Images in Cardiovascular Medicine

Malignant Fibrous Histiocytoma of the Heart

Hafize Yaliniz, MD
Orhan Kemal Salih, MD
Acar Tokcan, MD

A 28-year-old man presented with dyspnea, fatigue, and epigastric pain. On examination, his pulse and arterial pressure were normal. A grade 1/6 systolic murmur was heard at the lower-left sternal border. Chest radiography showed cardiomegaly.

Transthoracic echocardiography showed a polycystic mass in the pericardium over the left ventricle; however, the heart appeared to be structurally normal (Fig. 1). Contrast-enhanced helical computed tomography revealed an unresectable mass adjacent to the great vessels and the left ventricle (Fig. 2). The mass involved the left ventricular wall, the right ventricular outflow tract, and the pulmonary artery. An incomplete resection was performed; the patient's postoperative period was uneventful. Histopathologic examination revealed that the tumor consisted of malignant cells with marked atypia (Fig. 3). Necrosis was prominent. Immunohistochemistry showed a diffuse immunoreaction for vimentin and no focal positivity for CD68 desmin. The histopathologic diagnosis was malignant fibrous histiocytoma. The patient underwent radiotherapy.

Comment

Malignant fibrous histiocytoma is a sarcoma that typically occurs in the extremities, the torso, and the retroperitoneum.[1] This tumor constitutes less than 3% of primary

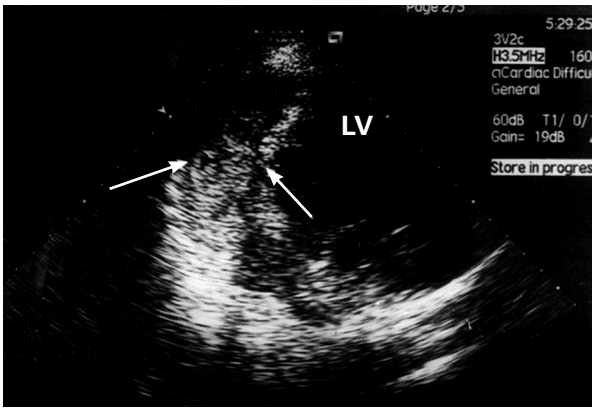

Fig. 1 *Transthoracic echocardiography (parasternal short-axis view) shows a polycystic mass (arrows) adjacent to the left ventricle (LV).*

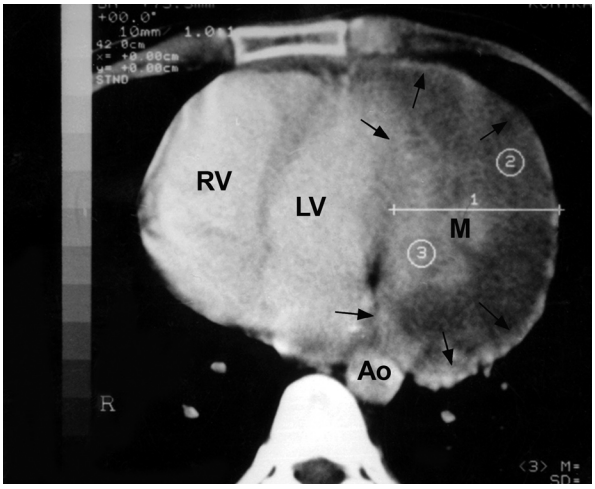

Fig. 2 *Helical computed tomography shows, at the patient's left, a mass that is mostly cystic at its caudal end. It lies adjacent to the great vessels and the left ventricle. The arrows delineate the borders of the mass.*

Ao = aorta; LV = left ventricle; M = mass; RV = right ventricle

Section Editor:
Raymond F. Stainback, MD, Department of Adult Cardiology, Texas Heart Institute and St. Luke's Episcopal Hospital, 6624 Fannin Street, Suite 2480, Houston, TX 77030

From: *Department of Cardiovascular Surgery, Medical Faculty, University of Cukurova, 01330 Balcali – Adana, Turkey*

Address for reprints:
Hafize Yaliniz, MD, Department of Cardiovascular Surgery, Medical Faculty, University of Cukurova, 01330 Balcali – Adana, Turkey

E-mail:
hafize101@yahoo.com

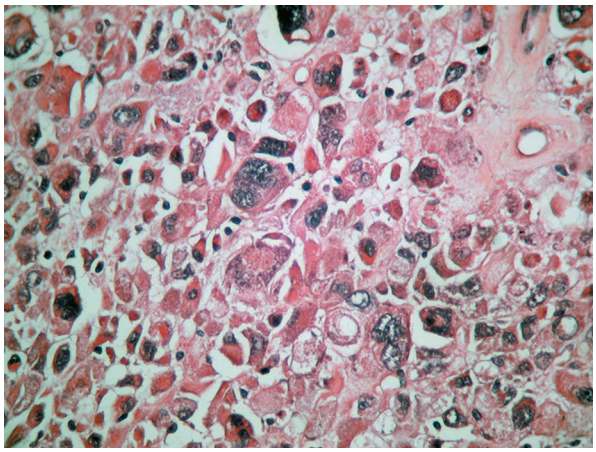

Fig. 3 *Photomicrograph shows multinuclear, markedly pleomorphic cells (H & E, orig. ×200).*

cardiac tumors. In 1987, Lee and colleagues[2] estimated that fewer than 20 cases had been reported since this tumor was first identified in the myocardium in 1964.

Cardiac malignant fibrous histiocytoma can be clinically and histologically confused with atrial myxoma, the most common primary cardiac tumor.[3,4] Although transesophageal echocardiography, helical computed tomography, and magnetic resonance imaging are highly reliable methods for examining intracardiac tumors, a definitive diagnosis of cardiac malignant fibrous histiocytoma requires either immunohistochemical or ultrastructural confirmation.

Patients with this tumor usually have a poor prognosis. Surgical resection is the mainstay of diagnosis and treatment.[5] Radiotherapy and chemotherapy can be beneficial, especially when only an incomplete resection is possible.

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