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Case Report

Early presentation of undifferentiated pleomorphic cardiac sarcoma ☆

Azin Alizadehasl, MD[a], *Hoda Hakimian, MD*[a], *Negar Dokhani, MD*[a], *Hamidreza Pouraliakbar, MD*[b], *Parisa Firoozbakhsh, MD*[c,*]

[a] *Cardio-Oncology Research Center, Rajaie Cardiovascular Medical and Research Center, Iran University of Medical Sciences, Tehran, Iran*

[b] *Rajaie Cardiovascular Medical and Research Center, Iran University of Medical Sciences, Tehran, Iran*

[c] *Cardiac Primary Prevention Research Center, Cardiovascular Diseases Research Institute, Tehran University of Medical Sciences, Tehran, Iran*

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ABSTRACT

Undifferentiated pleomorphic cardiac sarcomas are extremely rare, highly malignant mesenchymal cardiac neoplasms typically presenting in the sixth decade of life. Here, we have reported a rare case of undifferentiated pleomorphic cardiac sarcoma presenting with dyspnea in a young male. On further evaluations, a large, ill-defined, multilobulated mass was observed in left atrium (LA) with encasement of left circumflex artery, attachment to mitral valve annulus, and invasion through LA wall and pericardium.

Abbreviations: CMR, cardiovascular magnetic resonance; CPB, cardiopulmonary bypass; CPK, creatine phosphokinase; CRP, C-reactive protein; CT, computed tomography; ECG, electrocardiogram; ESR, erythrocyte sedimentation rate; IHC, immunohistochemical; LA, left atrium; LAVi, left atrium volume index; LCx, left circumflex; LDH, lactate dehydrogenase; MG, mean pressure gradient; MR, mitral regurgitation; MS, mitral stenosis; MV, mitral valve; PMVL, posterior mitral valve leaflet; TEE, transesophageal echocardiography; TTE, transthoracic echocardiography; WBC, white blood cell.

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* Corresponding author.
E-mail address: pfiroozbakhsh75@gmail.com (P. Firoozbakhsh).

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Introduction

Primary cardiac tumors are a rare entity with an incidence ranging from 0.001%-0.03% [1]. One out of 4 of these primary tumors are malignant, with sarcomas being the most frequent type accounting for approximately 20% of all primary cardiac neoplasms [2]. Undifferentiated pleomorphic cardiac sarcomas are extremely rare endocardial-based polypoid lesions with fibroblastic or myofibroblastic differentiation and marked cellular pleomorphism mostly found in left atrium (LA). They typically present in the sixth decade of life with dyspnea, malaise, chest pain, weight loss, and palpitation [3]. This tumor has an aggressive, locally invasive nature making complete resection unfeasible, leading to poor prognosis and low survival rates [4].

Here, we have reported a rare presentation of undifferentiated pleomorphic cardiac sarcoma with encasement of left circumflex artery (LCx) in a young male.

Case presentation

A 33-year old male without previous medical history presented with a recent-onset dyspnea on exertion function class II started 2 weeks prior to admission. His physical examination and electrocardiogram (ECG) findings were unremarkable. In laboratory tests, he had WBC count of 11200 cells/μl (76% neutrophil, 20.4% lymphocyte, 3.6% mixed monocyte and eosinophil), erythrocyte sedimentation rate (ESR) of 50 mm/hour (normal: <15 mm/hour), C-reactive protein (CRP) level of 39 mg/L (<6 mg/L), lactate dehydrogenase (LDH) level of 578 (<480 IU/L), and creatine phosphokinase (CPK) level of 223 IU/L (25-225 IU/L). The patient's hemoglobin, platelet, renal and thyroid function tests, serum levels of electrolytes and blood sugar were all within the normal range. On transthoracic echocardiography (TTE), he had severe LA enlargement (LAVi=54 mL/m^2), in addition to a large (5.6 × 3 cm) heterogeneous multilobulated mass with irregular borders and necrotic area in LA with invasion to the lateral and posterior LA walls and attachment to the base of posterior mitral valve leaflet (PMVL) leading to significant mitral stenosis (MS) (mean pressure gradient [MG]: 20 mmHg) and moderate eccentric mitral regurgitation (MR). A mild to moderate pericardial effusion with no hemodynamic effect was also detected (Fig. 1). For further evaluation of the cardiac mass, he underwent cardiovascular magnetic resonance imaging (CMR) which demonstrated a large, ill-defined multilobulated, semi-mobile mass with encasement of LCx, attachment to mitral valve annulus, and invasion through LA wall and pericardium leading to mild pericardial effusion (Fig. 2). The CMR findings were most compatible with primary cardiac sarcoma. Coronary CT-angiography also revealed encasement of LCX leading to significant stenosis at the distal portion.

Due to the large LA mass size, its critical location and its interference with mitral leaflet motion leading to significant MS and elevated MG, the patient underwent palliative surgery. Under general anesthesia and through median sternotomy, cardiopulmonary bypass (CPB) was initiated by aortic and bicaval cannulation. The large atrial mass (about 4 × 4 cm) was seen surrounding the external surface of LA's lateral wall adjacent to the auricle. After incising the LA, a large mass was seen attached to the posterior mitral annulus and LA's auricle. Tumor debulking was performed and MR was repaired using suture annuloplasty. The patient was successfully weaned from CPB and postoperative transesophageal echocardiography (TEE) demonstrated the remnant mass (2.2 × 1.2 cm) in the lateral of mitral valve (MV) attached to the base of PMVL. Repaired MV was also seen with normal motion, mild increased gradient (MG = 5 mmHg) and mild MR. The patient's CT scan images before and after palliative surgery are demonstrated in Fig. 3.

Histopathologic and immunohistochemistry (IHC) examination of the excised mass revealed malignant spindle-cell tumor with positive S100, CD68, and Ki67 (in 60%-70% of tumor cells), and negative desmin, Ckit, and Alk1, mostly compatible with undifferentiated pleomorphic sarcoma. The patient's symptoms relieved after surgery, and treatment continued with radiotherapy and chemotherapy using ifosfamide, docetaxel, and epirubicin.

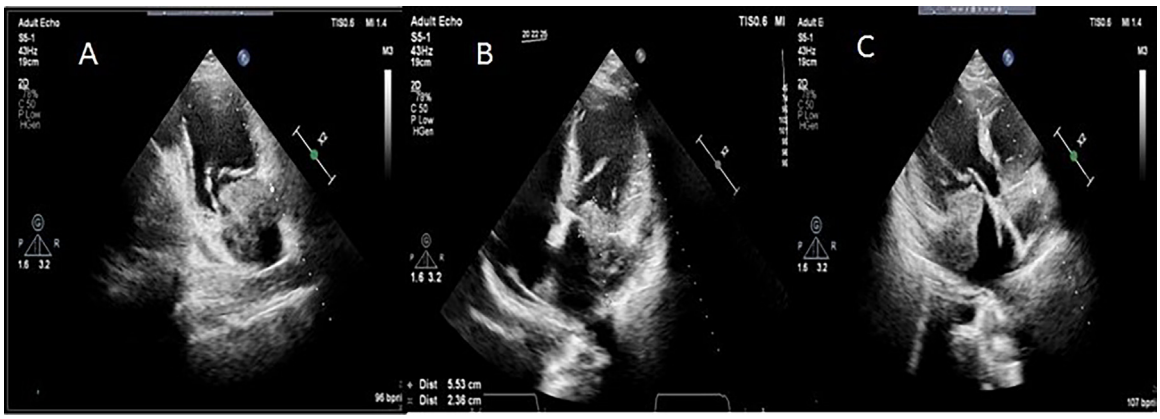

Fig. 1 – Trans-thoracic Echocardiography (TTE) demonstrating: a large, heterogenous, multilobulated, semi-mobile mass with attachment to left atrium's lateral wall and mitral valvue annulus in (A) 2-chamber, (B) 4-chamber, and (C) 3-chamber view.

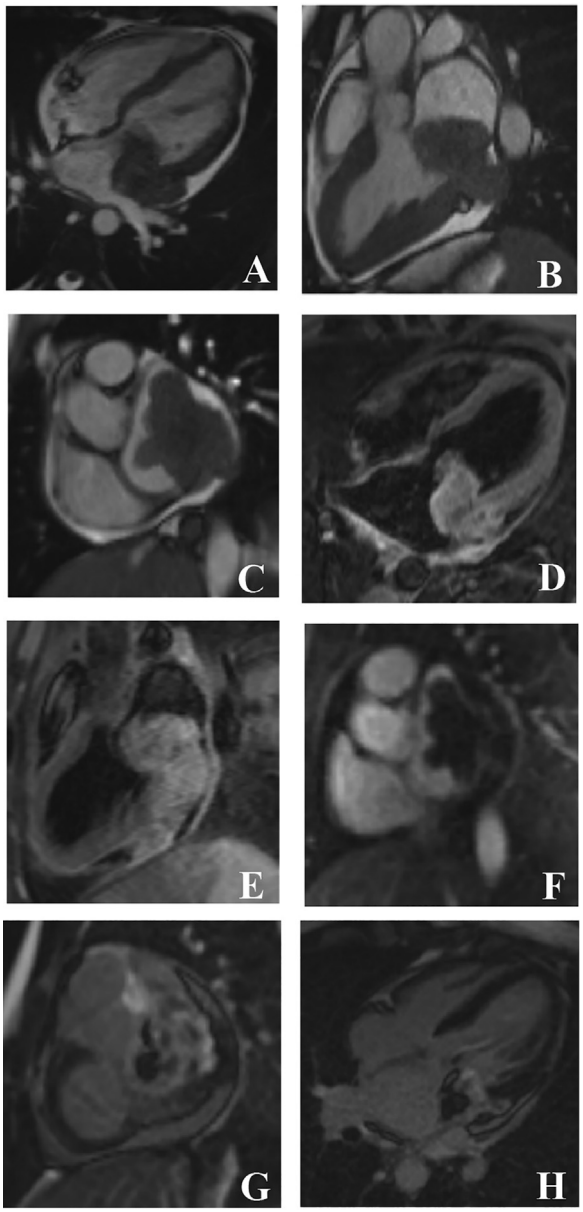

Fig. 2 – Cardiovascular Magnetic Resonance Images demonstrating: (A, B, C) 4-chamber, 3-chamber and short axis SSFP sequences showing iso-hyper signal tumor at the left atrioventricular groove with involvement of LCX and invasion in to the left atrium, base of the left ventricle and posterior mitral valve leaflet. (D, E) 4-chamber and 3-chamber STIR sequence demonstrating high signal T2 tumor. (F) 4-chamber first pass perfusion sequence reveals partially early enhancement of the tumor. (G, H) Short axis and 3-chamber late gadolinium enhancement (LGE) sequence showing heterogeneous enhancement of tumor.

Discussion

Primary cardiac tumors with an incidence rate of 0.017-0.019 are quite rare with one-fourth of them being malignant and 95% of those being malignant reported as sarcomas [5,6]. Undifferentiated pleomorphic cardiac sarcoma, a highly malignant mesenchymal cardiac neoplasm mostly arising from LA, especially from its posterior or lateral wall, accounts for a substantial number of cardiac sarcomas. It usually presents in the sixth decade of life with symptoms including dyspnea, palpitation, heart failure, or constitutional symptoms [3]. It can also cause complications including arrhythmia, embolic events, pericardial effusion, or distant metastases. It is also associated with a very poor prognosis and a median survival of less than a year [3,7]. Diagnosis is confirmed by observing epithelioid or spindle cell morphology in pathologic examination and after rule out of leiomyosarcoma or synovial sarcoma. Detecting a polypoid tumor with absent or minimal myxoid or fibrotic matrix in the LA can also help accurate diagnosis [3].

Cardiac tumors are usually hard to diagnose due to their rarity, nonspecific symptoms, and late presentation. Development of advanced multimodality imaging techniques, including echocardiography (whether trans-thoracic or transesophageal), computed tomography (CT) scan, and CMR have been a great aid in early and proper diagnosis [8]. Transthoracic echocardiography is the modality of choice for initial assessment of intracardiac masses and can properly demonstrate the tumor's location, dimensions, mobility, and involvement of cardiac structure. CMR allows qualitative and quantitative evaluation of the mass and can accurately depict cardiac structure and function, in addition to adjacent structures [9]. In cases of metastatic cardiac lesions, which are far more common than primary tumors, thoracoabdominopelvic CT scan would be helpful in detecting the primary source of the malignancy. CT coronary angiography is another imaging modality that helps to assess involvement of coronary vessels, and extra-cardiac extension of a tumor into adjacent structures. The contrast enhancement can appropriately illustrate the vascular nature of the mass [9,10]. In the current case, the tumor's size, appearance, invasion to adjacent structures, pathologic and immunohistochemical features and imaging characteristics helped us to rule out nonmalignant causes of cardiac masses, including thrombus, and benign neoplasms like cardiac myxoma or inflammatory myofibroblastic tumor [11].

Wide surgical resection is the mainstay treatment for cardiac sarcomas. Although complete tumor resection with negative margins can double the patients' life expectancy, it is not feasible in more than half of the cases [4,8]. Palliative surgery and debulking can lead to symptomatic and hemodynamic relief. Additionally, it can provide sample for histopathologic examination. Adjuvant polychemotherapy and radiotherapy are also widely used in the course of treatment, despite their indefinite beneficial effect [7,12].

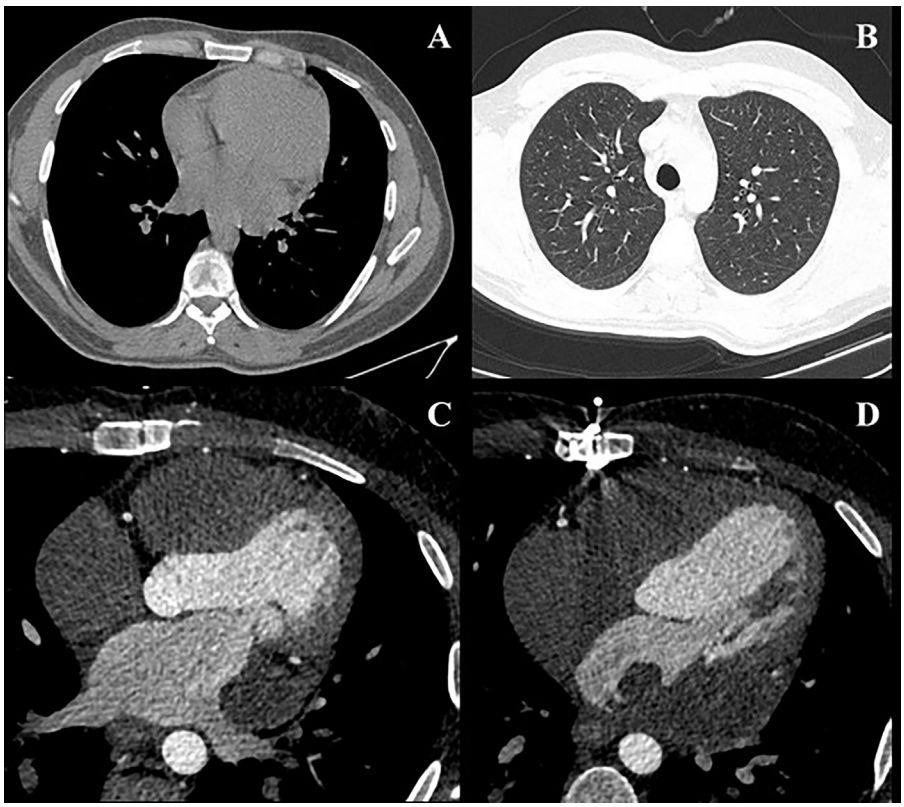

Fig. 3 – (A, B) Thoracic CT scan without IV contrast prior to surgery depicting a tumor in the left atrioventricular groove with encasement of the coronary artery. (C, D) Multidetector computed tomography of the heart with IV contrast, 2 months after surgery, demonstrating an infiltrative tumor in the left atrioventricular groove with invasion into the left atrium and encasement of the coronary artery.

Patient consent

Informed consent was obtained from the current patient to report his case and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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