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Primary Cardiac Myxosarcoma Invading the Mitral Valve

A 32-year-old woman presented with a 1-year history of palpitations and worsening exertional chest tightness. Cardiac auscultation revealed a moderate diastolic murmur and a grade 3/6 systolic murmur in the apical region. Transthoracic echocardiography (TTE) showed a hypoechoic mass measuring 2.8 cm × 2.3 cm attached to the atrial side of the anterior mitral valve leaflet (Figure 1A and B), prolapsing into the left ventricle during diastole causing mitral valve stenosis (Figure 1C), a peak flow velocity of 3.2 m/s, mean trans-mitral gradient of 41 mm Hg (Figure 1D), and severe mitral regurgitation in systole (Figure 1E). Cardiac computed tomography (CT) confirmed that the soft tissue mass was isolated to the mitral valve with no extension into other chambers or vessels (Figure 1F and G). The patient underwent surgical mass resection and mitral valve replacement;

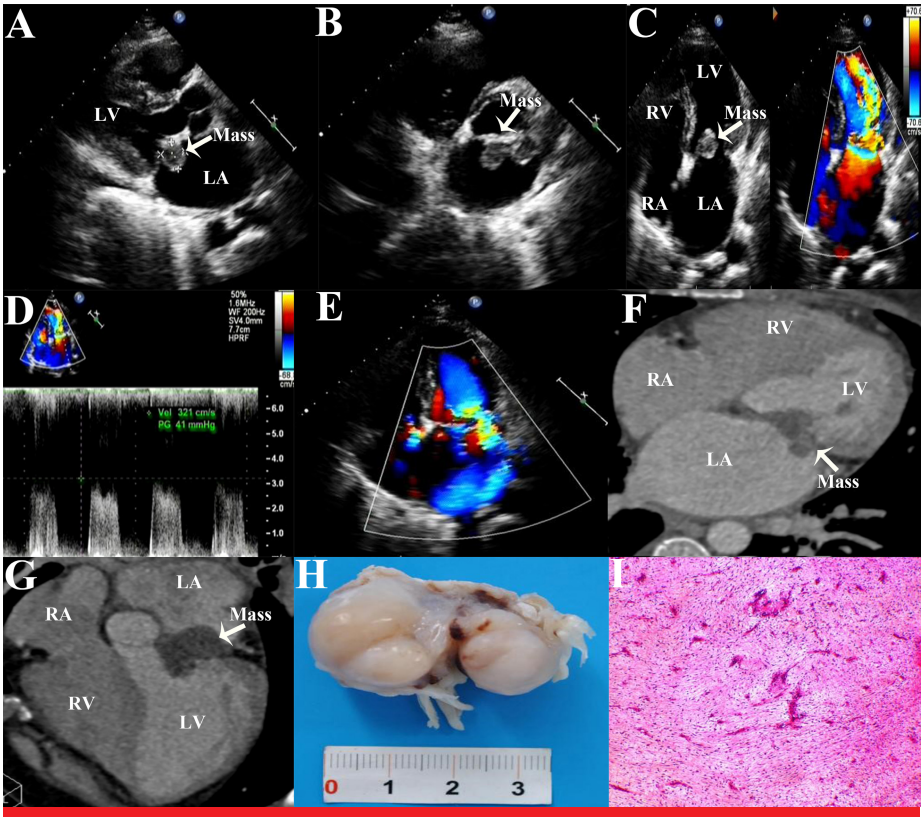

Figure 1. (A-E) Transthoracic echocardiography shows a hypoechoic mass attached to the atrial side of the anterior mitral valve leaflet (A and B), prolapsing into the left ventricle during diastole, causing mitral valve stenosis (C), a peak flow velocity of 3.2 m/s, mean trans-mitral gradient of 41 mm Hg (D), and severe mitral regurgitation in systole (E). (F and G) Cardiac CT confirms that the soft tissue mass is isolated to the mitral valve with no extension into other chambers or vessels. (H and I) Histopathology confirmed the diagnosis of myxosarcoma. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

E-PAGE ORIGINAL IMAGE

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the gross and histopathological examination confirmed the diagnosis of myxosarcoma (Figure 1H and I). The postoperative adjuvant chemotherapy with pazopanib hydrochloride was given. At 12 months of follow-up, there were no signs of local recurrence or metastases.

Primary cardiac myxosarcoma is a rare condition that most commonly originates in the left atrium. It is frequently involved in the pulmonary artery, pericardium, or pleura, and may also lead to distant metastasis.[1] Myxosarcomas that invade the mitral valve are extremely rare and are difficult to differentiate from the more common benign myxomas. The multimodality imaging technique, including TTE, transesophageal echocardiography (TEE), cardiac CT, cardiac magnetic resonance imaging (CMR), and ^{18}F-fluorodeoxyglucose positron emission CT, can provide important preliminary diagnostic information on the tumor size, boundary, location, and infiltration status, which is necessary to plan the treatment. The first-line imaging technique for classifying cardiac masses according to their size and border is TTE. Additionally, it offers details on ventricular function, abnormal pericardial thickness, valve involvement, and cardiac hemodynamics. A non-septal origin, extension into the pulmonary vein, multiple masses, extensive attachment to the left atrial wall, and a semisolid mass consistency within the left atrium are all possible characteristics of malignant lesions. When characterizing cardiac masses and surrounding structures, TEE is superior to TTE. The TEE is an intrusive test, though, so it might not be appropriate for every patient. Cardiac CT offers a high-resolution picture of the cancerous tumors and their surroundings. It is acknowledged as the best method for assessing extracardiac involvement in cardiac metastases. The CMR provides comprehensive details on the location of the tumor in relation to adjacent structures, motility, tissue characteristics, vascularization, and whether the tumor contains fibrous or necrotic tissue. It is also possible to employ PET-CT to identify distant metastases and differentiate between benign and malignant tumors.[2] Nevertheless, histopathologic and immunohistochemical examinations are required to confirm a diagnosis of cardiac myxosarcoma.[3] Early diagnosis and a multidisciplinary approach, including cardiac surgeons, oncologists, and critical care specialists, are crucial in the management of this disease.[4] At present, surgery is the preferred treatment option for cardiac myxosarcoma. Radiotherapy and chemotherapy may play a positive role in improving prognosis.

Informed Consent: The informed consent was obtained from the patient for this study.

Declaration of Interests: The authors have no conflicts of interest to declare.

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