Control Number: 11

Abstract Category: Clinical Case Challenge in Cardio-Oncology

Title: Challenges in Diagnosis of Cardiac Masses: Importance of Cardiac Imaging and Biopsy to Guide Treatment

ABSTRACT BODY

Background and Purpose

Primary cardiac tumors (PCT) are a rare clinical entity with an incidence of 200 per million. Only 25% of PCT are malignant and sarcoma accounts for 75% of malignant PCT. The diagnosis and management of PCT is often challenging given their low incidence and variable clinical presentation.

Case Description and Outcomes

Case 1: 44-year-old African American man presented with dyspnea and was diagnosed with cardiac tamponade. Pericardiocentesis was performed with removal of 1,350 ml of bloody fluid: cytology and flow cytometry were negative for malignancy. Two months later he reported worsening dyspnea, a cardiac MR (CMR) revealed an intrapericardial mass within the right atrioventricular groove with recurrent pericardial effusion (Figure 1). There was no evidence of extracardiac disease on staging scans. Pericardial fluid cytology from repeat pericardiocentesis was negative for malignancy and the patient underwent sternotomy for tissue biopsy. Pathology revealed high grade pericardial angiosarcoma (Figure 1). The extent and location of the invasive tumor precluded resection and systemic chemotherapy with ifosfamide and doxorubicin was recommended. Case 2: 66-year-old Hispanic woman presented with a 2-month history of fatigue and weight loss. A CT scan showed a large pericardial effusion and pericardiocentesis pericardial fluid was negative for malignancy. The fluid re-accumulated, she underwent extensive pericardiectomy, 2,000 ml of fluid were removed. The fluid collection, and the pericardial specimen were negative for malignancy. Her symptoms recurred and three months later a CMR that showed an infiltrating epicardial mass extending from the anterior base to the left ventricle into the septum and lateral wall (Figure 2). Biopsy revealed diffuse large B-cell lymphoma (Figure 2). She received six cycles of R-EPOCH chemotherapy and has remained disease free for seven years.

Discussion

Pericardial fluid evaluation is not sufficient to exclude malignancy. CMR is of critical importance to guide differential diagnosis and biopsy approach which may require sternotomy. Prognosis and treatment of PCT depend on histopathological diagnosis and staging (Table 1). A multidisciplinary approach is needed to assure optimal outcomes.

References

N/A

Table

Table 1: Disease Characteristics

	Primary cardiac sarcoma	Primary cardiac lymphoma
Epidemiology	Medias age is 46. Not associated to gender predominance	Median age of 63. Male/female ratio 1.94. More common in immunocompromised patients
Histology	Angiosacroma is the most common type	The most common type is diffuse large B-cell lymphoma
Prognosis	Small case series, median OS of 9-27 months	Median OS 12-30 months. Left ventricular involvement, evidence of extracardiac disease, arrhythmia and immunocompromised are associated with worse prognosis
Treatment	If patients have no evidence of metastatic disease, complete surgical removal of the tumor is the preferred treatment modality. If surgical excision is not possible, prognosis is significantly worse, consider chemotherapy and/or radiation	Standard lymphoma chemotherapy. Often favor regimens with infusional doxorubicin vs bolus due to concerns for anthracycline induced-cardiotoxicity

OS: overall survival

Image 1

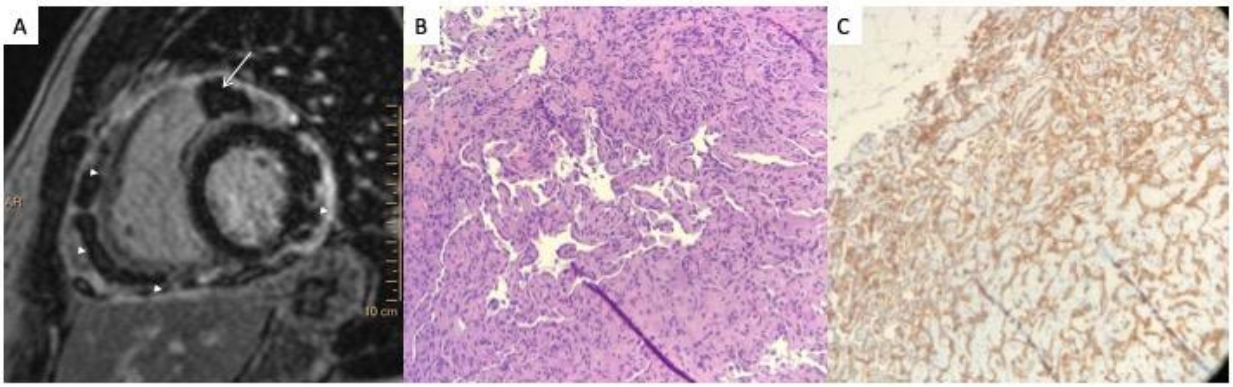

Figure 1: A) CMR showing large amount of complex material is contained in the pericardial space that likely represents a complex loculated pericardial effusion (arrowheads); however, a more mass like structure is noted within the right atrioventricular groove measuring 55 mm x 31 mm (arrow). B) H&E, 20X: cellular, infiltrative neoplasm consisting of anastomosing channels lined by pleomorphic spindle cells C) 20X: CD34 (pictured) and CD31 positivity confirm vascular origin.

Image 2

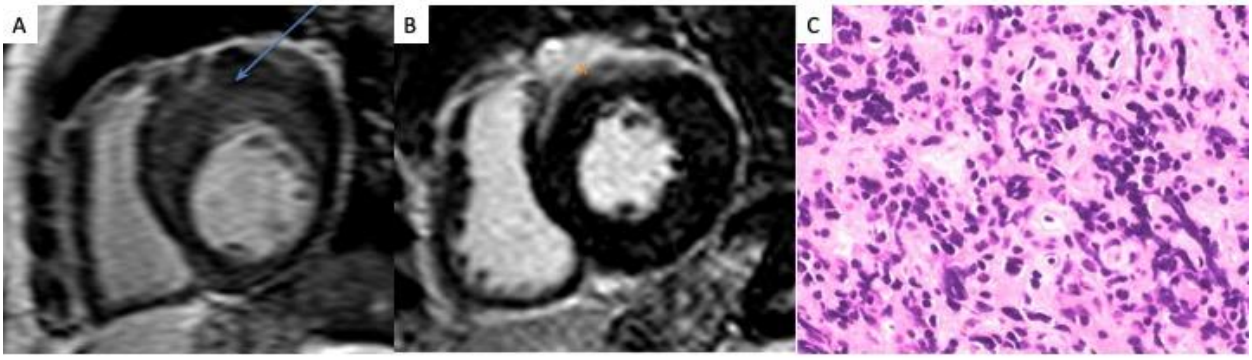

Figure 2: A) Mass noted in the pericardium (arrow) adjacent to the anterolateral, anterior and septal walls of the left ventricle and extending into the superior portion of the right ventricle and right ventricular outflow tract. There is infiltration of the left ventricular myocardium. B) After chemotherapy, mass was not longer detected on CMR. C) Dense atypical lymphoid infiltrate with several scattered and clusters of large cells. These large cells also percolate into the interstitium and myocytes. The neoplastic lymphoid cells were positive for CD20, BCL-2, BCL-6.