Biomedicine Advances

Biomedicine Advances. 2025;2(4):207-209
doi: 10.34172/bma.34
https://biomedad.ae

Case Report

High-grade undifferentiated pleomorphic cardiac sarcoma extending from the interatrial septum to both atria and the anterior mitral valve annulus

Abidin Emre Tirnaksiz[1*], Arda Guler[1], Ersin Kadirogullari[2], Sinem Aydın[3], Gonca Gul Gecmen[4], Gamze Babur Guler[1]

[1]Department of Cardiology, University of Health Sciences Istanbul Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training and Research Hospital, Istanbul, Turkey
[2]Department of Cardiovascular Surgery, University of Health Sciences Istanbul Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training and Research Hospital, Istanbul, Turkey
[3]Department of Radiology, University of Health Sciences Istanbul Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training and Research Hospital, Istanbul, Turkey
[4]Department of Pathology, Kartal Dr. Lutfi Kirdar City Hospital, Istanbul, Turkey

*Corresponding Author: Abidin Emre Tirnaksiz, Email: Emretirnaksiz@gmail.com

Summary
Cardiac undifferentiated pleomorphic sarcomas are rare primary malignant tumors of the heart and constitute a significant portion of cardiac sarcomas. We present the diagnostic approach in a case of high-grade undifferentiated pleomorphic cardiac sarcoma with an unusual location in a 70-year-old woman.
Keywords: Cardiac sarcoma, Undifferentiated pleomorphic sarcoma, Mitral valve annulus

Received: August 1, 2025, **Revised:** September 6, 2025, **Accepted:** September 26, 2025, **ePublished:** October 20, 2025

Introduction

Cardiac undifferentiated pleomorphic sarcomas most commonly arise from the left atrium, with less frequent involvement of other cardiac chambers.[1,2] Within the left atrium, the posterior or lateral wall is typically affected, while the left atrial appendage and mitral valve are less commonly involved. Rarely, these tumors originate from the left atrial septum and can fill the entire left atrial chamber.[1]

Case Presentation

A 70- year-old female who presented with progressive dyspnea and recurrent paroxysmal atrial fibrillation. Physical examination revealed no remarkable abnormalities. Transthoracic echocardiography revealed a hyperechoic, well-circumscribed mass with components in both the right and left atria, extending from the interatrial septum to the anterior leaflet of the mitral valve (Supplementary file 1, Video 1). Transesophageal echocardiography confirmed the mass's extension to the anterior mitral valve annulus (Supplementary file 2, Video2) and provided further anatomical detail with X-plane imaging (Supplementary file 3, Video 3).

Thoracic multislice computed tomography showed a heterogeneous, hypodense, solid lesion originating from the interatrial septum, extending into the mitral-aortic intervalvular fibrosa and projecting into the atrial cavity on the atrial side of the anterior leaflet of the mitral valve, forming a lobulated mass. The component in the right atrial fossa ovalis was 29×15 mm, the component in the left atrium was 25x23 mm, and the total dimensions were 45×25×12 mm (Figure 1A). In cardiac magnetic resonance imaging, the mass lesion was iso-mildly hyperintense in T1A series and hyperintense in T2A series. Mild heterogeneous contrast enhancement was observed in contrast enhanced series (Figure 1B).

After performing preoperative preparations of the patient and providing required pre-surgical patient instructions, he was submitted to median sternotomy under general anesthesia and right atriotomy under cardiopulmonary bypass following implementation of aorto-bicaval cannulation. A mass extending from the interatrial septum into the right atrium was observed and the septum was opened by removing the mass. The mass extended from the anterior annulus of the mitral valve to include the aortomitral junction, and the mass was removed, paying attention to the surgical margins (Figure 2A). The anterior mitral valve annulus was repaired using a bovine pericardial patch. TEE revealed severe aortic insufficiency. An aortotomy was performed, and a perceval sutureless aortic valve was replaced. Follow-up TEE revealed mild mitral regurgitation, but there was no aortic valve regurgitation. Drains were placed, and the sternotomy was appropriately closed. The patient was transferred to the ICU in stable conditions with low-dose dopamine supplementation. Undifferentiated pleomorphic sarcoma was established as

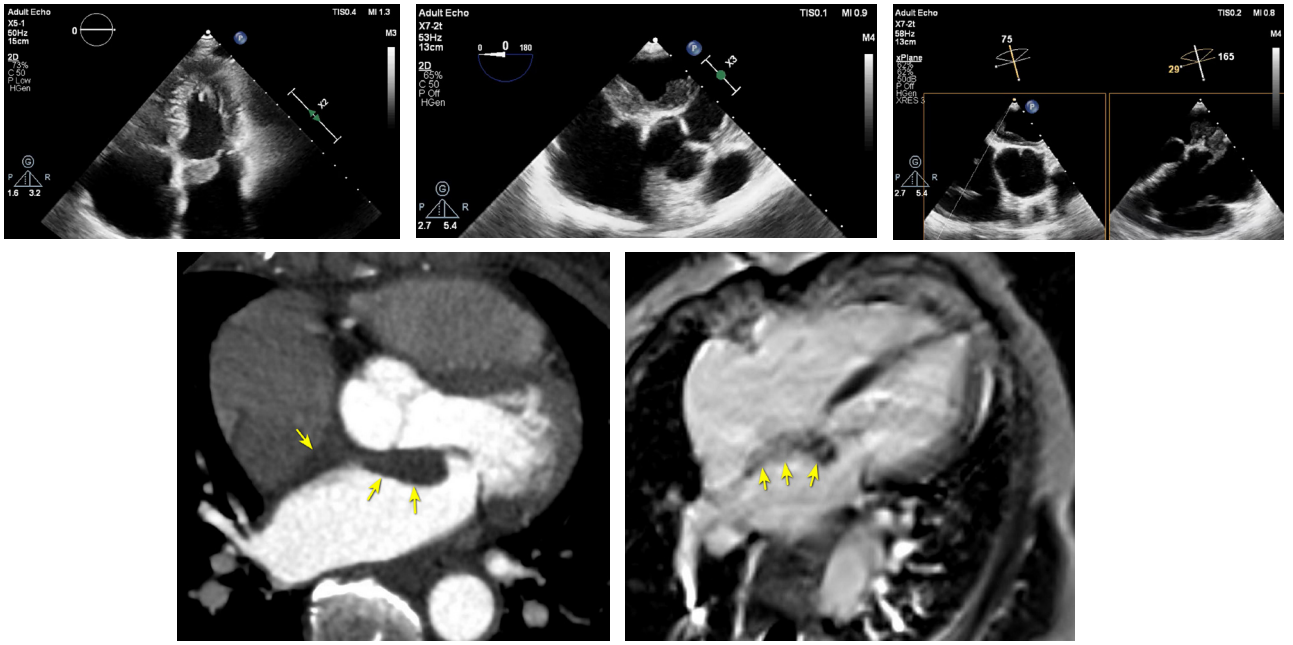

Figure 1. (A) Thoracic multislice computed tomography demonstrating a heterogeneous, hypodense, lobulated mass originating from the interatrial septum, extending into the mitral-aortic intervalvular fibrosa, and projecting into the atrial cavity. **(B)** Cardiac magnetic resonance imaging showing the mass as iso-to mildly hyperintense on T1-weighted imaging and hyperintense on T2-weighted imaging, with mild heterogeneous enhancement on contrast-enhanced sequences

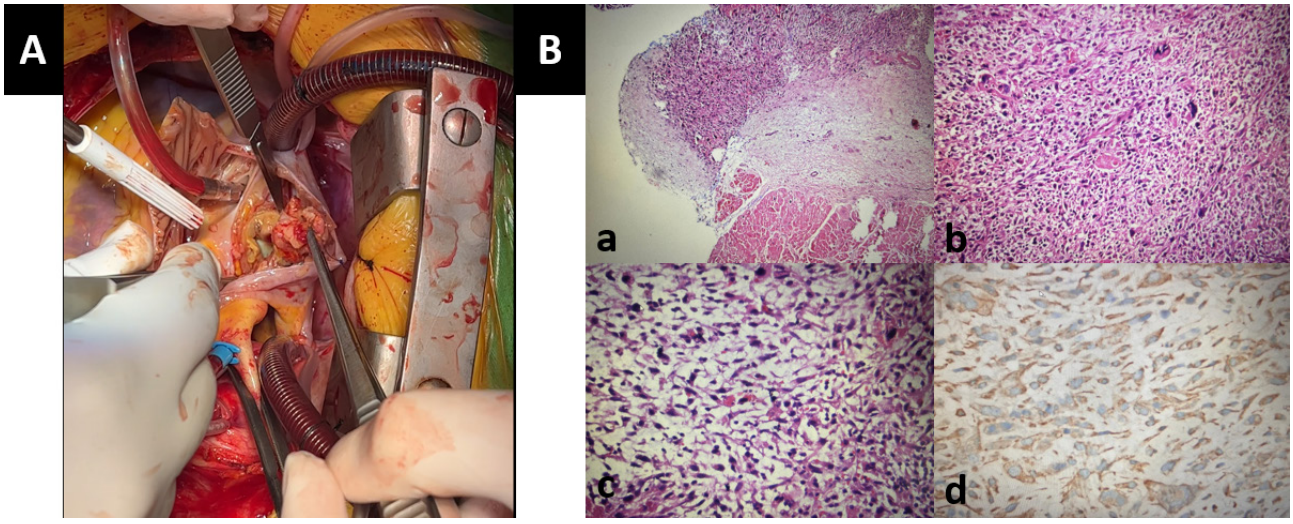

Figure 2. (A) Gross surgical specimen of the resected tumor displaying a lobulated, solid mass. **(B)** Histopathological examination confirming the diagnosis of undifferentiated pleomorphic sarcoma. **a)** Undifferentiated pleomorphic sarcoma with predominant spindled component. There was no invasion into the ventricular wall at the base of the figure H&E X10 **b)** The spindle cells are highly atypical and pleomorphic, there is no significant amount of stroma H&E X20 **c)** This area shows myxoid background H&E X40 **d)** At immunohistochemistry, the neoplastic cells appeared positive for Vimentin

the definitive pathological diagnosis (Figure 2B).

Discussion

Left atrial undifferentiated pleomorphic sarcoma is a rare subtype occurring in 1% of cases.8 Primary cardiac tumors often remain clinically asymptomatic until sequelae of mass effect or nonspecific symptoms (fatigue, fever, weight loss, night sweats) prompt evaluation; consequently, these tumors are a challenge to diagnose and treat.[3-6] These tumors are often mistaken for benign cardiac tumors such as atrial myxomas during initial evaluation because of their similar clinical and imaging presentations; however, undifferentiated pleomorphic sarcoma often presents with multiple masses rather than a single mass.[7,8] Left atrial undifferentiated pleomorphic sarcoma can invade the left atrial and ventricular walls, leading to mitral valve dysfunction and eventual left-sided heart failure with dyspnea, chest pain, or embolic phenomena.[3,6,7]

The diagnosis of left atrial undifferentiated pleomorphic sarcomas is also challenging because of their heterogeneous nature and the lack of specific diagnostic tests. Typically, a combination of imaging studies, including echocardiography, CT scan, MRI, and PET scan are used to detect the presence of a left atrial undifferentiated pleomorphic sarcoma.[4-9] Although echocardiography is often used as a first-line imaging

study for cardiac tumors, it has distinct disadvantages. Limited field of view, the inability to characterize tissue, and factors such as body habitus and patient cooperation limit the utility of echocardiography in the diagnosis and evaluation of cardiac tumors.[9]

Cardiac MRI offers distinct advantages over echocardiography, including excellent spatial resolution, wide field of view, the relationship of the tumor with structures outside the heart, and tissue characterization which allows for identification of cardiac tumors that do not need further evaluation or resection.[9] However, imaging studies may not be specific enough to distinguish between left atrial undifferentiated pleomorphic sarcoma and other cardiac tumors such as myxomas. [3] Specifically, the mass was isointense to the myocardium on T1-weighted imaging and hyperintense to the myocardium on T2-weighted imaging, suggestive of high fluid content and high interstitial content that are most consistent with left atrial myxoma vs other cardiac tumors. The increased T1 relaxation time was likely attributable to a combination of inflammation and fibrosis. Classic findings on multimodality imaging that favor malignancy include uptake of contrast, infiltration and invasion of normal anatomic boundaries, and the presence of necrosis.[8]

Conclusion

Echocardiography is the most readily available noninvasive imaging technique and thus remains the first-line diagnostic test. 3D echocardiography has further enhanced the role of US techniques in the assessment of cardiac masses, particularly in term of anatomic location, morphology, and functional impact. Cardiac CT is a commonly used second-line diagnostic modality for cardiac masses. Several technologic advances, including submillimeter detector arrays, half-scan postprocessing algorithms and ECG gating, have resulted in improved imaging of cardiac structure including cardiac masses. In this context, MRI offers higher temporal resolution and additional tissue characterization, not exposing patients to ionizing radiations. For all these reasons, a good knowledge and a subsequent correct use of multimodality imaging offer a chance to better differentiate between the different types of cardiac masses.

Authors' Contribution

Conceptualization: Arda Guler.
Data curation: Abidin Emre Tirnaksiz.
Formal analysis: Abidin Emre Tirnaksiz, Arda Guler.
Funding acquisition: Gamze Babur Guler.
Investigation: Abidin Emre Tirnaksiz, Arda Guler.
Methodology: Abidin Emre Tirnaksiz, Arda Guler.
Project administration: Gamze Babur Guler.
Resources: Ersin Kadirogullari, Sinem Aydın, Gonca Gul Gecmen.
Supervision: Gamze Babur Guler.
Validation: Abidin Emre Tirnaksiz, Arda Guler.
Visualization: Arda Guler, Ersin Kadirogullari, Sinem Aydın, Gonca Gul Gecmen.
Writing-original draft: Abidin Emre Tirnaksiz.
Writing-review and editing: Abidin Emre Tirnaksiz.

Competing Interests

The authors have no conflicts of interest to declare.

Ethical Approval

Written informed consent was obtained from the participant for anonymized patient information to be published in this article.

Funding

None.

Intelligence Use Disclosure

This article has not utilized artificial intelligence (AI) tools for research and manuscript development, as per the GAMER reporting guideline.

Supplementary Files

Supplementary File 1, Video 1. Transthoracic echocardiography showing a hyperechoic, well-circumscribed mass with components in both the right and left atria, extending from the interatrial septum to the anterior leaflet of the mitral valve."
Supplementary File 2, Video 2. Transesophageal echocardiography showing a hyperechoic mass extending from the interatrial septum to the anterior mitral valve annulus, involving both atrial chambers.
Supplementary File 3, Video 3. Transesophageal echocardiography (TEE) with X-plane imaging demonstrating the intracardiac mass.

References

1. WHO Classification of Tumours Editorial Board. Thoracic Tumours: WHO Classification of Tumours. World Health Organization; 2021.
2. Siontis BL, Zhao L, Leja M, McHugh JB, Shango MM, Baker LH, et al. Primary cardiac sarcoma: a rare, aggressive malignancy with a high propensity for brain metastases. Sarcoma. 2019;2019:1960593. doi: 10.1155/2019/1960593.
3. Ha JW, Kang WC, Chung N, Chang BC, Rim SJ, Kwon JW, et al. Echocardiographic and morphologic characteristics of left atrial myxoma and their relation to systemic embolism. Am J Cardiol. 1999;83(11):1579-82, a8. doi: 10.1016/s0002-9149(99)00156-3.
4. Pinede L, Duhaut P, Loire R. Clinical presentation of left atrial cardiac myxoma. A series of 112 consecutive cases. Medicine (Baltimore). 2001;80(3):159-72. doi: 10.1097/00005792-200105000-00002.
5. Zaragoza-Macias E, Chen MA, Gill EA. Real time three-dimensional echocardiography evaluation of intracardiac masses. Echocardiography. 2012;29(2):207-19. doi: 10.1111/j.1540-8175.2011.01627.x.
6. Sparrow PJ, Kurian JB, Jones TR, Sivananthan MU. MR imaging of cardiac tumors. Radiographics. 2005;25(5):1255-76. doi: 10.1148/rg.255045721.
7. Tyebally S, Chen D, Bhattacharyya S, Mughrabi A, Hussain Z, Manisty C, et al. Cardiac Tumors: JACC CardioOncology State-of-the-Art Review. JACC CardioOncol. 2020;2(2):293-311. doi: 10.1016/j.jaccao.2020.05.009.
8. Scheffel H, Baumueller S, Stolzmann P, Leschka S, Plass A, Alkadhi H, et al. Atrial myxomas and thrombi: comparison of imaging features on CT. AJR Am J Roentgenol. 2009;192(3):639-45. doi: 10.2214/ajr.08.1694.
9. Keeling IM, Oberwalder P, Anelli-Monti M, Schuchlenz H, Demel U, Tilz GP, et al. Cardiac myxomas: 24 years of experience in 49 patients. Eur J Cardiothorac Surg. 2002;22(6):971-7. doi: 10.1016/s1010-7940(02)00592-4.