A mobile mass on the tricuspid valve in a young female: The critical distinction between the vegetation and cardiac tumor

Yavuzer Koza, Uğur Kayab, Enise Armağan Koza

Ataturk University Faculty of Medicine, Department of Cardiology, Erzurum 25100, Turkey
Cardiovascular Surgery, Erzurum 25100, Turkey
Erzurum Regional Training and Research Hospital, Department of Anesthesiology, Erzurum, Turkey

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Papillary fibroelastoma (PFE) is a rare primary benign tumor that usually involves left-sided heart valves. They are the third most common cardiac tumors after myxomas and lipomas and account for less than 10% of all primary cardiac tumors. An accurate diagnosis and distinction from other intracardiac masses is essential because they are a potential cause of systemic emboli, stroke, myocardial infarction and sudden death. We report a case of a 34-year-old woman who experienced progressive dyspnea and fatigue. Transthoracic echocardiography revealed a pedunculated, homogenous echo-dense, and highly mobile mass attached to the tricuspid valve. The patient was diagnosed with cardiac PFE because of its size and mobility, and surgical resection was scheduled.

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Y. Koza, U. Kayab, E. A. Koza

PFEs are rare benign avascular cardiac tumors that typically attached to the left-sided valve leaflets with the aortic valve being the most common site. Tricuspid and pulmonary valve involvement is unusual. They are usually located on the ventricular side of the atrioventricular valves and the arterial side of the semilunar valves. PFEs are not causally associated with valve dysfunction.\(^1,2\)

Although the etiopathogenesis of PFE is unknown, various possible causes have been suggested in the literature. They have been considered as hamartomas, organized thrombi, true neoplasms or inflammatory foci due to unusual endocardial responses to infection or hemodynamic trauma.\(^3\)

The clinical presentation of PFE varies widely from asymptomatic to severe embolic complications. Right-sided PFE are asymptomatic and rarely cause pulmonary embolism. The differential diagnosis of PFE includes other cardiac tumors, thrombus, vegetation, and Lambli’s excrescences. One of the most considered differential diagnosis is cardiac myxoma.\(^1\) In the present case, the differential diagnosis included cardiac tumor and vegetation. Infective endocarditis was excluded based on the absence of fever, signs of valvular dysfunction, and a history of intravenous drug abuse. Blood cultures were also negative for microorganisms.

The echocardiographic characteristics of PFE include small size (usually <1.5 cm), high mobility, attachment to the endocardium through a pedicel or stalk, and refractive appearance. The distinction from the vegetations can be challenging due to their small sizes.\(^4\) Moreover, PFE may lack some of these findings whereas vegetations may represent many of these characteristics. For that reason, PFE can often be differentiated by clinical informations, blood cultures and laboratory tests. In this case, there was a history of infected intrauterine device extraction, so the clinical information was quite misleading. However, on MRI, in contrast to vegetations, PFEs usually have complete enhancement in late gadolinium enhancement as seen in this case. PFE MRI features include isointense T1-imaging and hyperintense T2-imaging.\(^1,4\)

The management of PFE depends upon its clinical presentation. Patients who are experiencing embolic events that are cardiovascular or neurologic in origin should undergo surgical resection. Although there are no firm guidelines on how to manage right-sided PFEs, surgery is shown to be curative, safe and well tolerated.\(^4\)

This report not only highlights atypical presentation of this tumor but also reminds us to keep this possibility in mind.
patients who present with a history and imaging findings considering infective endocarditis. In patients with a high clinical suspicion for a cardiac tumor, advanced imaging tools such as MRI and computed tomography should be also used.

Appendix A. Supplementary data
Supplementary data associated with this article can be found, in the online version.

References