Cardiac Papillary Fibroelastoma Originating from the Mitral Valve Chordae

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Abstract

Cardiac papillary fibroelastoma (CPF) is a rare benign primary cardiac neoplasm. In particular, CPF originating from the mitral valve chordae is extremely rare. A 74-year-old man was hospitalized for the evaluation of a cardiac mass in the left ventricle. Echocardiography revealed a mobile, spherical, pedunculated 2.1×2.1 cm mass at the mitral valve chordae with no mitral regurgitation. The patient underwent excision of the mass without repairing the mitral valve. A histological examination confirmed the mass to be a papillary fibroelastoma.

Key words: echocardiography, cardiac tumor, papillary fibroelastoma, mitral valve chordae


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Introduction

Primary cardiac tumors are rare across all age groups, with a reported incidence of 0.021% in one autopsy series (1) and 0.019% in one echocardiography series (2). Cardiac papillary fibroelastomas (CPF s) are common benign neoplasms of the cardiac valves. Their prevalence ranges from 4.4-8.0% of primary cardiac tumors (3, 4). CPF s can arise anywhere in the heart, but most are located on the cardiac valves. The aortic valve is the most common site (37-45%). In contrast, papillary fibroelastomas originating from the mitral valve chordae are extremely rare (5). We herein present a case of CPF originating from the mitral valve chordae, which was detected by echocardiography and treated by surgical resection.

Case Report

A 74-year-old man with a past medical history of hypertension was referred to our cardiology department for evaluation of an asymptomatic mass in the left ventricle, which had been found incidentally in the course of routine transthoracic echocardiography. The findings from a physical examination, routine laboratory studies, electrocardiography and a chest X-ray were unremarkable. Transthoracic echocardiography revealed a mobile, spherical, pedunculated mass. Transesophageal echocardiography confirmed that the mass was 2.1×2.1 cm in size and that it was located at the mitral valve chordae with no mitral regurgitation (Fig. 1A, B). Cardiac magnetic resonance imaging demonstrated the mass to have increased signal intensity on T2 weighted short-inversion-time inversion-recovery (STIR) black blood imaging and ring-enhanced signal on T1 weighted delayed enhancement imaging following the administration of gadolinium (Fig. 1C, D).

Computed tomography of the chest and abdomen and magnetic resonance imaging of the brain revealed no signs of embolism. No feeding arteries arising from the coronary arteries were visualized on coronary angiography. The patient was referred to the Department of Cardiothoracic Surgery. The patient underwent the surgical resection of the mass under cardiopulmonary bypass. The heart was exposed through a median sternotomy and the mitral valve was exposed via a superior transseptal approach. The mass was observed to be adherent to the anterior mitral valve leaflet chordae at its junction with the posterior papillary muscle. The tumor was attached to a single secondary chordae, with-
out involving the mitral leaflets (Fig. 2A). A complete tumor resection was performed and mitral valve preservation was achieved. A postoperative transesophageal echocardiography (TEE) showed the absence of residual tumor and normal mitral valvular function. There were no findings of mitral valvular regurgitation. The resected tumor was round and measured 2.1x2.1 cm in size. There was no evidence of a thrombus on its surface (Fig. 2B). The mass was diagnosed as a papillary fibroelastoma based on the histopathologic examination (Fig. 3). The postoperative course was uneventful, and the patient was discharged on postoperative day 9.

**Discussion**

With an incidence of up to 0.33% in one autopsy series, CPF is the third most common primary cardiac tumor, after myxoma and lipoma (6). Although most CPF patients are asymptomatic, CPF is often associated with severe complications. The correct diagnosis is therefore important. The most common clinical presentations include stroke, acute coronary syndrome, heart failure, syncope, mesenteric ischemia, pulmonary embolism, and sudden death due to embolization, either from thrombus or from the tumor itself (7). The clinical presentation is determined by location, size, and mobility of the tumor; systemic embolism is frequently seen with tumors arising from the left heart.

Over 80% of CPFs are found on the valvular endocardium (5). In particular, the aortic valve is the most common site. CPFs can also exist on the mitral, tricuspid, or pulmonary valves, right atrial endocardium, and endocardial surfaces in both ventricles, including the papillary muscles and interventricular septum. It occurs less frequently on the mitral chordae (5, 7, 8). The average size of a CPF is approximately 1 cm in diameter, but their size can range from 0.2-4.6 cm. Forty-four percent of CPFs have a 1-3 mm stem; this mobile type of CPF appears to be more likely to embolize (2, 5, 6, 9).

In the case of papillary fibroelastoma, a simple tumor excision is generally the primary surgical goal. However, valve replacement may be required when the tumor has invaded the valve or when there is associated degeneration of the valve (10). Fortunately, we were able to perform a complete tumor resection with mitral valve preservation in our patient. In a prospective study involving 45 patients with papillary fibroelastomas that were suspected based on transesophageal echocardiography, 6.6% of the patients went on to develop embolic symptoms during a one-year follow-up period (5). However, the risk of CPF-related complications must be weighed against the risk of surgery on a case-by-case basis. Patients with small, asymptomatic, or nonmobile CPFs in

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**Figure 1.** Echocardiography and cardiac magnetic resonance (CMR) imaging revealed a 2.1x2.1cm highly mobile round mass attached by a stalk to the anterior mitral valve chordae. A: Transthoracic echocardiography. B: Transesophageal echocardiography. C: Sagittal T2 weighted short-inversion-time inversion-recovery black blood CMR imaging. D: Sagittal T1 weighted delayed enhancement CMR imaging after administration of gadolinium.
the left heart require careful observation. In contrast, excision should be considered for patients with large (≥1 cm) or mobile papillary fibroelastomas (5, 11). No data are currently available to support the use of systemic anticoagulation to prevent the embolic complications of papillary fibroelastoma. The prognosis for patients with surgically resectable papillary fibroelastoma is excellent, and there have been no reported cases of recurrence to date (5).

In this report, we presented a case of CPF originating from the mitral valve chordae that was successfully removed by surgery.

The authors state that they have no Conflict of Interest (COI).

References
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Figure 2. A: An intraoperative view of the mass with a stalk attached to the anterior mitral valve chordae. B: The gross appearance of the tumor was round, with no apparent thrombus on the surface.

Figure 3. Histopathologic examination. A: The tumor had the appearance of a sea anemone [Hematoxylin and Eosin (H&E) staining]. B: The tumor consisted of multiple fronds surrounded by a superficial endothelial layer (H&E staining). C: Elastic fibers were present in the tumor, as demonstrated by the dark purple color (Elastica-Masson staining).


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