Scientific Research Publishing

Case Reports in Clinical Medicine, 2023, 12, 339-344
https://www.scirp.org/journal/crcm
ISSN Online: 2325-7083
ISSN Print: 2325-7075

Primary Left Atrial Intimal Sarcoma Disguised as Functional Mitral Stenosis: A Case Study

Zachary Visinoni[1*], Frances Patmon[1], Rishi Menon[1,2]

[1]Department of Graduation Medical Education Internal Medicine Residency, Sutter Roseville Medical Center, Roseville, USA
[2]Roseville Cardiology, Roseville, USA
Email: *zachary.visinoni@sutterhealth.org

How to cite this paper: Visinoni, Z., Patmon, F. and Menon, R. (2023) Primary Left Atrial Intimal Sarcoma Disguised as Functional Mitral Stenosis: A Case Study. *Case Reports in Clinical Medicine*, **12**, 339-344.
https://doi.org/10.4236/crcm.2023.129047

Received: July 26, 2023
Accepted: September 2, 2023
Published: September 5, 2023

Abstract

A 66-year-old female with a 1-month history of increasing fatigue, dyspnea on exertion, and palpitations presented with clinical signs of heart failure. Chest computed tomography (CT) revealed a large, mobile left atrial (LA) mass attached to the mitral valve causing severe mitral stenosis and mitral regurgitation. The mass was surgically debulked and the mitral valve was replaced. Pathology revealed a poorly differentiated malignant spindle cell neoplasm with diffuse nuclear positivity of MDM2 and multifocal positivity of CDK4, consistent with intimal sarcoma. This case seeks to describe an uncommon presentation of a rare malignancy, and the surgical and medical management of the disease.

Keywords

Intimal Sarcoma, Cardiac Tumor, Mitral Stenosis, Atrial Mass

1. Introduction

Malignant cardiac tumors have an especially aggressive course and carry a poor prognosis [1]. The discovery of primary cardiac tumors on autopsy is rare, with an approximate prevalence of between 0.001% - 0.030% [2]. Of primary tumors, 90% are benign with the majority being myxomas; cancerous tumors make up the remaining 10% [3]. The most common malignant cardiac tumors are sarcomas [4]. After analyzing 100 patients diagnosed with cardiac sarcoma, a 2014 study found that 42 had histopathology, consistent with intimal sarcoma [5]. Metastatic tumors—most commonly from melanoma, lung cancer, breast cancer, and lymphoma—are approximately 30 to 40 times more common than pri-

mary tumors [6]. Cardiac tumors cause symptoms based on their location and size, as opposed to their histopathology. Valvular dysfunction, myocardial infiltration, embolism, heart failure, electrical disturbances, and systemic symptoms are all possible [7]. Diagnosis requires imaging and biopsy. These tumors may invade multiple structures, including the myocardium, pericardium, valves, chordae, and great vessels, rendering complete resection difficult; this contributes to the highly aggressive nature of cardiac sarcomas [3]. Intimal sarcomas present a unique challenge to clinicians and surgeons due to their local destruction, critical involvement of nearby structures, and difficulty in obtaining complete surgical resection [8]. The mean survival for all patients diagnosed with primary cardiac intimal sarcoma is approximately 17 months. Patients who achieve complete resection have a mean survival of 38 months, compared to less than 12 months for those who do not undergo surgery [9]. We present one such example of a patient who presented clinically with heart failure from functional mitral stenosis and regurgitation.

2. Case Report

A 66-year-old female with a history of hypothyroidism presented to the emergency department (ED) with worsening shortness of breath for the past day. Over the last month, she developed progressive dyspnea on exertion, palpitations, and profound fatigue. She was evaluated by her primary care physician a week prior and was noted to have a new systolic murmur. In the ED, she had a blood pressure of 97/67 mmHg, a heart rate of 87 bpm, and an oxygen saturation of 99% on room air. A physical exam revealed clear lungs, an irregular rhythm, and a prominent systolic murmur at the apex. Labs were significant for hemoglobin 10.6 g/dL and an elevated D-dimer of 1.24. Renal and hepatic function were normal. Chest CT angiogram was negative for pulmonary embolism, but revealed cardiomegaly, a moderate pericardial effusion, bilateral pleural effusions, and a poorly identifiable LA mass (**Figure 1**).

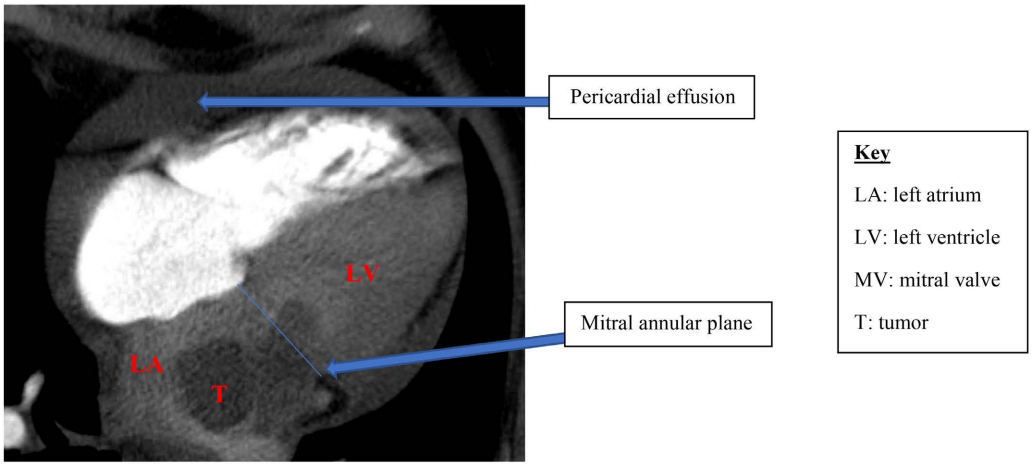

Figure 1. Four-chamber view showing presence of the LA mass extending across the mitral valve. A moderate pericardial effusion is also noted.

Echocardiography demonstrated a large, irregular, mobile mass occupying most of the LA (Figure 2). The mass was adherent to the LA appendage and caused severe mitral stenosis with a mean inflow gradient of 12 mmHg. There was associated severe mitral and tricuspid regurgitation (Figure 3). She underwent surgical debulking of the mass. A 6.5 × 5.2 × 4.0 cm white-tan mass was resected, however, a remaining 4.0 × 2.5 cm portion adherent to the LA appendage could not be excised. The native mitral valve appeared significantly thickened with poor movement of the posterior leaflet requiring replacement with St. Jude Epic porcine bioprosthesis. The tricuspid valve demonstrated severe insufficiency when tested intraoperatively and warranted repair.

Pathology of the mass revealed a highly pleomorphic spindle cell neoplasm with poorly differentiated morphology and nuclear positivity of MDM2 and CDK4 genes. This was most consistent with intimal sarcoma of cardiac origin with surface involvement of the resected native mitral valve.

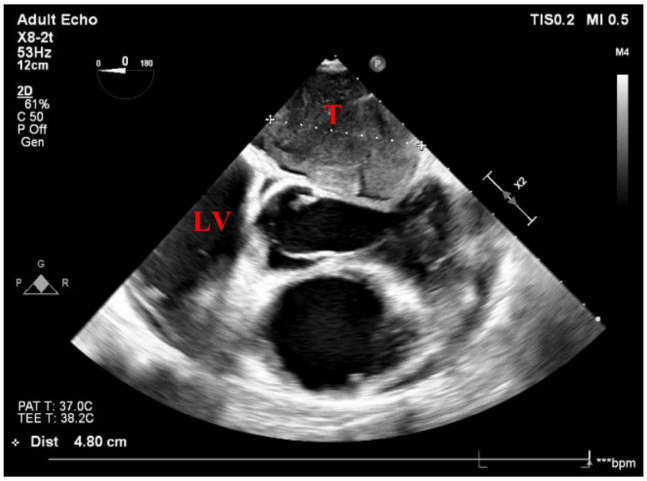

Figure 2. Single view on transesophageal echocardiogram (TEE) demonstrating near complete occupation of left atrium by tumor.

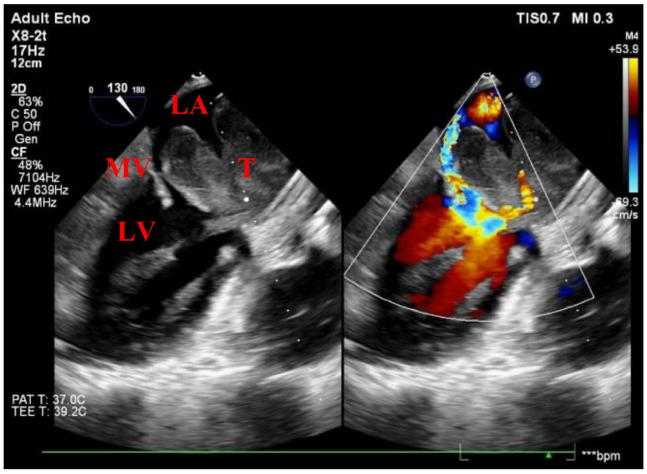

Figure 3. TEE color doppler revealing functional mitral stenosis limiting mitral inflow due to obstruction from mass.

Positron emission tomography (PET)/CT revealed heterogenous fluorodeoxyglucose (FDG) accumulation within the LA and LA appendage. There was no evidence of distant or extra-thoracic metastasis secondary to sarcoma. She was evaluated by a radiation oncologist who felt that although sarcomas are often resistant to radiotherapy, it would be the best option as complete resection was not possible. The case was discussed with specialists at Brigham and Women's Hospital and Stanford University; subsequent arrangements were made for her to be evaluated at a regional sarcoma center in the coming weeks.

However, before therapy could begin, she was readmitted with heart failure symptoms. Electrocardiogram revealed atrial fibrillation with rapid ventricular response and a chest X-ray was consistent with pulmonary edema. Unfortunately, she expired the next day from acute congestive heart failure.

3. Discussion

Primary left atrial intimal sarcoma is an especially rare tumor in an uncommon location. Intimal sarcoma is more commonly seen in the pulmonary trunk, pulmonary arteries, and aorta. The prognosis of cardiac intimal sarcomas is poor. Favorable prognostics include complete tumor resection, chemotherapy, lower tumor stage, and young age [10]. Left-sided tumors tend to carry a slightly better prognosis—possibly due to earlier detection. Poor prognostic features include incomplete surgical resection and evidence of tumor necrosis on biopsy [7]. The lungs are the most common site of metastasis, making up approximately 40% of all metastatic foci. Bone, brain, liver, peritoneum, and mesenteric lymph nodes are also common sites for seeding of tumor emboli [11].

Diagnosis of cardiac intimal sarcoma typically begins with echocardiography [7]. Cardiac magnetic resonance imaging (MRI), cardiac CT, and 3D echocardiography may provide additional diagnostic information, although a definitive diagnosis is done with resection and histological review [6]. The primary means of treatment is surgical resection of the mass; the benefit of chemotherapy and radiation therapy is questionable. Since cardiac intimal sarcoma is associated with metastasis 80% of the time at diagnosis, a palliative approach is often pursued [7].

Histological findings of cardiac intimal sarcoma reveal a poorly differentiated mesenchymal tumor with fibroblastic or myofibroblastic differentiation, often including atypical spindle cells and evidence of necrosis. Large myxoid areas and epithelioid-appearing tumor cells may be present. Positivity for vimentin, osteopontin, and MDM2 is also common [11].

The clinical manifestation of cardiac tumors is dependent upon the location, size, and appearance of cardiac imaging [12]. Due to the size and location of our patient's mass, she presented with symptoms related to severe mitral stenosis and mitral regurgitation. The hemodynamic effects of the tumor were especially striking. Mitral stenosis itself is a rare condition [13]. The etiology of mitral stenosis in our patient was even more unusual: primary LA intimal sarcoma [13].

Furthermore, the pathophysiological consequences caused by the tumor mass were particularly significant.

In patients with chronic mitral stenosis, the increased gradient between the LA and left ventricle (LV) during diastole requires LA compensation to maintain LV filling pressures. LA pressures must increase, which over time leads to cardiac remodeling and structural changes [14]. However, in our patient's case, the acuity of tumor growth did not allow for appropriate LA compensation; this led to heart failure symptoms. By the time she was evaluated in the ED, the mitral stenosis was severe enough to cause pulmonary venous congestion, pulmonary arterial congestion, and subsequent tricuspid insufficiency.

4. Conclusion

Primary cardiac intimal sarcoma presents several challenges to clinicians and surgeons due to its location, propensity for distant metastasis, and difficulty in achieving complete resection. Symptoms and manifestations of the disease are a result of the location and size of the tumor, which may affect nearby structures. In our patient's case, the interference with mitral valve function precipitated heart failure symptoms and rhythm disturbances.

Declaration of Patient Consent

The authors certify that the patient's spouse signed an informed consent form, which is available upon request.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

References

[1] Hamidi, M., Moody, J.S., Weigel, T.L. and Kozak, K.R. (2010) Primary Cardiac Sarcoma. *The Annals of Thoracic Surgery*, **90**, 176-181. https://doi.org/10.1016/j.athoracsur.2010.03.065

[2] Castillo, J.G. and Silvay, G. (2010) Characterization and Management of Cardiac Tumors. *Seminars in Cardiothoracic and Vascular Anesthesia*, **14**, 6-20. https://doi.org/10.1177/1089253210362596

[3] Bussani, R., Castrichini, M., Restivo, L., Fabris, E., Porcari, A., Ferro, F., Pivetta, A., Korcova, R., Cappelletto, C., Manca, P., Nuzzi, V., Bessi, R., Pagura, L., Massa, L. and Sinagra, G. (2020) Cardiac Tumors: Diagnosis, Prognosis, and Treatment. *Current Cardiology Reports*, **22**, Article No. 169. https://doi.org/10.1007/s11886-020-01420-z

[4] Siontis, B.L., Zhao, L., Leja, M., McHugh, J.B., Shango, M.M., Baker, L.H., Schuetze, S.M. and Chugh, R. (2019) Primary Cardiac Sarcoma: A Rare, Aggressive Malignancy with a High Propensity for Brain Metastases. *Sarcoma*, **2019**, Article ID: 1960593. https://doi.org/10.1155/2019/1960593

[5] Neuville, A., Collin, F., Bruneval, P., Parrens, M., Thivolet, F., Gomez-Brouchet, A., Terrier, P., de Montpreville, V.T., Le Gall, F., Hostein, I., Lagarde, P., Chibon, F. and Coindre, J.-M. (2014) Intimal Sarcoma Is the Most Frequent Primary Cardiac

Sarcoma. *American Journal of Surgical Pathology*, **38**, 461-469. https://doi.org/10.1097/PAS.0000000000000184

[6] Paraskevaidis, I.A., Michalakeas, C.A., Papadopoulos, C.H. and Anastasiou-Nana, M. (2011) Cardiac Tumors. *ISRN Oncology*, **2011**, Article ID: 208929. https://doi.org/10.5402/2011/208929

[7] Moeri-Schimmel, R., Pras, E., Desar, I., Krol, S. and Braam, P. (2020) Primary Sarcoma of the Heart: Case Report and Literature Review. *Journal of Cardiothoracic Surgery*, **15**, Article No. 104. https://doi.org/10.1186/s13019-020-01157-4

[8] Gupta, R., Meghrajani, V., Desai, R. and Gupta, N. (2020) Primary Malignant Cardiac Tumors: A Rare Disease with an Adventurous Journey. *Journal of the American Heart Association*, **9**. https://doi.org/10.1161/JAHA.120.016032

[9] Romanowska, A., Lewicka, E., Sławiński, G., Jankowska, H. and Zaucha, R. (2021) Case Report: Adjuvant Radiotherapy Can Be an Effective Treatment for Intimal Sarcoma of the Heart. *Frontiers in Oncology*, **11**, Article 621289. https://doi.org/10.3389/fonc.2021.621289

[10] Hammami, M.B. (2021) Incidence, Prognostic Significance and Survival Outcomes of Primary Cardiac Sarcoma: An Updated Population-Based Retrospective Study. *The Anatolian Journal of Cardiology*, **25**, 104-110. https://doi.org/10.14744/AnatolJCardiol.2020.78107

[11] Ibrahim, A., Luk, A., Singhal, P., Wan, B., Zavodni, A., Cusimano, R.J. and Butany, J. (2013) Primary Intimal (Spindle Cell) Sarcoma of the Heart: A Case Report and Review of the Literature. *Case Reports in Medicine*, **2013**, Article ID: 461815. https://doi.org/10.1155/2013/461815

[12] Nakagawa-Kamiya, T., Mori, M., Ohira, M., Iino, K., Kawashiri, M., Takemura, H. and Takamura, M. (2021) Intimal Sarcoma: An Extremely Rare Case of a Left Atrial Tumor with Partial Obstruction of the Mitral Orifice. *CASE*, **5**, 93-96. https://doi.org/10.1016/j.case.2020.12.009

[13] Carabello, B.A. (2005) Modern Management of Mitral Stenosis. *Circulation*, **112**, 432-437. https://doi.org/10.1161/CIRCULATIONAHA.104.532498

[14] Moise, B., Rosca, M., Alexandru, D. and Ginghina, C. (2017) Left Atrial Remodelling in Patients with Mitral Stenosis. *Romanian Journal of Cardiology*, **27**, 170-176.