Case Report

Mitral Valve Papillary Fibroelastoma as an Incidental Finding in a 62-Year-Old Man: A Case Report

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ABSTRACT

Background: Primary cardiac tumors are rare, with the majority being benign. Papillary fibroelastoma (PFE) represents the third most common type of primary benign cardiac tumor. While most patients with PFEs remain asymptomatic, the condition is incidentally discovered in approximately one-third of cases. Nonetheless, some patients initially present with embolic events, such as stroke, transient ischemic attack, myocardial infarction, or symptoms of cardiac obstruction, including syncope, heart failure, or even death due to coronary ostial obstruction. Although PFEs are benign, they can pose life-threatening risks if they lead to valve obstruction or systemic embolization.

Case Presentation: A 62-year-old man was evaluated for exertional dyspnea (functional class II) and found to have a large, well-defined, round, hypermobile echo-dense mass attached to the ventricular side of the anterior mitral valve leaflet. Pathological examination confirmed the diagnosis of PFE.

Conclusions: Given that PFEs are relatively common tumors, they should be included in the differential diagnosis of cardiac masses. Despite their benign nature, urgent surgical resection is not only lifesaving but also critical in preventing tumor-related vascular, embolic, or neurological complications. ***(Iranian Heart Journal 2025; 26(2): 88-96)***

KEYWORDS: Primary cardiac tumors, Papillary fibroelastoma, Cardiac mass

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Primary cardiac tumors are rare, with the majority being benign. Papillary fibroelastoma (PFE) is the third most common type of primary benign cardiac tumor. Because many of these tumors are identified only postmortem, their true incidence remains unclear. According to autopsy studies, their incidence can reach up to 0.33%. This tumor affects both men and women equally, with an average age of 60 years, and accounts for approximately 75% of all heart valve tumors. [1,2] PFEs predominantly involve the left side of the heart, with the aortic valve being the most common site (35%–63%), followed by the mitral valve leaflets (9%–55%), the tricuspid valve (6%–15%), and the pulmonary valve (0.5%–8%). [3, 4]

Although PFEs are benign, they can become life-threatening if they cause valve obstruction or systemic embolization. Notably, the first reported case of PFE in 1975 involved an embolic complication that resulted in myocardial infarction. [5]

We herein describe a 62-year-old man who was evaluated for exertional dyspnea (functional class II). Imaging revealed a large, well-defined, round, hypermobile echo-dense mass attached to the ventricular side of the anterior mitral valve leaflet (AMVL). Pathological examination confirmed the diagnosis of PFE.

Case Presentation

A 62-year-old man was intubated due to an exacerbation of chronic obstructive pulmonary disease (COPD). During the initial evaluation at another center, a cardiac mass was identified on transthoracic echocardiography (Fig. 1), prompting his referral to our tertiary center for further assessment. His medical history included COPD and hypertension. Prior to this recent exacerbation, he reported exertional dyspnea (functional class II) but denied any other symptoms. He had a history of smoking and opium addiction.

On clinical examination, the only notable finding was the presence of coarse crackles. ECG showed normal sinus rhythm with left axis deviation, left anterior hemiblock, and poor R-wave progression in the precordial leads (Fig. 2).

Transesophageal echocardiography performed at our center revealed the following: a left ventricular ejection fraction of 50%-55%, mild-to-moderate mitral regurgitation, and a large (1.8 × 1.2 cm), dense, hypermobile echogenic mass attached to the ventricular side of the AMVL via a stalk (Fig. 3).

Cardiac magnetic resonance imaging was conducted to further characterize the mass. The results demonstrated an iso-signal intensity mass attached to the ventricular side of the AMVL (Fig. 4).

Given that the imaging findings were consistent with PFE, a cardiac surgery consultation was conducted, and the patient was scheduled for surgery. The mass was successfully resected, and the pathology report confirmed the diagnosis of PFE (Fig. 5). The patient experienced no postoperative complications and was discharged in stable condition.

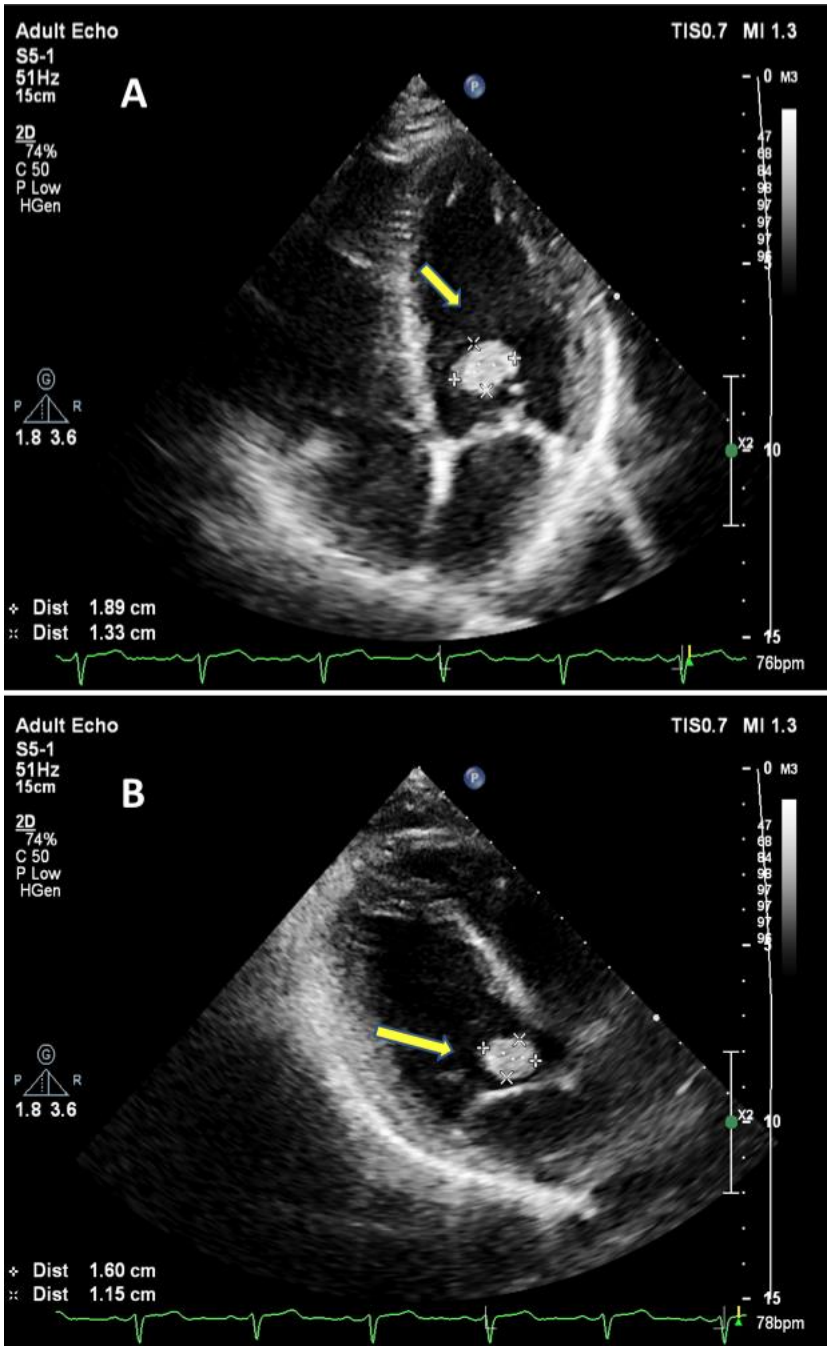

Figure 1. Transthoracic echocardiography in the 4-chamber (panel A) and 2-chamber (panel B) views illustrates the mass (yellow arrow) attached to the ventricular side of the anterior mitral valve leaflet.

Figure 2. The patient's ECG shows normal sinus rhythm with left axis deviation, left anterior hemiblock, and poor R wave progression in the precordial leads.

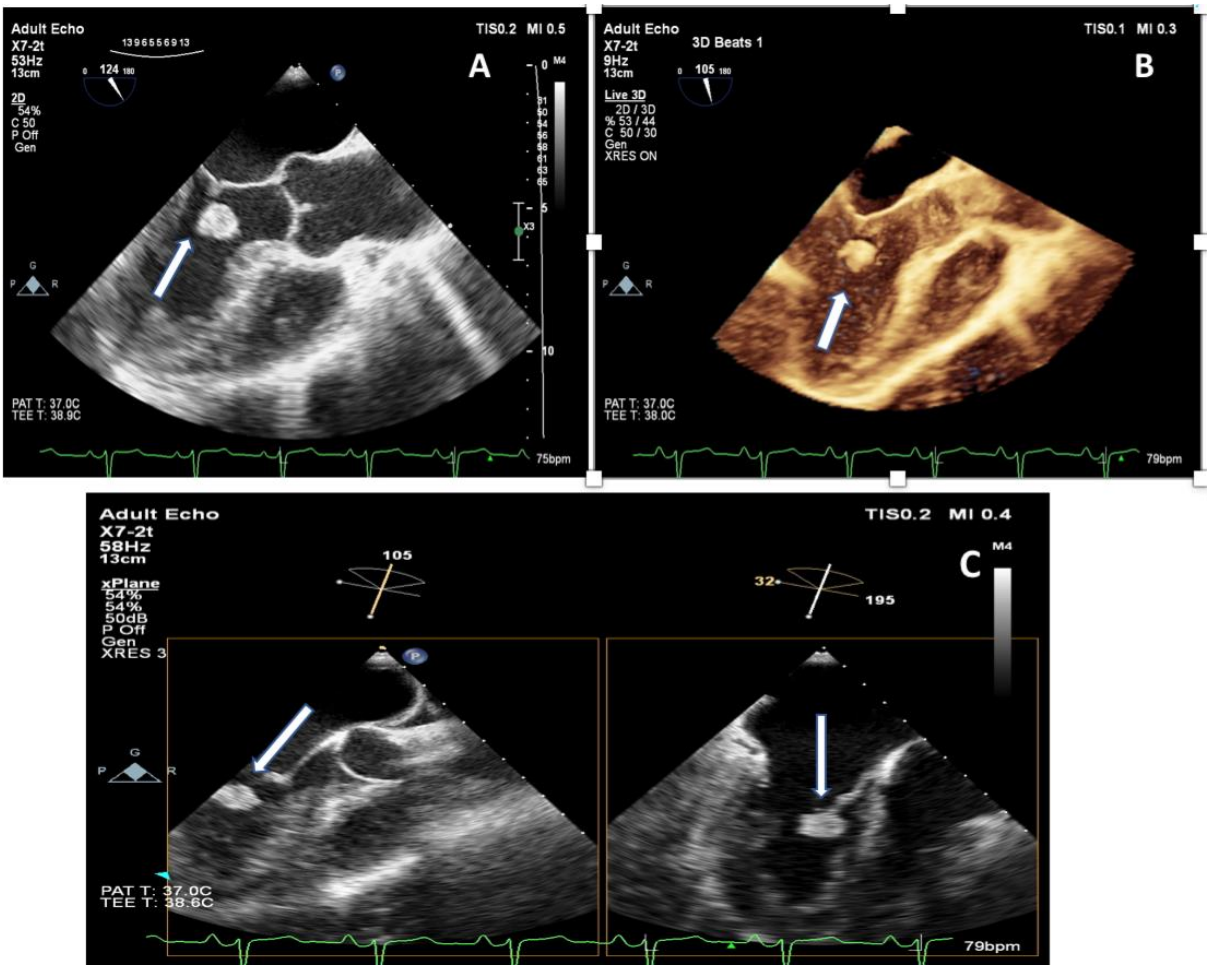

Figure 3. (A-C) Transesophageal echocardiography reveals a large, dense, hypermobile mass (white arrow) measuring 1.8 x 1.2 cm, attached to the ventricular side of the anterior mitral valve leaflet through a stalk.

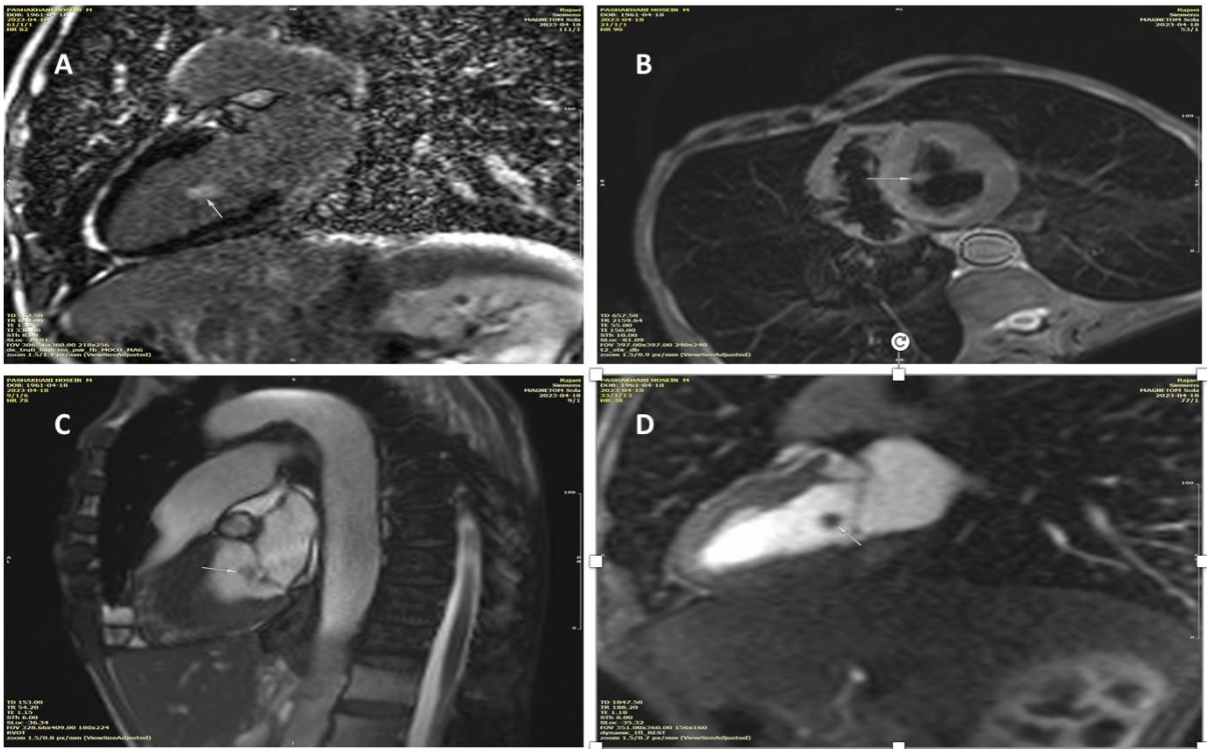

Figure 4. Late gadolinium enhancement images are presented herein. (A) The 3-chamber view displays pronounced homogeneous enhancement of the mass. (B) The T2 short tau inversion recovery (T2-STIR) image in the short-axis view shows iso-signal intensity of the mass. (C) The right ventricular outflow tract view steady-state free precession (SSFP) image reveals a small mass attached to the ventricular side of the anterior mitral valve leaflet. (D) The 2-

chamber view from the first-pass perfusion image demonstrates no significant vascularity.

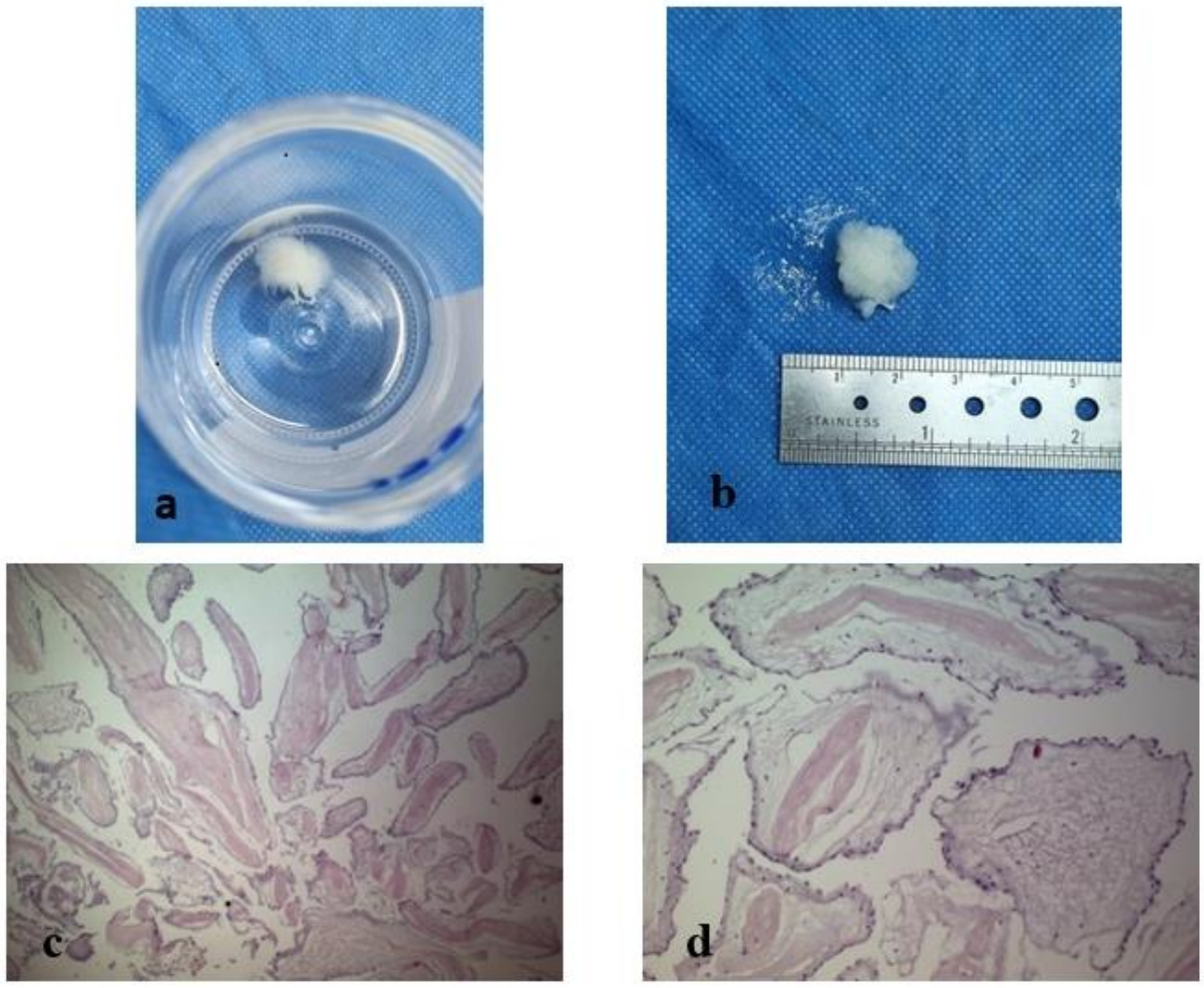

Figure 5. The typical gross appearance of a mitral valve mass shows multiple small fronds and a sea anemone appearance in a bowl of water (a & b). Microscopic examination reveals numerous papillary projections composed of paucicellular avascular fibroelastic tissue, lined by a single layer of endocardium, with areas of hydropic changes (c & d).

DISCUSSION

PFEs are rare endocardial tumors that most frequently involve the aortic valve, followed by the mitral valve, though a nonvalvular origin has been reported in approximately 16% of cases. [6] Primary mitral valve fibroelastomas are uncommon and often asymptomatic. Before the advent of echocardiography, which has revolutionized early detection, these lesions were rarely diagnosed prior to death. [7]

Most patients with PFEs are asymptomatic, and the tumors are incidentally discovered in about one-third of cases. Nevertheless, some patients initially present with embolic events, such as stroke, transient ischemic attack, myocardial infarction, or symptoms of cardiac obstruction, including syncope, heart failure, or even death due to coronary ostial obstruction. Stroke is the most common presentation, occurring in 30% of patients. [8] In our case, the only symptom was dyspnea on exertion (functional class II), which, given the patient's history of

COPD, could not be definitively attributed to the tumor.

PFEs are typically small, ranging from 2 mm to an average size of 9 mm, and are usually solitary. Some are mobile, though their maximum dimensions can extend to several centimeters. These tumors are classically described as resembling a "sea anemone," with a central stem and branch-like arms extending outward. Histologically, they consist of an outer layer of endothelium, a middle layer of connective tissue, and a central core composed of fibrin and mucopolysaccharides. [9]

Tumor mobility is recognized as an independent predictor of death or nonfatal embolization. Embolic fragments may arise from the tumor itself due to its soft and fragile tissue or from the formation of platelet-fibrin thrombi on its surface. [10]

Given their small size, typically ranging from 0.5 to 2.0 cm in diameter, and their frequent involvement of heart valves, PFEs can pose significant clinical challenges despite their benign nature.

PFEs can be challenging to differentiate from valvular vegetations. As a result, clinical information and laboratory tests, such as blood cultures, play a crucial role in differential diagnosis. Other potential diagnoses include myxoma, lipoma, rhabdomyoma, amorphous tumors, or endocarditis. Considering that symptomatic PFEs carry a significant risk of severe complications, aggressive surgical intervention is recommended, regardless of the tumor's size or the patient's symptom profile. [11]

Complete and successful resection of a PFE is curative, and the long-term postoperative prognosis is excellent. There have been no reported cases of tumor regrowth following resection. However, long-term transthoracic echocardiography follow-up is recommended to monitor for potential recurrence. For patients who are not surgical candidates, long-term oral anticoagulation may be considered, although its efficacy is not supported by randomized controlled data.

CONCLUSIONS

Since PFEs are relatively common tumors, they should be included in the differential diagnosis of cardiac masses. With a high index of suspicion, laboratory tests, blood cultures, and appropriate imaging modalities—such as echocardiography and a dedicated cardiac magnetic resonance protocol for comprehensive tissue characterization—can help establish the diagnosis. Despite the benign nature of these tumors, urgent surgical resection is not only lifesaving but also critical in preventing tumor-related vascular, embolic, or neurological complications.

Declaration

Availability of Data and Materials:

The data sets used and/or analyzed during the current study are available from the corresponding author upon reasonable request.

Authors' Contributions:

M.F. drafted the initial patient report, while S.S., M.H., and S.M. contributed to the investigation and data collection. A.A. finalized the research and discussion sections. M.S., S.F.H.J., and N.M. were responsible for drafting, revising, and editing the manuscript. All authors reviewed and approved the final manuscript for publication.

Ethics Declarations:

Ethics Approval and Consent to Participate: Not applicable.

Consent for Publication:

The patient provided informed consent for the publication of this case. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Conflict of Interest:
The authors declare no competing interests related to this study.

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