Concomitant Papillary Fibroelastoma and Non-Bacterial Thrombotic Endocarditis of the Tricuspid Valve in a Patient with Antiphospholipid Antibody Syndrome: A Case Report

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ABSTRACT

Primary cardiac tumors are rare and usually benign. Among these, papillary fibroelastoma is one of the most frequently encountered types. Although often asymptomatic, papillary fibroelastoma can lead some serious embolic complications. This case report presents a 42-year-old male patient diagnosed with antiphospholipid antibody syndrome, who developed nonbacterial trombotic endocarditis on a tricuspid valve papillary fibroelastoma. Histopathologically findings supported a diagnosis of papillary fibroelastoma with coexisting nonbacterial trombotic endocarditis, This case highlights the association between papillary fibroelastoma and nonbacterial trombotic endocarditis in the context of antiphospholipid antibody syndrome, emphasizing the importance of clinical vigilance regarding cardiac involvement in rheumatologic diseases and the embolic potential of valvular masses.

Keywords

Papillary fibroelastoma, nonbacterial trombotic endocarditis, antiphospholipid antibody syndrome, tricuspid valve replacement, cardiac tumor

ÖZET

Primer kalp tümörleri nadirdir ve genellikle iyi huyludur. Bunlar arasında papiller fibroelastoma en sık karşılaşılan tiplerden biridir. Genellikle asemptomatik olmasına rağmen, papiller fibroelastoma bazı ciddi embolik komplikasyonlara yol açabilir. Bu vaka raporunda, triküspit kapak papiller fibroelastomasında bakteriyel olmayan trombotik endokardit gelişen, antifosfolipid antikor sendromu tanısı konmuş 42 yaşında bir erkek hasta sunulmaktadır. Histopatolojik bulgular, eşlik eden bakteriyel olmayan trombotik endokardit ile birlikte papiller fibroelastoma tanısını desteklemiştir. Bu vaka, antifosfolipid antikor sendromu bağlamında papiller fibroelastoma ile bakteriyel olmayan trombotik endokardit arasındaki ilişkiyi vurgulayarak, romatolojik hastalıklarda kalp tutulumu ve kapak kitlelerinin embolik potansiyeli konusunda klinik uyanıklığın önemini vurgulamaktadır.

Anahtar Kelimeler

Papiller fibroelastom, nonbakteriyel trombotik endokardit, antifosfolipid antibody sendrom, triküspit kapak replasmanı, kalp tümörü.

Introduction

Primary cardiac tumors are extremely rare and are mostly benign. The most common types are myxomas and papillary fibroelastomas (PFE) (Devanabanda AR and Lee LS, 2023). PFE can develop on any endocardial surface. The distribution is most frequently on the aortic valve (44%), followed by the mitral (35%), tricuspid (15%), and pulmonary valves (8%) (Ramesh M Gowda and friends, 2003). Although the exact etiology remains unclear, it is generally associated with endothelial injury and is more commonly observed on valvular endothelial surfaces (Gowda RM and friends, 2003).

PFE is often asymptomatic but may cause complications such as vascular embolism, stroke, heart failure, or cardiac arrest (Devanabanda AR and Lee LS, 2023). On echocardiographically, papillary fibroelastomas can be similar findings seen in vegetations, endocarditis, trombus, Libman–Sacks endocarditis (a form of non-bacterial trombotic endocarditis - NBTE), or Lambl's endocarditis (Reda Bzikha and friends, 2021).

NBTE occurs as a result of endothelial injury followed by the deposition of sterile fibrin and platelet material, forming sterile vegetations (aseptic trombus). It develops secondary to hypercoagulability and endothelial damage (Şimşek-Yavuz S and friends, 2019). NBTE can be seen in valvular heart diseases associated with hypercoagulable states such as systemic lupus erythematosus (SLE), rheumatoid arthritis, and antiphospholipid antibody syndrome (APS), elderly patients, uremia, other chronic conditions, and after intracardiac catheter insertion (Şimşek-Yavuz S and friends, 2019).

In this case report, we present a patient with APS, which has NBTE on the tricuspid valve in the background of papillary fibroelastoma and underwent surgical treatment with tricuspid valve replacement.

Case Presentation

A 42-year-old male presented with dyspnea. He had been complaining of shortness of breath on exertion for approximately 1 month. His medical history included known immune trombocytopenic purpura and hypertension. There was no history of substance use. Physical examination was normal. Transthoracic echocardiographically revealed a 3x10 mm mobile lesion adherent to the tricuspid valve, protruding into the right atrium (differential: vegetation vs. myxoma), eccentric grade 2 tricuspid regurgitation (TR), and a pulmonary artery pressure of 39 mmHg. Transesophageal echocardiographically confirmed the findings: ejection fraction 50–55%, a trombus-like mass measuring 2.6x1.9 cm on the lateral leaflet of the tricuspid valve, and mild TR. Blood cultures were negative for infective endocarditis.

Following a multidisciplinary discussion with the cardiology and cardiovascular surgery team, surgical intervention was planned. Preoperative coronary angiography was normal. Laboratory findings revealed a platelet count of $95\times10^3/\mu L$, positive ANA and dsDNA, with other parameters within normal limits. Hematology consultation recommended proceeding with surgery while maintaining platelet count above $80\times10^3/\mu L$.

Median sternotomy and right atriotomy were performed. Vegetations were observed on the anterior and posterior leaflets of the tricuspid valve (Figure 1). The damaged leaflets were resected, and the vegetations were removed (Figure 2). A 33 mm Pericarbon bioprosthetic tricuspid valve was implanted. The patient was extubated 7 hours postoperatively and transferred to the inpatient servise on postoperative day 3. He was discharged on postoperative day 7 with medical treatment consisting of apixaban 5 mg twice daily and furosemide once daily.

Histopathologically examination of intraoperative specimens revealed fibrinous material consistent with vegetations, lined by a single-layered flattened epithelium, with scattered papillary-like structures and myxoid changes. These areas showed focal S100 positivity, while calretinin and CD34 were negative, ruling out a diagnosis of myxoma. In light of these findings, the diagnosis was consistent with papillary fibroelastoma and non-bacterial trombotic endocarditis.

Postoperative researches for trombotic vegetation and trombositopenia revealed positive anticardiolipin IgG and IgM, ANA positivity, confirming the diagnosis of antiphospholipid antibody syndrome (APS).

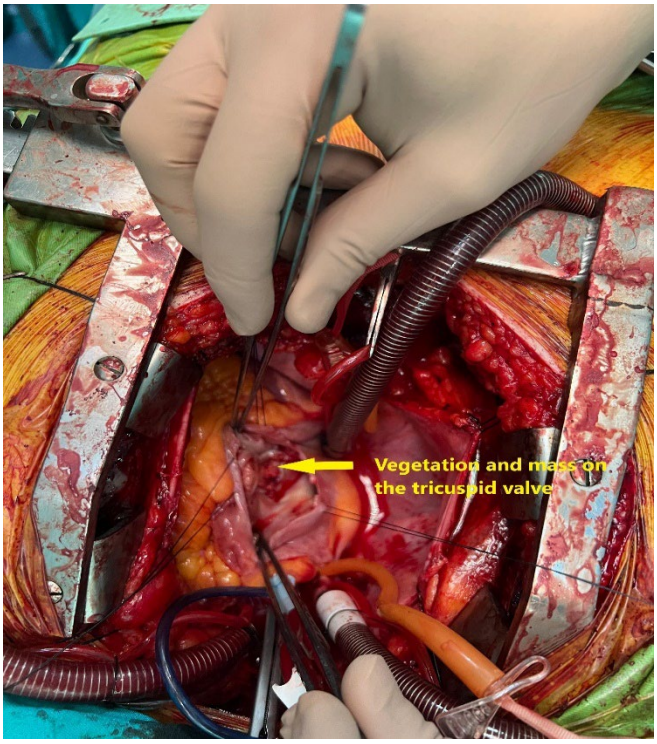

Fig 1: *Intraoperative image of the tricuspid valve mass*

Discussion

Histopathologically, PFE is characterized by papillary fronds composed of acellular matrix lined by a single layer of endothelial cells. Echocardiographically, it appears as a pedunculated, mobile, speckled mass, ranging from 2–40 mm in size (Devanabanda AR and Lee LS, 2023). The most common symptoms are dyspnea (37.5%), TIA (37.5%), angina (12.5%, especially in aortic valve involvement due to prolapse over coronary ostium), and syncope (12.5%) (Devanabanda AR and Lee LS, 2023).

Clinical presentation of PFE varies depending on tumor size, mobility, and degree of obstruction it causes on the mitral or tricuspid valves (Gowda RM and friends, 2003). PFE can rarely present in association with conditions like APS, thyroid dysfunction, and trombocytopenia (Devanabanda AR and Lee LS, 2023– Işık M and friends, 2024).

NBTE is more frequently associated with APS. Trombocytopenia is observed in 20–40% of patients with APS, usually mild-to-moderate in severity (70,000–120,000/mm³) (Şimşek-Yavuz S and friends, 2019). In one study, NBTE was associated with malignancy in 40.5% of cases, SLE in 33.3%, and APS in 35.7%. Stroke was the most common initial presentation (59.5%) (Zmaili MA and friends, 2021). However, the association of NBTE and PFE has been reported to be rare (Gaetano Thiene and Cristina Basso, 2006).

Treatment is primarily surgical resection; however, antiplatelet and anticoagulant therapy is essential regardless of whether surgery is performed (Zoltowska DM and friends, 2021).

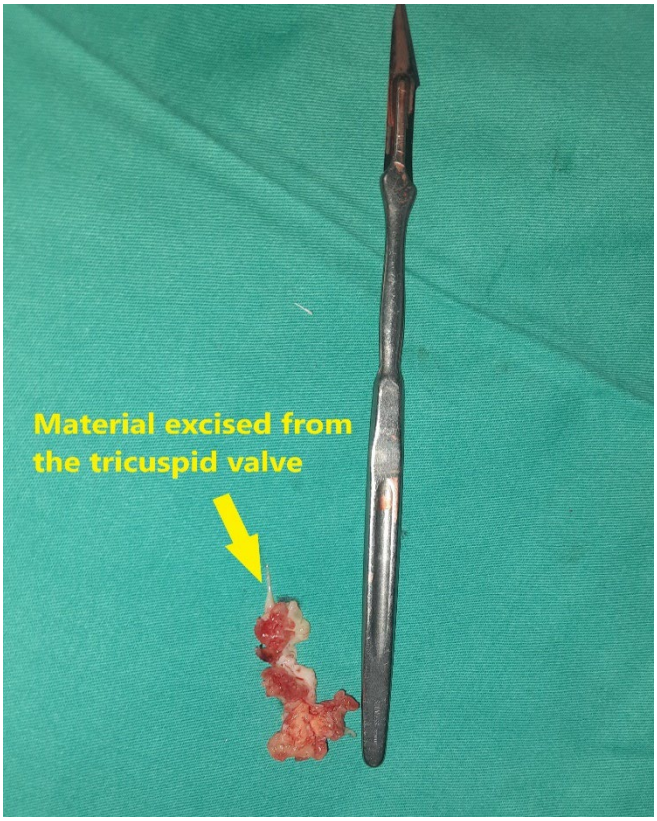

Fig 2: *Image of the excised mass*

Valvular lesions such as PFE may act as foreign bodies and serve as sources of tromboembolism. It is important to remember that PFE can be associated with rheumatologic diseases. As in our case, in the presence of NBTE and mild-to-moderate trombocytopenia, APS should be considered in the differential diagnosis. Early diagnosis and treatment are crucial to prevent potentially severe complications.

Conflict of Interest

There is no conflict of interest.

Financial Support

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