

Synovial Sarcoma of the Heart

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ABSTRACT

A young male presented with dyspnoea and was found to have a diastolic murmur at the left lower sternal edge. Transthoracic and transoesophageal echocardiograms found a large mass attached to the tricuspid valve, which was projecting into the right atrium and the right ventricle. The mass was causing significant obstruction of the right ventricular inflow. Emergency surgery had to be performed because the patient developed severe vomiting (of unknown cause) leading to haemodynamic compromise. This condition was not responding to fluid resuscitation as there was obstruction to right ventricular inflow. Intraoperatively the mass was removed and the tricuspid valve was repaired. The histological and immunohistochemical examination of the excised tissue confirmed the rare diagnosis of synovial sarcoma of the heart. Postsurgical imaging showed no metastases. The patient received postoperative radio therapy and chemotherapy. The tumour recurred after 6 months and the patient succumbed to his illness and expired.

Key words: *Synovial sarcoma. Heart. Tricuspid valve mass. Right ventricular outflow tract obstruction. Dyspnoea. Diastolic murmur.*

INTRODUCTION

Synovial sarcoma is an extremely rare malignant tumour of the heart and only a handful of cases have been reported in literature. There appears to be no genetic association, although immunohistochemical and molecular markers for its diagnosis are available. The clinical presentation of the tumour is no different from the other primary malignant tumours of the heart. Treatment modalities including surgery, chemotherapy and radiotherapy have been tried but the overall mortality outlook has not changed.

We present a case of this rare primary cardiac tumour that recurred after surgical resection and chemotherapy.

CASE REPORT

A 20 years old male, a resident of Gilgit presented to this hospital with a 2 months history of lethargy and progressively increasing dyspnoea. The patient began to feel un-well 2 months ago with insidious onset of lethargy, progressively increasing dyspnoea (NYHA class III at the time of presentation). Systemic enquiry was unremarkable for chest pain, palpitations, syncope, seizures, ankle swelling, and abdominal distention. Clinical examination was remarkable only for a diastolic murmur best heard at left lower sternal border without

any postural changes. The patient was haemodynamically stable. The jugular venous pressure was not raised and there were no signs of heart failure.

The ECG is shown in Figure 1. The transthoracic echocardiogram showed a large multi-lobular mass occupying most of the right atrial cavity, arising from the posterior leaflet of the tricuspid valve, and projecting into the right ventricle. Colour Doppler examination revealed minimal flow across the tricuspid valve (Figure 2). These findings were confirmed by a transoesophageal echocardiogram. A working diagnosis of cardiac tumour, possibly a myxoma, was made. Further evaluation with a cardiac CT was planned. However, urgent cardiac surgical consultation had to be called in since the patient's blood pressure had begun to drop because of unexplained vomiting. He was responding poorly to antiemetics and fluid replacement. Based on the hypothesis that the obstruction to the right ventricular inflow in the face of reduced intravascular volume (secondary to vomiting) was the cause of collapse; emergency cardiac surgery was carried out. Operative findings described a large mass in the right atrium arising from the posterior leaflet of tricuspid valve. The main body of the mass was in the atrium with a small portion projecting into the right ventricle. The tumour was excised along with a part of the posterior tricuspid valve leaflet, which was repaired. Grossly, the specimen consisted of a 7 x 5 x 5 cm fibrofatty mass. External surface was irregular. The cut surface was homogenous and light yellow in colour. The tumour was sent for histopathological examination and a diagnosis of synovial sarcoma of the heart was made (Figure 3). The immunohistochemical staining was focally positive for cytokeratin, EMA (epithelial membrane antigen), BCL-2 and CD-56.

The patient was referred to the Oncology Department for onwards management. Because of the surgery that was

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Received February 08, 2012; accepted May 10, 2012.

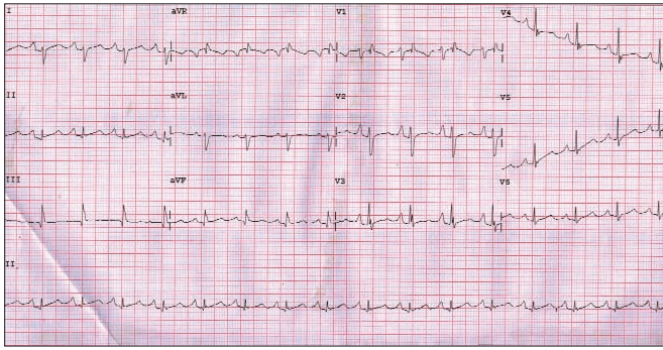


Figure 1: ECG showing right axis deviation.

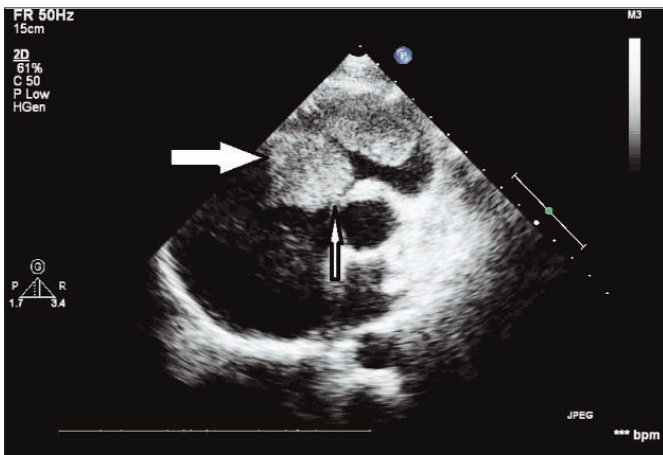


Figure 2: Transthoracic echocardiogram showing large bilobed mass (broad arrow) attached to the tricuspid valve (narrow vertical arrow).

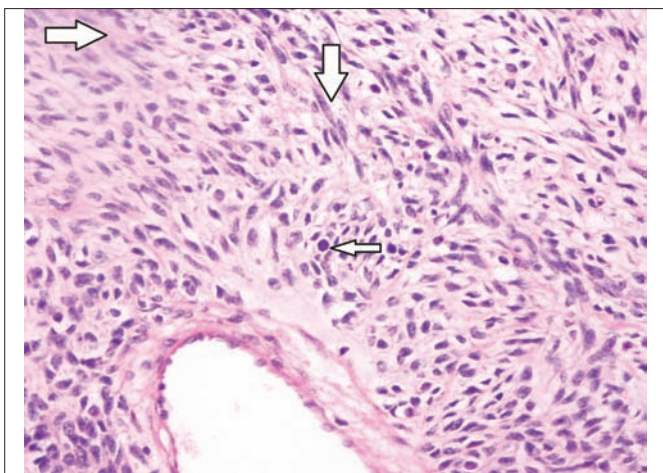


Figure 3: Histopathology haematoxylin and eosin stain of the tumour showing the clusters of spindle shaped cells (vertical arrow), mitotic figures (left arrow) on a mesenchymal background (right arrow).

urgently performed the screening for metastatic deposits was done after surgery. CT scan of the thorax, abdomen and bone scan revealed no metastatic disease. Post-operative echocardiography revealed complete resection of the tumour and moderate tricuspid regurgitation. He received postoperative radiotherapy and chemotherapy with a regimen including doxorubicin. The patient was discharged home and followed-up in the

Oncology Clinic. Six months later, the patient began to lose weight rapidly accompanied by NYHA class III dyspnoea. Transthoracic echocardiogram performed at that time revealed local recurrence of tumour at the original site as well as large pericardial metastatic deposits. Palliative radiotherapy and chemotherapy was started but the patient succumbed to his illness and died 10 days later.

DISCUSSION

Primary cardiac tumours are extremely rare and occur less commonly than metastatic disease of the heart. Distinction is made based on history, clinical examination, imaging findings and histological examination of the resected specimen. The different malignant tumours of the heart include angiosarcomas (most common), rhabdomyo-sarcoma, mesothelioma, fibrosarcoma, malignant schwannoma, synovial sarcoma. Attributable to their rarity, the threshold for their clinical diagnosis is high. Clinically these tumours can present as mimics of many cardiac disorders such as heart failure, ischaemic heart disease, valvular heart disease, arrhythmias, and embolic events. Local extension of the tumours causes signs and symptoms such as superior vena cava syndrome, haemoptysis and dysphonia. Transthoracic and transoesophageal echocardiography are both first line investigations for the evaluation of cardiac tumours. Chest X-ray may show cardiomegaly, features suggestive of congestive cardiac failure, pleural effusion, lung nodules, cardiac mass or left hemi-diaphragm paralysis. Electrocardiogram may show non-specific changes such as conduction block right ventricular hypertrophy, atrial fibrillation, paroxysmal atrial tachycardia etc. Search for metastasis is mandatory, as upto 80% patients may have systemic metastases at diagnosis, most commonly in the lung. The CT and MRI scans of chest and abdomen further complement echocardiography by showing the extra cardiac tumour spread and presence of metastases in addition to the anatomical details of the lesion. Cytology of fine needle aspiration and pericardial biopsies for pericardial infiltration have not shown very accurate results. Transvenous endomyocardial biopsy is helpful to yield a histological confirmation before operation.

Primary cardiac synovial sarcoma is a rare malignancy, comprising approximately 5% of cardiac sarcomas and less than 0.1% of all primary cardiac tumours.¹ It is more commonly seen in the limbs rather than the heart. Tumours of the limbs metastasizing to the heart have been described.¹ The tumour affects males more commonly, and is found more commonly in the right atrium.² Cases of left atrial and left ventricular¹ synovial sarcoma have been described. Clinically, the tumour may present with any of the manifestations mentioned above.

The tumour defies early detection because of its aggressive behaviour. Histologic sections are essential for the diagnosis of synovial sarcomas, however, FNA biopsy has been used to make the diagnosis; the biopsy sample shows cytomorphologic appearance typical of the tumour; usually a high grade tumour showing an intimate admixture of spindle and epithelial cells. An important tool for the confirmation of the diagnosis is the detection of the chimeric transcript SYT-SSX using reverse transcriptase-polymerase chain reaction (RT-PCR). This transcript is generated by reciprocal translocation between chromosomes X and 18, and is specific to synovial sarcoma that usually occurs in the extremities of young adults. The translocation between X and 18 is a characteristic one in synovial sarcoma of the lower extremities.³ In cases, where the pathological and immunohistochemical diagnosis of synovial sarcoma is challenging, this molecular biological technique using RT-PCR becomes a powerful method of confirmation of the diagnosis.⁴ Complex karyotype analysis using COBRA-FISH (Combined Binary Ratio labelling Fluorescence *in situ* hybridization) may demonstrate the derivative chromosomes with multiplex arrangements.⁵

Complete surgical excision is the only form of treatment. The prognosis in patients with malignant tumours remains unchanged despite surgical resection.⁶ The role of heart transplantation in patients with malignant tumours remains unclear.⁷ Postoperative doxorubicin did not produce satisfactory results.⁸ Cardiac auto-transplantation (cardiac explantation, *ex vivo* tumour resection with cardiac reconstruction, and cardiac reimplantation) has been described as a feasible modality of treatment with a reasonable operative morbidity and mortality in one study.⁹

As with other sarcomas, in synovial sarcoma wide surgical resection remains the linchpin of therapy.

Adjuvant radiation therapy to prevent local recurrence and chemotherapy for control of systemic disease may have some beneficial effect on overall survival, but the benefit is likely limited.¹⁰

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