

An insidious cardiac sarcoma presenting with progressive neurologic dysfunction

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Abstract

Central nervous system metastases as the presentation of primary cardiac sarcoma are a very rare entity, with only a few previously reported cases. Sarcomas specifically make up 10 to 20% of all primary cardiac tumors. Patients with primary cardiac tumors typically present with cardiac symptomatology that may include arrhythmias, obstruction to blood flow and valve function, or symptoms of heart failure. We report a unique case of a patient with a primary cardiac sarcoma who presented with progressive neurologic dysfunction secondary to brain metastases without any preceding cardiac symptoms. We describe our novel management of these unique cases and discuss the current medical and surgical approaches to treating neurologic metastases from cardiac sarcoma.

Introduction

Primary cardiac tumors are an exceedingly rare entity with an autopsy incidence of 0.001% to 0.03%.¹ Sarcomas specifically make up 10 to 20% of all primary cardiac tumors.² Patients with primary cardiac tumors typically present with cardiac symptomatology that may include arrhythmias, obstruction to blood flow and valve function, or symptoms of heart failure.^{3,4} We report a unique case of a patient with a primary cardiac sarcoma who presented with progressive neurologic dysfunction secondary to brain metastases without any preceding cardiac symptoms.

Case Report

A 45 year-old Caucasian woman, with no significant past medical history, developed new-onset headaches. She was initially treated conservatively with rest and analgesics over the course of several months. However, her symp-

toms gradually worsened and began to include nonspecific mental status changes such as forgetfulness, daytime somnolence and postural imbalance. She eventually sought care at a local emergency department where her review of symptoms at the time was negative for any cardiac or respiratory involvement. Physical examination revealed no abnormal findings in the chest on auscultation. During the course of her evaluation a magnetic resonance imaging scan revealed two brain lesions, one in the right frontal lobe measuring 2×1.7 cm with extensive edema and another in the left parietal lobe measuring 0.8×0.6 cm on the axial T1-weighted images (Figure 1, yellow and red arrow, respectively). She underwent a stereotactic biopsy of the right frontal lobe lesion that revealed a spindle cell sarcoma with high-grade features of necrosis, increased mitoses, and nuclear pleomorphism.

Subsequent staging computed tomography scans of the chest, abdomen and pelvis were not able to identify a primary tumor site. However, a PET scan revealed a FDG-avid in the left atrium. Further evaluation with a transesophageal echocardiogram showed a 2.5×1.8 cm left atrial mass at the base of the posterior mitral valve leaflet with mobile fronds (Figure 2, yellow circle). An MRI of the chest further demonstrated that this mass was irregular and extended into the appendage and down the anterolateral mitral annulus without interfering with mitral valve function. Shortly thereafter, she underwent stereotactic radiosurgery for the two cerebral metastases present that was later followed by an excision of the left atrial mass with a pericardial patch reconstruction of the atrium and pulmonary veins. Intraoperatively, the tumor was found to obliterate the left atrial appendage and extended into the mitral valve annulus (Figure 3, yellow arrow). Unfortunately, the mass proved to be larger than radiographic imaging had predicted making the goal of achieving clear margins unattainable, as a small rim of residual tumor was left behind in the atrioventricular groove. The final pathology confirmed a 5.5×5.1×2.1 cm high grade undifferentiated cardiac sarcoma.

During the subsequent month as she was recovering from her surgery, she began to have severe headaches again and an MRI of the brain revealed several new lesions. Moreover, a repeat CT of the chest also showed interval growth of the mass within the left atrium as compared to post-surgical CT studies. She underwent palliative whole brain radiation therapy but continued to experience headaches, visual disturbances, and numbness in the upper extremities. She eventually transitioned to palliative care in view of the rapid progression of disease in the brain.

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Discussion and Conclusions

Primary cardiac sarcoma with brain metastasis is exceedingly rare and nearly all patients exhibit cardiac symptomatology at time of diagnosis. The most common presenting symptoms are dyspnea on exertion (79%), chest pain (38%), cough (21%), and paroxysmal nocturnal dyspnea (12%).⁵ It can also cause

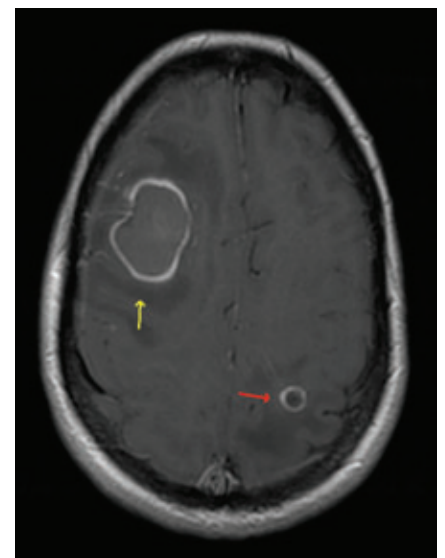


Figure 1. The magnetic resonance imaging scan reveals two brain lesions.

obstruction of blood flow, arrhythmias, or tumor embolization.⁶⁻⁹

Current understanding of cardiac sarcomas is based primarily on case reports and small institutional reviews given the limited number of patients. The origin of cardiac sarcomas is poorly understood, with the most current theory presuming an origin from pluripotent mesenchymal cells.⁷ Some of the most widely described sarcomas include leiomyosarcomas, angiosarcomas, and rhabdomyosarcomas

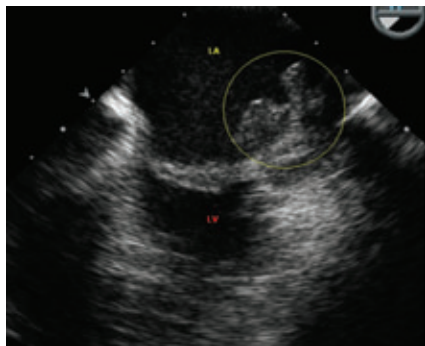


Figure 2. Transesophageal echocardiogram showed a left atrial mass at the base of the posterior mitral valve leaflet with mobile fronds.



Figure 3. Intraoperative image: the tumor was found to obliterate the left atrial appendage and extended into the mitral valve annulus (yellow arrow).

which preferentially present in the left atrium,¹⁰ right atrium,¹¹ and diffusely throughout the myocardium,³ respectively. In general, these tumors have a poor median survival of approximately six months,¹ with a lack of gender predilection and a median age of onset in the 40s.^{4,12} Metastasis at time of diagnosis have been reported at 29%⁶ and 26% in larger case series,¹³ and the site of metastases show predilection for the lung, liver, and mediastinum. The brain seems to be an uncommon site of metastasis, seen in only one patient in a large case series from our institution.⁶ Patients with metastatic disease at presentation and those with angiosarcoma *versus* other histologies have inferior survival.⁶

Primary cardiac sarcomas present as higher-grade tumors compared to non-cardiac sarcomas,¹ which is why they are considered particularly aggressive. Imaging with echocardiography can diagnose a cardiac mass in nearly 90% of patients,⁶ while cardiac MRI is particularly sensitive.¹⁴ However, these imaging techniques rarely define the true etiology of the cardiac mass and surgery is often required for tissue diagnosis. Surgery is both therapeutic for cardiac symptoms and bears a survival advantage, making it the mainstay of treatment.^{1,2,6} Unfortunately, prognosis remains poor and patients usually survive less than 1 year despite surgical excision of primary tumor and systemic chemotherapy.³ There is no consensus on the role of adjuvant chemotherapy and in cases where it is used, it usually has minimal benefit. For this patient, a therapeutic trial at surgical resection of the primary tumor and adjunctive chemoradiation for metastatic disease likely extended her life by perhaps a few months. This unusual case of a cardiac sarcoma with advanced metastases presenting like a primary brain tumor without cardiac symptoms shows our limited understanding of its pathogenesis and behavior. Further study is necessary to elucidate the optimal management of these rare and lethal tumors.

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