

CASE REPORTS

A rare presentation of a soft tissue sarcoma metastasized to the left ventricle

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ABSTRACT

Primary cardiac tumors are a rare diagnosis with a reported incidence of 0.001%-0.1% in the general population. They provide a diagnostic challenge as the patients may be asymptomatic or have a wide variety of symptoms. Because of this, cardiac tumors are often discovered incidentally by imaging modalities or during surgery. Secondary tumors that metastasize to the heart are more common with an incidence of 0.7%-3.5% in the general population and up to 9.1% in patients with known malignancies. Metastatic tumors most frequently involve the pericardium followed by the epicardium and myocardium. Intracavitary and endocardial metastasis has been rarely described in the literature. We present a case of a patient who had a known soft tissue sarcoma with widespread metastases who presented because of dyspnea. In her workup, a transthoracic echocardiogram revealed an intracavitary mass attached to the left ventricular side of the interventricular septum presumed to be a sarcoma. In our comprehensive literature search, a sarcoma has rarely been reported in the left ventricle. We have found that sarcomas that do metastasize to the myocardium often are highly differentiated and carry a poor prognosis. A cardiac tumor may be detected by transthoracic echocardiogram, transesophageal echocardiogram, magnetic resonance imaging (MRI), computed tomography (CT) or PET scan. The transthoracic echocardiogram is the preferred modality given its availability, cost and portability. Additional imaging modalities such as contrast echocardiography and MRI can help differentiate cardiac tumors from other cardiac masses such as thrombi or vegetations. Because of the inability to rely on physical signs and symptoms, there must be a high degree of suspicion when a patient with known metastatic disease presents with cardiac complaints. It is important for clinicians to be aware of how different imaging modalities can help guide their differential diagnosis of cardiac masses.

Key Words: Cardiac sarcoma, Echocardiography, Magnetic resonance imaging

1. INTRODUCTION

Cardiac tumors encompass a rare form of heart disease and the prognosis may be improved with a prompt diagnosis. Diagnosing cardiac tumors requires a strong clinical suspicion as the patient may present with a wide range of symptoms if they are symptomatic at all. Clinical history of malignancy may be suggestive and clue the clinician to conduct further investigation. There are several modalities to aid in

the diagnosis of a cardiac tumor. Using two-dimensional echocardiography may be the first step in diagnosis as it is noninvasive, portable and cost-effective. Masses can be further classified and differentiated with the use of intravenous contrast, magnetic resonance imaging, computed tomography or PET imaging. We describe a case of a 60-year-old female with known metastatic sarcoma who was found to have metastases to her left ventricular wall.

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2. CASE PRESENTATION

The patient is a 60-year-old female with a past medical history of hypertension, hyperlipidemia, thyroid cancer status post thyroidectomy and a left leg muscle sarcoma with known diffuse metastases who presented from her physician's office with the chief complaint of dyspnea. She had been conducting follow-up with her oncologist at an outside facility in regards to her malignancy. As part of her outpatient workup for dyspnea, a computed tomography (CT) scan of the chest was ordered. The patient was then informed while driving home that she had developed a pulmonary embolism. She was advised to go to the nearest hospital for further evaluation. In the Emergency Room, basic laboratory values were drawn and a discussion was had with her oncology physicians regarding her extensive medical history and the treatment plan as she was currently receiving chemotherapy. She was medically stabilized and empirically started on anticoagulation with enoxaparin. Upon initial examination, the patient was in minimal distress and only complaining of dyspnea and chronic abdominal pain.

Her initial vitals were significant for a temperature of 98.3°C, blood pressure 128/72 mmHg, heart rate of 122 beats per minute, respiratory rate of 15 respirations per minute and an oxygen saturation of 94% on room air. The patient was not in acute distress; sitting comfortably and speaking in full sentences. She did not have any significant jugular venous distension. The patient was tachycardic with a rather benign cardiac exam. Her lungs had slightly decreased breath sounds at the bilateral bases with scattered fine crackles. Her abdominal sounds were hypoactive with slight abdominal distention noted and she was diffusely tender to palpation without rigidity or guarding. Minimal right lower extremity edema was noted with the left lower extremity wrapped in a bandage.

Her initial laboratory data was significant for sodium 136, potassium 3.8, chloride 102, bicarbonate 24, glucose 98, BUN 23, creatinine 1.02, calcium 8.6, troponin 0.00. WBC 12.3, hemoglobin 8.3, platelet 283. INR 1.1. PTT 28.2.

At the time of presentation, a CT Chest/Abdomen/Pelvis was ordered which revealed lateral left lung base heterogenous pleural masses measuring up to 10 cm × 3.1 cm with calcified pleural plaques medially. These were noted initially on Chest X-Ray as well as CT (see Figures 1, 2). Additional masses were noted at the left costophrenic angle in the pleura measuring 1.2 cm × 2 cm, the left lower major fissure measuring up to 2.7 cm and a soft tissue mass 9.2 cm × 9.0 cm × 10.6 cm in the right medial lung base. Small bilateral pleural effusions and bilateral calcified pleural plaques were also noted. Multiple parenchymal nodules and masses were pres-



Figure 1. Chest X-Ray revealing large left pleural density with smaller right mid lung field density

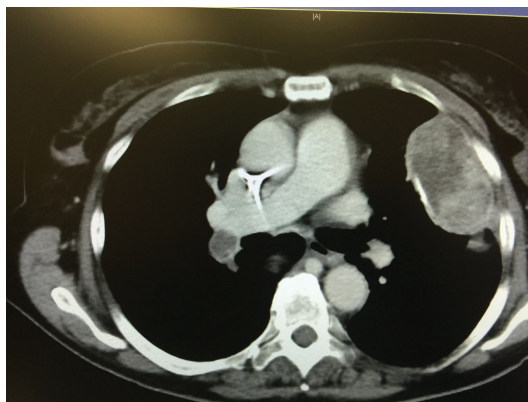


Figure 2. Chest CT revealing large left lung pleural mass ent bilaterally. The middle right lower lobe demonstrated a pulmonary embolism.

Abdominal portion of imaging revealed a large left hepatic lobe mass with central necrosis measuring 23.7 cm × 18.9 cm × 25.3 cm in size (see Figure 3). Multiple renal nodules were present with the largest measuring 3.0 cm × 2.3 cm. A left lower quadrant peritoneal or omental tumor was noted, measuring 5.7 cm × 5.3 cm × 4.4 cm. There was noted perihepatic, right paracolic gutter and pelvic ascites present. In the pelvis, the left common iliac vein was expanded with an irregular lumen suspicious for thrombus. There were centrally located left psoas mass and left external iliac adenopathy noted. The left groin was noted to have operative changes from sarcoma removal as evident in her left

proximal thigh. There were large masses in her left gluteus and right upper thigh.

The sarcoma pathology was done prior to admission at an outside hospital several years prior when she complained of a lump in her upper thigh found to be a solid mass on ultrasound. A CT scan revealed a 7.5 cm × 4.5 cm enhancing mass along the proximal adductor muscles suspicious for malignancy along with a hypodense right kidney lesion. The patient received a CT-guided biopsy of the mass which was consistent with high-grade myxoma sarcoma. Immunostain for OSCAR, S100, SMA and desmin were performed but were negative. The sarcoma exhibited muscular invasion with marked central necrosis and myxoid changes.



Figure 3. CT Abdomen revealing large left hepatic lobe mass with central necrosis

A two-dimensional echocardiogram was then performed at this admission to evaluate her dyspnea. The echocardiogram revealed a new 2.3 cm × 1 cm echogenic mass attached to the left ventricular basal septum (see Figures 4-6). Left ventricular wall motion was hyperdynamic with a left ventricular ejection fraction of 70%-75%. Grade 1 diastolic dysfunction was demonstrated as well. There was a maximum left ventricular outflow tract gradient of 10 mmHg. Pulmonary hypertension was not appreciated and there were no shunts detected on color Doppler. There were no other cardiac abnormalities detected on the echocardiogram.

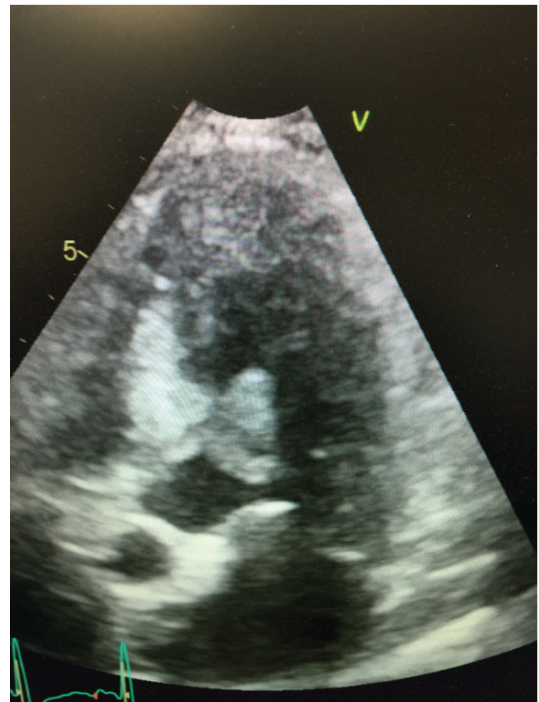


Figure 4. Cardiac sarcoma attached to the left ventricle seen in an apical 4-chamber view on transthoracic echocardiogram

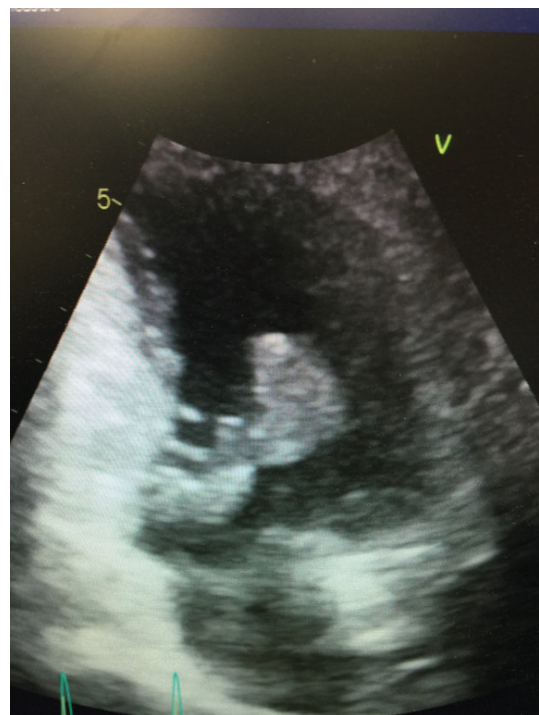


Figure 5. Another magnified view of a sarcoma attached to the left ventricle seen in an apical 4-chamber view on transthoracic echocardiogram

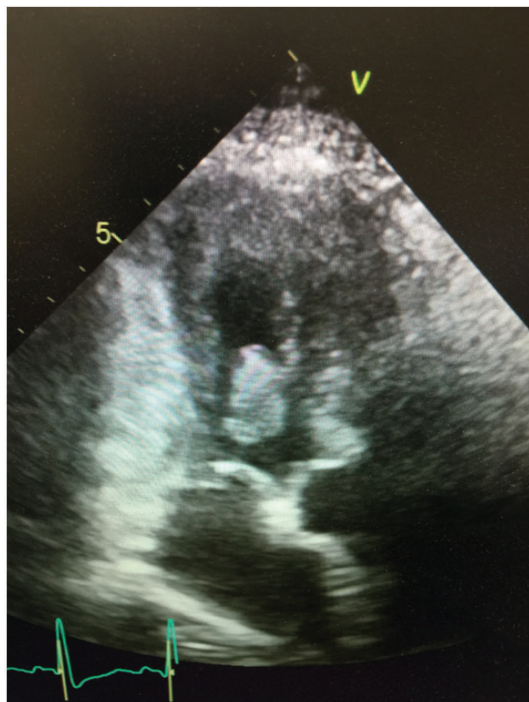


Figure 6. Cardiac sarcoma seen on an apical 2-chamber view on transthoracic echocardiogram

The patient remained hemodynamically stable throughout the hospital course and did not require oxygen. Her acute treatment options included anticoagulation for her pulmonary embolus and surgery. The patient did not have any symptoms consistent with the tumor causing compression of cardiac structures. Given her severe metastatic disease, surgical options were declined in favor of anticoagulation. She was to continue her chemotherapy regimen of ifosfamide and doxorubicin with monthly doses of neulasta were administered by her oncology team with her oncology team. Given the extent of her metastases, palliative care was consulted for pain management and goals of care. The patient was treated with anticoagulation using enoxaparin and her symptoms continued to improve. The patient eventually was stable enough to go home and she was scheduled to follow-up with her oncologist.

3. DISCUSSION

Cardiac tumors remain a rare condition with the diagnosis most often made after surgical pathology confirmation. The reported incidence is 0.001%-0.1% in the general population after postmortem analysis.^[1-4] Secondary tumors with cardiac metastasis are more common with an incidence of 0.7%-3.5% in the general population and up to 9.1% in patients with known malignancies. Several individual studies were conducted with higher incidence rates such as a study by Yu Kun *et al.*, 33,108 cardiac surgeries that were performed

for any indication were reviewed with 0.71% of cases (n = 234) having a cardiac tumor. In this study, there was a greater incidence of benign neoplasms compared to malignant neoplasms (90.6% vs. 9.4%, $P < .01$), similar numbers were reported by a study by Bruce CJ as well.^[2,5]

Clinical manifestations are nonspecific and are more characteristic of the tumor location and overall burden rather than histological type. If present, symptoms are quite variable. In a study by Yusuf *et al.* looking at 59 consecutive patients with intracardiac tumors, the most common presenting symptom for primary and secondary tumors was dyspnea (62.5%, 35% respectively). For secondary tumors, only 56% of patients were even symptomatic.^[3] Many different malignancies are implicated in secondary cardiac tumors as all malignancies can cause metastases by lymphatic, hematogenous, direct or transvenous extension. Melanoma was reported as having the highest rate of cardiac metastasis. Central nervous system tumors have not yet been reported as a cause for metastasis.^[4]

Cardiac tumors often are characteristically discovered in specific areas of the heart for which they most commonly metastasize. Bruce CJ *et al.* described in their study at Mayo Clinic of 323 consecutive patients undergoing resection of primary cardiac tumors that 94% of their cardiac tumors were confirmed to be benign with 50% of those being myxomas. Of the 10% of tumors that were malignant, 90% were sarcomas which most commonly were angiosarcomas. According to location, they report a predilection of the left atrium for myxomas, right atrium for angiosarcomas. Rhabdomyomas and fibromas were more often found in the ventricles. Yusuf *et al.* describe sarcomas as the most common primary tumor. They also discovered the right atrium to be the most frequent site affected. Sarcomas affecting the myocardium generally are well differentiated and have a very poor prognosis. They can often cause cardiogenic shock or outflow obstruction.^[3]

Because of the importance of early diagnosis, two-dimensional echocardiogram can be a quick and inexpensive way of finding a suspected cardiac mass. The differential of a cardiac mass includes intracardiac thrombus, vegetation and anatomical structures such as the crista terminalis and moderator band. Echocardiography can be a tool in determining the overall appearance as well as detecting pericardial invasion and characterizing morphological features which may help further classify cardiac masses. Both modalities of echocardiogram, transthoracic and transesophageal, can provide useful information in detection of a cardiac thrombus and aid in biopsies. There is high diagnostic sensitivity as well with transthoracic (93.3%) and transesophageal (96.8%) echocardiography. The location of the tumor may also offer some clue as to its morphological type.^[5-7] The echocardi-

graphic appearance of certain tumors may be pathognomonic such as a left atrial myxoma prolapsing across a mitral valve may not need any further workup for diagnosis.

Intravenous contrast can also be added to an echocardiogram to help further differentiate cardiac masses. Contrast echocardiography was able to distinguish qualitative and quantitative differences in perfusion of cardiac masses in relation to other cardiac structures. In a study by Kirkpatrick *et al.*, malignant or higher vascular tumors had greater enhancement compared to adjacent myocardium. Myxomas were found to have decreased enhancement and thrombi were found to have no perfusion at all. The use of microbubbles in tissue was able to give off measurable intensities which reflected the degree of blood flow. This is useful for malignant neovascularized tumors or vascular tumors which have a large amount of blood flow. The use of contrast allowed non-experienced readers to more accurately diagnose cardiac masses. The use of microbubble technology has been used in the liver to help differentiate benign from malignant tumors as well.^[8]

MRI provides advantages over echocardiography in distinguishing between thrombi and malignant tumors and benign tumors. Compared to echocardiography, MRI provides greater delineation of mass borders and can provide greater soft tissue characterization making it more specific. Its high spatial and temporal resolution allows MRI to accurately detect size, degree of involvement, vascularity, lesion mobility, fat content, and the presence of central necrosis. T1 and T2 weighted imaging enables more accurate definition of mass borders, water content and delineation between tissue and adjacent myocardium. The addition of gadolinium in certain patients can help further characterize inflammation, fibrosis, and scar within a tumor. CT is useful to detect tumor calcifications and an added utility for patients who cannot tolerate MRI.^[9] PET scans also have added utility as there is reported correlation between glucose accumulation in tumors that are malignant.^[10] Electrocardiography may detect conduction abnormalities but is rather nonspecific.

In our case, our patient had known metastases of her soft

tissue high-grade myxoid sarcoma. Given her complaint of dyspnea, it was appropriate to include a two-dimensional echocardiogram as part of her evaluation. In her case, she was found to have a sarcoma that had metastasized to the left ventricle (see Figures 4-6). In our comprehensive literature search, various cardiac tumors and metastases have been reported, but a sarcoma metastasized to the left ventricle has rarely been described. Many other cases of metastatic tumors have been reported specifically in the left atrium. The left ventricle most commonly holds rhabdomyomas, fibromas, hamartomas and purkinje cell tumors. Masses in the left ventricle may also be benign and include thrombus, apical hypertrophic cardiomyopathy and sub-aortic membranes. Normal or variant structures noted include calcified or multi-lobed papillary muscle, redundant mitral chordae, trabeculations, false tendons, and focal upper septal hypertrophy.^[5-7] Survival is dependent on the tumor classification, type of tumor histology and the patient's functional status. Sarcomas specifically are described as having a poor prognosis when they affect the myocardium with median survival reported at 6-12 months.^[2,3] The mechanism for metastases in our case is unclear given the large multiple tumor burden. There was no detected shunt, atrial or ventricular septal defect noted on echocardiogram. Solitary cardiac tumors, whether primary or secondary, are generally considered for surgical treatment. Complete surgical resection can be considered in isolated settings or with obstructive symptoms. Cardiac autotransplantation is an option that has been studied to be safe in left sided tumors but has not been studied in metastatic disease. This patient had undergone chemotherapy in the past, resumption of which could be considered along with chemotherapy.^[11,12] Many cases have been reported of cardiac tumors but little has been reported of sarcoma metastases to the left ventricle.

CONFLICTS OF INTEREST DISCLOSURE

The authors have declared no conflicts of interest.

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