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Review

Abstract

We report a very rare case of association between mitral valve fibroelastoma and myxomatous disease, in a patient with long history of asymptomatic myxomatous disease, and progressive severe mitral regurgitation. The tumour was an intraoperative echocardiographic transesophageal finding, and was confirmed during surgical evaluation. The most important differential diagnosis of echocardiographic image was infective endocarditis. This information is critical to the surgeon in planning the operative procedure and the preoperative management.

For Peer Review

Introduction

Primary cardiac tumours are extremely rare, with an incidence of 0.28% in the necropsy series. Papillary fibroelastomas (PFEs) are the third most frequent benign tumours of the heart (8-10%), after myxoma and lipoma¹. There are small tumours, with range in diameter from 0.2 to 7 cm^{2, 3}. A comprehensive analysis of 725 cases showed that valvular endocardium was the predominant location of tumour³. The most commonly involved valve is the aortic valve (30%), followed by the mitral valve (20-25%)^{3, 4, 5}. Highest prevalence is in the 8th decade of life. Males comprise 55% of patients³.

No clear risk factor for development of PFE has been reported. Some PFEs are congenital, but most lesions are probably acquired⁶.

Most often diagnosed incidentally, PFEs may embolize, mainly to the cerebral circulation. Clinically, PFEs were associated with transient ischemic attack, stroke, myocardial infarction, sudden death, peripheral embolism and blindness, syncope, pulmonary embolism, and heart failure^{3, 6, 7}. Because of their infrequent occurrence, an appropriate management strategy has been difficult to define.

Case Report

A 45-year old man was admitted to the cardio-surgery department for assessment of the opportunity for surgical correction of a previously known mitral regurgitation due to myxomatous disease. Patient was known to have a 10 years history of asymptomatic myxomatous mitral valve disease, with progressive mitral regurgitation. He was followed annually by transthoracic echocardiography (TTE) in order to establish the correct timing for surgical correction. On admission, he presented with class 2 NYHA symptoms. On examination, he had a grade IV/VI pansystolic apical murmur, without signs of congestion. There were no other pathological findings. Laboratory examination was unremarkable. The electrocardiogram displayed normal sinus rhythm. Lung fields and heart size were normal on chest X-ray examination. Repeated TTE examination found severe eccentric mitral regurgitation (regurgitant volume of 100 ml), in a myxomatous valve with prolapse of both leaflets, flail of the P2-P3 scallops, and mitral annular dilation. Left ventricle was dilated (EDD of 60 mm, ESD of 41 mm, EDV of 190 ml, and ESV of 90 ml), with normal ejection fraction (of 55%), without signs of elevated filling pressure ($E/E' = 10$, $E/Vp = 1.5$). There was no pulmonary hypertension, and good right ventricular systolic function (TAPSE of 27 mm, systolic velocity of the tricuspid annulus of 12 cm/s). He was therefore scheduled for mitral valvuloplasty.

An intraoperative transesophageal echocardiography (TEE), done just before the cardiopulmonary bypass, confirmed the presence, severity, and the mechanism of mitral regurgitation described by TTE (Video1), but also revealed a small, round shape, "phantomathic" image (7x7 mm), with an echo lucent centre, that appeared only in diastole, involving the atrial side of the anterior mitral valve leaflet (A1) (Video2). This image was considered to be either a bulge of the myxomatous anterior

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leaflet, or a tumoral tissue (Figure 1). We could not exclude completely a vegetant mass suggestive for endocarditis. A commercially available sytem was used (Vingmed Vivid 7, and Vivid i, General Electric). Data were obtained using a 3,5-Mhz transducer, and a multiplane transesophageal probe 6T-RS (2.2-8 Mhz).

Left atriotomy and visual exploration of the mitral valve revealed a small translucide tumoral mass on the atrial side of A1 scallop (Figure 2). Surgical procedure consisted of complete resection of the tumoral mass and partial resection of A1 scallop, associated with mitral annuloplasty with complete ring, and three Gore-Tex chordae on the P1, P2, and P3 scallops and both mitral commissures. Intra operative TEE after cardiopulmonary bypass showed a competent valve repair, with minimal residual regurgitation of the level of A1 scallop. Histopatological exam of the tumour revealed benign papillary fibroelastoma. Patient had an uneventful postoperative recovery and was discharged on the 8th postoperative day. Warfarin was recommended for 3 months.

Discussion

We reported a rare case of myxomatous mitral valve disease with severe regurgitation, associated with fibroelastoma. PFEs are rare, benign tumours. The incidence is not clear because most patients are asymptomatic. They usually occur in elderly patients (>65 years), and are seen rarely in children^{5, 8}. PFEs have a valvular predilection, and the mitral valve is the second common location. When the mitral valve is involved, the tumour is most often on the atrial mid portion side of the valve, away from the free edge or the annulus⁵. Particularly, in our patient, we found an uncommon location, on the A1 scallop of the anterior mitral leaflet, closely related to the antero-lateral commissure.

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15 In conclusion, we have reported a very rare association of PFE and myxomatous
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Figure 1 legend

TEE showed a round (7x7 mm), mobile tumour attached to the atrial side of the anterior mitral leaflet (A1 scallop). Image is visible only in diastole, as a camber of the anterior leaflet (arrow).

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Intraoperative exploration of the mitral valve showed a 10x10 mm translucent yellow tumoral mass on the atrial side of the A1 scallop (arrow), with a large base of implantation.

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The chronic trauma of the endocardium appears the most important mechanism in the pathogenesis of PFE⁴. In our case, we speculate that long-standing mitral disease, with turbulent blood flow and mechanical trauma, might be the mechanism for tumour development.

There are no cases reported in which PFEs was considered responsible for valvular dysfunction⁴. Similarly, in our patient, mitral regurgitation was produced by mitral valve prolaps, tumour having no implication.

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propensity to embolize. Tumoral resection should proceed on an emergency basis,
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After operation, it is common practice to put patients on warfarin. However, there are
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In conclusion, we have reported a very rare association of PFE and myxomatous
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Figure 1 legend

TEE showed a round (7x7 mm), mobile tumour attached to the atrial side of the anterior mitral leaflet (A1 scallop). Image is visible only in diastole, as a camber of the anterior leaflet (arrow).

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Intraoperative exploration of the mitral valve showed a 10x10 mm translucent yellow tumoral mass on the atrial side of the A1 scallop (arrow), with a large base of implantation.

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Figure 1
TEE showed a round (7x7 mm), mobile tumour attached to the atrial side of the anterior mitral leaflet (A1 scallop). Image is visible only in diastole, as a camber of the anterior leaflet (arrow).
160x106mm (96 x 96 DPI)



Figure 2

Intraoperative exploration of the mitral valve showed a 10x10 mm translucent yellow tumoral mass on the atrial side of the A1 scallop (arrow), with a large base of implantation.
183x149mm (96 x 96 DPI)