



EDITORIAL COMMENT

Cardiac papillary fibroelastoma: So small and yet so dangerous



Fibroelastoma papilar cardíaco: tão pequeno e no entanto tão perigoso

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Cardiac papillary fibroelastoma (PFE) is the third most frequent tumor of the heart after atrial myxoma and lipoma, and is the most frequent tumor of the cardiac valves.¹ Before the advent of echocardiography, these tumors were found incidentally at autopsy and called “small harmless bodies”. The first reported pre-mortem case was an incidental finding during surgical ventricular septal defect repair in 1979. However, while histologically benign, PFE is a dangerous condition due to the potential for cerebral and coronary embolization. While in around two thirds of cases diagnosis is made fortuitously by echocardiogram, one third are diagnosed following an embolic event that triggers further exploration.²

PFE are single in 90% of cases and more than 95% are located in the left heart. Once diagnosed, surgical resection is recommended to avoid embolic complications; this constitutes definitive treatment as recurrence is very rare.

PFE consists of an endocardial layer covering a dense mesh of fibroelastic bundles and a loose avascular connective tissue matrix. Its etiology is unknown and its histopathogenesis is the subject of debate, some theories favoring a hamartomatous origin while others suggest a virus-induced tumor mechanism. PFE are soft, white to tan, and friable, often with adherent thrombus, which explains their propensity to embolize.

In this issue of the *Journal*, Rodrigues et al.³ report a series of 26 patients with cardiac PFE in whom the tumor was the primary indication for surgery. The authors are to be congratulated for drawing our attention to this peculiar benign cardiac tumor, because of its potentially severe complications. The imaging characteristics of this tumor should be well known as surgical resection is simple, effective and the only method that prevents the dreadful risk of embolization.

With the increasing use of transthoracic echocardiography (TTE) for screening purposes and for a multitude of other reasons, the detection of anomalous intracardiac masses has increased. It is therefore important to know the imaging features that can help establish the differential diagnosis with other intracardiac tumors and masses.

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Multimodality imaging plays a key role in the assessment of cardiac masses in general and of PFE in particular, allowing its diagnosis and often establishing the differential diagnosis, based on location, attachment, size, borders, mobility, enhancement, vascularity and metabolism, among other factors.⁴ Because of their availability, low cost, and absence of radiation or contrast, together with their good match between spatial and temporal resolution, TTE, and especially transesophageal echocardiography (TEE), are the most important imaging techniques in the assessment of PFE.⁵ The typical morphological appearance of the mass (like the fronds of a sea anemone, with a shimmering border, a diameter of 5-40 mm, with a short stalk, usually attached to a valve in the left heart valve, and independently mobile), is usually well depicted by two-dimensional images. Additionally, the use of three-dimensional technology often adds further anatomical information, such as correct spatial orientation and the attachment point, that is useful in surgical planning.⁶

Despite the better spatial resolution of cardiac magnetic resonance (CMR) and cardiac computed tomography (CCT), the potential advantages of these techniques in the assessment of valvular PFE are reduced by their low temporal resolution, an important limitation when assessing rapidly moving structures like PFE. However, tissue characterization by CMR (with late gadolinium enhancement in T1- and T2-weighted sequences) and CCT may be useful to differentiate PFE from other cardiac masses, helping to provide a more accurate differential diagnosis. In rare cases, when differential diagnosis is needed between a PFE and a malignant valve tumor, positron emission tomography (PET)/CMR or PET/CCT often provides the correct diagnosis, differentiating benign from malignant tumors based on different metabolic and anatomical data.⁷

In the series by Rodrigues et al., patients were identified from their center's cardiovascular surgery and pathology databases, and so false positive and false negative results of TTE are unavailable.³ We therefore do not know how many patients had masses that were undetected, which would have been of interest. Healed vegetations of infective endocarditis, myxomas, Lambl excrescences, and even mobile thrombi, can mimic the echocardiographic findings of a cardiac PFE. It is also important to rule out a history of bacterial or non-bacterial endocarditis, antiphospholipid syndrome and lupus erythematosus, particularly in older patients with associated multiple morbidities, including atrial fibrillation, and with a higher baseline stroke risk. In a large Mayo Clinic series, a quarter of patients had cardiac PFE detected by TEE but not by TTE, reinforcing the crucial role of TEE when exploring the cause of embolic phenomena.⁸ Unlike most series, which report the aortic valve as the most common PFE location, Rodrigues et al. observed 14/25 cases (55%) of PFE on the mitral valve, 9/26 (35%) on the aortic valve and only two that did not involve a cardiac valve.³

Although cardiac PFE are benign, they can have a malignant behavior due to their embolic potential.^{3,8} In the present report, PFE presented with neurological deficits in eight cases (who had undergone echocardiography to exclude a cardiac embolic source). The authors could not identify any imaging characteristic differentiating between the eight patients who presented with stroke and the

18 patients who were asymptomatic, except that the former were significantly younger (42 ± 17 years vs. 54.3 ± 18.4 years for the overall population), confirming that size, mobility or location of the tumor cannot be relied upon to exclude stroke risk.⁸ This may be due to their small sample size, as a review of 725 cases showed that tumor mobility was the only independent predictor of PFE-related death or non-fatal embolization.⁹ As the tumor is usually single and still attached to the cardiac structures after the neurological event, the mechanisms of stroke or transient ischemic attack (TIA) in patients with PFE are poorly understood; they may be related to tumor fragments or thrombi attached to the tumor. It would have been interesting to know the clinical characteristics of neurologically symptomatic patients (type and severity of stroke or TIA, and the timing of surgery after stroke).

The authors were able to perform valve-sparing operations in all 24 patients with a valvular tumor location. During a median follow-up of around five years, there were no deaths or recurrence of embolic events. However, as there was no systematic echocardiographic follow-up, it is uncertain whether there was tumor recurrence. Although rare, recurrence has been reported in 1.6% of cases, stressing the importance of TTE follow-up.⁸ There is also evidence that the risk of subsequent cerebrovascular accident (CVA) is greater in patients with echocardiography-identified but unoperated PFE, compared with an age- and gender-matched population, and that excision substantially decreases CVA risk and even mortality from PFE.⁸

In summary, it should be stressed that echocardiography is not 100% accurate in making a tissue diagnosis of intracardiac masses, highlighting the role of a multimodality imaging approach. Young patients without significant comorbidities and a high diagnostic suspicion of cardiac PFE should be offered surgical resection, independently of symptoms, particularly if the tumor is mobile. The surgical risk is low, valve repair can be achieved in virtually all patients and the risk of embolic stroke is reduced. For older or sicker patients, antiplatelet or anticoagulant therapy have been proposed, but supporting data are limited and therefore the final decision should be based on discussions by the heart team.

Conflicts of interest

The authors have no conflicts of interest to declare.

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