CLINICAL IMAGE

Primary sarcoma of the heart

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A 74-year-old woman presented with progressive dyspnea and exercise intolerance. The past history revealed a weight loss of 5 kg over 2 months. A loud systolic murmur in the left second intercostal space was noted. A transthoracic echocardiography showed pericardial effusion with a large mass protruding to the right ventricular outflow tract (RVOT) and pulmonary artery (FIGURE 1A), leading to a severe RVOT obstruction and pulmonic stenosis with a maximum pressure gradient of 120 mm Hg. Black-blood magnetic resonance images showed a large mass (52×50×71 mm) located in the RVOT (FIGURE 1B). The tumor almost completely obstructed the right ventricular inflow and outflow tracts (FIGURE 1B and 1C). A delayed enhancement technique revealed areas of heterogeneous enhancement due to regional variations in vascularity and distribution volumes (ie, areas of necrosis) within the tumor and infiltration of the myocardium and epicardium (FIGURE 1D). Endomyocardial biopsy was performed. Histological examination with immunohistochemical staining confirmed the diagnosis of undifferentiated sarcoma. Due to the extent of the tumor, its invasiveness and the presence of metastases, the patient was not considered to be a suitable candidate for cardiothoracic surgery and was referred for palliative chemotherapy.

Primary cardiac tumors represent only a small fraction of all cardiac tumors, the most prevalent being pseudotumors. The estimated frequency of primary tumors of the heart ranges from 0.0017% to 0.33%,[1] of which 75% are benign. Primary malignant cardiac tumors are predominantly sarcomas. Angiosarcomas and undifferentiated sarcomas are the most prevalent.[1] Metastases to the heart are far more common, and metastastic cardiac tumors originate mainly from the lung, breast, and renal cancer, melanomas, lymphomas, and leukemias.[1] The term "undifferentiated sarcoma" is used to describe tumors with no definable line of cellular differentiation and microscopic features of fibroblasts or myofibroblasts (or both). An undifferentiated sarcoma predominantly affects the left atrium, typically presenting with a locally advanced tumor. It rarely occurs in the right ventricle, with only dozen cases reported in the literature.[2]

Clinical presentation mainly depends on the size of the tumor and its anatomical location,

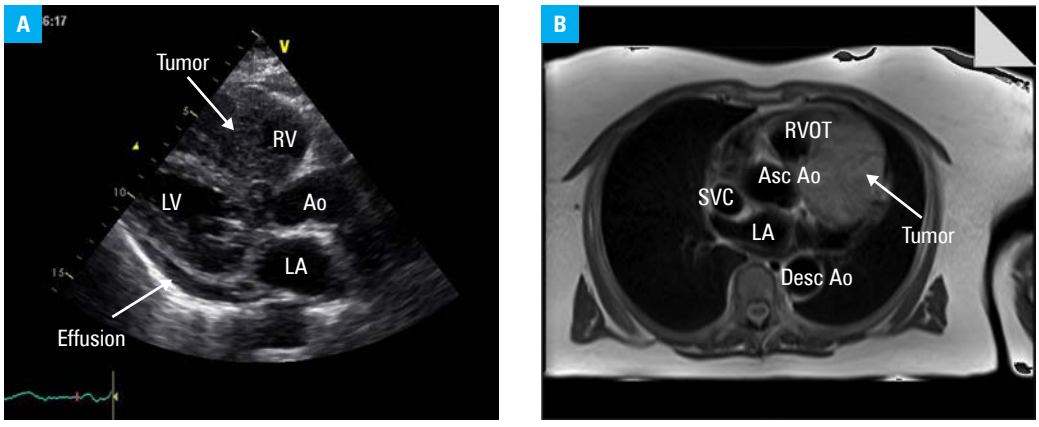

FIGURE 1 A – a transthoracic echocardiography showing pericardial effusion with a large mass protruding to the right ventricular outflow tract (RVOT); B – axial, T1-weighted black-blood magnetic resonance images showing a large mass located in the RVOT

Abbreviations: see the next page

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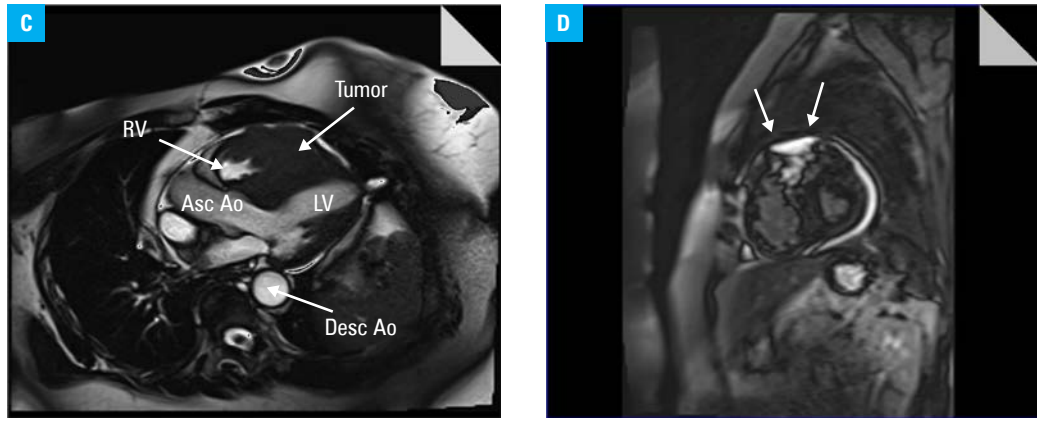

FIGURE 1 **C** – dynamic gadolinium perfusion study showing a large tumor almost completely obstructing the RVOT with no first-pass enhancement; **D** – magnetic resonance imaging (short-axis view) showing the tumor infiltrating the myocardium and epicardium. A delayed enhancement technique revealed areas of heterogeneous enhancement due to regional variations in vascularity and distribution volumes (ie, areas of necrosis) within the tumor.

Abbreviations: Ao, aorta; Asc Ao, ascending aorta; Desc Ao, descending aorta; LA, left atrium; LAD, left anterior descending artery; LV, left ventricle; RV, right ventricle; SVC, superior vena cava

rather than on its histological type.[3] Diagnostic modalities include echocardiography, computed tomography, magnetic resonance imaging, and cardiac catheterization with coronary angiography or endomyocardial biopsy (or both). The imaging features suggestive of cardiac malignancy include right atrial location, involvement of more than 1 cardiac chambers, size exceeding 5 cm, hemorrhagic pericardial effusion, broad-base attachment, and extension to the mediastinum or great vessels.[1,4]

Surgical resection is the treatment of choice in cardiac sarcomas.[4] This goal is rarely achievable in practice, given that these tumors are aggressive and infiltrative and that the diagnosis is usually made late in the course of the disease. Adjuvant chemotherapy should always follow surgical excision.[4] In case of inoperable disease, palliative chemotherapy should be offered, although in some cases palliative surgical debulking may be undertaken to relieve rapidly progressing symptoms (cardiac tamponade, blood flow obstruction).[4] Younger patients with no metastatic disease may be considered suitable for an orthotopic heart transplant.[1]

Prognosis in all primary cardiac sarcomas is dismal.[4] However, it is slightly better if tumors present with the following features: left atrial location, low mitotic count, no necrotic regions, and the absence of metastases at the time of diagnosis. The median survival for cardiac sarcomas is 6 months, whereas for noncardiac sarcomas, it is 93 months. Patients with primary cardiac sarcomas who undergo surgical resection have a median survival of 12 months, while those who undergo only palliative chemotherapy have a median survival of 1 month.[5]

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