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Intimal Sarcoma in the Left Atrium: A Rare Cause of Subacute Heart Failure

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Abstract

Background: Primary cardiac sarcomas are rare, highly aggressive tumors with nonspecific clinical presentations, making early diagnosis challenging. Intimal sarcoma, a subtype of undifferentiated sarcoma, is characterized by high metastatic potential and poor prognosis. **Case Summary:** A 23-year-old woman with no prior medical history presented to the emergency department with a 20-day history of progressive palpitations and dyspnea. Initial evaluation revealed tachycardia, hypotension, and a systolic mitral murmur. Laboratory tests demonstrated elevated D-dimer and high-sensitivity troponin I levels. Pulmonary thromboembolism was initially suspected; however, chest CT angiography revealed a left atrial mass, subsequently confirmed by transthoracic echocardiography as a heterogeneous mass causing dynamic mitral obstruction and moderate eccentric mitral regurgitation, with associated right ventricular dysfunction. The patient underwent emergency surgical resection, including removal of the left atrial appendage and partial atrial roof, followed by reconstruction of the interatrial septum. Histopathology confirmed high-grade intimal sarcoma with MDM2 overexpression. Postoperative management included adjuvant chemotherapy (Doxorubicin and Ifosfamide) and planned radiotherapy. Despite early intervention, follow-up MRI five months later revealed cerebral metastases, necessitating neurosurgical resection. **Discussion and Conclusion:** This case highlights the rapid progression and aggressive nature of cardiac intimal sarcoma, even in young patients with timely diagnosis and multidisciplinary care. Early recognition, prompt surgical intervention, and coordinated postoperative management remain critical to optimizing outcomes in this rare and high-risk disease.

Keywords: Sarcoma; Heart failure; Heart atria

Introduction

Primary cardiac neoplasms are rare, with an incidence of 1.38 per 100 million people [1,2]. Malignancy occurs in up to 10% of cases and is more common in young adults, regardless of sex [1,3].

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Sarcoma is the most frequently reported primary malignant cardiac tumor reported in the literature, accounting for approximately 65% of cases. Clinical presentation can be asymptomatic (incidental finding) or non-specific, with symptoms such as dyspnea, palpitations, chest discomfort, or syncope [1,3,4]. Prognosis is poor, with an average survival of 3-12 months after diagnosis, and fewer than 15% of patients surviving beyond five years [3,5,6].

Intimal sarcoma is a subtype of undifferentiated sarcoma arising in the heart or the intimal layer of the great vessels. Its aggressiveness is attributed to delayed diagnosis and high metastatic potential. Overexpression of the MDM2 gene is a key contributing factor [2,7].

Surgical resection is typically indicated and may improve prognosis, particularly in the absence of metastasis. Combined treatment with chemotherapy and radiotherapy can reduce the risk of recurrence, which is common [3-5].

Case Presentation

Patient Information, History of Present Illness and Physical Examination:

A 23-year-old woman with no known comorbidities presented to the emergency department complaining of palpitations and dyspnea that had started approximately 20 days earlier, with progressive worsening.

On initial assessment, she reported dyspnea even at rest and persistent palpitations. Physical examination revealed tachycardia (116 bpm) and hypotension (90/70 mmHg). Cardiac auscultation showed a regular rhythm with normal heart sounds and a systolic murmur at the mitral focus graded 4/6, radiating to the left midaxillary line. Pulmonary auscultation revealed vesicular breath sounds bilaterally, without adventitious sounds.

Investigations and Differential Diagnosis:

Among the initial complementary tests, the electrocardiogram showed sinus rhythm at 125–130 bpm, without repolarization abnormalities, and a possible S1Q3T3 pattern. Laboratory tests revealed elevated D-dimer (5.11 µg FEU/mL) and high-sensitivity troponin I levels, measured in two serial samples one hour apart (1.5 ng/mL and 1.39 ng/mL), both values approximately ten times above the upper reference limit. Point-of-care ultrasound was not performed at this stage because it was unavailable. With pulmonary thromboembolism as the leading diagnostic hypothesis, a chest CT angiography was performed (Figure 1). The report ruled out embolism but revealed an image in the left atrium measuring approximately 60 mm in its longest axis, suggestive of a thrombus.

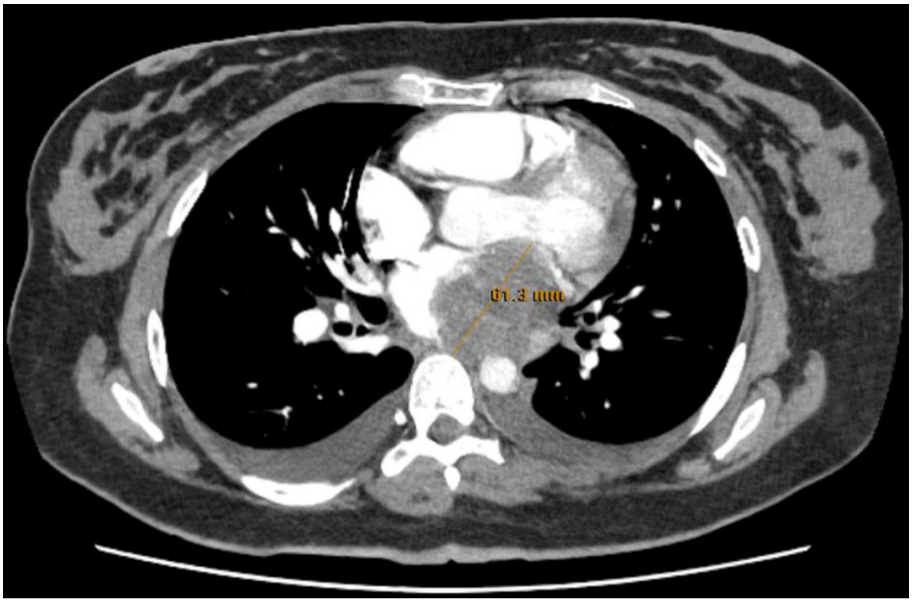

Figure 1: Axial section of chest CT angiography showing a mass in the left atrium.

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A transthoracic echocardiogram (Figure 2) was performed for further evaluation the CT finding. It revealed significant left atrial enlargement (indexed volume 49 mL/m²) and a heterogeneous mass with regular borders in close contact with the posterior leaflet and lateral wall of the left atrium, measuring 62 × 34 mm. This mass caused dynamic obstruction (mean gradient 19 mmHg) and moderate eccentric mitral regurgitation on Doppler, directed toward the interatrial septum. Right ventricular enlargement with systolic dysfunction was also noted (TAPSE: 15 mm — RV ≥ 17 mm; FAC: 24% — RV ≥ 35%), with a positive McConnell sign (hypokinesia of the mid and basal segments with preserved apical motion of the right ventricular free wall). Pulmonary artery systolic pressure was estimated at 75 mmHg. The possibility of a cardiac tumor was raised.

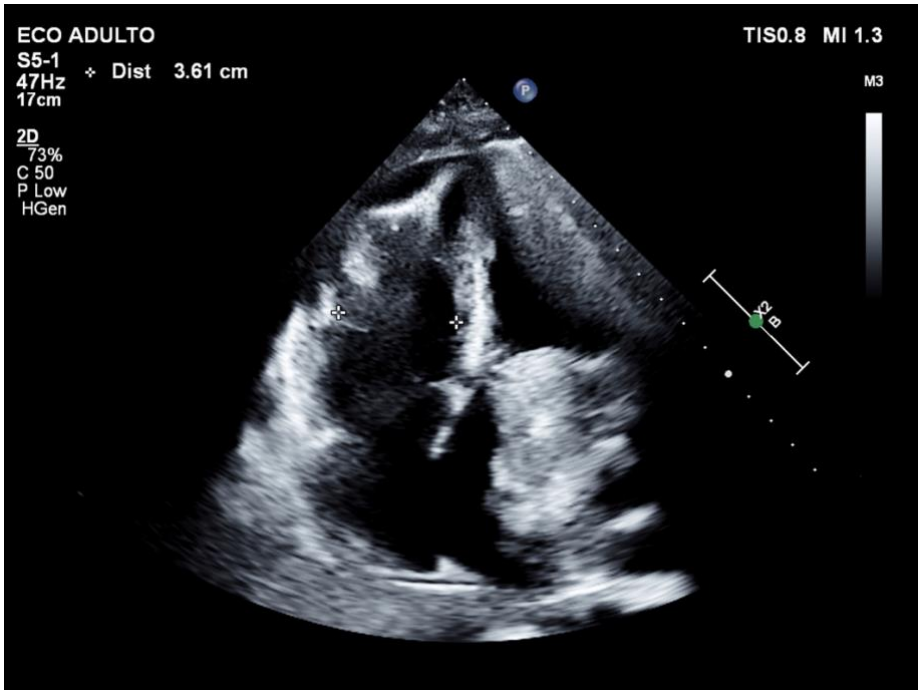

Figure 2: Transthoracic echocardiogram, apical four-chamber view showing a mass in the left atrium.

Intervention / Treatment:

The patient was referred to the cardiac surgery team for evaluation and underwent emergency surgical resection. Cardiac magnetic resonance imaging was considered, but the patient's clinical condition did not allow the examination. Moreover, surgery was indicated due to the hemodynamic impact of the mass obstruction, regardless of its invasive nature.

Intraoperatively, a heterogeneous mass with regular borders was found in the left atrium, occupying most of the cavity and adherent to the posterior and lateral walls. The mass protruded into the left ventricular inflow tract, maintaining contact with the atrial aspect of the mitral valve leaflets and causing obstruction to mitral inflow (mean diastolic gradient 19 mmHg on intraoperative echocardiography). There was no attachment to the atrial septum, but points of adherence were observed along the posterior aspect of the left atrium, in continuity with the right inferior pulmonary vein. Pulmonary artery systolic pressure was estimated at 35 mmHg. The left atrial appendage and a segment of the atrial roof were resected to allow complete tumor removal (Figure 3), followed by reconstruction of the interatrial septum. The procedure was completed without complications. The surgical margin status was R1, indicating microscopic residual tumor.

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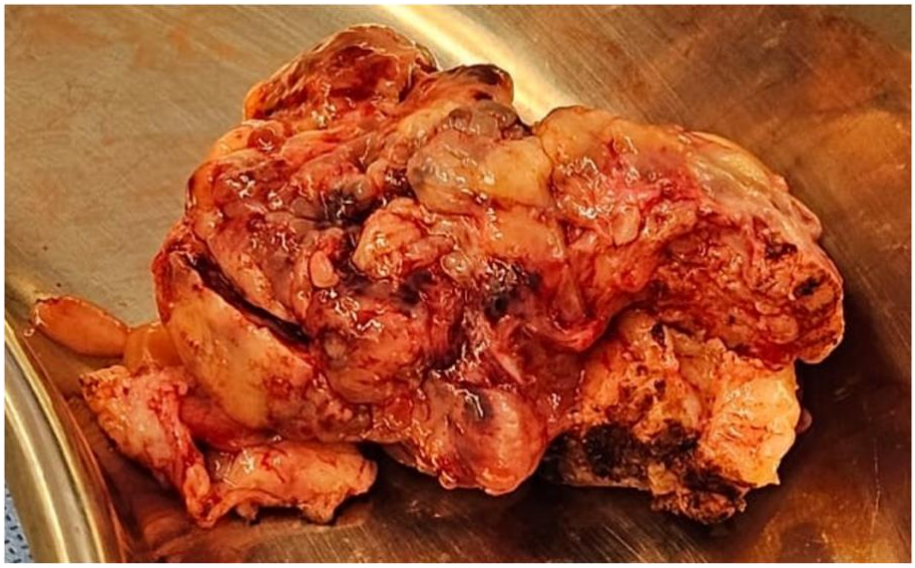

Figure 3: Excised surgical specimen from the left atrium.

Histopathology revealed a high-grade spindle cell sarcoma (Grade 3, National Federation of Cancer Centers [NFCC] classification), with MDM2 gene overexpression, consistent with intimal sarcoma (Figure 4). Confirmation of MDM2 status by FISH was requested but not performed due to financial constraints. Ki-67 was positive in about 40% of the neoplastic cells. Given the early stage and preserved functional status, the oncology team recommended adjuvant chemotherapy (Doxorubicin and Ifosfamide) followed by radiotherapy.

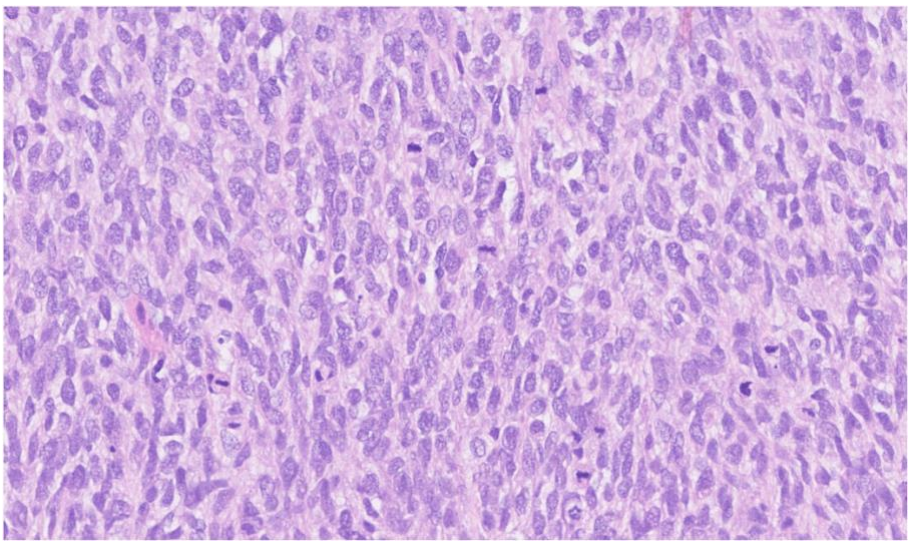

Figure 4: High-grade spindle cell sarcoma with numerous mitoses (H&E stain, magnification ×40).

Follow-Up:

The patient was discharged 20 days postoperatively and remained under outpatient follow-up, showing good adherence to adjuvant therapy and symptomatic improvement. Approximately five months after diagnosis, a screening brain MRI revealed supra- and infratentorial intra-axial lesions, the largest located in the left cerebellar hemisphere and right frontal lobe, consistent with metastatic disease. The patient underwent surgical resection of the left cerebellar and right fronto-parieto-temporal lesions.

This case highlights the aggressiveness of high-grade cardiac sarcoma, demonstrated by early central nervous system metastases within months of initial presentation.

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Discussion

This case is consistent with the literature regarding the patient's age, nonspecific clinical presentation, and the primary malignant cardiac tumor identified by histopathology.

Symptoms such as dyspnea and palpitations in the emergency department generate a broad differential diagnosis. The initial clinical suspicion of pulmonary thromboembolism was appropriate, supported by elevated D-dimer and troponin levels.

The key turning point was chest CT angiography, which excluded pulmonary embolism but revealed a left atrial mass, subsequently better characterized by transthoracic echocardiography.

Timely diagnosis and rapid intervention are critical in modifying the natural course of this highly aggressive disease. In this case, the interval between diagnosis and surgical intervention was only 24 hours, and the integration among care teams - from the patient's arrival at the emergency department to the operation - demonstrated exemplary coordination and efficiency in patient management.

Conclusions

Cardiac sarcoma is a rare and aggressive disease, often presenting with absent or nonspecific symptoms, which makes early diagnosis challenging. Surgical resection combined with adjuvant chemotherapy and/or radiotherapy remains the mainstay of treatment. Despite timely intervention, prognosis is generally poor, with average survival rarely exceeding 12 months, underscoring the importance of rapid diagnosis and coordinated multidisciplinary care.

Acknowledgements: To the patients, the reason of our being; especially to the patient of the history presented in this case report.

Conflict of Interest Statement: The authors declare that they have no conflicts of interest regarding the publication of this case report.

Patient Consent Statement: Written informed consent was obtained from the patient for the publication of this case report and any accompanying images.

REFERENCES

1. Tyebally S, Chen D, Bhattacharyya S, et al. Cardiac Tumors: JACC Cardio Oncology State-of-the-Art Review. JACC Cardio Oncol. 2020; 16: 293-311. doi: 10.1016/j.jaccao.2020.05.009.
2. Diamond JE, Mi MY, VanderLaan PA, et al. An Unusual Cause of Functional Mitral Stenosis: A Left Atrial Intimal Sarcoma. JACC Case Rep. 2021; 3: 829-833. doi: 10.1016/j.jaccas.2021.02.043.
3. Siontis BL, Leja M, Chugh R. Current Clinical Management of Primary Cardiac Sarcoma, Expert Review of Anticancer Therapy. 2020; 20: 45-51. doi: 10.1080/14737140.2020.1711738.
4. Santos RR, Abecasis J, Gomes DA, et al. Um Achado Acidental de um Sarcoma Cardíaco. Arq. Bras. Cardiol. 2022; 119: 349-352. doi: 10.36660/abc.20210703.
5. Butany J, Nair V, Naseemuddin A, et al. Cardiac Tumours: Diagnosis and Management. Lancet Oncol. 2005; 6: 219-228. doi: 10.1016/S1470-2045(05)70093-0.
6. Magarakis M, Salerno TA. Commentary: Cardiac Sarcoma-Can we win this battle? J Thorac Cardiovasc Surg. 2021; 162: 118-119. doi: 10.1016/j.jtcvs.2020.01.033.
7. Reynoso-Hermosillo M, Sandoval-García J, López-Rosales B, et al. Malignant Primary Tumor in Left Atrium with Mitral Valve Invasion, Pulmonary Veins and Cerebral Metastasis. A Case Report. Cir Cir. 2020; 88: 91-94. doi: 10.24875/CIRU.19000909.

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