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Cardiac Papillary Fibroelastoma Result in Acute Left Main Coronary Artery Disease: A Case Report

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ABSTRACT

Background: Cardiac papillary fibroelastoma is a rare, benign primary cardiac tumor that remains asymptomatic. Severe complications have been reported in some cases. The only effective therapy is surgical excision.

Case presentation: Here, we report a case of cardiac papillary fibroelastoma with the initial symptom of chest pain and a first diagnosis of acute left main coronary artery disease. This patient eventually underwent tumor excision surgery and recovered well.

Conclusion: For patients with symptomatic cardiac papillary fibroelastoma, we provide a series of comprehensive data from before, during, and after surgery. This might be helpful for the future diagnosis and treatment of these tumors.

INTRODUCTION

Cardiac papillary fibroelastoma is classified as a rare, benign primary cardiac tumor, accounting for approximately 10% of all cardiac tumors [Edwards 1991]. Although most patients remain asymptomatic, the tumor might occasionally result in severe complications. The diagnosis of such tumors usually is incidental, and the only effective therapy is surgical excision. Here, we present a patient with non–ST-segment elevation myocardial infarction (NSTEMI) of acute left main coronary disease secondary to fibroelastoma on the aortic valve. This patient successfully was treated with surgical intervention.

CASE PRESENTATION

A 75-year-old female with chest pain and palpitations was admitted for acute coronary syndrome from the emergency department. The initial electrocardiogram was negative for ischaemia, along with mild elevation of the initial serum troponin T level of 61.5 ng/L. After further episodes of chest pain and ST depression in leads V_{2-6}, I, II, III, and $_aV_F$, and ST elevation in lead $_aV_R$, the patient was considered to have left main trunk (LMT) obstruction and referred for cardiac catheterization. No coronary stenosis was observed on coronary angiogram (Figure 1). However, during angiography, this patient developed shortness of breath and decreased blood pressure. Therefore, atropine and dopamine were administered to treat the hypotension. When her discomfort resolved, she was transferred to the cardiac care unit for special care and more examination.

Further blood tests showed a peak serum troponin T level of 4852.0 ng/L. Repeated electrocardiogram revealed further widespread ST-segment depression. Moreover, the patient felt exhausted, and she found it difficult to lie flat. Thus, echocardiography and computed tomographic angiography (CTA) were considered to identify whether she had heart failure, aortic dissection, or pulmonary embolism. Unexpectedly, echocardiogram demonstrated an isodense mass on the upper margin of the right coronary cusp, while CTA demonstrated a nodular filling defect on the aortic valve (Figure 2).

Due to angina, a history of myocardial infarction and the presence of tumors, the patient received surgical intervention. Transesophageal echocardiography (TEE) also was performed both during and after the operation to reveal the tumor, guide the excision, and evaluate valvular function. The long axis (at the level of the left ventricle) and short axis (at the level of the aortic valve) during the operation demonstrated an 18×12 mm pedunculated, isodense mass attached to the left coronary cusp margin of the aortic valve, which moved along with the opening and closing of the aortic valve. There also was mild regurgitation of the aortic valve (Figure 3). Aortotomy identified an approximately 2×1 cm pedunculated mass arising from the left coronary cusp margin

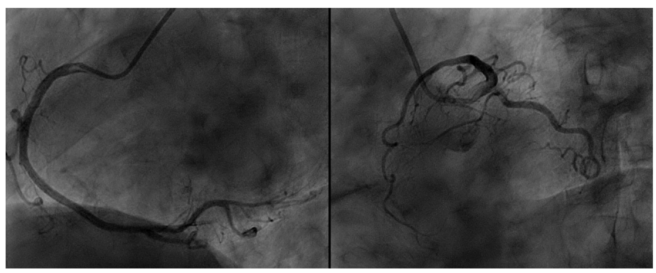

Figure 1. There was no obstruction in either the left or right coronary artery.

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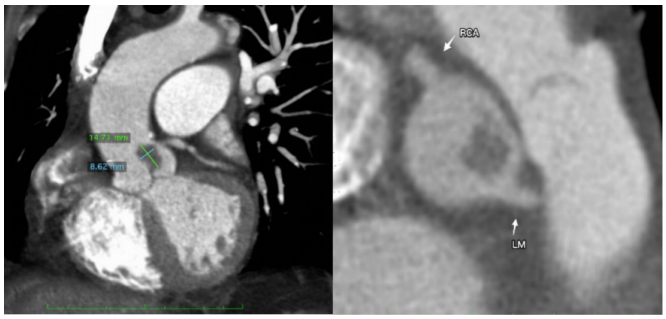

Figure 2. An isodense mass on the upper margin of the right coronary cusp. CTA demonstrated a nodular filling defect on the aortic valve approaching the left coronary ostia.

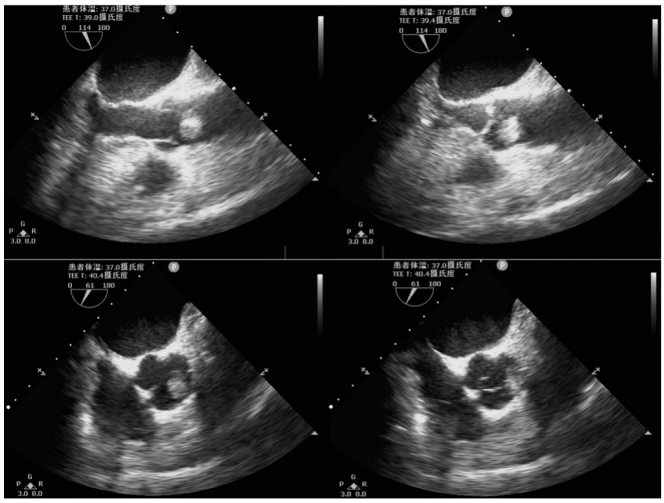

Figure 3. Intraoperative echocardiography showed an 18×12 mm pedunculated, isodense mass attached to the left coronary cusp margin of the aortic valve, which moved with the opening and closing of the aortic valve.

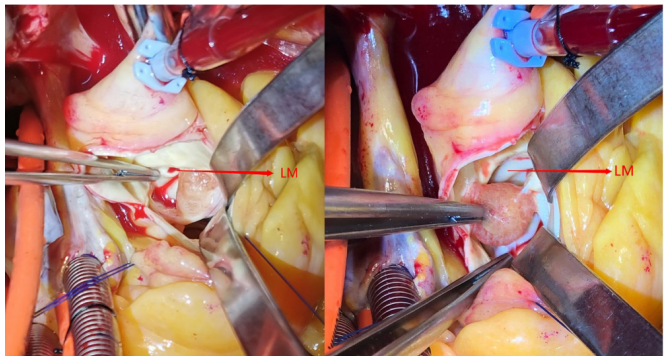

Figure 4. An approximately 2×1 cm pedunculated mass resembling a sea anemone arose from the left coronary cusp margin of the aortic valve near the boundary between the left and right cusp.

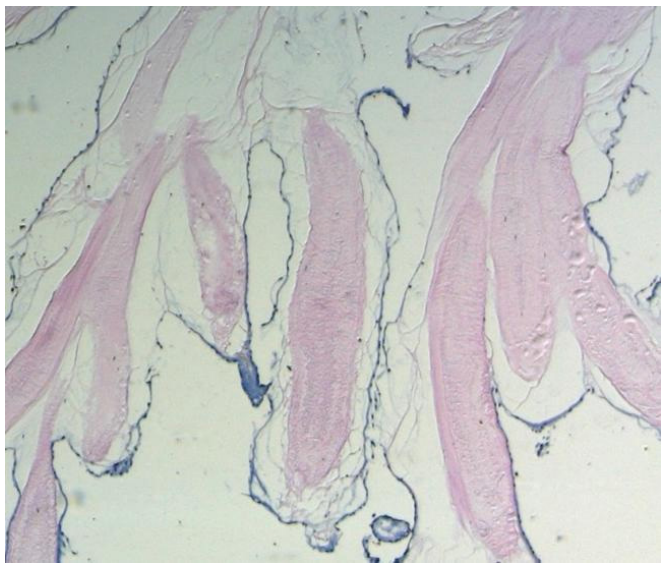

Figure 5. Microscopic appearance of the "tumor" showing a mass of villous projections of myxoid connective tissue with endothelial cell proliferation.

of the aortic valve near the boundary between the left and right cusp. It was freely mobile with multiple fronds, resembling a sea anemone (Figure 4). There was no thrombus on its surface, and it was suspected to intermittently obstruct the left coronary ostia by its movement. During the surgery, the tumor was dissected from the valve, and the aortic valve was repaired by suturing the left coronary cusp margin. Postoperative TEE demonstrated that valvular function was good. Histologic characteristics of the tumor specimen included a center core of collagen and elastin and the characteristic papillary fronds of a cardiac fibroelastoma (Figure 5). The patient did well postoperatively and was discharged home on the 10th postoperative day (Figure 6).

DISCUSSION AND CONCLUSION

Cardiac papillary fibroelastoma is classified as a rare, benign primary cardiac tumor accounting for approximately 10% of all cardiac tumors [Reynen 1996]. The etiology of this tumor still is unclear. There are many hypotheses of the original mechanism, including neoplasms, hamartomas, organized thrombi, and unusual endocardial responses to infection or hemodynamic trauma [Almagro 1982; Grandmougin 2000; Kurup 2002]. The histological feature is described as an inner vascular core of connective tissue covered by endothelium that surrounds a layer of acid mucopolysaccharide, which often has a peduncule or stalk, resembling a sea anemone [Sun 2001].

As a past report presented, cardiac papillary fibroelastoma can be found anywhere inside the heart, such as valvular surfaces, papillary muscles, chordae tendineae, ventricular septum, or endocardial surface [Gowda 2003]. Frequently, it appears on the aortic valve or mitral valve, especially on the aortic valve [Gowda 2003; Anastacio 2012]. The location of the tumor may be associated with the occurrence of complications.

Although most patients are asymptomatic, serious complications have been reported, such as neural symptoms, angina,

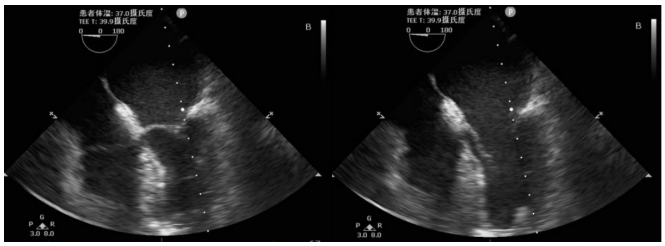

Figure 6. The echocardiogram after the operation showed that the function of the aortic valve was normal.

myocardial ischaemia, and distal embolic events [Alawi 2002; Alozie 2011; Gilbert 2009; Zamora 1995]. The mechanism is thought to be intermittent obstruction of the coronary ostia or valve by the mobile tumor mass and embolization of the tumor mass or platelet thrombi on the tumor into the coronary arteries or distal arteries [Deodhar 1997]. It also can cause sudden death by occlusion of the coronary artery, as reported on autopsy [Butterworth 1973]. As a result, the tumor can pose potential risks to patients who have yet to be symptomatic.

In living patients, diagnosis was rare before the introduction of echocardiography [Topol 1986]. With the development of radiological technology, computed tomography scans (CT) or magnetic resonance imaging (MRI) has shown limited use for diagnosing this tumor [de Visser 2008; Binhas 2019]. Of all these techniques, echocardiography is described as a convenient and noninvasive way to diagnose this tumor and should be the first choice when searching for cardiac papillary fibroelastoma, which can be demonstrated as a pedunculated, mobile, and echodense mass. The use of TEE provides a more visualized image to identify the location and guide excision. CT and MRI can be supplements to echocardiography.

Surgical excision is the only therapy for cardiac papillary fibroelastoma [Mariscalco 2010]. For symptomatic patients, excision is recommended regardless of the size. For asymptomatic patients, mobile lesions or lesions in specific locations, such as the coronary ostia, should also be surgically excised because of the higher risk of obstructive and thromboembolic complications. Valvuloplasty or valve replacement often needs to be performed after cutting the tumor. Other patients with nonmobile lesions less than 1 cm in diameter can be managed expectantly or with anticoagulation. In addition, recurrence of cardiac papillary fibroelastoma is very low [Sun 2001].

In this case reported here, the patient presented with angina and was admitted for the primary diagnosis of acute coronary syndrome and paroxysmal atrial fibrillation. However, the coronary angiogram was normal, while the definite symptoms, electrocardiogram, and enzymatic test suggested this patient had a myocardial infarction. Subsequent echocardiography and computed tomography (CT) scans both revealed a mass on the left coronary cusp margin of the aortic valve. Due to persistent symptoms and the presence of an aortic valvular mass, surgical excision was performed. The location of this mass was identified intraoperatively. Histological examination confirmed that the mass was cardiac papillary fibroelastoma. According to other reported cases, the patient's myocardial infarction may have been caused by embolization of tumor fragments or thrombotic material, but it is more possible that the tumor blocked the coronary ostia, causing coronary ischaemia. To our knowledge, this might be the most comprehensive case of cardiac papillary fibroelastoma, which is why we meticulously have recorded the patient's primary symptoms and their enzymatic, electrocardiogram, angiogram, echocardiogram, CT, and histological test results before and after the operation. We believe the findings in this patient might be helpful for the diagnosis and treatment of these tumors.

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Ethics approval and consent to participate: Written informed consent was obtained from the patient for publication of this case report and any accompanying images. The ethics committee at West China Hospital has approved the case report to be submitted.

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