

Case Report

## Primary Mediastinal Synovial Sarcoma

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### ABSTRACT

Synovial sarcoma occurs predominantly in the soft tissues of the extremities, but is exceedingly rare in the mediastinum. It has overlapping histological and immunophenotypic features with other tumours in the differential diagnosis. We report a case of a patient who had an incidental finding of such a tumour. Because of the rarity of this tumour in the mediastinum, optimal therapy is unknown and the prognosis remains guarded.

**Key Words:** mediastinal tumour, sarcoma

### INTRODUCTION

Synovial sarcoma is a malignant neoplasm predominantly affecting soft tissues of the extremities of adolescents and young adults<sup>1</sup>. It occurs in less than 10% of patients over 60 years old, in which this diagnosis is often not considered<sup>2</sup>. Its occurrence as a primary tumour in the mediastinum is rare and was first described in the medical literature in the mid 1990's<sup>3</sup>. Herein, we report a further case of a mediastinal sarcoma that required a pneumonectomy to excise completely. We utilised positron emission tomography and computed tomography (PET-CT) to aid in the staging.

### CASE HISTORY

A 59 year old male smoker presented to his local casualty department with non specific abdominal pain. During his workup, a chest X-ray was performed which revealed a large mass in the left thoracic cavity, adjacent to the mediastinum (Fig 1). Computed tomography (CT) scan revealed a 12.4cm mass within the left upper zone of the chest. It was closely related to the brachio-cephalic vein and aortic arch, but there was a clear plane of cleavage. However as the mass progressed inferiorly, it appeared to be in close contact with the main pulmonary artery and the left pulmonary artery.

A flexible bronchoscopy was subsequently performed which demonstrated extrinsic compression of the left upper lobe bronchus. However, bronchial washings were negative for malignant cells. To gain a tissue diagnosis, we performed a left anterior thoracotomy which also allowed us to examine the mediastinum for resectability. This revealed the tumour to be free from the visible lung but adherent to the mediastinum. Histopathological analysis found it to be a poorly differentiated sarcoma. PET-CT scan was performed to exclude any metastatic disease. This demonstrated an 18F-Fluorodeoxyglucose (18F-FDG) avid mass with a standardised uptake value (SUV) of 16 with no other foci that may represent metastatic disease (Fig 2).

At thoracotomy, the mass was seen arising from the left hilum, adherent to the left pulmonary artery, left superior pulmonary vein, phrenic nerve and adjacent pericardium and required an intrapericardial pneumonectomy and resection of the latter two structures to achieve complete resection. Histopathological analysis revealed a biphasic synovial sarcoma. The primary site for this tumour was uncertain as the tumour was located in the lateral portion of the mediastinum, close to both the pleura and pericardium. The patient made an uneventful recovery and at 18 months post-operative follow-up was disease free and his quality of life had returned to normal.



Fig 1. Chest radiograph demonstrating a mass abutting the mediastinum within the left hemithorax.

### PATHOLOGY

The tumour measured 13 x 9 x 6cm and was adherent to the hilar surface of the left lung. Sectioning revealed a uniform consistency and it was sharply demarcated from the lung parenchyma. Histologically the tumour was composed of spindle cells and epithelioid cells (Fig 3) although the spindle cells were predominant. Numerous mitotic figures (more than 4 per 10 high power field) were seen and there were focal areas of necrosis. Immunohistochemistry demonstrated strong positivity for Vimentin, Cytokeratins, and focal positivity for EMA. Mesothelial markers (CK5/6, Calretinin) and

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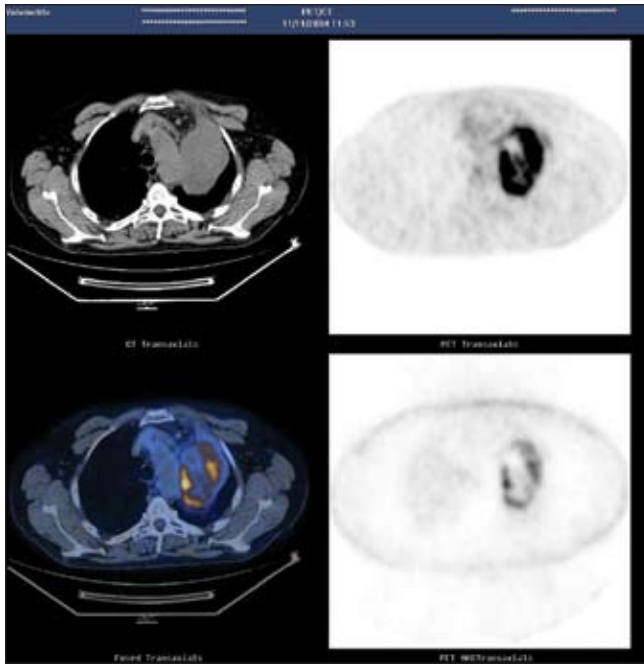


Fig 2. Transaxial image showing tumour with plane of cleavage from arch of aorta. Small volume of pre-vascular/superior mediastinal nodes on CT are not FDG avid.

primary pulmonary tumour marker (TTF-1) were negative. These findings would be consistent with a biphasic synovial sarcoma.

## DISCUSSION

The mediastinum is host to a vast array of both primary and metastatic neoplasms, the differential diagnosis of which is extensive and depends upon the mediastinal compartment involved, clinical history, presentation, and age of the patient. The case illustrated demonstrates the diagnostic difficulty encountered when the patient's age and tumour location are taken into account, as synovial sarcomas are rare mesenchymal neoplasms that primarily affect the deep soft tissues of the extremities and predominates in adolescents and young adults<sup>1</sup>. Although synovial sarcoma has been reported to metastasise to the mediastinum, its occurrence as a primary neoplasm in this location is rare and has only recently been recognised<sup>3</sup>. Patients have presented with dyspnoea<sup>4</sup>, chest pain<sup>1</sup>, cardiac tamponade<sup>5</sup> and as an incidental finding<sup>1</sup>.

Classically, synovial sarcoma is a biphasic tumour composed of spindle shaped fibroblast-like cells and epithelioid cells, but the majority are actually monophasic spindle cell in type. A poorly differentiated variant composed of small round cells has also been described.

The differential diagnosis for thoracic synovial sarcomas includes mesothelioma, fibrosarcoma, malignant peripheral nerve sheath tumour, smooth muscle tumours, thymoma, blastoma, and sarcomatoid carcinoma<sup>6</sup>. However, the diagnosis can be difficult to achieve in an unusual location, overlapping histological and immunohistological features, or if a small sample size is present<sup>6</sup>. Immunohistochemistry can be very helpful. Synovial sarcomas are positive for Vimentin, EMA, Cytokeratins, CD99 (~60%) and S100 (~30%)<sup>7</sup>. The

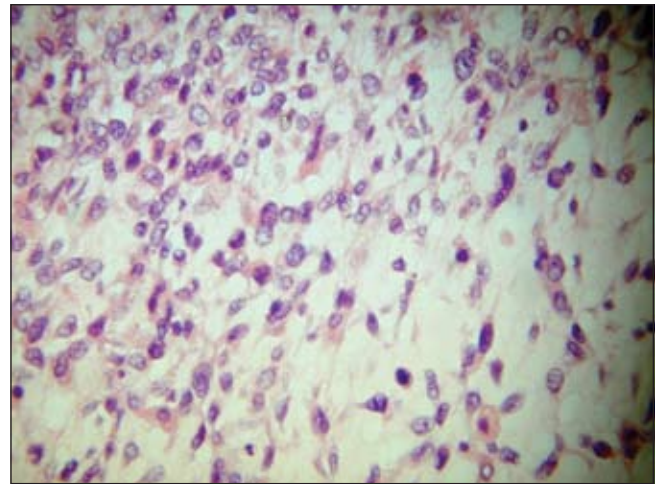


Fig 3. Histology demonstrating spindle and epithelioid cells.

availability of molecular genetic identification (fluorescent in situ hybridization or polymerase chain reaction) of the t(X;18) has improved diagnostic specificity as this translocation is found in over 90% of synovial sarcomas<sup>1</sup>. This translocation involves the SYT gene on chromosome 18 and either the SSX1 or SSX2 gene on the X chromosome<sup>1</sup>.

Accurate staging of the disease is important for appropriate patient management and requires the evaluation of the primary tumour and assessment for distant disease. Magnetic resonance imaging (MRI) has been the traditional imaging tool for assessment of soft tissue masses, but more recently soft tissue sarcomas are well recognised as showing increased 18F-FDG uptake in Positron Emission Tomography (PET)<sup>8</sup>.

PET-CT combines the strengths of two imaging modalities, exploiting the recognised greater sensitivity of CT in demonstration of small volume lung metastases (sensitivity, specificity 100%, 96.4% vs. 86.7%, 100%) and enhancing 18F-FDG PET imaging by allowing precise anatomical localisation of functional abnormalities<sup>8</sup>. MRI frequently struggles to differentiate the inflammatory and fibrotic changes which occur following surgery and adjuvant therapy from recurrent disease, whilst 18F-FDG PET allows accurate differentiation of locally recurrent disease from healing/post-surgical change<sup>8</sup>. Combined PET-CT imaging is likely to become the imaging modality of choice for follow-up of patients with these rare tumours.

Broad surgical resection is the cornerstone of therapy. Complete resection of the tumour was the overwhelming factor in determining survival in a review of primary mediastinal sarcomas<sup>9</sup>. Radiotherapy is recommended with positive margins<sup>10</sup>. The place for chemotherapy of this tumour is not well defined, and in none of the other case reports was preoperative chemotherapy given<sup>5</sup>. Combination of adriamycin and ifosfamide has been used as an adjuvant therapy and for recurrences<sup>5</sup>. Prognosis used to be poor with survival lasting two months<sup>4</sup>, but with aggressive multimodal therapy, survival up to 14 years has been documented<sup>5</sup>.

In summary, though mediastinal sarcoma is a rare entity, if aggressively treated with complete resection and or multimodal therapy with intensive follow-up, the survival of such patients can improve.

The authors have no conflict of interest.

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