

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

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Background: 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 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 undifferentiated pericardial pleomorphic angiosarcoma (Figure 1). The extent and location of the invasive tumor precluded resection and systemic chemotherapy with ifosfamide and doxorubicin was recommended.

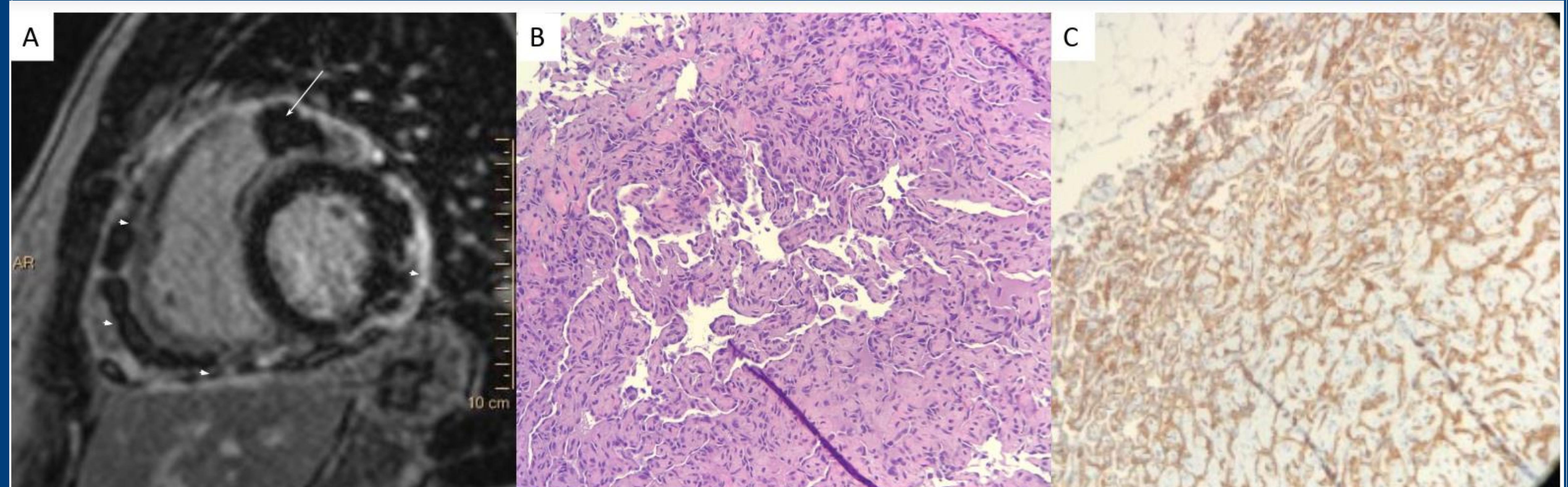


Figure 1: A) CMR showing large amount of complex material is contained in the pericardial space that likely represents a complex loculated pericardial effusion (arrowheads); however, a more mass like structure is noted within the right atrioventricular groove measuring 55 mm x 31 mm (arrow). B) H&E,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

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 pericardectomy, 2,000 ml of fluid were removed. The fluid cytology and pericardial specimen were negative for malignancy. Her symptoms recurred and three months later a CMR 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.

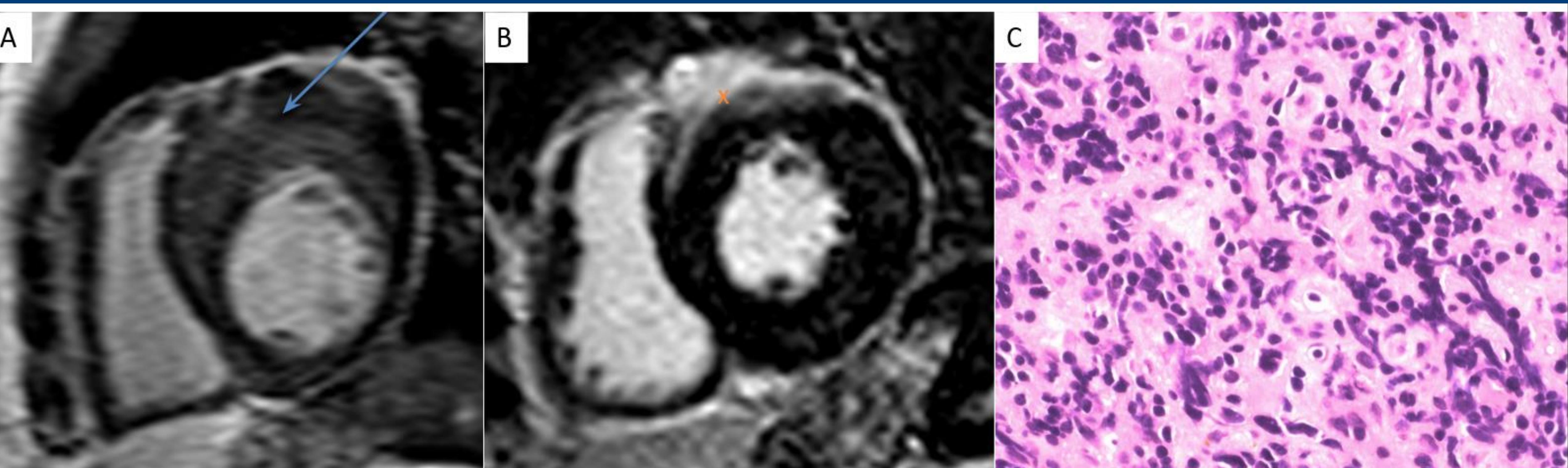


Figure 2: A) Mass noted in the pericardium (arrow) adjacent to the anterolateral, anterior and septal walls of the left ventricle and extending into the superior portion of the right ventricle and right ventricular outflow tract. There is infiltration of the left ventricular myocardium. B) After chemotherapy, 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.

Conclusions: 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.

Clinical Implications: High level of suspicion is needed in patients presenting with pericardial effusion to pursue advanced cardiac imaging and obtain early tissue diagnosis.