Hypertrophic cardiomyopathy and left atrial myxoma

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Patients with a cardiac tumor may present with cardiovascular or constitutional symptoms, but, in general, a cardiac mass is detected incidentally on imaging examination performed for other reasons.¹ Hypertrophic cardiomyopathy (HCM) is also often diagnosed incidentally, and it can present with symptoms of blood flow obstruction mimicking cardiac myxoma.² Primary cardiac tumors are rare and usually benign (approximately 75% of the cases); nearly half of the cardiac tumors are myxomas.¹

A 46-year-old woman was admitted to the hospital owing to a tumor in the left atrium incidentally diagnosed on echocardiography after ischemic stroke. The medical history revealed ischemic stroke that occurred 9 months earlier and led to motor aphasia and paralysis of the right upper limb that resolved over time. Vascular brain damage was detected by magnetic resonance imaging (MRI). The patient also had hypertension, and hyperlipidemia. On admission, auscultation revealed a systolic murmur over the apex and aortic valve. The blood pressure was 120/70 mmHg. Coagulation tests, troponin levels, and tumor markers were within normal ranges. An electrocardiogram, chest X-ray, and carotid

FIGURE 1 A patient with a myxoma in the left atrium; A – cardiac magnetic resonance imaging, T₂-weighted image (2-chamber view); arrows indicate a myxoma; B – cardiac magnetic resonance imaging, T₁-weighted image (2-chamber view); arrows indicate a myxoma; C – cardiac magnetic resonance imaging, first pass sequences (2-chamber view); D – cardiac magnetic resonance imaging, late gadolinium enhancement image (2-chamber view); E – cardiac magnetic resonance imaging, asymmetric hypertrophy of the left ventricle (short-axis view)
Doppler ultrasound showed no abnormalities. Transthoracic echocardiography showed enlargement of the left atrium (56 mm) and hypertrophy of the posterior wall (12 mm), interventricular septum (26 mm), anterolateral wall, and apex (HCM type III, on the basis of Maron BJ), mild mitral and tricuspid insufficiency, E wave/A wave (0.7), additional balloting echo in the left atrium prolapsing into the mitral valve and attached to the interatrial septum, which suggested a myxoma. The ejection fraction was preserved.

To confirm HCM and a cardiac tumor, cardiac MRI was performed with a standard protocol and showed a balloting mass (27 × 16 × 37 mm) within the left atrium. High signal intensity in T2-weighted images, intermediate signal intensity in T1-weighted images, hypoperfusion in first-pass images, and heterogeneous intense enhancement of the lesion with the areas of delayed enhancement peripherally due to necrosis in late gadolinium enhancement images confirmed a myxoma (FIGURE 1A–D). Asymmetric hypertrophy of the anterior wall and interventricular septum (basal segments up to 27 mm; middle segments up to 22 mm) with a typical pattern of intramural late gadolinium enhancement confirmed HCM (FIGURE 1E). Coronary angiography showed no abnormalities.

Considering partial obstruction of the mitral valve and a history of stroke, we decided to schedule the patient for urgent tumor removal. Histopathology confirmed a myxoma.

Myxoma is the most common primary cardiac tumor. A myxoma coexistent with HCM is extremely rare, and, to our knowledge, as few as 2 such cases have been described. HCM and left atrial myxomas have been observed in patients with progressive cardiomyopathic lentiginosis (LEOPARD syndrome)—a rare autosomal dominant disease affecting multiple organs and systems, mostly the skin and skeletal and cardiovascular systems. However, LEOPARD syndrome was not diagnosed in our patient.

The tumor was first incidentally detected by echocardiography, but it was later confirmed by cardiac MRI. Echocardiography is the best first-line imaging modality because it is simple, non-invasive, widely available, and cost-effective, and shows tumor structure and location as well as hemodynamic alterations induced by a tumor. Cardiac MRI is a complementary technique that is not associated with radiation risk and that provides information on tumor vasculature and tissue appearance.

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