Metastatic synovial sarcoma to the left atrium

A management dilemma

Atif W. Qubbany, MBBS, *Abdulhalim J. Kinsara,* FACC, FRCP.

ABSTRACT

We present a rare case of synovial sarcoma with minimal symptoms that metastasized and threatened to embolize the heart in a 26-year-old male admitted to our hospital for evaluation of low back pain. The tumor made a direct extension to the left atrium and moved freely in the left ventricle outflow tract. We discuss the potential therapeutic modalities in this difficult scenario.

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From the Division of Cardiology, Department of Medicine, King Khalid National Guard Hospital, Jeddah, Kingdom of Saudi Arabia.

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Address correspondence and reprint request to: Dr. Atif W. Qubbany, Division of Cardiology, Department of Medicine, King Khalid National Guard Hospital, PO Box 10021, Jeddah 21433, Kingdom of Saudi Arabia. Tel. +966 (2) 624-0000 Ext. 22070. Fax. +966 (2) 642-0000 Ext 22765. E-mail: qubbany@gmail.com

Sarcomas are rare, malignant tumors that arise from mesenchymal tissue at any body site. They have broad histopathologic spectrum. However, the World Health Organization classified most soft tissue sarcomas according to the presumptive tissue of origin, such as liposarcoma, synovial sarcoma, leiomyosarcoma, and so forth.[1] Synovial sarcomas are a rare type of tumor that can arise from any site in the head and neck, although called synovial sarcoma, they do not arise from synovium. They behave more aggressively than other tumors, and the prognosis is generally less favorable. We describe a rare case of metastatic synovial sarcoma with direct extension to the left atrium, and moving freely to the left ventricle outflow tract in a 26-year-old male who was admitted to our hospital for evaluation of low back pain.

Case Report. A 26-year-old, non-smoker, Saudi male, presented with low back pain for 6 months prior to presentation. In the preceding 3 weeks, he developed left thigh numbness and heaviness, and started to complain of decrease of appetite and had lost 6 kg in weight. There was no history of fever, night sweats, and shortness of breath. There were no cardiovascular symptoms, cough or gastro-urinary symptoms. He is a known case of neck hemangiopericytoma, treated previously with surgical excision and radiotherapy. He was seen in another hospital where MRI of the spine was carried out, which showed destructive heterogeneous expansible lesion in S1 extending into the sacral ela, associated with soft tissue component extending to the spinal canal posteriorly and presacral area anteriorly. On physical examination, he looked depressed with low-grade fever (38.3°C); pain scale of 4/10, no pallor, jaundice, cyanosis, or lymphadenopathy, with normal jugular venous pressure. He had a scar on the right shoulder from previous surgery. Chest examination revealed a left breast nodule with decreased air entry in the entire left lung and right lung base. On cardiovascular examination, he had pan systolic murmur at the apex with no organomegaly, and normal neurological examination. The working diagnosis was a spinal mass either a primary tumor, metastasis, or of other pathology. Complete blood count revealed white blood cell 14.8 (normal range [NR] 4.0-11.0 10^9/L), hemoglobin 12 (NR: 13.0-18.0 g/dl), and platelet 553 (NR: 150-450 10^9/L), normal urea electrolytes and calcium, liver function test with mild rise in gamma-glutamyltransferase 255 (NR: 2-30 IU/L) and lactate dehydrogenase 335 (NR: 100-190 IU/L). He had a very high erythrocyte sedimentation rate 117 (NR: 0-15 MM/HR) and raised C-reactive protein at 220 (NR: <5.0 mg/L). His chest x-ray showed extensive consolidation of whole left lung. His electrocardiography (ECG) test was normal, and the echocardiogram showed a large tumor with

central necrosis, which was attached to the posterior left atrial wall and protrudes through mitral valve towards the left ventricle (**Figure 1**). His cardiac MRI showed solidification of the left lung due to involvement by a solid mass. The mass extended from the left lung into the left atrium with the same signal intensity and texture. The intra-arterial portion is approximately 5x3x3 cm (**Figure 2**). The CT of the chest, abdomen, and pelvis showed large osteolytic lesions involving the sacrum, associated with right lung metastasis and left lung large mass lesion, which was extending to the heart, mainly in the left atrium, and also associated with mediastinal lymphadenopathy and rib lesion. A CT guided biopsy showed undifferentiated malignant neoplasm, consistent with synovial sarcoma. The dilemma was either to opt for surgery of the metastasis in which case there was a high risk of fatal embolization or to seek other less aggressive management and therapy, such as radiation and chemotherapy. The different modalities were discussed, unfortunately he refused any treatment when he became aware of the prognosis.

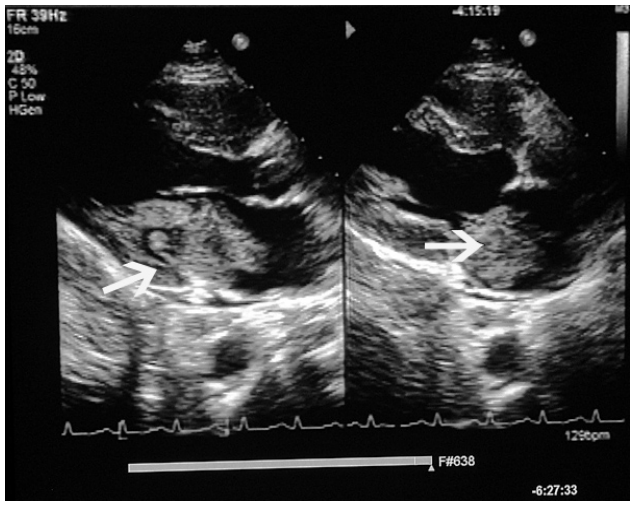

Figure 1 - Echocardiogram showing a large tumor with central necrosis protruding through the mitral valve, towards the left ventricle.

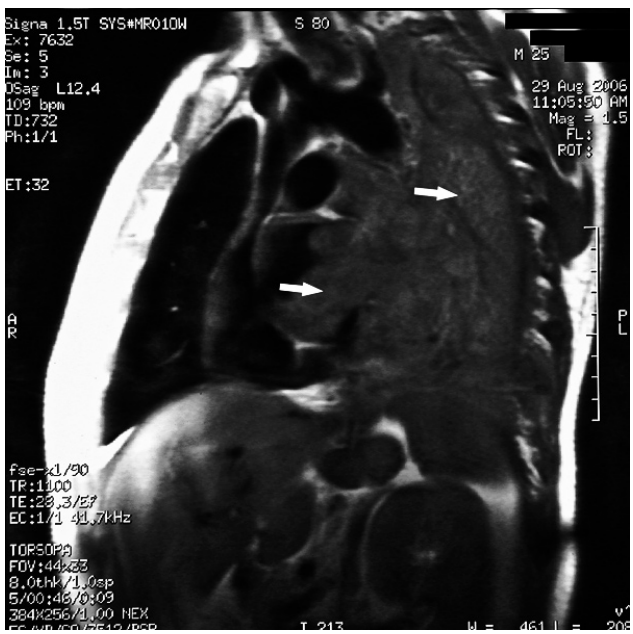

Figure 2 - Cardiac MRI showing solidification of the left lung, due to the involvement by a solid mass extending to the left atrium cavity.

Discussion. In contrast to the primary malignant cardiac tumors, metastatic involvement of the heart is relatively common. Cardiac involvement may arise from hematogenous metastases, direct invasion from the mediastinum, or tumor growth into the vena cava, and by extension into the right atrium. Malignant melanomas are particularly likely to metastasize to the heart. Others include lung and breast cancer, soft tissue sarcomas, renal and hepatocellular carcinoma, esophageal cancer, and thyroid cancer, with high prevalence of secondary cardiac involvement in leukemia and lymphoma. Cardiac or pericardial metastasis should be considered whenever a patient with known malignancy develops cardiovascular symptoms, a new or changing heart murmur, ECG conduction delay, or arrhythmia. Emboli thought to originate in the heart should also raise the possibility of cardiac involvement with tumor. Hence, it is a clinical challenge to consider the possibility of a cardiac tumor.

The most useful diagnostic tool is the echocardiogram, which locates and defines the tumor extent. The echo may also allow accurate prediction of the tumor type, whether it is malignant or benign. The MRI serves as the next most important test where the density of T1 and T2 images may allow tumor cell type identification.[2] Limited treatment modalities are available, and in very carefully selected patients, resection of cardiac metastases has been used to provide symptom palliation and prolongs life.[3] Sarcomas normally proliferate rapidly. Although complete resection with wide margin surgery is the treatment of choice, the recurrence rate is high. Most of the malignant tumors cannot be resected completely. Transplantation may be an option for those with extensive local disease. Although long-term survival has been reported with complete resection, the median survival is typically 6-12 months.[3] Autotransplantation had been used in primary, however, no data in metastatic tumors is available.[4,5] For patients with unresectable sarcomas, radiation and chemotherapy may be used, however, without great

expectation of successful results.[6] Death occurs through widespread infiltration of the myocardium, obstruction of blood flow, and distant metastases.

In summary, although cardiac metastases are rare, a high index of suspicion and an open mind at all times is needed to pick them up when they occur, especially when the symptoms are subtle. Prognosis is usually bad, however aggressive therapy if possible might be beneficial.

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