Turkish Journal of Thoracic and Cardiovascular Surgery 2025;33(1):117-120

Brief Report / Kısa Rapor

Could papillary fibroelastoma in the tricuspid valve cause thrombocytopenia?

Triküspit kapaktaki papiller fibroelastom trombositopeniye neden olabilir mi?

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ABSTRACT

Cardiac myxoma and papillary fibroelastoma are the most common benign primary cardiac tumors in adults. In this brief report, we described a male patient with a mass in the tricuspid valve (papillary fibroelastoma as a result of pathology), who presented with thrombocytopenia and underwent tricuspid valve replacement. The patient was followed and treated by the hematology clinic due to thrombocytopenia, but a definitive diagnosis could not be made. The patient was incidentally diagnosed while being investigated for urinary stones. Thrombocytopenia improved in the postoperative follow-up. The successful surgical procedure, along with echocardiography and pathology images of the patient, was shared.

Keywords: Papillary fibroelastoma, thrombocytopenia, tricuspid valve replacement.

ÖZ

Kardiyak miksoma ve papiller fibroelastoma yetişkinlerde en sık görülen iyi huylu primer kalp tümörleridir. Bu kısa raporda, triküspit kapakta kitlesi (patoloji sonucu papiller fibroelastom) olan, trombositopeni ile başvuran ve triküspit kapak replasmanı yapılan bir hasta sunuldu. Trombositopeni nedeni ile hematoloji kliniği tarafından takip ve tedavi edilen hastaya kesin tanı konulamamıştı. Üriner sistem taşı nedeniyle tetkik edilirken hastaya rastlantısal olarak tanı konuldu. Cerrahi sonrası takiplerde trombositopeni düzeldi. Hastanın başarılı cerrahi işlemi ile birlikte ekokardiyografi ve patoloji görüntüleri paylaşıldı.

Anahtar sözcükler: Papiller fibroelastom, trombositopeni, triküspit kapak replasmanı.

In adults, cardiac myxoma and papillary fibroelastoma (PFE) are the most common benign primary heart tumors.[1] In order of site of involvement, PFE is most commonly localized to the aortic valve (35 to 63%), mitral valve (9 to 55%), tricuspid valve (6 to 15%), and pulmonary valve (0.5 to 8%).[2]

The exact etiology of PFEs is unknown. They mostly originate from the valvular endocardium and occur in areas of valvular endothelial damage.[2] Although PFE is mostly clinically asymptomatic, it can cause stroke and embolism. Rarely, it can cause thrombocytopenia. Surgical treatment is prioritized as tumors on the left side of the heart may be a source of embolism. Treatment for right-sided pathologies is unclear.

This brief report described a male patient with a mass in the tricuspid valve (PFE due to pathology), who presented with thrombocytopenia and underwent tricuspid valve replacement. The case was shared due to the rarity of the tricuspid valve pathology, the incidental diagnosis, and the notable echocardiographic and pathological images.

BRIEF REPORT

A 42-year-old male patient complained of dyspnea on exertion. Except for a systolic murmur in the tricuspid annulus, the physical examination findings were normal. The patient had been investigated and treated for thrombocytopenia at the hematology clinic for about 10 years (steroid, intravenous immunoglobulin, and platelet suspension), but no definitive diagnosis could be made. While being examined for urinary stones, a tricuspid valve mass was discovered at the urology clinic. Echocardiography showed Grade 2 tricuspid regurgitation, pulmonary artery pressure of 50 mmHg, and a 2.6×1.9 cm hyperechogenic mass with a thrombus image on the lateral leaflet of the tricuspid valve (Figure 1, Video 1). Preoperative hemogram and biochemistry analyses were normal, except for thrombocytopenia (85,000 uL). A written informed consent was obtained from patient.

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Doi: 10.5606/tgkdc.dergisi.2024.25719

Received: November 03, 2023
Accepted: July 05, 2024
Published online: September 20, 2024

Cite this article as: Işık M, Fındık S, Tatar S, Kara Ş, Görmüş N. Could papillary fibroelastoma in the tricuspid valve cause thrombocytopenia? Turk Gogus Kalp Dama 2025;33(1):117-120. doi: 10.5606/tgkdc.dergisi.2024.25719.

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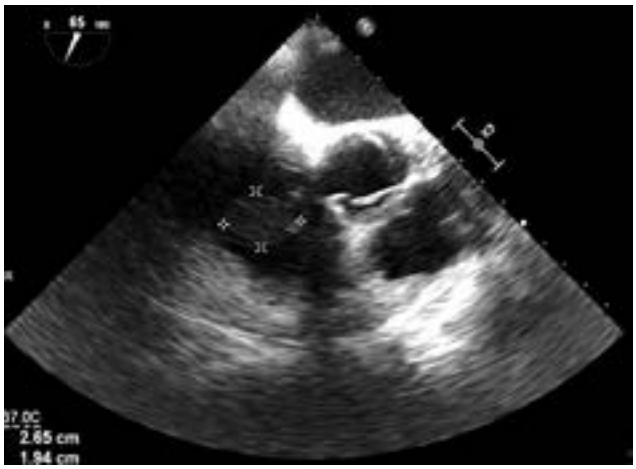
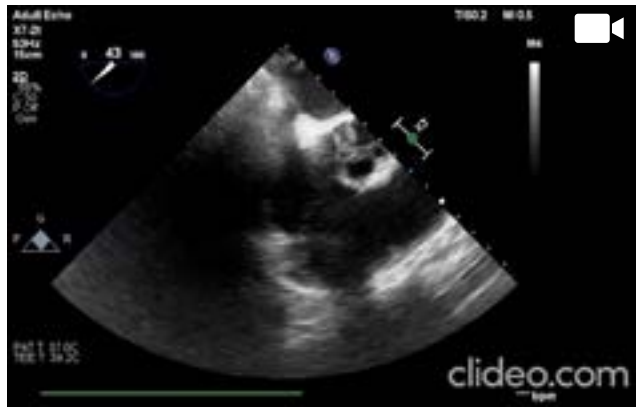


Figure 1. Echocardiography image.



Video 1. Echocardiography video image.

Median sternotomy was performed under general anesthesia. Aortic and bicaval venous cannulations were completed, and cardioplegia was administered. Right atriotomy was performed. Intraoperative examination revealed a thrombus/vegetation-like structure in the posterior and anterior leaflet of the tricuspid valve. A number 33 bioprosthetic valve replacement was performed. The pathology report revealed fibrin material without significant inflammatory cell infiltration, as well as highly hypocellular structures with myxoid changes forming superficial scattered

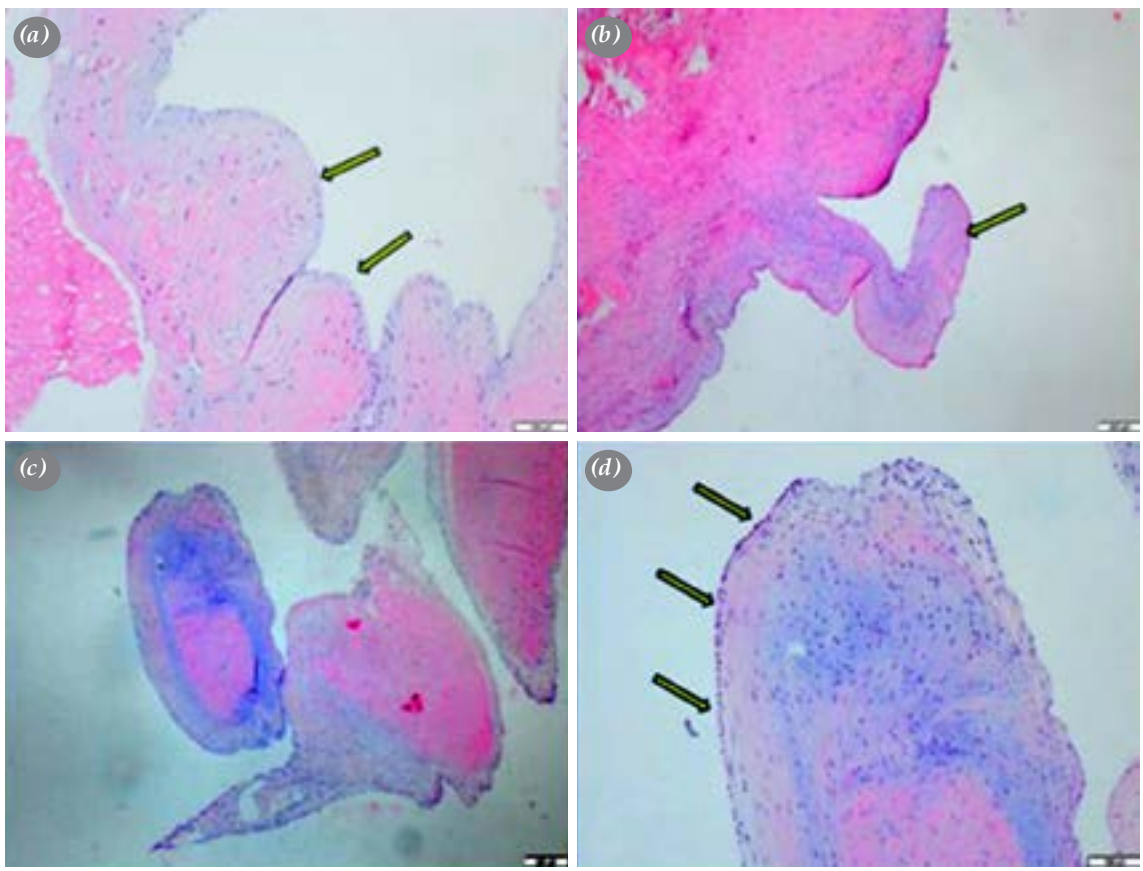


Figure 2. Microscopic image of the material excised from the tricuspid valve. **(a)** Papillary fronds (arrows; H&E, ×100), **(b, c)** Papillary fronds (arrows; H&E, ×40), **(d)** Paucicellular, avascular fibroelastic tissue lined by a single layer of endocardium (arrows; H&E, ×100).

papillary-like structures in focal areas with a single layer of flattened epithelium. In these areas, calretinin and CD34 were negative, and S100 was focally positive. The negativity of calretinin did not support the diagnosis of myxoma (Figure 2).

The platelet value was 81,000/uL on the day of the operation and 114,000/uL on the seventh postoperative day after discharge. In outpatient follow-up, platelet values were between 138,000 and 193,000/uL.

DISCUSSION

Although PFE can originate from all endocardial structures, the most common site of involvement is the heart valves (>80%).[3,4] Papillary fibroelastoma is a condition with no specific clinical findings and is mostly detected incidentally. Since more than 95% of PFE is located on the left side of the heart, systemic embolism and embolization-related symptoms have been reported.[3,4] Antiphospholipid syndrome, thyroid dysfunction, and thrombocytopenia were reported in a rare case.[3] In this case, there was no history of previous embolism. It was incidentally detected while being examined for urinary system stones and was consistent with the literature.

There are no clear guideline recommendations for the treatment of PFE. Total excision of the tumor along with the endocardium is the preferred method. Deterioration of the heart valve structure during excision may require valve repair or replacement. Tamin et al.[1] recommend surgical resection of all left-sided tumors, regardless of size, mobility, or symptoms. Surgical resection of right-sided tumors is controversial. Surgical treatment is recommended for patients with very large, mobile, pedunculated tumors that cause hemodynamically significant flow obstruction or embolization, particularly in the presence of patent foramen ovale and significant right-to-left shunt.[5] In our case, the indications for the removal of the mass were its considerable size, the presence of material that gave the appearance of a thrombus, and the presence of second-degree tricuspid regurgitation. Valve repair could have been an alternative to replacement. However, in this case, total excision of the valve was performed because there was a vegetation-like structure on the leaflet and the diagnosis was unclear. Definitive diagnoses of cardiac masses are based on pathology results. Preoperative echocardiography may cast doubt on the diagnosis. However, since PFE cases are not common, it is difficult to master specific echocardiography findings. Therefore, it is challenging to distinguish benign or malignant with preoperative echocardiography and decide on surgery or follow-up.

Thrombocytopenia may be due to many mechanisms, including decreased platelet production (hereditary thrombocytopenia, anemia, and suppression by physical/chemical agents), increased destruction (autoimmune disease, infection, drugs, and disseminated intravascular coagulation) and impaired distribution (hypersplenism). Our patient had a 10-year history of isolated thrombocytopenia. He was followed up by the hematology clinic, laboratory and bone marrow investigations were performed, but no diagnosis could be made to explain the current situation. The patient was treated with steroids and intravenous immunoglobulin, but no response was obtained. On the seventh postoperative day, the patient's platelet count was 117,000/uL, and platelet counts remained within the normal range during outpatient follow-up. One of the conditions that could have caused thrombocytopenia in this case was infective endocarditis. However, the absence of inflammatory cells in the pathology report ruled out this diagnosis. The return of the platelet count to the normal range after excision of the mass suggests that antibodies against tumor antigens may cause autoimmune platelet disorders. However, the lack of response to immunosuppressive treatment reduces the possibility of an immunologic event. Prosthetic heart valves are known to cause thrombocytopenia.[6] We believe that the mechanism may be similar in the current patient. The tumor likely caused turbulent flow in the right atrium and a coaptation defect in the valve, increasing platelet consumption through mechanical effects.

In conclusion, a cardiac origin should be kept in mind in cases of unexplained thrombocytopenia.

Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

Author Contributions: Analysis and interpretation of data, writing the article, review and approval: M.I.; Writing the article, review and approval: S.F., S.T.; Ş.K.; Analysis and interpretation of data, writing the article, review and approval: N.G.

Conflict of Interest: The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding: The authors received no financial support for the research and/or authorship of this article.

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