CASE REPORT

Right atrial spindle cell sarcoma as a rare cause of tricuspid stenosis

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SUMMARY

Spindle cell sarcomas of the right atrium are extremely rare primary cardiac tumours with very few cases reported in the medical literature. The prognosis of this malignant neoplasm remains poor due to delay in diagnosis, early metastasis and few available therapeutic options. It responds poorly to chemotherapy and radiotherapy. Surgical excision is the mainstay of treatment. We report a 42-year-old man, who presented to emergency room with dyspnoea and fatigue (New York Heart Association classification II) since 1 month. Transthoracic echocardiogram showed massive pericardial effusion with tamponade. The patient underwent emergency pericardiocentesis; postpericardiocentesis echocardiogram showed large right atrial mass involving the right ventricle and the tricuspid valve causing tricuspid stenosis. Findings were confirmed by cardiac CT scan. Percutaneous transvenous biopsy was carried out which revealed spindle cell sarcoma.

BACKGROUND

Cardiac tumours may be either primary or secondary. Primary cardiac tumours are rare with reported incidence of 0.001–0.03% while secondary tumours are far more common (100–1000 times).¹ Among the secondary cardiac tumours, the most common primary site is lungs.² Majority of primary cardiac tumours are benign (75%). Myxomas account for around 50% of all primary benign tumours and others like rhabdomyoma, lipoma and papillary fibroelastoma constitute 10% each.³ About 25% of primary cardiac tumours are malignant of which sarcomas are most common entities, while lymphomas are rare (5%).¹ Among cardiac sarcomas, angiosarcoma (about 40%) is the most common form while others include undifferentiated sarcoma (24%), malignant fibrous histiocytoma (14–24%), leiomyosarcoma (8%) and osteosarcoma (6%), while the least reported malignant cardiac tumour is spindle cell sarcoma.^{1 4} Many sarcomas are misdiagnosed as myxomas because of their similar clinical presentation. However, both these types of cardiac tumours behave quite distinctly. Myxomas can be resected completely and therefore have an excellent prognosis while sarcomas have a poor prognosis, in spite of surgical resection.^{5 6} Thus accurate diagnosis is extremely important for deciding the plan of management and prognosis.

CASE PRESENTATION

A 42-year-old man presented with history of dyspnoea and fatigue (New York Heart Association classification II) since 1 month. There were no associated symptoms of angina, syncope or cough. Symptoms were not related to posture. There were no constitutional symptoms like fever, loss of appetite or weight loss.

Clinical examination revealed a pulse rate of 102 bpm, low volume and regular with pulsus paradoxus. His blood pressure was 104/68 mm Hg. Jugular venous pressure (JVP) was elevated with absent 'y' descent. Cardiovascular auscultation revealed muffled heart sounds without any additional sounds or murmurs. Respiratory system, abdominal and neurological examinations were unremarkable.

INVESTIGATIONS

Erythrocyte sedimentation rate was elevated (63 mm/hour). All other haematological and serological investigations were within normal limits. An ECG showed sinus tachycardia. Transthoracic echocardiogram (TTE) showed massive pericardial effusion with tamponade. Emergency pericardiocentesis was carried out and 500 mL of haemorrhagic pericardial fluid was withdrawn. The patient was stabilised haemodynamically after pericardiocentesis, but raised JVP with absent 'y' descent persisted. On analysis, the pericardial fluid was exudative with predominant lymphocytes and was negative for malignant cells. Adenosine deaminase levels were within normal range. Repeat TTE revealed a large mass in right atrium (RA) with minimal pericardial effusion ([figure 1](#), [videos 1](#) and [2](#)). Transesophageal echocardiogram (TEE) showed heterogeneous large solitary mass attached

Figure 1 Transthoracic echocardiogram showing large mass in right atrium. BPM, beats per minute; LA, left atrial LV, left ventricle; RA, right atrial; RV, right ventricle.

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to RA free wall with frond-like projections. It was occupying nearly the entire RA and extending to right ventricle involving the tricuspid valve and causing tricuspid stenosis with gradient of 17/8 mm Hg (figure 2, videos 2–5). Cardiac CT scan

corroborated the TEE findings and also revealed multiple foci of metastasis in both the lungs (figure 3).

Percutaneous transvenous biopsy of the RA mass was carried out in the catheterisation laboratory under fluoroscopy and TEE

Video 1 Apical four-chamber view (transthoracic echocardiogram) showing a large mass in the right atrium.

Video 3 Transesophageal echocardiogram at 0° showing large heterogeneous mass in the right atrium involving the right ventricle through the tricuspid valve.

Video 2 Colour flow Doppler showing turbulence across the tricuspid valve.

Video 4 Bicaval transesophageal view showing a right atrial mass with frond-like projections protruding through the tricuspid valve.

Figure 2 Continuous wave Doppler across tricuspid valve showing tricuspid stenosis with peak gradient of 17.3 mm Hg.

Video 5 Bicaval transesophageal view showing a huge right atrial mass occupying almost the whole of right atrium and involving the superior vena cava–right atrial junction.

guidance from right femoral vein approach and adequate tissue sample was obtained using a biptome. Histopathological examination showed myocardial infiltration with bundles of spindle cells separated by myxoid stroma (figure 4A). The tumour cells had large, pleomorphic nuclei with prominent nucleoli and

frequent abnormal mitotic figures were identified (figure 4B); all these features supported the diagnosis of high-grade spindle cell sarcoma.

OUTCOME AND FOLLOW-UP

Surgical excision was planned, but patient refused for surgery. On fifth day after discharge, patient died of sudden cardiac arrest.

DISCUSSION

The earliest report of tumour in the heart dates back to mid-16th century, by Columbus.⁷ Primary cardiac angiosarcoma is the most common primary cardiac sarcoma in adults and rhabdomyosarcoma in children. Cardiac spindle cell sarcomas are classified as undifferentiated sarcomas. Spindle cell sarcomas are commonly encountered in large arterial blood vessels and extremely rare in the heart.

Depending on the cardiac chamber involved, sarcomas can present with intermittent or positional symptoms, dyspnoea, cough, chest discomfort, orthopnoea or syncope. They may present with heart failure, pericardial effusion with/without tamponade, arrhythmias, valvular abnormalities or obstructive symptoms. Brain is the most common site of embolism, sometimes pulmonary embolism can be the presenting feature.⁶ As compared to myxomas, only 10% of cardiac sarcomas have non-specific constitutional symptoms.

Figure 3 CT axial section showing large heterogeneous mass occupying whole of right atrium (horizontal arrow).

Figure 4 Histopathological examination of right atrial mass biopsy showing myocardial infiltration with bundles of spindle cells separated by myxoid stroma (A) and tumour cells with large, pleomorphic nuclei and prominent nucleoli (B).

The diagnosis is usually established using non-invasive imaging modalities such as TTE or TEE. TEE is the preferred modality as it provides a better resolution of cardiac structures. CT scan and MRI are used for complete assessment of the tumour in the form of extracardiac extent and presence of metastases, myocardial infiltration and compression of the cardiac chambers along with pericardial and great vessel involvement.⁸ Features such as broad-based lesion, infiltration of myocardium, pericardium or adjacent structures, lesions occupying entire cardiac chamber, metastasis and necrosis are suggestive of the malignant potential of the tumour.

Definitive diagnosis is made by tissue biopsy, which may be obtained through percutaneous biopsy, endoscopy, mediastinoscopy or thoracotomy. Cardiac spindle cell sarcomas are poorly differentiated cells of mesenchymal origin, consisting of atypical spindle cells with variable degrees of atypia, mitotic activity, necrosis and nuclear polymorphism commonly affecting the large blood vessels. They lack a specific histology, with negative immunohistochemistry for desmin, factor VIII and keratin. They

are extremely rare in heart, with only few cases reported so far.^{9 10} Prognosis depends on the grade of the tumour, which is based on the mitotic activity, necrosis and degree of cellular differentiation. Individual subtypes of cardiac sarcomas do not appear to affect the overall clinical outcome.

Treatment of cardiac sarcomas remains challenging due to their rare incidence and aggressive nature. Cardiac sarcomas have poor prognosis with a mean survival of <12 months. In rare cases, survival up to 11 years has been reported.¹⁰ Neoadjuvant therapy has a limited role. Surgical resection with negative margins forms the mainstay of treatment.

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Learning points

- ▶ Spindle cell sarcoma is one of the rare primary cardiac malignancies.
- ▶ Its diagnosis can be challenging owing to non-specific and varied presentations.
- ▶ Transthoracic echocardiogram and transesophageal echocardiogram are extremely useful for prompt provisional diagnosis, while definitive diagnosis is made by tissue biopsy.
- ▶ CT scan and MRI help in delineating intracardiac as well as extracardiac extensions and metastasis to other organs.
- ▶ Involvement of the tricuspid valve by the spindle cell tumour can be a rare cause of tricuspid stenosis.
- ▶ Owing to aggressive nature of the tumour, high index of suspicion is vital for prompt diagnosis and proper management of such patients.

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