Left Ventricular Metastasis presenting 15 years after excision of Right Thigh Myxoid Liposarcoma

Sanchit Kundal[1], Hong Yu Wang[1], Samuel Tan[1], Wenchy Tan[1], Maria Riasat[1], and Misra Deepika[1]

[1]Mount Sinai Beth Israel Hospital

March 12, 2024

Title Page

Left Ventricular Metastasis presenting 15 years after excision of Right Thigh Myxoid Liposarcoma

[Author: Sanchit V. Kundal[1], M.D., Hong Yu Wang[2], M.D., Samuel Tan[2], Wenchy Tan[2], Maria Riasat[2], M.D., Deepika Misra[1], M.D.]

Institutional Affiliations [1. Department of Cardiology, Icahn School of Medicine at Mount Sinai, Mount Sinai Beth Israel 2. Department of Medicine, Icahn School of Medicine at Mount Sinai, Mount Sinai Beth Israel]

Author's contributions:

SK was responsible for concept of the article and drafted and revised the manuscript. HW was responsible for the conception of the article and revised the manuscript. ST, WT, and MR contributed revising of the manuscript. DM contributed to the conception of the article, the drafting and revising of the manuscript.

Conflict of Interest Disclosures: There are no conflicts of interest disclosure for all authors.

Acknowledgement: N/A

Abstract/Introduction

While both primary and secondary cardiac tumors are extremely rare, tumors found in the heart are more likely to be metastatic lesions from other organs. Myxoid liposarcoma is second most common type of liposarcoma in adults, usually presenting in deep tissue of the lower extremities. Myxoid liposarcoma is known to have an unusual tendency for extrapulmonary metastasis. However, the link between myxoid liposarcoma and cardiac metastasis are rare. We report a case of a 49-year-old female with myxoid liposarcoma of the right thigh with metastasis to left ventricle of the heart fifteen years after initial excision.

Keywords : Cardiac tumor, Liposarcoma, Myxoid liposarcoma, Cardiac metastasis

Abbreviations :

Transthoracic echocardiography (TTE)

Cardiac magnetic resonance imaging (MRI)

Positron emission tomography–computed tomography (PET/CT)

Case Presentation/Examination

49-year-old female with past medical history of hypertension, tobacco and marijuana use, presented to the ER with complaints of intermittent palpitations for 2 weeks. On the day of presentation the palpitations were more frequent and associated with new onset shortness of breath along with lightheadedness. Initial physical examination and vital signs were unremarkable. EKG showed sinus rhythm, premature ventricular contractions with T-wave inversion in leads V3-V6 and II, III, aVF.

Patient reported being diagnosed with sarcoma of right thigh in May 2007, and its subsequent removal in August 2007. She followed up with oncology afterwards and remained in remission. Her clinical history also relevant for hypertension, tobacco and marijuana smoking and gastro-esophageal reflux disease. Patient denied any significant family history of cardio-pulmonary disease.

Methods

Differential Diagnosis

Given patient's history of sarcoma of the right thigh, neoplastic process was considered. The differential diagnosis were primary benign tumors, primary malignant tumors, and secondary distant metastasis cardiac tumors.

Investigations

Initial Laboratory workup and chest x-ray were unremarkable (Table 1). TTE demonstrated a 4.16 x 2.5 cm mass attached to the apical septum of the left ventricle with echogenicity similar to the myocardium and smooth borders which partially enhanced with perflutren sonographic contrast (Figure 1).

Cardiac magnetic resonance imaging (MRI) was done to further evaluate and characterize the mass. Cardiac MRI revealed an apical mass of the left ventricle measuring approximately 4 cm with probable extension outside of the expected left ventricular cavity with imaging features most compatible with soft tissue tumor, potentially containing internal fat (figure 2). A Positron Emission Tomography–Computed Tomography (PET/CT) demonstrated a mixed density mass in the apical region of the left ventricle with mildly increased FDG uptake with SUV max of 2.4, with the size of the mass measuring 2.6 x 2.4 x 3.7 cm (figure 3). It also demonstrated markedly enlarged fibroid uterus with focal intense radiotracer uptake. A subsequent pelvic MRI confirmed the numerous large uterine fibroids.

Treatment

The patient underwent an incisional biopsy with intra-operative findings demonstrating tumor burden extending through the chamber wall to the external surface of heart, including invasion into septum. Frozen section demonstrated a low-grade neoplasm suspicious for myxoid liposarcoma. Cytopathology later confirmed a low-grade myxoid neoplasm which was consistent with metastatic liposarcoma (Figure 4).

Conclusion and Results

Outcome and Follow-up

Due to the widespread local disease, complete excision of the mass was not possible. Patient was discharged on metoprolol succinate 25 mg and amiodarone 200 mg daily and was scheduled to follow up with oncology and cardiology.

Discussion

Primary Cardiac tumors are extremely rare. Comparatively, metastatic cardiac tumors are far more common than primary cardiac tumors.[1] Sarcomas are malignant tumors that are of mesenchymal origin, comprising less than 1 percent of malignancies in adult.[2] Liposarcomas are malignancies of soft tissue and have been described into three main subtypes: well-differentiated/dedifferentiated, myxoid/round cell, and pleomorphic.[3] The myxoid liposarcoma is the second most common subtype of liposarcoma and account for approximately 10% of all adult soft tissue sarcomas.[3,4,5]

Myxoid liposarcoma is the second most common subtype of liposarcoma. It is characterized by a recurrent translocation (12;16)(q13;p11) that results in FUS-CHOP gene fusion, which occurs in more than 95% of cases. In other rare cases, a similar variant has been characterized (12;22)(q13;q12) which fuses EWSR1 to DDIT 3. These translocations are believed to be the primary oncogenic event stimulating the proliferation of tumor cells of myxoid liposarcoma.[3] Myxoid liposarcomas can arise in any part of the body but have a propensity to occur in the lower extremities and buttocks. They have a peak incidence in patients in their fourth and fifth decades of life.[6,7] Myxoid liposarcoma also known to have unusual metastasize to extrapulmonary sites such as retroperitoneum, intra-abdominal cavity, and bone.[5,6,7]

Aforementioned case describes the primary tumor for the patient was found in right thigh. Myxoid liposarcoma is also known to have unusual metastasize extrapulmonary sites such as the retroperitoneum, intra-abdominal cavity, and bone.[5,6,7] Cardiac metastases of myxoid liposarcoma are extremely rare, with approximately thirty cases reported previously.[9] The time interval between the primary lesion and cardiac metastasis is relatively long, ranging from 1 to 25 years.[8,9] Our patient's primary tumor presented in 2007, and the cardiac metastasis presented 15 years after the onset of primary tumor.

The symptoms of cardiac tumors are determined by the location of the tumor in the heart. Only 10% of patients with cardiac metastasis have been reported symptoms.[10, 12] Left ventricle tumors in particular may present with arrhythmias or conduction defects. If intracavitary, the tumor can cause outflow obstruction, syncope, and left ventricular failure.[12] Our patient presented with cardiac arrhythmia and symptoms of frequent palpitations.

Furthermore, the diagnostic workup for secondary cardiac tumors is generally aimed to ascertain whether or not cardiac tumor is present. Patients with a known history of malignancy that manifest a change in their clinical status associated with arrythmias or murmurs may warrant further investigations.[12] TTE and cardiac MRI was used in our case as part of the initial investigation and diagnosis. These imaging studies provided critical information regarding location, tumor homogeneity and relation to surrounding structures. As cardiac metastases generally represent widespread disease, FDG-PET/CT was also used in the staging workup of liposarcoma to detect distant metastases.

The treatment of cardiac metastasis is very challenging. Surgery is an option, and it has been shown to improve symptom palliation and prolong life.[11] However, in our patient, because the widespread local disease, the tumor was not amenable to resection. The adjuvant chemotherapy and radiotherapy for secondary cardiac tumor are avenues still in need of further investigation.

Little in-depth investigation exists at this time regarding cardiac metastasis. This is in part due to the rarity of the cases. Currently there is a lack of screening regimens, and targeted therapy for sarcoma and cardiac metastases has not been rigorously studied. However, because of the severity of the conditions and the high potential for morbidity and mortality associated with these lesions, we hope to contribute data to allow further investigation into targeted screening of cardiac metastasis leading to earlier diagnosis, treatment and hopefully a better outcome.

g. References

1. Lam KY, Dickens P, Chan AC. Tumors of the heart. A 20-year experience with a review of 12,485 consecutive autopsies. Arch Pathol Lab Med. 1993;117(10):1027-1031.
2. Siegel RL, Miller KD, Fuchs HE, Jemal A. Cancer statistics, 2022. CA Cancer J Clin. 2022 Jan;72(1):7-33. doi: 10.3322/caac.21708. Epub 2022 Jan 12. PMID: 35020204.
3. Conyers R, Young S, Thomas DM. Liposarcoma: molecular genetics and therapeutics. *Sarcoma* . 2011;2011:483154. doi:10.1155/2011/483154
4. Henze J, Bauer S. Liposarcomas. Hematol Oncol Clin North Am. 2013;27(5):939-955. doi:10.1016/j.hoc.2013.07.010
5. Nishida Y, Tsukushi S, Nakashima H, Ishiguro N. Clinicopathologic prognostic factors of pure myxoid liposarcoma of the extremities and trunk wall. Clin Orthop Relat Res. 2010;468(11):3041-3046. doi:10.1007/s11999-010-1396-3

6. Moreau LC, Turcotte R, Ferguson P, et al. Myxoid\round cell liposarcoma (MRCLS) revisited: an analysis of 418 primarily managed cases. Ann Surg Oncol. 2012;19(4):1081-1088. doi:10.1245/s10434-011-2127-z
7. Estourgie SH, Nielsen GP, Ott MJ. Metastatic patterns of extremity myxoid liposarcoma and their outcome. J Surg Oncol. 2002;80(2):89-93. doi:10.1002/jso.10093
8. Silvestri F, Bussani R, Pavletic N, Mannone T. Metastases of the heart and pericardium. G Ital Cardiol. 1997;27(12):1252-1255.
9. Ikuta K, Sakai T, Koike H, Okada T, Imagama S, Nishida Y. Cardiac metastases from primary myxoid liposarcoma of the thigh: a case report. World J Surg Oncol. 2020;18(1):227. 2020. doi:10.1186/s12957-020-02009-0
10. Xu G, Shi X, Shao G. An unusual case of metastasis of a pulmonary undifferentiated pleomorphic sarcoma to the right ventricle: a case report. J Med Case Rep. 2013;7:165. Published 2013 Jun 27. doi:10.1186/1752-1947-7-165
11. Messner G, Harting MT, Russo P, et al. Surgical management of metastatic melanoma to the ventricle. Tex Heart Inst J. 2003;30(3):218-220.
12. Coller JM, Parente P, Esmore D, et al. "Large left ventricular metastasis causing left ventricular outflow tract obstruction and haemolysis". *Eur J Echocardiogr* . 2009;10(3):456-458. doi:10.1093/ejechocard/jen340

Hosted file

`MLS_Figures.docx` available at https://authorea.com/users/754203/articles/724147-left-ventricular-metastasis-presenting-15-years-after-excision-of-right-thigh-myxoid-liposarcoma

Hosted file

`Table 1.docx` available at https://authorea.com/users/754203/articles/724147-left-ventricular-metastasis-presenting-15-years-after-excision-of-right-thigh-myxoid-liposarcoma