Case Report

Large Left Atrial Myxoma in an Oligosymptomatic Young Woman

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We present the case of a young female with a large sporadic left atrial myxoma. Interestingly, despite the tumor's large size, this patient had only mild exertional dyspnea without any embolic events or constitutional symptoms.

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Primary tumors of the heart are rare, with a reported incidence of 0.0017% to 0.19% in the general population.[1] Atrial myxoma is the most frequent primary tumor of the heart, representing 30-50% of all cardiac masses.[1,2] Here we present a case of a large left atrial myxoma in a young woman with minimal symptoms.

Case presentation

A 35-year-old woman presented to our outpatient clinic complaining of mild new-onset exertional dyspnea of one week's duration. She denied fever, chills, weight or appetite changes, chest pain, palpitations, orthopnea, paroxysmal nocturnal dyspnea, dizziness, presyncope or syncope. She had no significant past medical history, but she had a positive family history of early cardiovascular disease. She was of normal weight (body mass index 23 kg/m^2) and had been smoking 15 cigarettes per day for the last 15 years (~11 pack/years), but denied use of illicit drugs, alcohol or medications.

At presentation, the patient was afebrile and had normal vital signs (pulse 80 bpm, blood pressure 105/65 mmHg, oxygen saturation 97% while breathing room air). Her lungs were clear on auscultation. Cardiac examination revealed a regular rate and rhythm, normal S_1 and S_2 heart sounds, and a soft 1-2/6 systolic murmur along with a low frequency diastolic rumble that were audible at the cardiac apex. No additional heart sounds or pericardial friction rub were detected. The remainder of the physical examination was unremarkable. The chest X-ray showed a normal lung parenchyma and a normally sized cardiac silhouette. A 12-lead electrocardiogram revealed sinus rhythm with nonspecific minor ST-T abnormalities. Transthoracic and subsequently transesophageal echocardiography were notable for the presence of a large mass in the left atrium, which was attached to the interatrial septum by a thick stalk and prolapsed across the mitral annulus into the left ventricular inflow tract during diastole (Figure 1A,B). The mass contained an area of high acoustic density along with a larger echolucent region. Mild mitral regurgitation and mild to moderate functional mitral stenosis (mean gradient ~5 mmHg) were also detected.

Laboratory tests were remarkable for mild normocytic anemia (hemoglobin 10.4

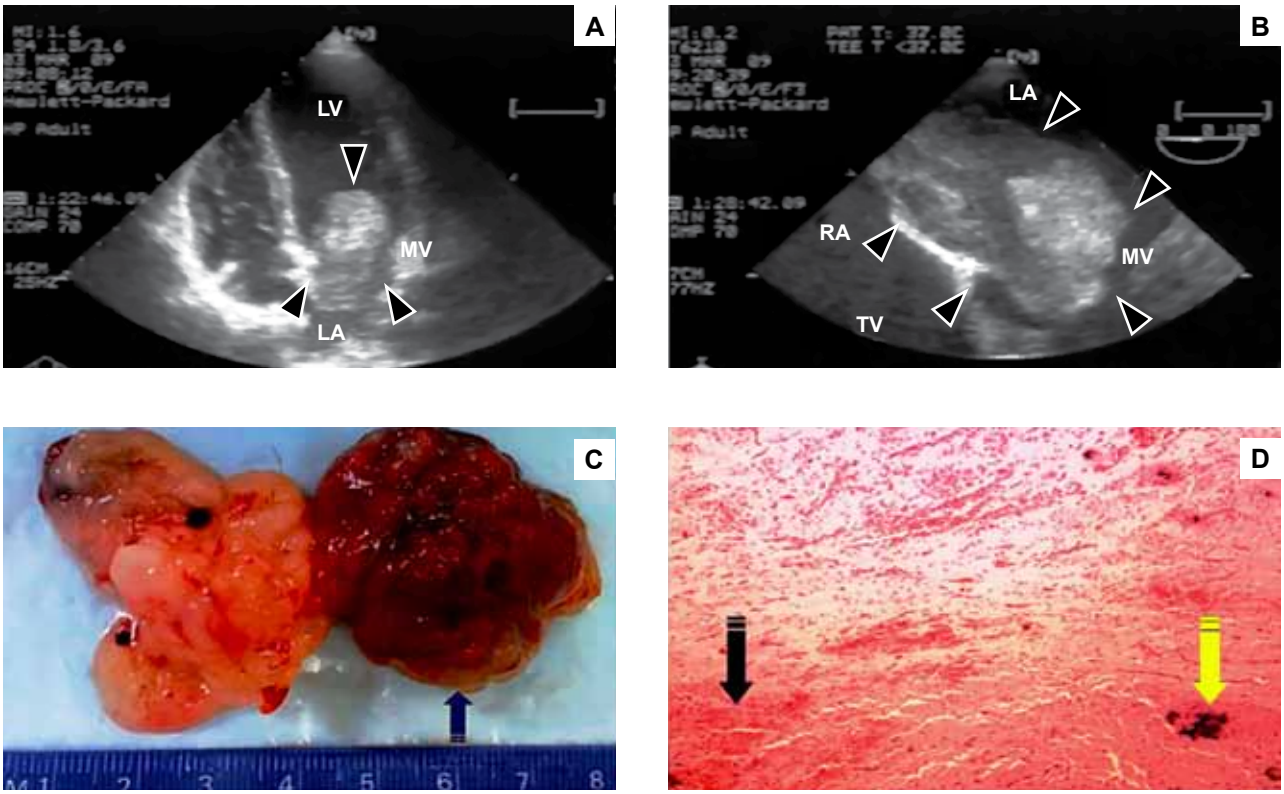

Figure 1. Transthoracic (panel A) and transesophageal echocardiography (multiplane view at mid-esophageal level and at 0°, panel B) showing a large lobular left atrial mass attached by a broad stalk to the interatrial septum and prolapsing during diastole into the left ventricular inflow tract. Arrowheads delineate the cardiac myxoma. Panel C: Surgical specimen of the resected mass, size 7.5 × 3.5 × 3.0 cm, with gelatinous appearance and hemorrhage in its free part (blue arrow). Panel D: Histological specimen in the hematoxylin–eosin stain (magnification ×20) showing spindle cells in myxoid stroma with foci of hemorrhage (yellow arrow) and calcifications (black arrow). LA – left atrium; RA – right atrium; LV – left ventricle; MV – mitral valve; TR – tricuspid valve.

g/dL, mean corpuscular volume 88 fL), elevated interleukin-6 (IL-6: 6.6 pg/mg, normal values <1 pg/ml), and markedly elevated high-sensitivity C-reactive protein (hsCRP) levels (50.0 mg/L, normal values <1.0 mg/L). Troponin-I and BNP levels, as well as the rest of the blood tests, were all within normal range.

The patient was referred for surgery, and a 7.5 × 3.5 × 3.0 cm mass was resected (Figure 1C). Histology of the mass showed spindle cells in a myxoid stroma with foci of hemorrhage and calcifications, compatible with the diagnosis of myxoma (Figure 1D). The calcified areas and those with hemorrhage corresponded with the echocardiographic area of high acoustic density and the echolucent area, respectively. The postoperative period was uneventful and the patient was discharged on the fifth postoperative day in excellent condition. The postoperative cardiac ultrasound was unremarkable. The patient was in excellent clinical condition without recurrence at two-year follow up.

Discussion

Cardiac myxoma is the most common primary intracardiac tumor.[1] It is more frequently detected in females (~57%), and the mean age of appearance of sporadic cardiac myxomas is around 50 years.[3,4] Occasionally, some cases of myxoma are familial, and then the tumor usually appears at a younger age (mean 24 years) and is frequently associated with Carney complex which, apart from cardiac myxomas, comprises myxomas in other locations (e.g. skin, breast), spotty pigmentation of the skin, endocrinopathy, and both endocrine and non-endocrine tumors (either benign or malignant).[1,3,4] Our patient, however, despite her relatively young age, had no family history of myxoma and she had no symptoms or physical signs suggestive of Carney complex.

Atrial myxomas present clinically with the classical triad of cardiac obstructive symptoms, constitu-

tional symptoms and embolic events.[3,4] Obstructive symptoms are reported in 60-70% of patients with left atrial myxoma, and include dyspnea (either exertional or paroxysmal), orthopnea, arrhythmias, syncope and sudden death.[3,4] Constitutional symptoms, such as fever, malaise, weight loss, anorexia, anemia, and arthralgias are noted in 10-45% of patients with myxomas.[3,4] Finally, embolic events have been described in about 35% of left-sided and 10% of right-sided myxomas, whereas multi-organ embolism in the same patient has also been described in rare cases.[3,5,6] In a series of 64 cases with atrial myxoma, the mean value of the tumor's longest dimension was 5.8 cm.[3] In the case described here, despite the large tumor size, our patient had only mild obstructive symptoms, without embolic events or constitutional symptoms.

Echocardiography is the gold standard for the detection of cardiac myxomas. Transthoracic echocardiography has a sensitivity of about 95% for myxoma detection and can be used for the determination of the location, size, shape, attachment and mobility of the tumor.[1,7] Myxomas are solitary in 90% of cases and in ~75% are located in the left atrium, with the interatrial septum at the border of the *fossa ovalis* being the usual site of attachment.[8,9] Other possible attachment points are in the left atrial appendage, mitral valve, and left atrial roof.[8] In 15-20% they originate in the right atrium and in rare instances in the ventricles.[9,10] It should be noted that there is a high suspicion of myxoma when an intra-cardiac mass is detected attached to the interatrial septum in the proximity of the *fossa ovalis*, just as in the reported case.[8,11] Myxomas are often pedunculated and may potentially prolapse in the inflow tract of the corresponding ventricle.[8,9,11] Left atrial myxomas may cause incomplete closure of the mitral valve in systole and may interfere with left ventricular filling in diastole, causing mitral regurgitation and functional mitral stenosis, respectively. Similarly, tricuspid regurgitation and stenosis can be detected by Doppler in right-sided atrial myxomas.[11]

Transesophageal echocardiography has a sensitivity of 100% for myxoma detection and provides more details regarding its morphologic features, revealing intra-tumoral areas of calcification, hemorrhage, necrotic foci, and importantly, satellite tumors.[1,7,8] In the present case, transesophageal echo detected echodense and echolucent areas, which, as the pathological study revealed, accounted for areas of calcification and hemorrhage/necrosis respectively.

Cardiac computed tomography (CT) and magnetic resonance imaging (MRI) can also be used to detect cardiac tumors.[1,10] With the above modalities, myxomas appear as spherical or ovoid masses with a lobular or smooth contour. The attenuation of myxomas in cardiac CT is usually less than or equal to the attenuation of myocardium.[3] Furthermore, there may be a heterogeneous enhancement of myxomas after contrast administration. Cardiac MRI can demonstrate heterogeneous signal intensity in the majority of myxomas, due to intra-tumoral areas of calcification and hemorrhage.[3,12] In addition, because of their vascularization, myxomas show a moderately high contrast enhancement after intravenous administration of a paramagnetic contrast agent.[3,12]

The elevated hsCRP and IL-6 levels observed in this case are common in patients with myxomas.[13] IL-6 levels correlate with the tumor size and are associated with embolic events.[13] However, despite the large dimensions of the tumor and high levels of IL-6, our patient had only mild exertional dyspnea and anemia without embolic phenomena or constitutional symptoms.

The treatment of choice for myxomas is surgical removal, which is usually curative.[1] Operative mortality ranges from 0% to 3% and recurrence rates are between 1% and 3% for sporadic myxomas.[14]

In conclusion, myxomas may occasionally present in young adults without a familial history of a cardiac tumor. Even patients with large myxomas may be oligosymptomatic. A high index of suspicion is essential for diagnosis.

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