choice. This is often difficult because of the proximity of large joints and adjacent vital structures, and hence, conservative surgery and adjunctive radiotherapy is often the favoured treatment. The reported five-, ten- and 15-year survival rates with this approach are 76%, 63% and 57%, respectively. The lower ten- and 15-year survival rates reflect the relatively high rate of late metastasis.[7] The role of adjunctive chemotherapy has not been clearly defined but phase two trials suggest a beneficial role.[7,8]

The role of palliative chemotherapy for unresectable or advanced disease is not clear, but this is common practice.[9] Our patient declined palliative chemotherapy and opted for best supportive care. Although most pericardial effusions are due to non-neoplastic causes, the possibility of a primary pericardial involvement by soft tissue sarcoma must be considered in large effusions, as an aggressive work-up will facilitate early diagnosis, thus allowing for optimal surgical removal, followed by chemotherapy.

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