**Case
Report**

Cardiac Papillary Fibroelastoma: Report of Three Cases

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Primary cardiac tumors are uncommon, and papillary fibroelastoma is considered relatively rare compared to myxoma and lipoma in the primary cardiac tumors. We experienced three cases of fibroelastoma. The patients' age was 28–75 years, and one patient was female. Two patients were presented with cerebral infarction and cardiac ischemia although, the other patient had no symptoms. Echocardiography revealed a hyperechoic mass in the left atrium, ventricle and on the aortic valve and helpful for differential diagnosis from myxoma. Surgical excision of the tumor was successfully performed in all patients and post-operative course was uncomplicated.

Keywords: cardiac tumor, papillary fibroelastoma, cerebral infarction, cardiac ischemia

Introduction

Primary cardiac tumors are rare, with a reported incidence of 0.021% in autopsy series¹⁾ and 0.019% in echocardiographic series.²⁾ Papillary fibroelastoma accounts for 4.4% to 8% of primary cardiac tumors,^{3,4)} and may be asymptomatic, or may cause severe complications after cerebral or coronary artery embolism. It is difficult to distinguish papillary fibroelastoma and myxoma although, surgical strategy might be different between these two tumors. We report three cases of papillary fibroelastoma with pathognomonic echocardiography.

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Case Report

Patient 1

A 28-year-old man presented to our hospital with acute onset of chest pain. He did not have any significant past medical history and was not taking any medication. He smoked one packet of cigarettes daily, but denied other cardiac risk factors. Physical examination was unremarkable. Laboratory testing showed a white blood cell count of 15100 cells/mL. Electrocardiography showed normal sinus rhythm and ST elevation in II, III, aVF, and V2–V6. A radiograph of the chest showed no evidence of congestive heart failure. Transthoracic echocardiography revealed severe apical akinesis. There was a hyperechoic mass on the atrial surface of the anterior mitral valve leaflet. Selective coronary angiography revealed total occlusion of the left anterior descending artery. Percutaneous transarterial angioplasty of the left anterior descending coronary artery was performed with good results, and embolism was suspected. Transesophageal echocardiography showed a homogenous, round, mobile mass measuring 8 × 7 mm in diameter. Three-dimensional transesophageal echocardiography demonstrated the mass attached to the anterior leaflet of the

mitral valve without a stalk. The patient underwent urgent resection of the tumor. Median sternotomy was performed and cardiopulmonary bypass was established via aorto-bicaval cannulation. An incision was made in the right side of the left atrium, and a 10 × 8 mm yellow-white mass was located on the atrial surface of the anterior mitral valve leaflet. On intraoperative frozen section examination, the tumor was suspected to be a myxoma. The tumor was excised with a clear resection margin. Mitral valve repair was attempted but was unsuccessful, and the patient underwent mitral valve replacement. Postoperative histological examination showed that the mass had avascular papillary fronds lined by endothelial cells, resulting in a final diagnosis of papillary fibroelastoma. The patient's postoperative course was uneventful, and he was discharged 26 days after surgery.

Patient 2

A 75-year-old man was referred to our hospital for investigation and treatment of a cerebral infarction. Magnetic resonance imaging revealed multiple cerebral infarctions, and magnetic resonance angiography showed no evidence of cerebrovascular or carotid artery disease. His infarctions were suspected to be due to embolization. Electrocardiography showed normal sinus rhythm. Transthoracic echocardiography revealed a nodular, mobile, pedunculated mass in the left ventricular outflow tract. Echocardiography did not show any left or right ventricular systolic dysfunction. The heart valves appeared normal, with no signs of regurgitation or stenosis. Transesophageal echocardiography showed an oval, mobile, gelatinous mass (maximum diameter 14 × 10 mm) attached by a thin stalk to the left ventricular outflow tract (Fig. 1). After establishment of cardiopulmonary bypass, the aorta was opened with an oblique incision. A 10 mm mass was located in the left ventricular outflow tract and was resected. Histological examination showed that the mass had avascular papillary fronds lined by endothelial cells, compatible with papillary fibroelastoma. The postoperative course was uneventful and the patient was discharged 22 days after surgery. He has been free of cerebrovascular events since discharge.

Patient 3

A 67-year-old woman presented to our hospital for the treatment of arteriosclerosis obliterans. Transthoracic echocardiography revealed a mobile mass on the non-coronary cusp of the aortic valve, measuring 6.9 ×

Fig. 1 Transesophageal echocardiography, demonstrating a mobile, round, gelatinous tumor measuring 14 × 10 mm in the left ventricular outflow tract.

Fig. 2 When the excised tumor was immersed in saline solution, it resembled a sea anemone.

6.9 mm. We advised surgery, but the patient refused. After percutaneous transluminal angioplasty of the left external iliac and femoral arteries, she was monitored closely. After 6 months, she agreed for surgery. There were no signs of embolism during the time she was monitored. Intraoperative transesophageal echocardiography showed a homogeneous, mobile, speckled mass. Three-dimensional echocardiography confirmed it was attached to the ventricular side of the non-coronary cusps of the aortic valve by a short stalk. The operation was performed under normothermic cardiopulmonary bypass, using ascending aortic and bicaval cannulation. After cardiac arrest using antegrade cardioplegia, the

Table 1 Patient characteristics

	Age (yr)	Sex	Location	Tumor size (mm)	Echocardiographic characteristics	Symptom	Procedure	Recurrence
Case 1	28	M	Mitral valve	8 × 7	Homogenous, round mobile mass without a stalk	Chest pain	Valve replacement	None
Case 2	75	M	Left ventricular outflow tract	14 × 10	Oval, mobile, gelatinous mass with a thin stalk	Hemiplegia	Simple resection	None
Case 3	67	F	Aortic valve	6.9 × 6.9	Homogenous, mobile, speckled mass with a short stalk	None	Simple resection	None

aorta was opened with an oblique incision. A 10 mm gelatinous mass was found on the non-coronary cusp of the aortic valve, and was resected together with its stalk. When immersed in water, the resected mass had a sea anemone-like shape (**Fig. 2**). Histological examination of the resected tumor revealed papillary proliferation with collagenous tissue and fibroblast infiltration, and an endothelial cell lining. The patient's early postoperative period was uncomplicated, and she was discharged on postoperative day 14.

Discussion

Primary cardiac tumors are uncommon, and three-fourths of them are benign. Papillary fibroelastoma accounts for 4.4% to 8% of primary cardiac tumors^{3,4)} and is the third most common cardiac tumor after myxoma and lipoma.⁵⁾ Papillary fibroelastoma most often arises from the valvular endocardium (60%–84%)^{2,6)} and is the most common cardiac valvular neoplasm. Gowda and colleagues⁶⁾ reported that the valvular surface was the predominant location (84% of 611 cases): the tumor was located on the aortic valve in 44% of cases, the mitral valve in 35%, the tricuspid valve in 15%, and the pulmonary valve in 8%. In our cases, two tumors were found on the valvular surface, and the other was in the left ventricular outflow tract (**Table 1**). Most papillary fibroelastomas are relatively small, and they commonly have a stalk. The shape varies, with some tumors having a well-developed head and others having elongated strand-like projections.

Patients with cardiac papillary fibroelastoma are seldom symptomatic, except for embolic events which are reported to occur in about 35%.²⁾ Although the tumors are benign, left-sided tumors may cause life-threatening complications such as stroke, angina, myocardial infarction, transient ischemic attack, mesenteric ischemia, renal infarction, or limb ischemia.^{2,5,7)} The most common

clinical symptoms are neurological, accounting for 53% of symptoms,⁸⁾ and even very small emboli may cause stroke.⁹⁾ In contrast, right-sided tumors are usually asymptomatic until they become large enough to interfere with intracardiac blood flow.¹⁰⁾ In our cases, all the tumors were left-sided. Although the tumors were small, two of them presented after embolization. Even very small tumors may cause embolic events.

Echocardiography is useful for differentiating from vegetations, thrombi, and myxoma.

Recently, three-dimensional echocardiography has been successfully employed in papillary fibroelastoma diagnosis.^{11,12)} In our cases, it gave us details of shape and mobility and was helpful especially for the location and the relationships of the tumor. Papillary fibroelastoma typically forms a round, mobile, oval or irregular, well-demarcated, homogenous, pedunculated mass, which may have a speckled appearance.^{2,7)} In contrast, myxoma has heterogeneous appearance with hypoechoic and hyperechoic foci and a broad-based pedicle attached to interatrial septum and exhibit little mobility.^{12,13)}

On intraoperative frozen section examination, it can be difficult to differentiate between myxoma and papillary fibroelastoma because myxoma may be solid and have a similar appearance to papillary fibroelastoma. However, it is important to differentiate between these tumors pre- and intraoperatively because the surgical approach may differ depending on the tumor type.

As recurrence has rarely been observed after surgical resection of fibroelastoma,^{6,7)} shaving of the tumor to preserve valve function is usually recommended.^{8,14)} In contrast, excision of myxoma requires a clear surgical margin because recurrence occurs in about 5% of patients. These different surgical approaches emphasize the importance of determining the type of tumor pre- and intraoperatively. In Patient 1, the intraoperative pathological diagnosis of myxoma caused us to excise the tumor with wide margins and subsequently perform

mitral valve replacement. Mitral valve plasty might have been possible if fibroelastoma had been diagnosed intra-operatively.

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