European Journal of Echocardiography (2008) **9**, 181–183
doi:10.1093/ejechocard/jem062

Mitral valve myxomas: an unusual entity

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Received 14 June 2007; accepted after revision 29 July 2007

KEYWORDS
Mitral valve;
Myxoma;
Echocardiography;
Transoesophageal echocardiography

Primary tumours of the heart are uncommon entities, cardiac myxomas being the most frequent. However, mitral valve myxomas are exceptionally rare. In the last 12 years, there have been 25 myxomas diagnosed at our institution, with only two of them originating from the mitral valve. Both patients were female, the first, 25, and the second, 72 years old. The younger patient was very symptomatic with a large mass, 4 cm long, which involved both leaflets causing significant obstruction to the left ventricular inflow. The second one had a smaller mass located at the atrial side of the posterior leaflet that only produced some flow divergence. Neither of them had constitutional nor embolic symptoms. Both patients were submitted to emergent surgical resection that in the first case involved the mitral valve and replacement with mechanical prosthesis. The macroscopic appearance of these tumours suggested a malignant aetiology which may represent somewhat different features of the myxomas when originating from the cardiac valves. Both patients are well reflecting the good prognosis of this illness after resection, although the younger patient was re-operated because of prosthetic valve obstruction and suspicion of recurrence that was not confirmed. Because of the illustrative images and different presentations, we found it interesting to report and discuss them together.

Introduction

Myxomas are the most common primary tumours of the heart. However, their occurrence on a valve is extremely rare. We report two cases that correspond to 8% of all myxomas diagnosed between 1994 and 2006 in our department.

Case 1

A 25-year-old female patient with complaints of sudden onset of anxiety and dyspnoea was referred to our tertiary hospital with the diagnosis of severe mitral valve disease, presumably of rheumatic origin.

The patient was anxious, dyspnoeic, and tachycardic. Blood pressure was normal and rales were heard in the lower halves of both lungs. A diastolic rumble was heard in the precordium. The transthoracic echocardiogram (TTE) showed a diffusely thickened mitral valve, with an echogenic mass arising from both leaflets to the lateral wall of the left ventricle. The mitral valve mean gradient was 14 mmHg and functional mitral valve area was 1 cm^2. Transoesophageal echocardiogram (TEE) confirmed the presence of a mass on the mitral valve (*Figure 1*, clip 1) which prevented an adequate left ventricular filling. Emergent surgical resection was performed, the operatory diagnosis being a malignant tumour of the mitral valve. Because of the mitral valve involvement by the mass, both were excised, and a 27 mm Carbomedics prosthesis was inserted. Pathological analysis showed 'destruction of the posterior leaflet of the mitral valve by a vegetating mass measuring 4×2 cm, which involved the chordae tendinae with an organized blood clot superimposed'. The final histological diagnosis was myxoma.

One year later, the patient developed severe heart failure. At that time, she was six months pregnant and during the first trimester she was prescribed low molecular weight heparin. Both TTE and TEE showed an echogenic mass in the posterolateral wall of the left ventricle in connection with the prosthesis. The suspicion was of tumour relapse with an associated thrombus. She was submitted to new surgery that showed that the valve obstruction was due to fibrous ingrowth and thrombosis. The prothesis was replaced by a St Jude 27 mm valve.

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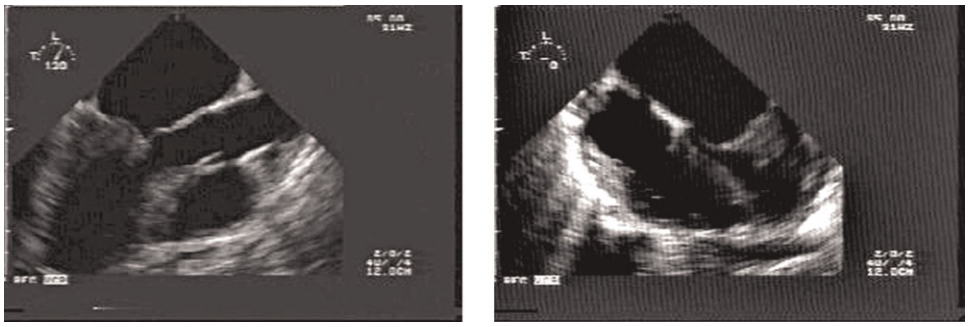

Figure 1 Horizontal (right panel) and 120° (left panel) transoesophageal echocardiogram views showing a very homogeneous echogenic mass coming from both leaflets of the mitral valve and occupying much of the left ventricular inflow tract.

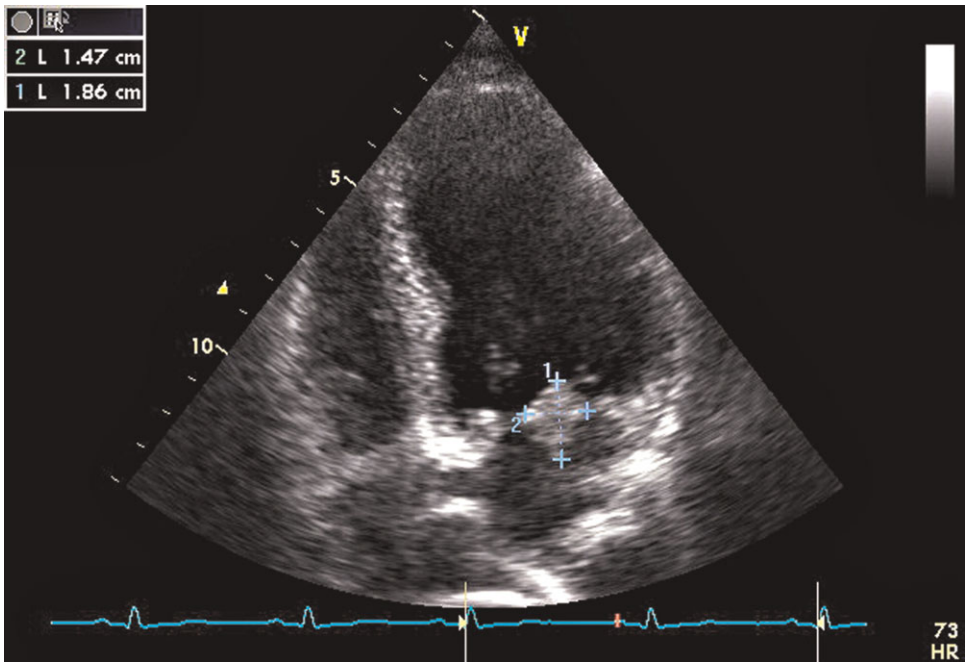

Figure 2 Transthoracic apical four-chamber view showing a rounded homogeneous mass located at the level of the mitral valve.

Ten years thereafter, the patient is doing well, in NYHA class II, with normally functioning mitral prosthetic valve and no recurrence of the tumour.

Case 2

A 72-year-old female patient with episodes of dyspnoea was referred for transthoracic echocardiography at our centre with suspicion of aortic valve stenosis. Her past history was of chronic obstructive lung disease that was compensated at that time. She had no constitutional or neurological symptoms and her physical examination was unremarkable, except for a soft systolic ejection murmur in the second right intercostal space.

Transthoracic echocardiogram revealed an echogenic mass located near the mitral valve annulus that prolapsed into the left ventricle in diastole (*Figure 2*, clip 2). For better characterization, TEE was performed and showed a sessile round mass with regular contours and homogeneous appearance, measuring 2 × 1.1 cm, attached to the atrial side of the posterior mitral valve leaflet and, although it produced some turbulence in the left ventricle inflow, it caused no significant obstruction or regurgitation of the mitral valve (*Figure 3*, clip 3). The echographic features were of myxoma. It was surgically removed with preservation of the mitral valve and, although there was intraoperatory suspicion of malignancy due to the hard consistency, the pathological analysis confirmed the first hypothesis of myxoma. The patient was discharged without any complications and remains well after 12 months of follow-up.

Discussion

Primary cardiac tumours are unusual with a reported incidence between 0.001 and 0.28%, and 75% are benign.[1,2] The most common is cardiac myxoma, typically located in the left atrium at the level of the fossa ovalis.[3,4] Valvular myxomas are extremely rare, the mitral valve being the most frequently involved structure, followed by the tricuspid, the aortic, and the pulmonary valves. Myxoma are usually located at the atrial side, as in our second patient,

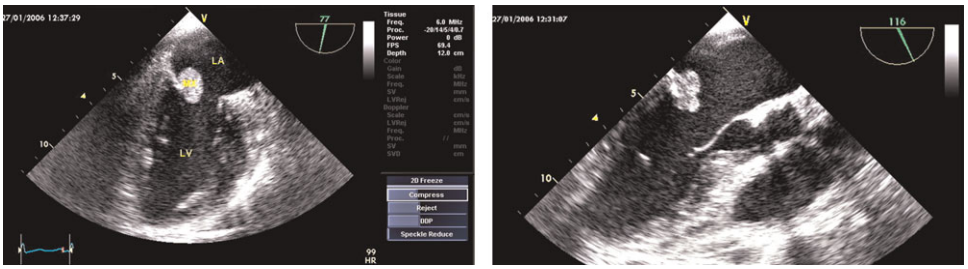

Figure 3 Two-chamber (left panel) and 120° (right panel) transoesophageal views showing a round, lobulated, homogeneous mass arising from atrial side of the posterior leaflet of the mitral valve.

with both leaflets equally affected, being the maximal dimension described, 3.5 cm long.[5,6] In our two cases, the first measured 4 cm, and the second 2 cm. The main differential diagnosis is made with papillary fibroelastoma, which is generally much smaller in size and usually pedunculated. Both our valvular myxomas were sessile, involving mostly the posterior leaflet.

The most common symptoms of cardiac myxoma are dyspnoea and orthopnoea, which these two patients reported. When located at the mitral valve, the obstruction to the left ventricular inflow caused by the tumoural mass simulates mitral stenosis, as seen in our first case. Frequently, patients may have peripheral and/or cerebral embolism, which our patients did not have.[3,4]

The 'gold standard' for non-invasive diagnosis of cardiac myxoma is transthoracic echocardiography, but the transesophageal approach provides a more complete characterization.[4,5] In our series, myxomas represent 74% of all diagnosed cardiac tumours, with 8% of them originating from the mitral valve. It was the only valvular location found during this period. Both patients were female, and at the time of surgery, there were doubts about the benignity of the condition, which may represent the different macroscopic aspect, with harder consistency of the tumour when it involves the valvular structures. Because of the potential for life-threatening complications, urgent complete resection is recommended,[4,6] and it was performed in both cases with excellent results and so far no recurrence. Because of the valvular destruction and tumour adherence to nearby structures, in one patient there was a need of valve replacement. As these two cases had different features, we considered it interesting to report them together.

Conflict of interest: none declared.

Supplementary material

Supplementary data associated with this article can be found in the online version.

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