A Rare Primary Cardiac Malignancy: Spindle Cell Sarcoma

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ABSTRACT

Spindle cell sarcoma (SCS) is the rarest primary cardiac tumour, with only a few cases reported in the literature so far. We hereby report a case of left atrial SCS diagnosed and managed by a multidisciplinary approach. A 50-year male presented with symptoms of shortness of breath with high-grade fever. Echocardiography showed an atrial mass with pericardial effusion. Contrast-enhanced CT scan, further validated by biopsy, showed it to be malignant SCS. These tumours are often missed and presented late with advanced disease due to the non-specific symptoms and extreme rarity. Better understanding of the disease and early diagnosis are essential to its management.

Key Words: *Primary cardiac malignancy, Spindle cell sarcoma, Left atrium.*

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INTRODUCTION

Cardiac tumours are either primary or secondary, with primary tumours being rare having 0.017- 0.019 as incidence range.[1] Secondary tumours are 100 to 1,000 times more common with the majority being of pulmonary origin.[2] Amongst the primary cardiac tumours, 75% are benign with the myxomas accounting for about 50%, lipomas for 10%, papillary fibroelastomas for 10%, and rhabdomyomas for 10%.[1] About 25% are malignant tumours. Sarcomas are the commonest malignant tumours (about 95%), with a high percentage of angiosarcomas (37%) followed by undifferentiated sarcoma in about 24%, malignant fibrous histiocytomas in 11-24%, leiomyosarcomas in 8-9%, and osteosarcoma in 3-9% cases.[1] The rarest cardiac tumours are spindle cell sarcomas (SCSs). In children, malignant cardiac tumours are <10% of the tumours with rhabdomyosarcoma being the most common. There is often a delay in the diagnosis due to no symptoms in early disease or non-specificity of the symptoms. Hence, the majority of these tumours are present at the advanced stages. Symptoms are usually wide-ranging from both pulmonary and cardiac symptoms including dyspnoea, chest discomfort, fever, arrhythmias, and changes of fluid overload. In the advanced stages of disease, patients may present with cardiac arrest and multisystem failure.

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With advanced imaging techniques including contrast-enhanced ultrasound imaging, echocardiography (transesophageal echocardiogram being more sensitive than transthoracic echo), computed tomography (CT) scans, and cardiac magnetic resonance (CMR), primary cardiac tumours are being diagnosed at an early stage at a potentially treatable stage of the disease. We hereby present a rare case of primary cardiac tumour which went undiagnosed initially due to non-specificity of the symptoms and presented at an advanced stage.

CASE REPORT

A 50-year male presented with complaints of shortness of breath and high-grade fever. The patient was initially diagnosed as a case of atrial myxoma with pericardial and pleural effusion based on an echocardiography performed at an outside facility. He also underwent pericardiocentesis for symptomatic relief and received a 4-month course of antituberculous medication a few months earlier.

On admission, the patient initially underwent echocardiography which showed a large mass in the right atrium arising from the interatrial septum compressing the inferior-posterior left ventricle wall. Ejection fraction was 50%. CT chest without contrast reported a predominantly cystic mass with internal hyperdense areas, inferior and posterior to the heart, likely within the pericardial cavity, exerting mass-effect on the left heart chambers. Considering imaging appearances, differential possibilities of primary cardiac tumour or cardiac pseudoaneurysm were given. Another consideration was chronic haematoma in the setting of the prior pericardiocentesis. However, as the study was non-contrast-enhanced, the evaluation was limited. Furthermore, diffuse-body wall oedema and pleural effusions were suggestive of fluid overload. Enlarged epicardial, lower cervical, supraclavicular, axillary, and mediastinal lymph nodes were also found.

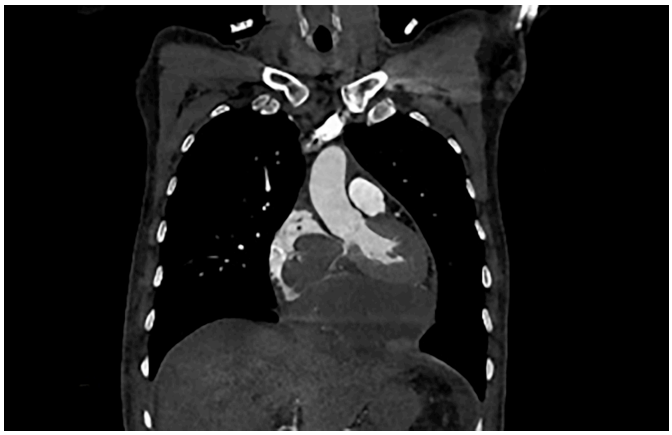

Figure 1: Coronal reformatted image of the large lesion in the arterial phase.

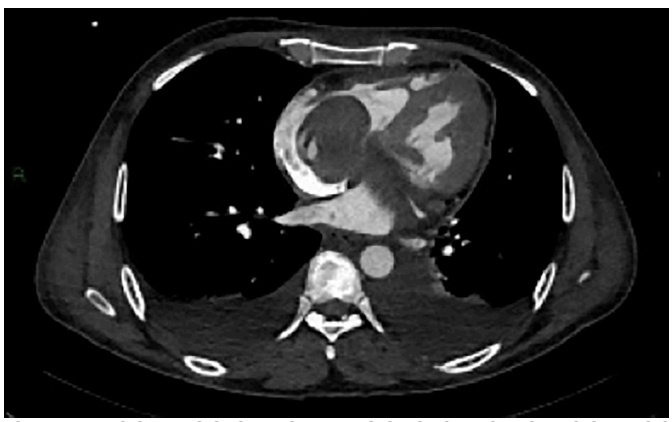

Figure 2: Axial arterial phase image of the lesion showing right atrial extension.

A follow-up contrast-enhanced CT chest, abdomen, and pelvis, performed a couple of days later, stated a large solid-cum-cystic lesion with heterogeneous enhancement having both intra- and extra-myocardiac components (Figure 1). Extra-cardiac bulk was postero-inferior to the left atrium causing a significant mass-effect on the left heart. The lesion was extending through the inter-atrial septum into the right atrium with a wide attachment of about 19 mm with the inter-atrial septum (Figure 2). It was seen bulging into the right atrioventricular groove with an opened mitral valve. Along with the enlarged local lymph nodes noted on the previous scan, prominent abdominal lymph nodes were also seen. Considering the imaging findings, a diagnosis of the cardiac angiosarcoma / undifferentiated sarcoma was given with lymphoma as a less probable differential diagnosis.

There was reflux of contrast into the inferior vena cava (IVC) and hepatic veins as well as significant dilation, suggesting circulatory compromise by the tumour. Diffuse hepatic congestion, nutmeg appearance of the liver, changes of cholecystopathy, diffuse omento-mesenteric congestion, and mild abdomino-pelvic ascites were ancillary findings.

The patient then underwent an endomyocardial biopsy of the lesion by a cardiac surgeon. Microscopic findings were cellular tumour comprising spindle-shaped cells with elongated nuclei. Blood vessels were seen between these cells. There were frequent mitoses and moderate pleomorphism. Immunohistochemistry was positive for TLE1, focal positive for ERG, CKAE1/AE3 and negative for *CD34*, S-100, Desmin, SOX 10, and ASMA. So, the biopsy report concluded the lesion to be SCS. The patient was unfortunately lost to treatment. After two months, the patient died of disease progression.

DISCUSSION

The discovery of the first cardiac tumour dates back to 1562 by Columbus.[3] The first primary cardiac tumour in a living patient was diagnosed by Barnes in 1936 by using electrocardiogram and biopsy data from a metastatic peripheral lesion.[3] Cardiac primary SCSs are extremely aggressive, infiltrating and involve all the layers of the heart and metastasise rapidly, with a mean survival of three months to one year.[4] Macroscopically, cardiac intimal sarcomas appear as polypoidal masses attached to the inner vessel surface, resembling thrombi and sometimes extending distally. They have mesenchymal origin with myofibroblastic and fibroblastic differentiation, predominantly found in large arterial blood vessels and very rare in the heart, with only a few cases reported in the literature so far. The pulmonary artery is more commonly involved than the aorta (165 *versus* 100 reported patients), more commonly affecting the pulmonary trunk (80%), main pulmonary arteries (50–70%), or both (40%).[5,6] Tumours involving the heart are more common in the left atrium. Metastasis to the lungs is observed in 40% of the cases and metastasis to extra-thoracic structures including the kidneys, brain, skin, and lymph nodes is observed in up to 20%. Distant metastasis by tumour emboli is common involving the peritoneum, bone, mesenteric lymph nodes, and the liver. The location of the tumour varies with the tumour type with angiosarcoma involving the pericardium and right atrium, malignant fibrous histiocytomas (MFH) showing a predilection for the left heart and rhabdomyosarcoma favouring valves. Primary osteogenic cardiac sarcoma, fibrosarcoma, and leiomyosarcoma are mostly seen in the left atrium.[7]

SCSs often show positive immunoreactivity for osteopontin, MDM2, and vimentin. Alpha smooth muscle actin, *CD68, CD117, p53,* and *bcl-2* show variable degrees of positivity. *CD34*, *CD31*, and factor VIII are typically negative. In the present case, the spindle cells had a diffuse expression of TLE-1, focal positivity for ERG, and CKAE1/AE3, and were immuno-negative for desmin, *CD34*, S100, SOX10, and ASMA.

Depending on the location, infiltration, and compromise of cardiac output, tumours manifest in various forms. Some nonspecific symptoms include dyspnoea, chest discomfort, heart failure, syncope, pericardial effusion, and tamponade. With valve involvement, they mimic valvular abnormalities. Neural pathway involvement leads to conduction abnormalities including arrhythmias. If detached, the tumour can manifest as a stroke. Lymphomas can present with an increase in temperature, weight loss, cough, exhaustion, and leukocytosis.

With advancements in imaging modalities, diagnosing cardiac tumours has been revolutionised. Echocardiography is inexpensive, non-invasive, and provides better resolution in diagnosing cardiac lesions with transesophageal echocardiography being a superior modality to transthoracic echocardiography. Although a chest x-ray has no significant diagnostic value, it can help in identifying ancillary findings such as, pulmonary infiltrates, pleural effusion, cardiomegaly, or cardiac mass. CT and MRI scans help in characterising soft tissue lesions, extracardiac involvement, metastasis, and the involvement of great vessels. Tumour volume, burden, mediastinal involvement, and treatment response is better evaluated with cardiac MR. 18F-fluorodeoxyglucose positron-emission tomography / computed tomography (FDG-PET/CT) has a sensitivity of about 90% in categorising benign and malignant tumours. CT coronary angiography can diagnose the involvement of coronary arteries.

Complete extensive surgical excision increases postoperative survival, but it is possible in less than 50% of cases.[8] Postoperative therapy increases survival after surgical excision of cardiac sarcoma; however, there is no significant difference in 5-year survival rates compared to surgery alone.[9] Survival rates according to different locations of tumour were studied by Ramlawi *et al*.[10] Overall, the median survival time was 20 months and the mean of 33.4 months.[10] Palliative surgery is effective in providing diagnostic samples and some relief from the symptoms. For inoperable tumours with no evidence of metastasis, cardiac transplantation is considered.

SCSs are often missed and presented late with advanced disease due to non-specific symptoms, aggressive nature, and extreme rarity. In this aspect, modern investigation methods including contrast-enhanced echocardiography and contrast MRI are of great importance. Surgical excision is the mainstay of the treatment with limited value of adjuvant therapy. A better understanding of the disease and early diagnosis is essential for its management.

PATIENT'S CONSENT:

Informed consent was obtained from the patient's attendants for the publication of this case report.

COMPETING INTEREST:

The authors declared no conflict of interest.

AUTHORS' CONTRIBUTION:

NK: Concept, literature review, and manuscript writing.
MS: Concept, literature review, and critical review.
SA: Analysis and interpretation of the data and critical review.
MA: Critical review and literature review.
All authors approved the final version of the manuscript to be published.

REFERENCES

1. Butany J, Nair V, Naseemuddin A, Nair GM, Catton C, Yau T. Cardiac tumours: Diagnosis and management. *Lancet Oncol* 2005; **6(4)**:219-28. doi: 10.1016/S1470-2045(05)70093-0.
2. Ekmektzoglou KA, Samelis GF, Xanthos T. Heart and tumors: Location, metastasis, clinical manifestations, diagnostic approaches and therapeutic considerations. *J Cardiovasc Med (Hagerstown)* 2008; **9(8)**:769-77. doi: 10.2459/JCM.0b013e3282f88e49.
3. Perchinsky MJ, Lichtenstein SV, Tyers GF. Primary cardiac tumors: Forty years' experience with 71 patients. *Cancer* 1997; **79(9)**:1809-15. doi: 10.1002/(sici)1097-0142(19970501)79:9<1809::aid-cncr25>3.0.co;2-0.
4. Ibrahim A, Luk A, Singhal P, Wan B, Zavodni A, Cusimano RJ, *et al*. Primary intimal (spindle cell) sarcoma of the heart: A case report and review of the literature. *Case Rep Med* 2013; **2013**:461815. doi: 10.1155/2013/461815.
5. Bode-Lesniewska B, Zhao J, Speel EJ, Biraima AM, Turina M, Komminoth P, *et al*. Gains of 12q13-14 and overexpression of mdm2 are frequent findings in intimal sarcomas of the pulmonary artery. *Virchows Arch* 2001; **438(1)**:57-65. doi: 10.1007/s004280000313.
6. Burke AP, Virmani R. Sarcomas of the great vessels. A clinicopathologic study. *Cancer* 1993; **71(5)**:1761-73. doi: 10.1002/1097-0142(19930301)71:5<1761::aid-cncr2820710510>3.0.co;2-7.
7. Mayer F, Aebert H, Rudert M, Konigsrainer A, Horger M, Kanz L, *et al*. Primary malignant sarcomas of the heart and great vessels in adult patients - A single-center experience. *Oncologist* 2007; **12(9)**:1134-42. doi: 10.1634/the oncologist.12-9-1134.
8. Shanmugam G. Primary cardiac sarcoma. *Eur J Cardiothorac Surg* 2006; **29(6)**:925-32. doi: 10.1016/j.ejcts.2006.03.034.
9. Hendriksen BS, Stahl KA, Hollenbeak CS, Taylor MD, Vasekar MK, Drabick JJ, *et al*. Postoperative chemotherapy and radiation improve survival following cardiac sarcoma resection. *J Thorac Cardiovasc Surg* 2021; **161(1)**:110-9.e4. doi: 10.1016/j.jtcvs.2019.10.016.
10. Ramlawi B, Leja MJ, Abu Saleh WK, Al Jabbari O, Benjamin R, Ravi V, *et al*. Surgical treatment of primary cardiac sarcomas: Review of a single-institution experience. *Ann Thorac Surg* 2016; **101(2)**:698-702. doi: 10.1016/j.athoracsur.2015.07.087.