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## Case Report

# Primary chondroblastic osteosarcoma of the mitral valve without metastasis ☆☆☆

Muhammad Umair, MD<sup>a,\*</sup>, Stefan L. Zimmerman, MD<sup>b</sup>, Elliot K. Fishman, MD<sup>b</sup>

<sup>a</sup>Northwestern University, Feinberg School of Medicine, Department of Radiology, 676 N St Clair St, Chicago, IL 60611, USA

<sup>b</sup>Johns Hopkins Hospital, Russell H. Morgan Department of Radiology and Radiological Science, Baltimore, MD, USA

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## ABSTRACT

**Abstract:** Primary osteosarcomas of the heart are extremely rare entities, with common subtypes including osteoblastic, chondroblastic and fibroblastic subtypes. We describe a case of a 53-year-old female with history of treated breast cancer who presented with progressive dyspnea on exertion. A pulmonary artery protocol CTA demonstrated an anterior mitral leaflet lobulated mass without an osteoid or chondroid matrix. Additional cross-sectional imaging demonstrated no evidence of distant metastasis. The mass was surgically excised with pathology demonstrating a malignant neoplasm with spindle cells, bone, cartilage and rare osteoid, most consistent with a high grade chondroblastic osteosarcoma. After the expected postsurgical recovery, the patient was initiated on adjuvant therapy consisting of ifosfamide and etoposide and is currently disease free for 9 years now. Review of literature demonstrates that cardiac primary osteosarcomas typically involve the left atrium. Imaging usually shows a lobulated or irregular mass with heterogenous attenuation/enhancement and, counterintuitively, a lack of a calcified matrix. Complete surgical excision is challenging leading to poor prognosis, even in cases undergoing post-surgical chemotherapy. Median survival has been reported as about 20 months.

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## Introduction

Primary cardiac osteosarcomas are extremely rare primary tumors arising within heart [1]. The most common histologi-

cal subtypes of skeletal osteosarcomas include osteoblastic, chondroblastic and fibroblastic osteosarcomas; all three have been reported as primary cardiac tumors [1]. The most common site of involvement is the mitral valve [2]. Histopathology

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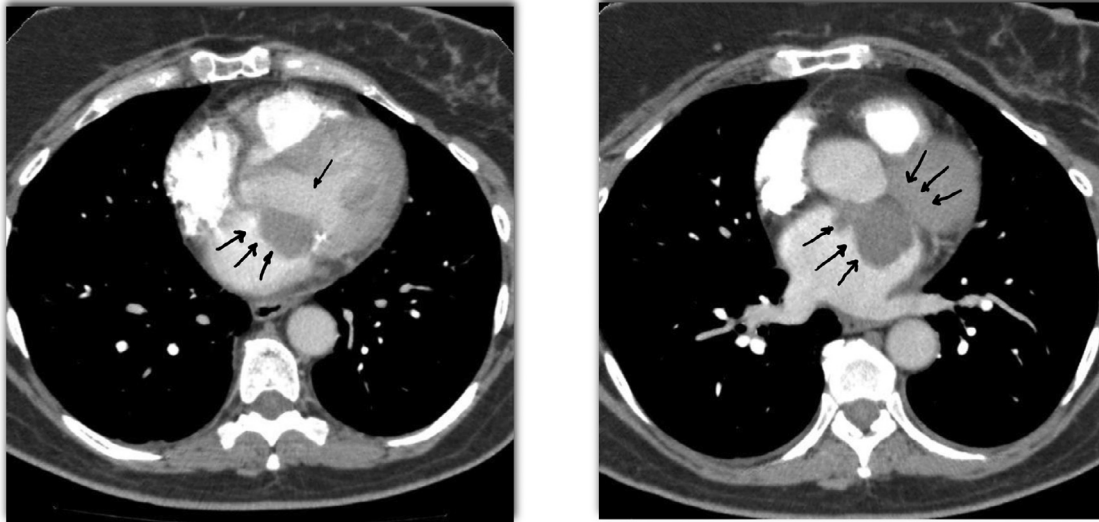
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\* Corresponding author.

E-mail address: [Muhammad.umair@northwestern.edu](mailto:Muhammad.umair@northwestern.edu) (M. Umair).

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**Fig. 1 – CT of chest demonstrates a mildly lobulated hypo-enhancing lesion arising from the anterior leaflet of the mitral valve. This mass was surgically excised to completion and demonstrated a malignant neoplasm with spindle cells, bone, cartilage and rare osteoid, most consistent with a chondroblastic osteosarcoma**

is usually confirmed on surgical specimen as imaging findings are not very specific, and prognosis remains poor due to incomplete resection even with adjuvant chemotherapy [1]. We describe a case of chondroblastic osteosarcoma of the anterior annulus/leaflet of the mitral valve without metastasis in a 53-year-old female.

#### Case description

A 53-year-old female with history of breast cancer, treated with radical mastectomy and chemotherapy at an outside hospital presented with progressive dyspnea on exertion while training for a marathon. The patient began having shortness of breath with minimal activity. The dyspnea progressed to a point where she felt short of breath carrying a small package. She also experienced chest tightness with the most recent episode and presented to the emergency department for treatment. A CTA of chest with pulmonary embolism protocol was performed for further evaluation.

The imaging demonstrated a mildly lobulated mass in the left atrium with epicenter around the anterior annulus and anterior leaflet of the mitral valve (Fig. 1). The mass was hypo-enhancing compared to the myocardium and the skeletal muscles. There was no osteoid or chondroid matrix identified on the CT images. There were no additional masses or other significant findings elsewhere on the CT of chest, and a subsequent CT abdomen, and pelvis with IV contrast. A bone scan was performed, and no sites of pathological radiotracer uptake were seen in the osseous structures to suggest a bone metastasis.

The patient was then taken to the operating room for an on-pump open heart surgical excision of the left atrial mass. A preoperative echocardiogram was performed demonstrating normal left ventricular size and function with EF of 60%-65%. The right ventricle was assessed to be moderately dilated. A 3.9 cm mass was seen arising from the atrial side of the ante-

rior mitral leaflet causing restricted mobility. There was mild mitral regurgitation. Intraoperatively, the mass was excised piecemeal and remaining portion dissected off the atrial wall seeming to shell out along with the surrounding scar tissue. The left atrium did not appear to have any retained tissue in it. The surgical pathology demonstrated a malignant neoplasm with spindle cells, bone, cartilage and rare osteoid. This was deemed most consistent with a high grade chondroblastic osteosarcoma. A post-surgical echocardiogram demonstrated a 60%-65% ejection fraction. The mitral valve was assessed to be bi-leaflet with good mobility, structurally intact, without stenosis and with a mild regurgitation. The right ventricular size and function was assessed to be normal on post-operative echocardiogram. She was discharged from the hospital after recovery from the surgery in a stable condition and was subsequently followed up in oncology office for initiation of adjuvant therapy consisting of ifosfamide and etoposide. She has been free of recurrent disease for the past 9 years now.

#### Discussion

Primary osteosarcomas of the heart are extremely rare entities, with descriptions in the literature limited to only case reports. Three common subtypes of osteosarcoma arise in the osseous structures, each of which has been reported in the heart as well, including osteoblastic, chondroblastic and fibroblastic subtypes [1]. Based on a meta-analysis of 53 cases of primary cardiac osteosarcomas [1], involvement of the left atrium is typical [2, 3]. Most cases are in their fourth or fifth decade of life, and imaging features on CT scan, when available, usually demonstrate lobulated or irregular masses with heterogenous attenuation and enhancement with a typical lack of a calcified matrix [4, 5], consistent with the demographics and the findings in this case. The differential considera-

tions given the findings in this case include cardiac metastasis with other primaries, atrial myxoma or lymphoma. This is in contrast with the skeletal osteoblastic or chondroblastic osteosarcomas where presence of a calcified matrix is key to the diagnosis. This case demonstrates an unusual subtype of cardiac osteosarcoma that presents without a calcified matrix, which makes imaging diagnosis difficult. The prognosis [1] is poor, with complete surgical excision usually un-achievable due to infiltration of vital structures. Prognosis remains dismal even among cases undergoing post-surgical chemotherapy even after a complete surgical excision, the median survival among cases diagnosed antemortem remained ~ 20 months in above case-series [1]. A unique feature of this case is the long period of recurrence free survival, atypical in patients with these tumors.

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### Patient Consent Statement

Special efforts were made to exclude any patient identifiers during the preparation of this manuscript.

Per Institutional Review Board (IRB), no patient consent is needed for publications of case reports if all patient identifiers are removed.

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