Tumoration in Anterior Leaflet of the Mitral Valve

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INTRODUCTION

This is the case of a 47-year-old male who developed mild dyspnea on great exertion. Routine transthoracic echocardiography (TTE) revealed a slightly thickened mitral valve with anterior leaflet prolapsed and mild incompetence. There was an echogenic image suggestive of an irregular and mobile tumor adhered to the ventricular face of the anterior mitral valve leaflet, measuring about 17 mm by 10 mm. The movement of the tumor contributed to a dynamic increase in the left ventricular outflow gradient, estimated at 33 mmHg. The transesophageal echocardiogram (TEE) performed 14 days later showed prolapse of the anterior mitral valve leaflet, which presents a movable filamentary mass, measuring 13 mm by 3 mm, located at its end on the ventricular surface, and which protruded during systole via the left ventricular outlet, causing dynamic obstruction with peak systolic gradient of 59 mmHg. Doppler revealed mild to moderate reflux (Figure 1). Surgery for mitral replacement or repair with tumor resection was performed with replacement of the mitral valve by bioprosthesis 29 (Figure 2). The fragment of the anterior leaflet was submitted to histological examination and immunohistochemical study (Figure 3).

QUESTIONS

A) What are the diagnostic possibilities and their characteristics?
B) Regarding imaging techniques, were they the most suitable for diagnosis?
C) What was decisive for surgical treatment? Does the surgical indication depend on the presence of symptoms?
D) Is there a need for anticoagulation and association of arrhythmias in this type of tumor?

Discussion of Questions

QUESTION A. The main diagnostic possibilities would be cardiac myxoma (CM), chordae rupture, papillary fibroelastoma, vegetation, and the redundant cusp itself. CM should be suspected in patients with chronic dyspnea, history of peripheral or systemic embolization, acute neurological disorder, and family history of CM⁵. However, approximately 3.2% to 46.4% of patients with CM are asymptomatic⁶-⁹. CM is a rare benign cardiac tumor, and its incidence is observed in approximately 0.5-1 case per 1 million people per year².⁷-¹⁰. Among primary
cardiac tumors, myxoma is responsible for 50% of cases\(^6\), whose average age range varies from 42 to 66 years\(^{11,12}\).

**QUESTION B**. The echocardiogram is the main modality of diagnostic imaging in the morphological evaluation of intracardiac tumors. TTE is defined as a first-line imaging modality for assessing valve anatomy, severity of mitral regurgitation, hemodynamics, and ventricular consequences. When TTE does not add value to the diagnosis, TEE is always advocated as an enhancement for additional diagnosis\(^{13}\). However, the three-dimensional (3D) echocardiogram provides a better characterization and morphological classification according to the microscopic appearance of the surface of these intracardiac masses, with good correlation with surgical and histopathological findings\(^{13}\), but the need for greater training of operators, cost, and high dependence on image quality currently limit the wide application of 3D in the oncology scenario\(^{14}\). Studies have shown that two-dimensional and 3D techniques are equally reliable, and that 3D TEE has an advantage in locating the disease\(^{15-18}\).

**QUESTION C**. For symptomatic patients, surgical exeresis is the treatment of choice, always trying to preserve the valve tissue and its function. In asymptomatic individuals, surgical management is controversial, with tumor mobility being the determining factor for surgical indication, as it is an independent predictor of embolization and death\(^{19-21}\). Due to malignant potentials, timely surgical resection is guaranteed in all patients, without contraindication when making the diagnosis, as the tumor’s behavior cannot be predicted and does not depend on how large and fragile it is.

**QUESTION D**. The follow-up of asymptomatic patients who do not undergo surgery should include echocardiographic monitoring and anticoagulation therapy, although its

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**Fig. 1** - (A) Mitral valve partially closed with hyperechogenic image (red arrow) adhered to its anterior leaflet (esophageal image with long axis at 120 degrees); (B) mitral valve during ventricular systole with a hyperechogenic image (red arrow) adhered to its anterior leaflet, partially obstructing the LVOT (120-degree long-axis esophageal image); (C) acceleration of systolic flow due to partial obstruction of the LVOT (long-axis esophageal image at 120 degrees); (D) hyperechogenic image (red arrow) adhered to the anterior mitral valve cusp partially obstructing LVOT in systole (esophageal image of 4 chambers at 0 degrees). AO=aorta; LA=left atrium; LV=left ventricle; LVOT=left ventricular outflow tract.

**Fig. 2** - (A) Mitral ring exposed by the left atriotomy, separate vertical “U” suture to fix the bioprosthesis; (B) wires passed through the bioprosthesis; (C) final aspect of the bioprosthesis fixed in the correct position.
effectiveness in protecting against embolic phenomena is controversial\textsuperscript{[23,24]}\textsuperscript{a}. Embolization is more likely to occur in mobile papillary cardiac tumors, which are smaller in size, and in patients who have preoperative atrial fibrillation, and it is necessary to treat embolic events with anticoagulation to avoid serious events such as stroke, an embolism in the peripheral vasculature, or an embolism in the pulmonary artery, caused by detached tumor tissue or mobilization of thrombotic deposits. Usually the occurrence of arrhythmias, mainly extrasystoles and atrial fibrillation, is due to some obstruction of the physiological blood flow\textsuperscript{[23,24]}\textsuperscript{a}.

**BRIEF CONSIDERATION OF THE CASE REPORTED**

Primary cardiac tumors are rare, with a prevalence between 0.0017% and 0.19% of the unselected autopsy studies. About 75% are benign tumors and almost half are myxomas. The rest are divided among rhabdomyomas, lipomas, and fibroelastomas.

Due to its predominant involvement in cardiac valves, nonspecific symptoms, and characteristics of its shape and size, the chordae rupture, papillary fibroelastoma, vegetation, and the redundant cusp itself have become suggestive in this clinical case.

It is important to note that obstruction of the heart valve caused by CM, for example, can lead to catastrophic consequences, including sudden death. A fragment of the tumor can embolize to the left or right coronary ostium and result in acute coronary syndrome, and surgical resection has a low rate of complications and mortality. This detail was also important for the decision for surgical treatment.

**Fig. 3** - Anatomopathological study (A, 4× magnification; B, 40× magnification) – Chronic degenerative valve disease, at the expense of hyalinized fibrosis and myxoid degeneration of the stroma. *Fibrosis (pink color), myxoid degeneration of the valve stroma (blue color). Immunohistochemistry of valve tissue (C, D, E, and F) – chronic valve disease with fibrosis and stromal myxoid disorder. *CD3, rare T lymphocytes; CD20, rare B lymphocytes; CD34, stromal positivity; CD68, negative; Ki-67 antigen, low index of cell proliferation.

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**REFERENCES**


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