



## Cardiac Tumors are not always Myxomas

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### Abstract

There are multiple etiologies that are responsible for an intracardiac mass, either benign or malignant, among them intracardiac abscess or a primary tumor. There is little evidence of diagnostic and treatment approaches for intracardiac masses. We report a case of a 60-year-old male diagnosed with a left thigh leiomyosarcoma that developed two left cardiac metastases. The current literature of cardiac metastases are mainly case reports and case series, there is an unmet need of more research on this area to come to the best diagnostic and treatment approach for this kind of patients for a better overall outcome.

### Introduction

There are multiple etiologies that are responsible for an intracardiac mass, either benign or malignant, among them intracardiac abscess or a primary tumor. There is little evidence of diagnostic and treatment approaches for intracardiac masses. We report a case of a 60-year-old male diagnosed with a left thigh leiomyosarcoma that developed two left cardiac metastases. The current literature of cardiac metastases are mainly case reports and case series, there is an unmet need of more research on this area to come to the best diagnostic and treatment approach for this kind of patients for a better overall outcome.

### Case Presentation

The patient is of a 60-year-old male with past medical history of hypothyroidism managed with levothyroxine, with past surgical history of tonsillectomy and knee surgery, family history of a not specified cancer in his mother and a penicillin allergy. The patient was diagnosed with left thigh leiomyosarcoma in 2002 after a gait disturbance associated with left thigh pain. The patient underwent biopsy that showed positivity for smooth muscle antigen, desmin and EMA, and stained negative for keratin, S-100 and CD34 supporting the diagnosis of malignant spindle cell neoplasm consistent with leiomyosarcoma. He underwent multiple surgeries followed by IMRT radiation therapy for local control for the first 10 years. Later he presented with a lump on the left lateral chest wall and a PET scan was ordered and results demonstrated bulky hypermetabolic hilar adenopathy, a left chest wall hypermetabolic mass in the mid axillary line, a soft tissue mass in his right psoas muscle and a hypermetabolic left retroperitoneal mass below left kidney. The patient underwent radiation therapy and was started on palliative chemotherapy treatment with gemcitabine/docetaxel. PET scans done for disease assessment showed improvement in areas of metastatic disease except for a new area in the left occipital bone for which he received radiation. The patient received 6 cycles of chemotherapy and then underwent PET scan evaluation which showed several areas of uptake compatible with Disease Progression (PD). A discussion took place with the patient and it was decided to continue with palliative chemotherapy with trabectedin. After 3 cycles, the patient had PD again in the right lung for which the chemotherapy regimen was changed to doxorubicin, ifosfamide and mesna (MAI). He completed 6 cycles and underwent CT scan which showed Disease Stabilization (DS). Molecular testing was done which showed no actionable mutations.

After one year, a PET scan for assessment showed a new lesion on the left Gluteal Musculature (PD) and a biopsy was done which showed malignant spindle cell neoplasm consistent with recurrent leiomyosarcoma.

The patient was referred to radiation oncology and completed treatment for local control with IMRT. Next Generation Sequencing (NGS) done in plasma showed non actionable mutation, being negative for EGFR, ALK, ROS-1, KRAS, MET, RET, BRAF and PD1/PLD1.

The patient underwent another PET scan after radiation which showed a mass on the Left Ventricle (LV) (Figure 1) and also a good response to radiation therapy of the previous lesions.

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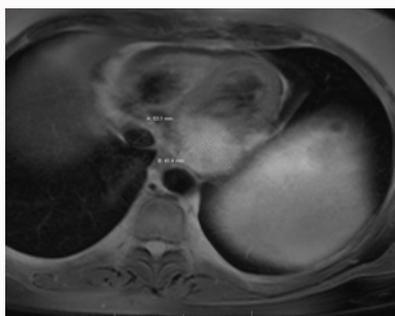
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**Figure 1:** PET scan showing a cardiac mas in the left ventricle.



**Figure 2:** Cardiac MRI showing the leiomyosarcoma metastasis in the left ventricle.

An echocardiogram was order and showed an Ejection Fraction (EF) of 63%, a left ventricle not well visualized a grade II (Pseudo-normal) abnormal left ventricular diastolic function. Elevated left atrial pressure. A cardiac MRI was ordered to better assess the cardiac mass and showed a centrally necrotic- approximately 5.3 cm-myocardial metastatic lesion located in the basal LV inferior wall and the mid RV posterior wall adjacent to the RV-LV insertion. A smaller adjacent exophytic 2 cm mass centered in the posterior aspect of the mid right ventricular myocardium adjacent to the inferior RV-LV insertion. A normal left ventricular size with mildly reduced global systolic function and hypokinesis to akinesis of the basal mid inferior and inferolateral segments, EF=47%, normal right ventricular size with normal systolic function and a small pericardial effusion.

The cardiac MRI with IV contrast showed small pericardial effusion and a left ventricular myocardial mass in the basal to mid inferior and inferolateral walls that measured 5.3 cm × 4.1 cm × 4 cm (Figure 2).

It was T2 hyperintense, T1 hypointense, and contained no internal fat. There was no significant enhancement on first pass perfusion but there was thick peripheral and heterogeneous internal enhancement on delayed arterial and venous phases. There was also a smaller adjacent mass centered in the posterior aspect of the mid right ventricular myocardium adjacent to the inferior right ventricle insertion, measuring 2 cm × 1.7 cm and demonstrating similar signal characteristics.

Because the extensive history of recurrence with chemo and radiation treatment, and the characteristics of the mass, it was considered to be metastases from his primary tumor. He was then scheduled for radiation therapy of the mass, but because the patient decided to move to a different state and its being considered for a clinical trial.

## Discussion

Our patient presented with an asymptomatic metastasis detected by PET scan from a leiomyosarcoma. The patient had already being treated with systemic therapy and failed: docetaxel/gemcitabine, trabectedin and he has maximized his adryamicin dose with 6 cycles of MAI regimen. Due to the fact that the cardiac metastasis was the only site of disease the decision was to proceed with IMRT.

The most common primary cardiac tumor is the atrial myxoma [1]. Cardiac primary tumors and metastases are uncommon and less common coming from soft tissue sarcomas. Metastatic sarcomas to the heart from a distant site, is much more common than a primary cardiac sarcoma. In one autopsy series, 25% of 120 patients dying of metastatic soft tissue sarcoma were found to have metastases in the heart and it was noted that survival was better in patients with left-sided tumors and those who received postoperative chemotherapy and/or radiation therapy [2].

According to the Atlas of Tumor Pathology, published by the Armed Forces Institute of Pathology (AFIP), the most prevalent primary cardiac tumor of 124 surgical and autopsy cases was myxoma, followed by sarcoma, rhabdomyoma, papilloma, and fibroma. The sarcomas were rhabdomyosarcoma, angiosarcoma and other sarcomas [3].

Tumors metastatic to the heart are among the least known and highly debated issues in oncology. Although primary cardiac tumors are extremely uncommon (various postmortem studies report rates between 0.001% and 0.28%), secondary tumors are not, and at least in theory, the heart can be metastasized by any malignant neoplasm. In general, cardiac metastases are considered to be rare; however, when sought for, the incidence ranges from 2.3% to 18.3%. Although no malignant tumors are known to preferentially metastasis to the heart, some do involve the heart more than others, for example melanoma and mediastinal primary tumors [4]. Tumors can spread to the heart through four pathways: By direct extension, through the bloodstream, through the lymphatic system and by intracavitary diffusion by the IVC or the pulmonary veins.

Patients presenting with functional decline and signs of congestive heart failure, may be ameliorated by excision of primary or metastatic cardiac sarcoma. Pericardial window, pericardiectomy, or valve replacement may also help although management experience is limited by the rarity of these malignancies.

It is known that chemotherapeutic agents like anthracyclines and radiation therapy are cardiotoxic thus they must be used with caution and close follow up.

In general, although a focal lesion secondary to the myocardium may result in unclear symptoms that may go undetected, tumors spreading more extensively to the pericardium or to other cardiac sites may produce dramatic clinical patterns, causing medical emergencies. The most typical scenario would be a patient with lung or breast cancer presenting with increasing shortness of breath, hypotension and tachycardia and clinical findings suggestive of tamponade. Neoplastic pericardial effusions are indeed among the most feared complications. Although they may result in no symptoms, they are commonly the ultimate cause of death.

In the case of secondary tumors located in the myocardium, the clinical scenario will be proportional to the degree of myocardial infiltration, or to the wall infiltration site. Typical presentation

includes arrhythmias, such as atrial flutter or fibrillation or ventricular arrhythmias, complete atrioventricular blocks, especially if the conduction system has been infiltrated. Myocardial metastases can involve any one of the heart chambers but there is a preferential involvement of the right or left ventricle has been suggested. Endocardial metastases, usually localized to the right heart, are rare and usually associated with tumors with endovascular growth such as renal, liver and uterine cancers [5].

Echocardiography is the best diagnostic tool for intracavity and mural cardiac tumors. Recent advances in echocardiography have led to a determination of both the precise localization and size of the tumors. Computed Tomography (CT), Magnetic Resonance Imaging (MRI), and/or angiography further add to the diagnostic information. For intrapericardial tumors, CT and MRI are helpful for clarifying the anatomy of the tumor and invasiveness into the cardiac structures. These methods even reveal the vascularity of the tumors and are superior to echocardiography [6].

Other cases that require surgery include benign tumors causing hemodynamic dysfunction, arrhythmia, pericarditis or cardiac tamponade. For malignant tumors, surgery is indicated to debulk the tumor as either a palliative procedure or for tissue diagnosis [5]. Malignant primary cardiac neoplasms often require palliative surgery because patients can present with a mechanical blood flow obstruction and in such cases, emergency surgery is indicated. The most common clinical presentation is heart failure, followed by symptoms caused by peripheral emboli to the cerebral, systemic and coronary arterial circulatory systems [7].

Regarding leiomyosarcoma, it is treated differently depending on whether the tumor originates from the uterus or extrauterine sites. The limited literature on the subject of cardiac metastases from a distant sarcoma origin, suggests that a surgical approach is the best option; when this is not possible, the combination of chemotherapy and radiotherapy seems to be better than chemotherapy alone [8].

Cases of leiomyosarcoma with cardiac metastases have not been reported so far in the literature and it is important to make the medical field aware of this type of cases because there could be a significant number of patients with cardiac metastases that could be assert and possibly offer a better treatment strategy. Although leiomyosarcoma is a rare entity, and cardiac metastases even more, this type of pathology should be assessed by a multidisciplinary team consisting of oncologist, cardiologist, CT surgeons, radiation oncologist and radiologists.

This case illustrates the importance to have a broad differential diagnosis when it comes to patient with soft tissue sarcomas with cardiac metastases. Recognition of this kind of presentation is critical to institution for the appropriate diagnosis and evaluation for future surgeries and medication adjustments.

## Conclusion

Cardiac metastases from leiomyosarcoma are a very rare entity. Patients benefit from early treatment, but because of its low prevalence, there aren't studies regarding a standardized treatment approach for cardiac metastases, chemotherapy vs. radiation, chemotherapy followed by radiation, radiation followed by chemotherapy, time of chemotherapy administration since diagnosis, or other treatment approaches with surgery. We hope that with reports like this, the interest for finding the answer to the best treatment approach is for a better overall outcome.

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