Rheumatic Mitral Valvopathy and Papillary Fibroelastoma An Unusual Association

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Introduction

Papillary fibroelastoma (PF) is the second most common primary cardiac tumor, predominantly affecting the cardiac valves and accounting for three-quarters of all cardiac valvular tumors.[1] Most patients with PF are asymptomatic and the evolution of echocardiography has allowed for an earlier detection and better characterization of PF. Although rare and benign, PF may result in multiple complications, such as stroke and arterial embolisms.[1]

Echocardiography is fundamental for the follow-up of asymptomatic patients with PF. Surgery to remove these tumors should be indicated when the patient reports symptoms, progresses with some complication or when the tumor becomes mobile.[1]

This report describes an unusual association between PF of aortic valve found in a patient with mitral valve rheumatic disease. When we analyze the literature, very few cases are observed with such association, which increases the importance of publications related to this topic in order to call attention to a careful evaluation of the echocardiograms.[2]

Case Report

A 52-year-old woman diagnosed with rheumatic mitral valvular disease, atrial fibrillation and previous history of a surgery (mitral valve repair in 1998), followed up at the valvopathic outpatient clinic of Instituto do Coração do Hospital das Clínicas da Faculdade de Medicina da Universidade de São Paulo (USP), had been evolving with dyspnea on minimal exertion (New York Heart Association – NYHA – functional class III), associated with lower limb edema. The patient was taking furosemide, digoxin and warfarin.

Physical examination showed heart rate of 66 bpm, blood pressure of 110/70 mmHg, irregular pulse, presence of jugular turgency with venous pulsation showing a giant V wave, second-sound hyperphonesis (B2), opening sound of the mitral valve, regurgitant mitral systolic murmur (1+/6+) and diastolic murmur in a mitral rhythm (4+/6+) with the presence of pre-systolic reinforcement.

Keywords

Neoplasms; Mitral Valve Insufficiency; Rheumatic Fever; Echocardiography, Transesophageal; Mitral Valve Stenosis.

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Transthoracic echocardiography revealed classic findings of rheumatic mitral valve disease: commissural fusion, thickening of the cusps, and reduction of the valve opening. Maximum diastolic gradient between the left atrium and left ventricle estimated at 30 mmHg and medium at 11 mmHg. The area of the effective valve orifice was estimated to be 0.8 cm^2. The atria presented significant dilatation. There were signs of marked pulmonary hypertension (systolic pulmonary artery pressure estimated at 90 mmHg) and moderate to severe tricuspid regurgitation. In association, a filamentary image was identified on the aortic side of the aortic valve (Figure 1). The aortic valve was functionally normal.

Intraoperative transesophageal echocardiogram confirmed the presence of filamentous image on the aortic side of the aortic valve, measuring approximately 6 mm, with PF as one of the diagnostic possibilities. The finding of the intraoperative anatomical piece, identified by the surgeon, was consistent with this possibility (Figure 2). Both valves (aortic and mitral) were replaced by biological prostheses. Anatomopathological examination confirmed the mitral rheumatic valvular disease and showed a pedunculated papillary lesion on the free edge of an aortic semilunar, measuring 4 mm, diagnosed as papillary fibroelastoma (Figure 3). The aortic valve did not present any rheumatic lesions.

The patient evolved well in the postoperative period and was discharged asymptomatic.

Discussion

PF is a rare type of endocardial papilloma that predominantly affects the heart valves, with the aortic valve being most frequently affected, followed by the mitral valve and rarely affecting the right valves.[1] Primary heart tumors are rare,[3,4] and PFs account for about 4.4% to 8% of primary cardiac tumors,[5,6] and may be asymptomatic or cause serious complications through cerebral or coronary embolism.[7]

In symptomatic individuals presenting angina, stroke, heart failure, syncope, blindness or other symptoms, surgical resection is recommended to avoid future thromboembolic events.[8] When such tumors are found incidentally in a preoperative evaluation for surgery, they are removed to avoid unexpected complications in the future.[8] However, the primary surgical indication in asymptomatic individuals is still controversial.[8]

There are several mechanisms that try to explain the development of papillary fibroelastomas, considering them as hamartomas, a product of the organization of microthrombi that coalesce like the Lambl excrescences, or a non-specific reaction process to the aggression of the valvular endothelium. PFs have two layers, a hyperplastic external endothelial layer and a dense central one consisting of an acellular fibrous axis that is rich in elastic fibers, continuous to the leaflet valve.[9]

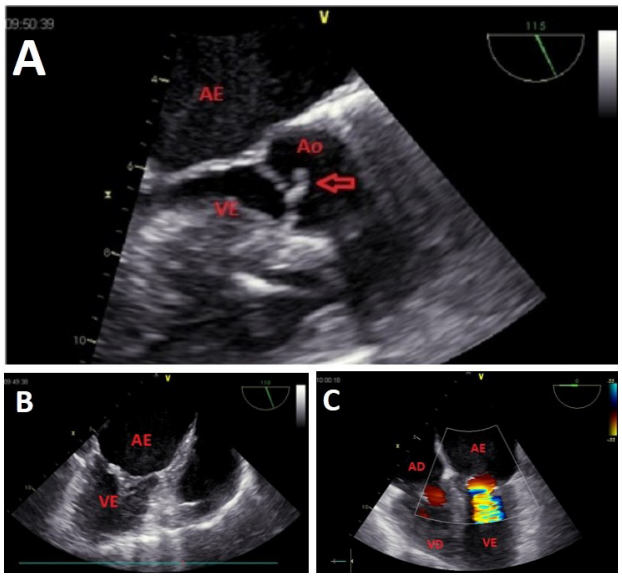

Figure 1 – *Intraoperative transesophageal echocardiography. (A) Image suggestive of papillary fibroelastoma in the aortic valve (arrow). Severe rheumatic mitral stenosis is demonstrated. (B) severe valve thickening. (C) Acceleration of transvalvar flow on color Doppler.*

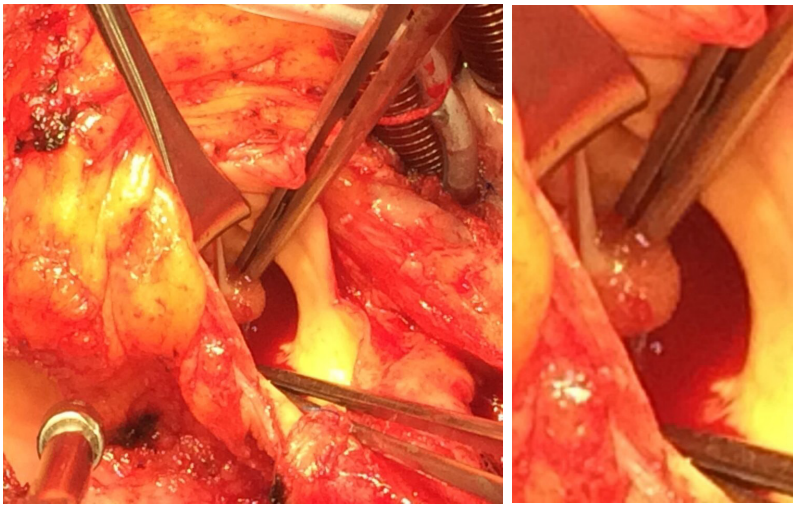

Figure 2 – *Surgical exploration of the aortic valve, identifying the fibroelastoma. On the right, zoomed in.*

Surgical treatment can be performed conservatively, with excision of the tumor, sparing the valve, or in a non-conservative way, by performing a plastic surgery or valve replacement with a prosthesis implant.[10] Valve preservation surgery is usually safe and curative.[10] However, in some cases, especially when there is significant impairment of the valve, one can choose the exchange.[10] Among the patients affected, an 83% rate of exclusive tumor resection has been reported, without valve surgery. In the other cases, which are the minority, tumor excision can be performed with concomitant valve repair or valve replacement with a prosthesis implant.[10]

Three-dimensional transesophageal echocardiography can help in the proper characterization of the tumor, allowing a more appropriate therapeutic decision.[11]

Conclusion

This report described an unusual association between papillary fibroelastoma of aortic valve found in a patient with rheumatic mitral valve disease. Surgical excision of the papillary fibroelastoma was chosen and the mitral and aortic valves were replaced, with good postoperative evolution.

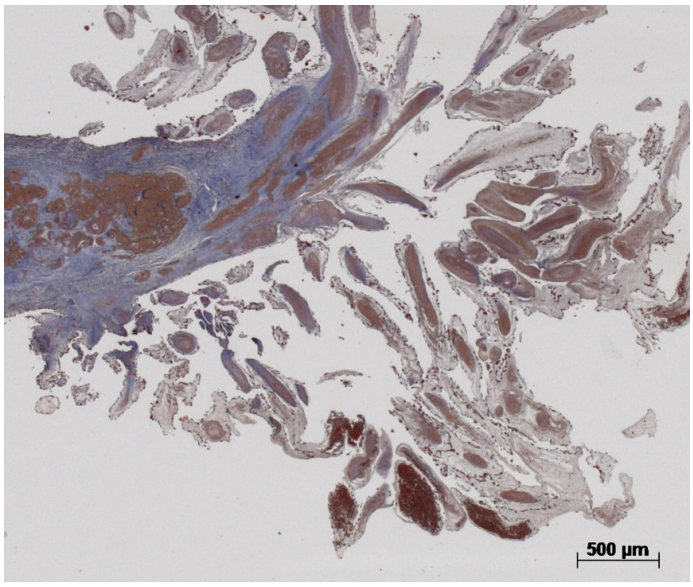

Figure 3 – *Histological section of the aortic valve lesion stained by Masson's trichrome showing papillary projections coated by hyperplastic endothelial cells, typical of papillary fibroelastoma.*

Authors' contributions

Data acquisition: Mustafé RM, Veronese ET, Brandão CMA, Lima MSM; Data analysis and interpretation: Lima MSM; Manuscript drafting: Mustafé RM, Lima MSM; Critical revision of the manuscript as for important intellectual content: Mustafé RM, Lima MSM.

Potential Conflicts of Interest

There are no relevant conflicts of interest.

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Academic Association

This study is not associated with any graduate programs.

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