Papillary Fibroelastoma of the Tricuspid Valve: Case Report and Review of the Literature

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Abstract

A 70-year-old man presented with exertional dyspnea and lower extremity edema. Cardiac examination was unremarkable. Transthoracic echocardiography disclosed a large mobile mass on the tricuspid valve. The patient was referred to surgery for the excision of the tumor. Histological examination revealed papillary fibroelastoma ***(Iranian Heart Journal 2008; 9 (2):62-64).***

Key words: papillary fibroelastoma ■ heart tumor ■ tricuspid valve ■ echocardiography

Papillary fibroelastomas are rare, primary benign cardiac tumors most frequently located on the cardiac valves.[1] In order of frequency, they are the third most common primary cardiac tumors after myxomas and lipomas. Fibroelastomas are usually found by chance in post-mortem examinations. However, their prompt detection is of great importance because they are potential causes of systemic emboli, stroke, myocardial infarction, and sudden death.[2-4] Right-sided localization is even more rare.[5]

Case report

A 70-year-old man presented with exertional dyspnea (functional class II) and lower extremity edema. The symptoms had begun 2-3 months previously. Physical examination was normal and laboratory tests showed no abnormality. Transthoracic and transesophageal echocardiography showed a large (25x18 mm) mobile, non-obstructing prolapsing mass on the atrial side of the anterior leaflet of the tricuspid valve with mild regurgitation (Fig. 1). Right ventricular function was normal.

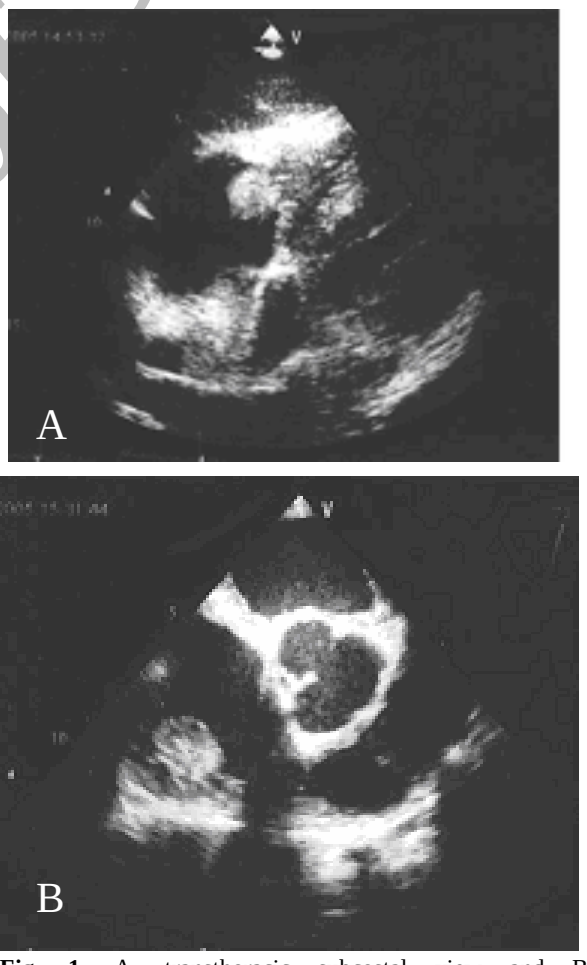

Fig. 1. A, transthoracic subcostal view and B, transesophageal RV inflow-outflow view

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The patient underwent the surgical excision of the mass. Gross pathology revealed a trabeculated tumor attached to the anterior tricuspid valve leaflet. Microscopic examination showed multiple avascular papillary frond-like structures lined by endothelial cells, compatible with papillary fibroelastoma (Fig. 2).

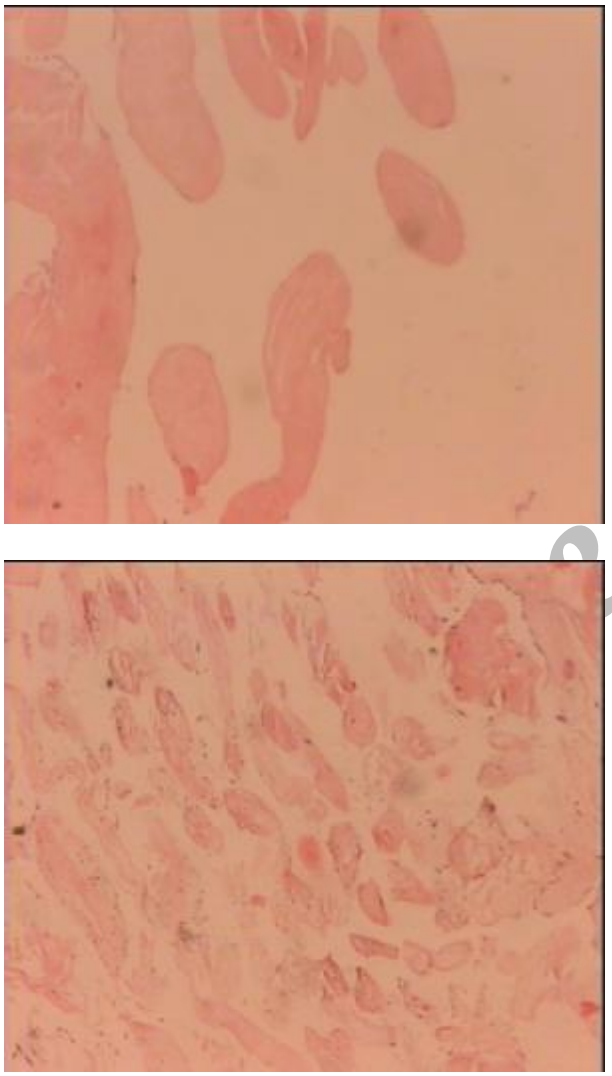

Fig. 2. Histopathology of resected tumor **s**howing multiple frond-like papillary structures with coverage of endothelium.

Discussion

Papillary fibroelastomas represent 7.9% of benign primary cardiac tumors in adults.[1] Papillary fibroelastomas are the third most common primary tumors of the heart.[6] Approximately 90% of primary fibroelastomas arise from valvular tissue, most commonly from the aortic or mitral valves.[7,8] In a study including 162 patients with papillary fibroelastomas,[9] the age ranged from 5-86 (mean 60±16 years). They may be single or multiple, occurring more frequently on the ventricular surface of the semilunar valves and on the atrial surface of the atrioventricular valves.[10,11] The tricuspid valve is most affected in children and the mitral and aortic valves in adults.[11] On gross anatomical examination, they resemble a sea anemone, consisting of multiple fingerlike fronds radiating from a stalk. Microscopically, each frond consists of a collagenous elastic core, surrounded by a mucopolysaccharide matrix and covered by endothelial cells.[9, 12]

The pathogenesis of papillary fibroelastomas remains unclear, but several possible explanations have been reported, including previous mechanical damage to the endothelium, iatrogenic factors, organizing thrombi, and a latent infectious mechanism due to cytomegalovirus.[7,12,13] With the advent of echocardiography, an increasing number of papillary fibroelastomas have been diagnosed. Typical echocardiographic features include the following:

1. The tumor is round, oval, or irregular in appearance, with well-demarcated borders and a homogenous texture.
2. Nearly half of them have small, mobile stalks.[9, 14]

Sun et al. found that 99% of papillary fibroelastomas were less than 20 mm in the largest diameter.[9] The largest reported one was 53 mm.[15] They often cause systemic embolic events, such as cerebrovascular stroke and more rarely, myocardial infarction.[16] This occurs because of their very friable and soft texture, as well as the creation of thrombi on their surface which may later embolize. Surgical removal of right-sided papillary fibroelastomas in asymptomatic patients is indicated only for large, mobile

tumors. The presence of a patent foramen ovale with a sizable right-to-left shunt is an additional consideration for surgery of the right-sided fibroelastomas. Asymptomatic patients with small, left-sided, non-mobile (no stalk) fibroelastomas are usually observed. However, fibroelastomas more than 1cm in size, especially if mobile, should be considered for excision, not least in patients with other cardiovascular diseases, young patients whose surgery would carry a low risk and those with a high cumulative risk for embolization.[9]

We describe a patient with a tricuspid valve fibroelastoma: an uncommon occurrence inasmuch as it was detected in an unusual location (tricuspid valve) and exceeded the usual size of less than 20 mm.

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