

A Case Report of a Papillary Fibroelastoma Arising from the Left Ventricular Outflow Septum Just Beneath the Aortic Valve

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Abstract

Cardiac papillary fibroelastomas are rare benign tumors with frond-like growths that typically involve the native valve tissue. Papillary fibroelastomas originate less commonly in the ventricular septum. We report a rare case of fibroelastoma arising from the left ventricle. (J Nippon Med Sch 2008; 75: 239–241)

Key words: papillary, fibroelastoma

Introduction

Papillary fibroelastomas (PFEs) are, after myxomas and lipomas, the rarest type of benign cardiac benign tumor¹. They are usually derived from the aortic or mitral valve endocardium^{1,2}. PFEs less commonly originate from the ventricular septum. A highly mobile round mass in the ventricular septum just beneath the aortic valve was detected incidentally with echocardiography during a medical workup in an 81-year-old man. We decided to remove the tumor to prevent any potential embolic complications.

Case Report

An 81-year-old man was referred for evaluation of chest discomfort. His medical history included implantation of a pacemaker because of bradycardia-related atrial fibrillation and hypertension since 2005. We performed transthoracic echocardiography as a cardiac function test. Echocardiography revealed

normal valve architecture and a highly mobile round mass approximately 10 mm in diameter in the left ventricle. Transesophageal echocardiography revealed more detailed information and a clearer image of the mass. A highly mobile pedunculated mass with a homogeneous echodensity was found in the left ventricular outlet septum just beneath the aortic valve (**Fig. 1**). It was not possible to determine with only the preoperative examination whether this mass was a tumor, thrombus, or vegetation. The patient did not exhibit pyrexia or any evidence of endocarditis. There were no findings suggesting malignant tumors metastatic from other organs. Coronary angiography revealed severe stenosis of the left anterior descending artery. After receiving informed consent regarding of the risk of embolism, we performed coronary artery bypass grafting and an urgent operation to remove the tumor.

A cardiopulmonary bypass was established after cannulation of the right atrium with a single two-staged cannula. A left atrial venting cannula was inserted from the right upper pulmonary vein. The ascending aorta was cross-clamped during induction

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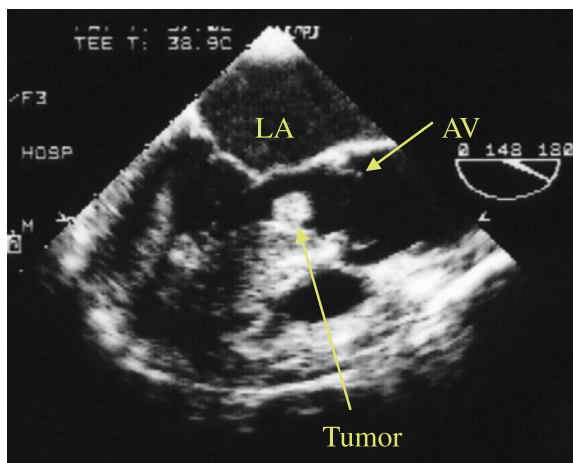


Fig. 1 A highly mobile pedunculated mass with a homogenous echodensity located in the left ventricular septal outlet just beneath the aortic valve. AV: aortic valve, LA: left atrium

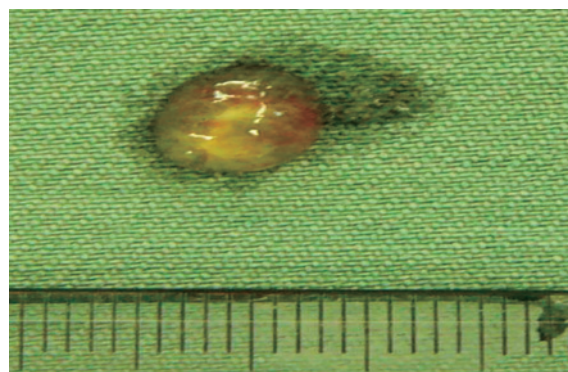


Fig. 2 The tumor appeared as a friable, tan, gelatinous mass that measured 0.7 cm in its greatest dimension.

of electrical ventricular fibrillation. Myocardial protection was assured with antegrade and retrograde blood cardioplegia after infusion of a crystalloid cardioplegic solution was started. Through the aortic valve, a fragile, gelatinous mass was easily grasped with forceps from the ventricular septum (Fig. 2, 3). Cryoablation was applied to the tip of the pedunculation site to prevent recurrence. Then the left internal thoracic artery was anastomosed to the left anterior descending artery.

Histopathologic examination of the mass confirmed the diagnosis of PFE (Fig. 4).

Discussion

Primary cardiac neoplasmas are rare, with a prevalence at autopsy of 0.002% to 0.33%^{1,3}. PFEs are the third most common tumors after myxomas and lipomas and account for less than 10% of all primary cardiac tumors¹. They are often detected with echocardiography, during autopsy, or because of systemic embolisms, causing cerebral infarction and myocardial infarction, and pulmonary embolisms^{4,5}. PFEs are uncommon but are increasingly recognized as a cause of embolic phenomena.

PFEs can arise from almost any endocardial surface, but they most often originate from the valvular endocardium¹⁴. Gowda et al. have reported that 84 of their cases originated from the cardiac

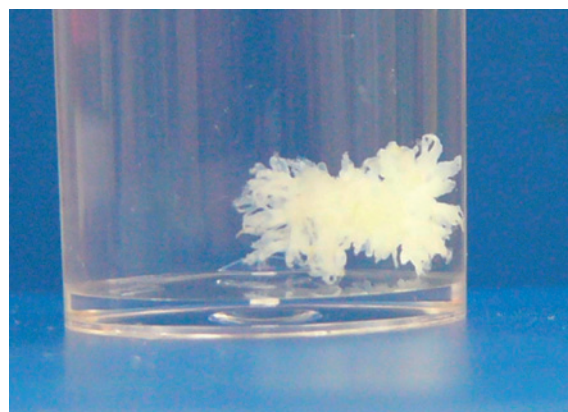


Fig. 3 Multiple papillary fronds arise from a central stalk. The tumor resembles a sea anemone.

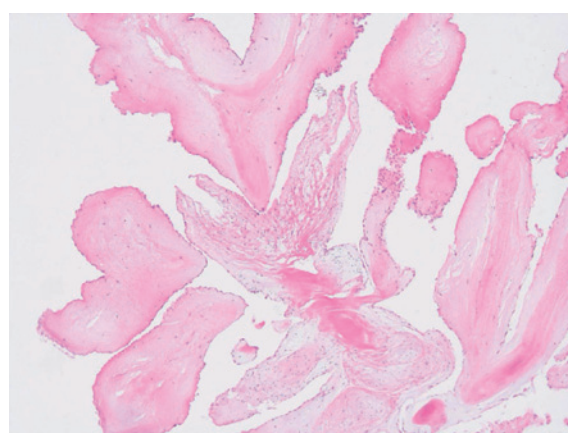


Fig. 4 Microscopic examination disclosed typical findings of a PFE including multiple papillary fronds consisting of a thin endocardial cell layer covering a loose stroma and a central core of connective tissue (hematoxylin and eosin staining).

valves: the aortic valve was involved in 44% of the cases, the mitral valve in 35% of cases, and the pulmonary valve in 8% of cases. The etiology of the PFE is unknown. One explanation is that they result from mechanical trauma, because they are more common in elderly patients with long-standing heart disease, including rheumatic disease, and are rarely seen in children¹.

Most PFEs are discovered incidentally with echocardiography during medical workup examinations. The most common presenting symptoms of PFE are cerebrovascular in origin, with 53% of the patients presenting with transient ischemic attacks or strokes. The next most common presenting symptoms are of cardiac origin⁵. The patient did not show any signs of embolic conditions such as cerebral or myocardial infarction. The only complaint was chest discomfort.

The tumors are benign but can cause embolisms. Embolisms may be caused by either a thrombus on the tumor or by the tumor itself. Anticoagulation therapy with heparin is recommended to prevent thrombui, but its efficacy has not been established. Because of the risk of embolism, surgical excision is the recommended treatment for primary cardiac tumors, regardless of the size or symptoms^{6,7}.

In conclusion, PFEs are uncommon but are becoming an increasingly recognized cause of embolic phenomena. Prompt surgical excion is

indicated to prevent any potential embolic complications.

The use of preoperative transesophageal echocardiography is recommended to determine the exact site of attachment and to guide tumor excision.

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