

The Importance of Imaging Multimodality in the Diagnosis of a Rare Case of Papillary Fibroelastoma in the Left Ventricular Apex

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Introduction

Papillary fibroelastoma (PF) is a rare, benign primary cardiac neoplasm, initially described as an incidental finding in autopsies or surgeries, and ranks third among the most common cardiac tumors. It mainly affects the heart valves, accounting for 75% of all tumors in this region and is often diagnosed incidentally.¹

Its clinical presentation can vary widely, ranging from asymptomatic cases to situations in which systemic embolization occurs, caused by adherent thrombi as well as by fragmentation of the tumor itself. Currently, PF is no longer a tumor diagnosed only in autopsies and can now be detected with new cardiovascular imaging modalities, thus enabling early treatment and preventing complications.²

Our study presents a clinical case report of a patient with PF located at the apex of the left ventricle, in which the diagnosis was suggested during a transthoracic echocardiogram. Hence, our work demonstrates the importance of the multimodality of cardiovascular imaging in identifying primary cardiac tumors, such as PF. Furthermore, the importance of differential diagnoses, propaedeutic aspects, and the therapeutic approach to PF was addressed.

Case Report

Our study presents the case of a 53-year-old female patient with a previous history of transient ischemic attack (TIA). The clinical examination was normal, and the electrocardiogram was in a proper sinus rhythm. In the preoperative assessment of lumbar denervation, a transthoracic echocardiogram (TTE) was requested, which revealed an oval mass, slightly more echogenic than the myocardium, homogeneous, with a smooth, mobile surface, located at the apex of the left ventricle, which could correspond to a thrombus or tumor (Figure 1). The TTE was complemented with ultrasound contrast to better characterize the mass, ruling out the possibility of a

thrombus, confirming that it was an apical, pedunculated tumor, with homogeneous contrast distribution, measuring 0.9 cm x 1.0 cm, with a peduncle of 0.6 cm in length and 0.2 cm in thickness (Figure 2), suggestive of PF. Subsequently, a cardiac magnetic resonance imaging (CMRI) scan was performed, which identified a homogeneous pedunculated mass at the apex of the left ventricle, measuring 1.0 x 0.8 cm, isointense on T1 and T2, with no fat saturation, without first-pass perfusion, showing late peripheral enhancement “in a peripheral halo” (Figure 3), compatible with PF. The patient underwent heart surgery, and an oval, pedunculated tumor was identified at the apex of the left ventricle (Figure 4). After surgical excision of the tumor, the histopathological study revealed that it was a neoplasm comprised of avascular bundles with fibroelastic content and endocardial lining, consistent with the diagnosis of PF.

Discussion

The diagnosis and treatment of cardiac masses are a real challenge, although they are rarely found in clinical practice.³ The incidence of primary cardiac tumors is approximately 0.017% to 0.20%, half of which are myxomas and nearly 75% are benign.² PF can be found in different regions of the heart, representing around 10% of all cardiac tumors. Typically, this type of tumor affects the heart valves, and is more common in the valves on the left side of the heart, especially the aortic valve (29%) and the mitral valve (25%), compared to the pulmonary valve (13%) and the tricuspid valve (17 %). However, it can appear on any endocardial surface, including non-valvular regions,⁴ as observed in the case described above.

The occurrence of PF in the apex of the left ventricle is considered to be extremely rare. According to a study conducted by Sovic et al.,⁵ from 1997 to 2018, only 13 cases of PF in the apical region were described in the medical literature.

In most cases, the tumor is single, pedunculated, and avascular, and has an average size of approximately 1 cm in diameter, but it can vary between 0.2 and 4.6 cm, and is attached to the endocardium by a short, thin rod. Microscopically, this pathology consists of avascular connective tissue covered by an endothelium, with an appearance similar to “sea anemone”.⁶

In the study by Saleh et al.,⁷ 14 patients were found with 18 lesions (mean age of 60.5 ± 12.3 years), resembling the age of the patient in this report. Eleven patients (79%) were symptomatic. Most lesions were solitary, with a diameter of ≤1.5 cm, half involving the left side of the heart.

Generally, PF is asymptomatic and, in most cases, is diagnosed incidentally during imaging tests or surgeries for other causes. The clinical manifestation occurs mainly due

Keywords

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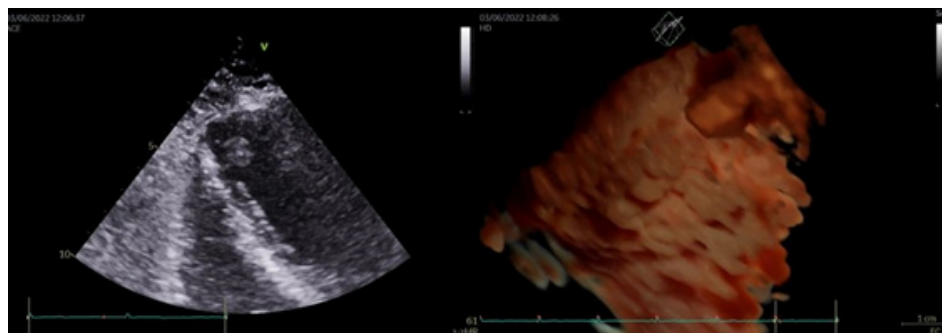


Figure 1 – Two-dimensional and three-dimensional transthoracic echocardiogram (TTEcho): a rounded structure was identified, slightly more echogenic than the myocardium, homogeneous, measuring 1.2 x 1.2 cm, with a smooth, mobile surface, located at the apex of the left ventricle, with a velvety appearance, which may correspond to a thrombus or a tumor.

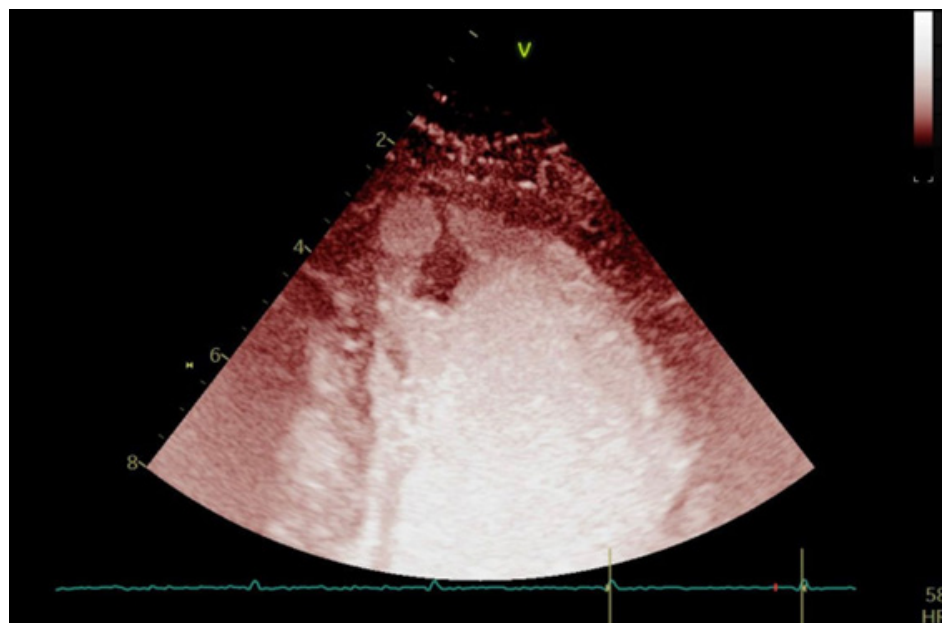


Figure 2 – Contrast echocardiogram, showing the presence of a tumor at the apex of the left ventricle, with benign characteristics, suggestive of papillary fibroelastoma.

to embolization, often cerebrovascular. In the case reported herein, the patient had a previous episode of TIA. PF can cause peripheral embolisms and an obstruction of the coronary ostia, which can begin as a myocardial infarction, especially in the aortic valve. Differential diagnosis must be made with clots, calcium, vegetation, or other foreign bodies.⁸

Symptomatic patients or those with pedunculated tumors should undergo surgical resection as the treatment of choice, aiming to prevent embolic phenomena. In asymptomatic individuals, surgery is controversial, with tumor mobility being the determining factor for surgical intervention. Therefore, asymptomatic patients with non-mobile tumors can be

monitored in periodic clinical evaluations. Recurrence is rare, and has been only described in a few cases, highlighting the relevance of monitoring with TTE for identification.^{2,9}

Advances in non-invasive diagnostic imaging techniques have played a key role in evaluating and identifying an increasing number of intracardiac tumors more quickly and more effectively, even in the case of those that are poorly defined and small in size. Among these techniques, TTE stands out as a non-invasive method with greater access and high accuracy in detecting cardiac tumors, presenting a specificity of 87.8% and a sensitivity of 88.9%.² Ultrasound contrast allows for a better visualization of the structures

Case Report

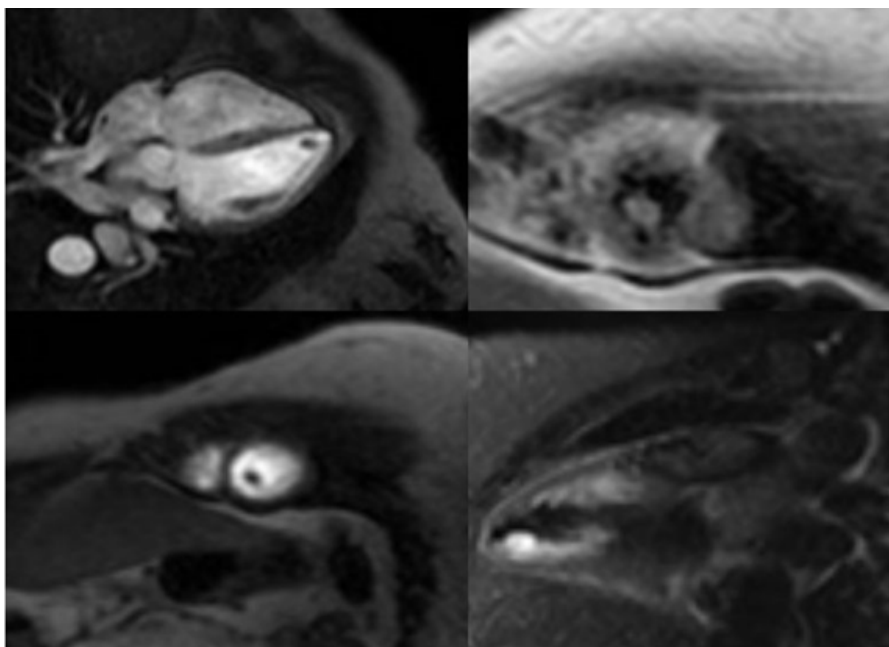


Figura 3 – Cardiac magnetic resonance imaging (CMRI), showing a homogeneous pedunculated mass at the apex of the left ventricle, measuring 1.0 x 0.8 cm, isointense on T1 and T2, with no fat saturation and no first-pass perfusion, showing late peripheral enhancement “in a peripheral halo”.

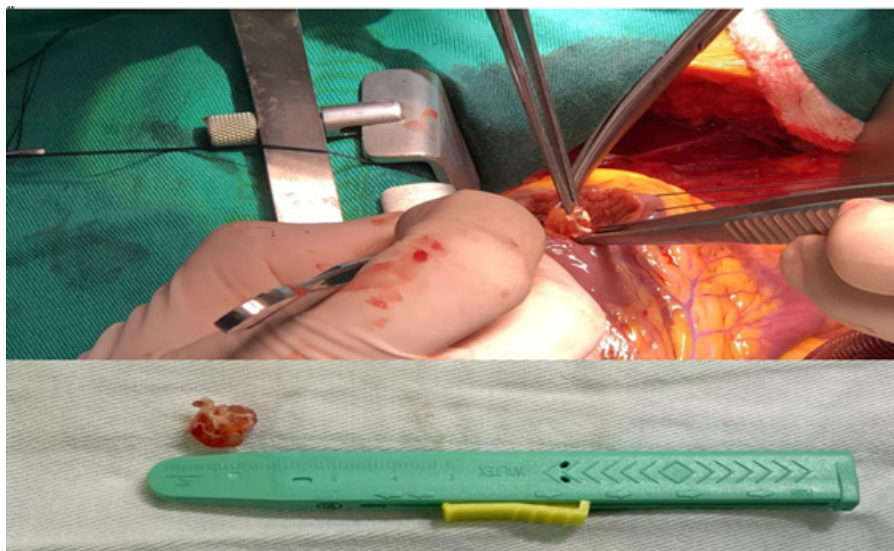


Figure 4 – Oval tumor, measuring 1.0 x 0.8 cm, pedunculated at the apex of the left ventricle.

within the cavity and the assessment of their vascularization. The difference in blood flow in cardiac masses can help distinguish between vascular or non-vascular thrombi or tumors through the contrast distribution pattern.¹⁰ CMR evaluates key characteristics, including morphology,

dimensions, location, extension, homogeneity, and presence of the infiltration of surrounding tissues. Furthermore, it also analyzes signal characteristics that can assist in histopathological characterization, such as fatty infiltration, necrosis, hemorrhage, calcification, and vascularization.¹⁰

Conclusion

This report shows that PF in the apex of the left ventricle is rare, as evidenced by the scarcity of reports in the medical literature. The diagnosis is generally made incidentally. Although it is benign and asymptomatic in most cases, it is important to perform differential diagnoses in order to choose the appropriate treatment and prevent complications, such as embolic events. Non-invasive cardiovascular imaging techniques have been very useful in diagnostic evaluations and play an important role in identifying PF. The present case highlights the importance of using different imaging modalities to diagnose and plan PF surgery.

Author Contributions

Conception and design of the research: Pereira MM, Pereira LSM, Melo Filho JX; acquisition of data: Pereira MM, Melo Filho JX, Juliano MTH; analysis and interpretation of the data and writing of the manuscript: Pereira MM, Pereira LSM; critical revision of the manuscript for intellectual content: Pereira MM, Melo Filho JX, Juliano MTH, Melo RJL, Nina VJS; patient management during hospitalization: Juliano MTH; cardiac MRI: Melo RJL; surgical treatment: Nina VJS.

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Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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Study Association

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Ethics Approval and Consent to Participate

This study was approved by the Ethics Committee of the Hospital e Maternidade São Domingos under the protocol number 6.482.701. All the procedures in this study were in accordance with the 1975 Helsinki Declaration, updated in 2013. Informed consent was obtained from all participants included in the study.



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