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CASE REPORT

A Rare Case of Large Nonvalvular Papillary Fibroelastoma Manifesting as Vertigo and Exertional Dyspnea

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Abstract

Background: **Papillary fibroelastoma (PFE) is an uncommon benign cardiac tumor, typically arising from valvular structures. Nonvalvular PFEs are exceedingly rare and may present with diverse symptoms.**

Case presentation: **We present a case of a 46-year-old woman with positional vertigo and exertional dyspnea. Transthoracic echocardiography revealed a mobile oval-shaped mass in the left ventricle. Cardiac magnetic resonance imaging suggested a large papillary fibroelastoma. The tumor was successfully resected, and histopathological examination confirmed the diagnosis.**

Conclusion: **This case highlights the importance of thorough imaging and surgical intervention in managing large, nonvalvular PFEs, which, although rare, can lead to significant complications.**

Keywords: Papillary fibroelastoma, Cardiac tumor, Cardiac mass, Echocardiography, Cardiac magnetic resonance, Case report

1. Introduction

Cardiac tumors are rare, with a prevalence of 0.02% in autopsy studies [1]. According to a comprehensive systematic review and meta-analysis conducted by Rahouma et al., benign primary cardiac tumors (PCTs) constitute the vast majority of cardiac tumors, accounting for approximately 84.6% of all cases analyzed and the prevalence of malignant primary cardiac tumors (PMCTs), in contrast, was reported to be significantly lower, comprising only 10.83% of all PCTs [2]. Papillary fibroelastomas (PFEs) and cardiac myxomas are the most prevalent types of primary cardiac tumors [3].As PFEs progress, they may increase in size and have the potential to dislodge, resulting in various complications, including stroke, heart attack, arrhythmia, sudden cardiac death, or obstruction within the heart, leading to symptoms like fainting, heart failure, or sudden cardiac death [4,5]. However, PFEs commonly manifest without symptoms and are typically detected incidentally through procedures like echocardiography, computed tomography scans, open-heart surgeries, or post-mortem examinations. They are generally fragile and exhibit slow growth [5].

Surgical removal of PFEs is advised for asymptomatic patients, irrespective of their location within the heart, particularly when the tumor exceeds 1 cm in size, displays a pedunculated structure, and exhibits significant mobility. This approach aims to minimize the risk of potential embolization [6]. Studies have shown that minimally invasive approaches can be effective for the resection of cardiac tumors, including PFEs, with outcomes comparable to those of traditional open surgeries [7]. In this

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case, the traditional open surgery approach was used due to the tumor's size and attachment, but minimally invasive techniques could be considered in cases where the tumor's location and size allow for such an approach.

While PFEs typically originate from the aortic valve, they can also occur on the mitral leaflets [3,8]. However, occurrences in multifocal and nonvalvular sites are exceptionally uncommon [8]. In this report, we describe a case of PFE in the left ventricle (LV) that manifested with symptoms of exertional dyspnea and positional vertigo.

Abbreviation list

PFEs	Papillary fibroelastomas
LV	left ventricle
CT	computed tomography
TTE	Transthoracic echocardiography
CMR	Cardiac magnetic resonance
SSFP	steady-state free precession
STIR	short-tau inversion recovery
FSE	fast spin-echo
PCTs	benign primary cardiac tumors
PMCTs	malignant primary cardiac tumors

2. Case presentation

A 46-year-old woman with a medical history of arterial hypertension, dyslipidemia, and kidney angiomyolipoma presented to our department with a 48-h history of transient positional vertigo and exertional dyspnea. Neurological examination and laboratory tests showed no abnormalities, and initial investigations, including a computed tomography (CT) scan of the brain, yielded normal results. Considering her low risk for stroke, she was discharged for routine outpatient follow-up and echocardiography.

The patient's hypertension and dyslipidemia were well-managed with antihypertensive therapy (losartan 25 mg daily) and statin therapy (atorvastatin 20 mg daily), resulting in normal laboratory test results at the time of presentation and a blood pressure of 120/70 mmHg. Laboratory analysis indicated no signs of increased inflammation or coagulation abnormalities. Sedimentation rate returned unremarkable results. Chest radiography revealed no abnormalities. The electrocardiogram displayed left anterior hemiblock, which was not previously documented in the patient's medical history.

Preoperative Transthoracic echocardiography (TTE) revealed a normal ejection fraction of 60%, with no evidence of associated valve disease or significant impairment by the mass. The mitral valve function appeared normal on echo, without regurgitation or stenosis and the presence of a mobile oval-shaped mass within the LV, probably attached to the chordae of the anterolateral papillary muscle, suggestive of either a papillary mass, papillary muscle hypertrophy, or thrombus (Fig. 1).

CT angiography was performed and showed the presence of the mass within the LV (Fig. 2).

Cardiac magnetic resonance imaging (CMR) was arranged and demonstrated that a round, well-demarcated, slightly mobile mass measuring 23.5× 19.6 × 16.5 mm was seen in the LV, attached to the papillary muscle. No evidence of endocardial or myocardial involvement was seen. No evidence of interference with mitral valve function was noted. The mass appeared homogeneously hypointense on steady-state free precession (SSFP) cine images and homogeneously hyperintense on short-tau inversion recovery (STIR) images. On fast spin-echo (FSE) sequences, the mass was homogeneously isointense and did not exhibit fat suppression properties. After gadolinium administration, the mass did not enhance in first-pass perfusion imaging. Early post-contrast images showed scattered patchy areas of enhancement within the mass, while late post-contrast images revealed diffuse homogeneous contrast uptake and gadolinium enhancement. According to CMR tissue characterization criteria, a benign avascular intracavity LV mass was noted, highly suggestive of a large intracavity PFE (Fig. 3). We have also provided a comparison of the CMR criteria for benign versus malignant cardiac tumors, which is detailed in Table 1.

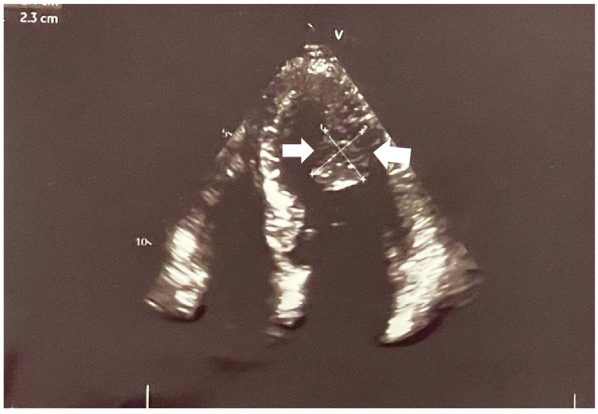

Fig. 1. TTE revealed a 22 × 20 mm mass attached to the papillary muscle of the LV.

The papillary muscle exhibited mobility due to the attachment of the mass to the chordae tendineae. Multiple imaging modalities were employed to accurately characterize the mass's size, mobility, and

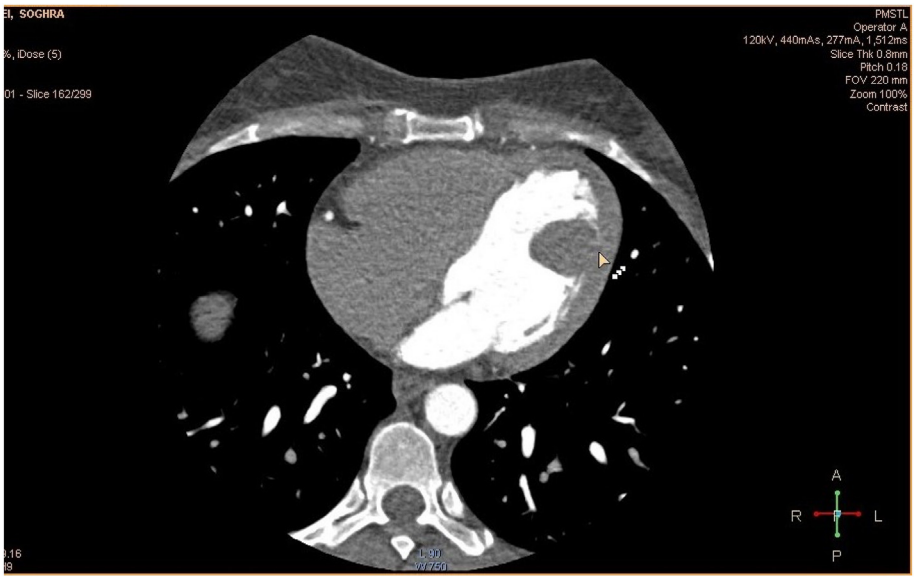

Fig. 2. CT angiography showed a mass attached to the mid lateral wall of the LV.

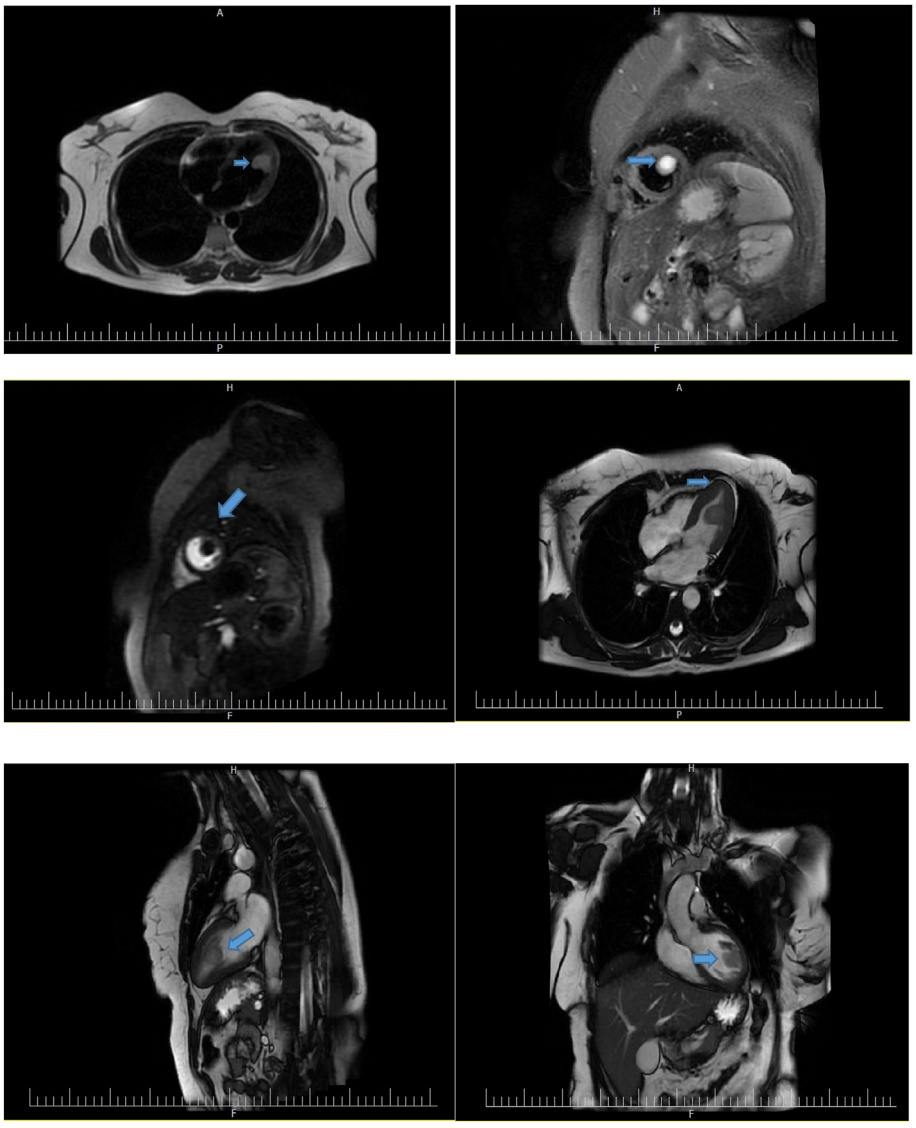

Fig. 3. CMR revealed tissue characteristics consistent with a papillary fibroelastoma.

Table 1. CMR criteria of benign vs malignant cardiac tumors.

Criteria	Benign Tumors (e.g., PFE)	Malignant Tumors
Growth Rate	Slow	Rapid
Borders	Well-demarcated Infiltrative,	poorly defined
Mobility	Often mobile	Typically less mobile
Tissue Characteristics	Homogeneous, often hypointense or isointense on CMR	Heterogeneous, necrotic, or hemorrhagic
Gadolinium Enhancement	No or minimal enhancement	Significant enhancement, often irregular

benign nature, which was crucial for planning surgical resection.

The patient subsequently underwent surgical excision of the mass. The heart was accessed via a median sternotomy, and following appropriate cannulation, cardiopulmonary bypass was initiated. The left atrium was then opened parallel to Sondergaard's groove, revealing a multilobular yellowish mass attached to the chordae of the anterolateral papillary muscle of the mitral valve, consistent in size with the echocardiographic and CMR findings. Through a transmitral approach, the tumor was completely excised from the chordae of the mitral valve without any injury to the chordae or mitral valve leaflets. The cardiopulmonary bypass time was approximately 90 min, with a cardiac arrest time of 45 min. Total operative time was 150 min, with an estimated blood loss of 600 mL (Fig. 4). The patient was successfully weaned off cardiopulmonary bypass and experienced an uneventful recovery in the postoperative period. Follow-up TTE confirmed the absence of residual tumor and normal mitral valve function, with no signs of mitral valve regurgitation. Histopathological examination confirmed the diagnosis of PFE. At the 6-month follow-up, the patient remained asymptomatic, and repeat TTE showed a normally functioning mitral valve with no evidence of regurgitation or tumor recurrence.

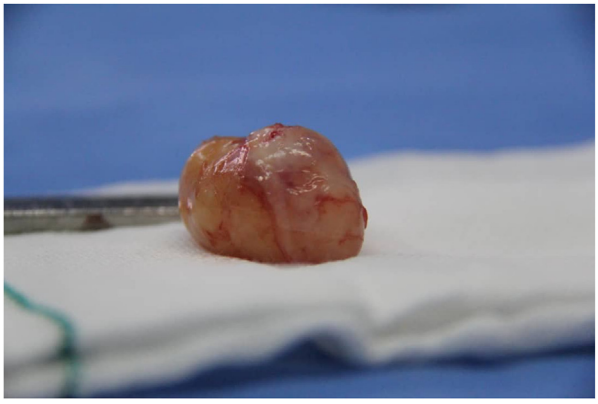

Fig. 4. Complete resection of papillary fibroelastoma: macroscopic appearance of the soft, yellowish, lobulated tumor.

3. Discussion

Nonvalvular PFEs are exceptionally rare, with most PFEs being valvular in origin. A previous case of PFE located in the right ventricular outflow tract presenting with vertigo has been reported [9], Although our case differs in the location of the tumor, both cases underscore the potential for nonvalvular PFEs to cause neurological symptoms. The unusually large size of the tumor in our case adds to its rarity and highlights the importance of thorough imaging for accurate diagnosis. The use of multiple imaging modalities, including TTE, CT angiography, and CMR, was crucial in confirming the tumor's benign nature and its precise attachment, ensuring a successful surgical outcome. Surgical resection remains the definitive treatment, especially for large, mobile masses to prevent embolic complications.

Ethics information

Ethical approval was obtained from the institutional review board Chamran Cardiovascular Research Education Hospital prior to the commencement of the study. Informed consent was obtained from all individual participants included in the study.

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Author contribution

Conception: MB, MM. Literature review: MB, PA. Methodology: MM, PA. Software: PA. Analysis and/or interpretation: PA. Investigation: PA. Resources: PA. Data collection and/or processing: MB, PA. Writer-original draft: MB, PA. Writing- review & editing: PA. Visualization: EA. Supervision: MM. Project administration: MP.

Conflict of interest

The authors declare no conflict of interest.

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