A Patient with Rheumatic Mitral Stenosis and an Atrial Myxoma

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A 34-year-old woman with exertional dyspnoea and clinical diagnosis of mitral stenosis underwent echocardiographic evaluation. Moderate to severe rheumatic mitral stenosis was confirmed but an unsuspected mass lesion in the dilated left atrium attached to inter-atrial septum in fossa ovalis region was found. Although the attachment was in favour of myxoma, it was difficult to differentiate with certainty between left atrial myxoma and thrombus. Warfarin was prescribed but after 1 year the mass lesion remained unchanged. Both the severity of the mitral stenosis and the presence of the mass lesion led to our decision to proceed with surgery. Pathology showed typical histology of a myxoma.

Key Words: myxoma; mitral stenosis; left atrium.

Introduction

Myxoma is the most frequent cardiac tumour. It usually originates from the fossa ovalis region in the left atrium (75%) but may arise from other locations as well. These tumours are attached to the endocardium by a broad base, and are usually pedunculated, polypoid and friable. They appear as a soft, gelatinous, mucoid, usually grey-white mass, often with areas of haemorrhage or thrombosis[1]. Left atrial thrombus is relatively common in mitral stenosis with a frequency of about 10–25%[2]. The association of left atrial myxoma and rheumatic mitral stenosis is an extremely rare condition and very few cases have been reported[3–5]. We report here a patient with left atrial myxoma and rheumatic mitral stenosis.

Case Report

A 34-year-old woman with exertional dyspnoea and physical diagnosis of mitral stenosis was referred for echocardiography assessment. The left atrium was enlarged. Mitral valve leaflets and chordae tendinea were thickened and their mobility was reduced. Valve area was 1.4 cm² by planimetry and the Doppler study revealed a transmitral mean and a peak diastolic pressure drop of 11 and 17 mmHg, respectively. There was also mild aortic regurgitation. A mass lesion with a broad base attached to the inter-atrial septum in the fossa ovalis region was seen in the left atrium (Fig. 1). Although the site of attachment suggested a myxoma, the diagnosis of a left atrial thrombus, which is common in mitral stenosis and left atrium dilatation was also considered.

Warfarin was started with the intention of dissolving of the mass lesion. One year later, echocardiography showed a slightly larger mass lesion and the mitral valve had become more stenotic. The transmitral mean and peak diastolic pressure drop had increased to 25 and 35 mmHg, respectively (Fig. 2).

Surgery was performed and the mitral valve was replaced with a size 27 mechanical prosthetic Carbomedics valve. The mass lesion was found attached to the inter-atrial septum and excised. Histology showed typical features of myxoma (Fig. 3). Postoperative course was without any complication and the patient was discharged after 1 week. She is doing well and follow-up echocardiography studies showed good prosthesis function and no lesion in the left atrium.
Rheumatic mitral stenosis is one of the most common acquired heart diseases worldwide and is usually diagnosed by physical examination. Echocardiography is the method of choice for assessment of its severity and pre-operative evaluation.

Left atrial thrombus is relatively common in mitral stenosis[2]. It is most often located in the atrial appendage but is occasionally attached to the inter-atrial septum and may mimic a myxoma, which is a rare condition[1]. A small myxoma cannot be diagnosed clinically but a larger myxoma can prolapse into the mitral orifice and be mistaken for mitral stenosis[6].

Echocardiography is the principal method to diagnose an intra-cavitary mass lesion[1,6,7] but the echocardiographic features of a thrombus and a myxoma can be similar[1].

Association of a left atrial myxoma and rheumatic mitral stenosis is extremely rare and very few cases have been reported[1–5]. Co-existing left atrial thrombus and myxoma have also been reported in mitral stenosis[5].

Figure 1. The stenotic mitral valve and mass lesion attached to inter-atrial septum are shown in the apical four-chamber view.

Figure 2. Continuous-wave Doppler recording showing severe mitral stenosis.
Figure 3. Histology of the mass lesion shows individual globular to elongated cells (myxoma cells) within a mucopolysaccharide-rich extracellular matrix.

References


