AZJCVS
AZerbaijan Journal of CardioVascular Surgery

AZJCVS 2026;7(2):49-51
DOI: 10.5455/azjcvs.2026.03.03

Case Report

Mehmet Burak Gulcan[1], Muhammed Aydin[2], Mehmet Ali Kaygin[2]

[1]Yenimahalle Research and Training Hospital, Department of Cardiovascular Surgery, Ankara, Türkiye
[2]Erzurum City Hospital, Department of Cardiovascular Surgery, Erzurum, Türkiye

Received: 30 March, 2026
Accepted: 31 July, 2026
Published: 31 July, 2026

Corresponding Author: Mehmet Burak Gulcan, Yenimahalle Research and Training Hospital, Department of Cardiovascular Surgery, Ankara, Türkiye
Email: drmburakgulcan@gmail.com

CITATION
Gulcan MB, Aydin M, Kaygin MA. Ozaki Procedure and Mitral Valvuloplasty in a young female patient, a case report. AZJCVS. 2026;7(2):49-51.

Primary fibroelastoma of heart: A rare clinical entity

Abstract

Papillary fibroelastomas are rare benign primary cardiac tumors that most commonly arise from the aortic and mitral valves, whereas tricuspid valve involvement is uncommon. A 71-year-old woman hospitalized for vaginal bleeding was referred for palpitations and elevated blood pressure. Transthoracic and transesophageal echocardiography revealed a mobile right atrial mass measuring up to 12x12 mm, associated with the tricuspid valve and initially considered to be an atrial myxoma. Surgical excision was planned because of the tumor's mobility and size. A short-stalked mass was completely excised through a right atriotomy under cardiopulmonary bypass without requiring tricuspid valve repair or replacement. Histopathological examination confirmed a papillary fibroelastoma. The patient's postoperative course was uneventful, and she was discharged on postoperative day 7. This case highlights the importance of considering papillary fibroelastoma in the differential diagnosis of right-sided intracardiac masses and contributes to the literature by demonstrating the successful valve-sparing excision of a rare tricuspid valve-associated fibroelastoma.

Keywords: Cardiac papillary fibroelastoma, mitral valve, cardiopulmonary bypass, aortic valve

INTRODUCTION

Primary cardiac tumors are extremely rare, with a reported incidence of less than 0.3% (1). Most primary benign cardiac tumors are myxomas. Primary cardiac tumors may be asymptomatic, and approximately 10% are discovered incidentally during the investigation of another condition (1). The patient's symptoms depend on the location and extent of tumor invasion. Papillary fibroelastomas constitute less than 10% of all primary cardiac tumors; however, they are the most common valvular tumors and the third most common cardiac tumors after myxomas and fibromas (1). Papillary fibroelastomas are most commonly observed in men around 60 years of age. They tend to arise from the cardiac valves, most commonly the aortic valve, followed by the mitral valve (1). They may rarely arise in the right side of the heart, particularly from the tricuspid valve (2). These tumors are commonly less than 20 mm in diameter and have a sea-anemone-like appearance, consisting of a central stalk with multiple papillary fronds arising from it (1). Although they generally do not impair valvular function, surgical excision is recommended because of the risks of embolization and thrombus formation (1). This report describes a cardiac tumor that was histopathologically diagnosed as a papillary fibroelastoma. The tumor was incidentally detected in an asymptomatic female patient during hospitalization in the gynecology department. We completely excised the sea-anemone-like mass, and histopathological examination confirmed our preoperative diagnosis. Our aim is to demonstrate the potential benefits of the complete surgical excision of benign cardiac masses. We believe that the tumor's unusual location and our surgical approach may contribute valuable information to the existing literature.

CASE REPORT

In this case report, an intracardiac tumor was identified in a 71-year-old female patient. The patient was hospitalized in the Department of Obstetrics and Gynecology because of vaginal bleeding and was referred to the Department of Cardiology for palpitations and elevated blood pressure. Transthoracic echocardiography (TTE) revealed a left ventricular ejection fraction (LVEF) of 55%, a pulmonary artery pressure (PAP) of 20 mmHg, no valvular pathology, mild right atrial dilatation, and a 1.1 × 1.1 cm mass in the right atrium that extended into the right

ventricle during diastole and was considered to represent either a vegetation or a tumor. Transesophageal echocardiography (TEE) demonstrated no tricuspid stenosis or regurgitation and revealed a 12x12 mm, mobile, smooth-contoured mass associated with the tricuspid valve, consistent with an atrial myxoma. Electrocardiography (ECG) showed sinus rhythm. Following the detection of the intracardiac mass, the patient was admitted to our department and scheduled for surgery.

The procedure was initially approached through a minimally invasive right thoracotomy. However, because of severe adhesions in the right hemithorax, conversion to a median sternotomy was required. Under cardiopulmonary bypass (CPB) and cardiac arrest, a right atriotomy was performed. A short-stalked, 1x1 cm tumor resembling a myxoma was identified within the right atrial cavity and completely excised (Figures 1 and 2). The right atrium and right ventricle were irrigated with isotonic saline, and the right atriotomy was closed. The CPB and aortic cross-clamp times were 70 and 45 minutes, respectively, and the patient was cooled to 30°C during the procedure. Her postoperative vital signs remained stable. The patient had an uneventful intensive care unit stay and was discharged on postoperative day 7. Histopathological examination revealed a papillary fibroelastoma..

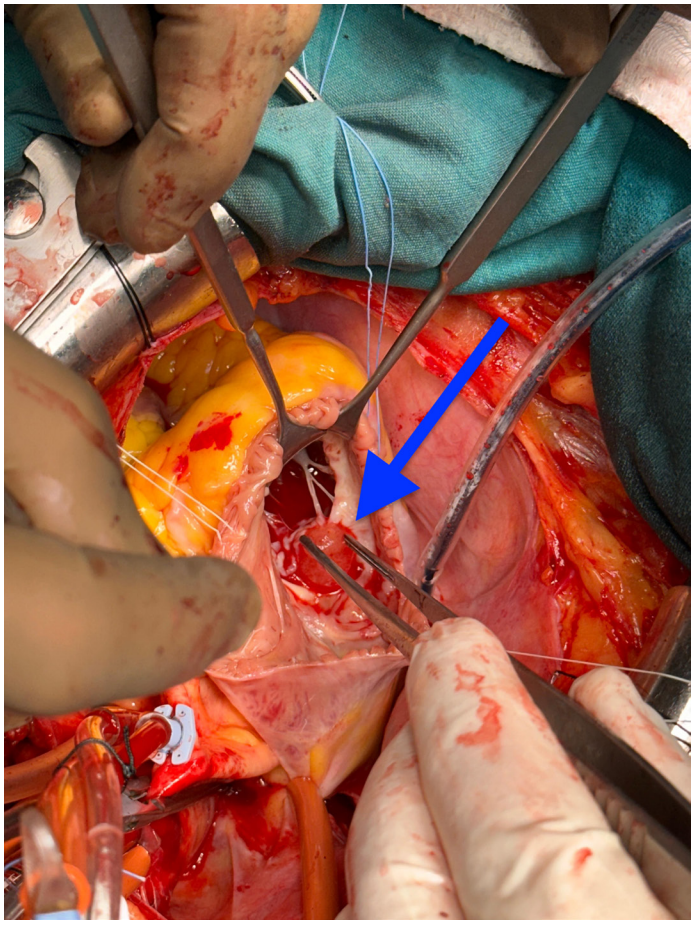

Figure 1. Arrow shows tumor next to tricuspid valve

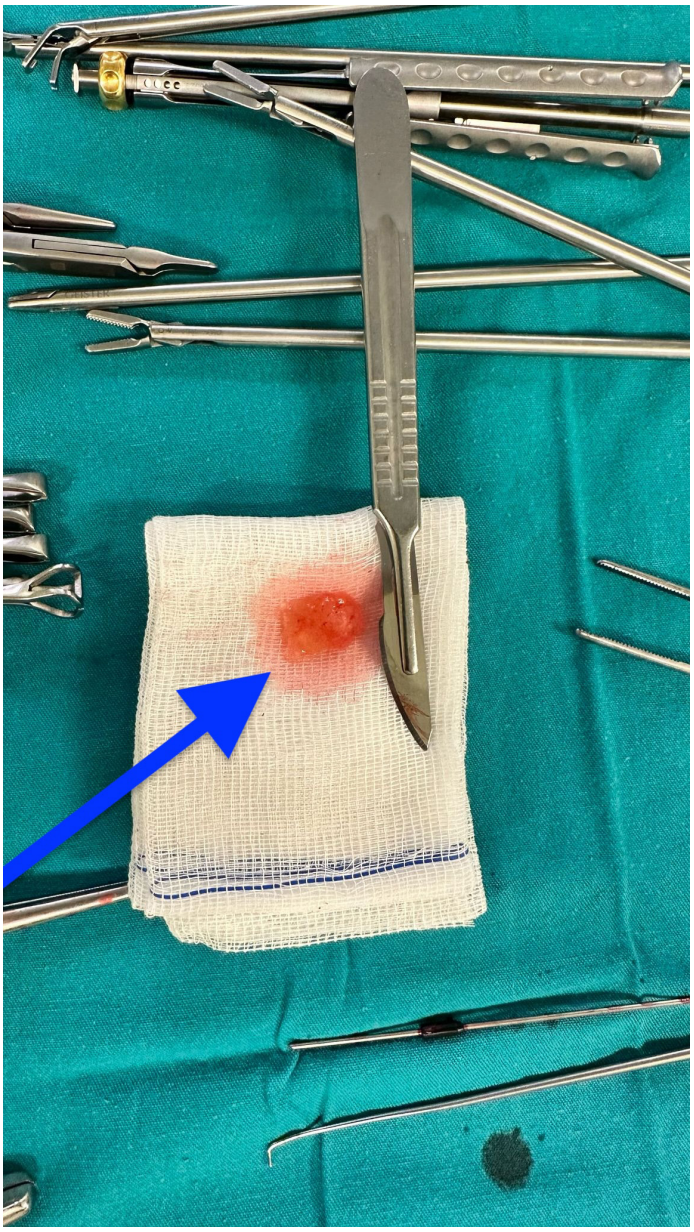

Figure 2. Arrow shows excised tumor

DISCUSSION

Papillary fibroelastomas (PFEs) are the second most common primary cardiac tumors in adults and originate from the valvular endothelium (3,4). Distinguishing PFEs from Lambl's excrescences using echocardiography can be challenging (1). Lambl's excrescences are most commonly observed on the aortic valve in older men (4). High mechanical stress at the coaptation sites of the left-sided heart valves triggers fibrin deposition, resulting in the formation of these excrescences (5). Lambl's excrescences are not neoplasms but may cause thromboembolic events (5). Therefore, dual antiplatelet therapy is considered the first-line treatment (5). In contrast, surgical excision is the gold-standard treatment for PFEs(3,4) Myxomas and PFEs can be differentiated according to their locations, as myxomas do not

typically arise from the cardiac valves (4). Although PFEs are benign cardiac tumors, they can cause serious complications, including stroke, systemic embolism, sudden cardiac death, acute valvular dysfunction, and myocardial infarction(6).

For left-sided PFEs, surgical excision is recommended because of the risk of embolization (3). However, for right-sided PFEs, surgery is recommended when the tumor is larger than 1 cm in diameter (3). For smaller tumors, the benefits of surgery remain unclear when weighed against the associated operative risks (3). Given the tumor's size and histopathological type, our surgical approach appears reasonable, particularly considering its atypical location. Although PFEs may arise from any endocardial surface (3), they are most commonly found on the aortic and mitral valves (1,4). In our patient, however, the mass was located adjacent to the tricuspid valve, making it an exceptionally rare cardiac tumor For example, in the study by Ngaage et al.(7), only 5 of the 88 patients who underwent surgery for PFE had tumors located near the tricuspid valve. Histopathological examination confirmed that the mass was a benign cardiac tumor that could be treated with surgical excision alone (8). n contrast, malignant tumors, such as sarcomas, generally require chemotherapy or radiotherapy in addition to surgery (8). Furthermore, valve-sparing surgery is usually sufficient for the surgical treatment of PFEs(7). Only approximately 20% of patients require valve repair or replacement in patients underwent surgery for PFE (7).

Several cases similar to ours have been reported in the literature. Watanabe et al. (9) operated on a 68-year-old male patient with a PFE arising from the tricuspid valve. Their approach was similar to ours, as they recommended surgical excision despite the tumor's right-sided location because of the risk of embolization (9). Their patient also had an uneventful postoperative course, similar to that of our patient. Nara et al. (10) operated on an 83-year-old female patient with a giant PFE arising from a tricuspid valve leaflet. The tumor's size and location made their case more surgically challenging than ours; therefore, tricuspid valve repair was required following the complete excision of the 3-cm tumor(10).

CONCLUSION

In conclusion, papillary fibroelastomas are rare benign cardiac tumors, and involvement of the tricuspid valve is particularly uncommon. Despite their benign histology, these tumors may cause serious thromboembolic and cardiovascular complications. Complete surgical excision can generally be achieved using a valve-sparing approach with favorable postoperative outcomes. This case contributes to the literature by demonstrating the successful surgical management of a rare tricuspid valve-associated papillary fibroelastoma.

Conflict of Interests: The authors declare that there are no conflicts of interest.

Financial Disclosure: The authors declare that there is no financial support.

Patient Informed Consent: Written informed consent was obtained from the patient for the publication of her clinical information and accompanying surgical images.

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