Multiple papillary fibroelastoma in left ventricle associated with obstructive hypertrophic cardiomyopathy

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Abstract

A 68-year-old man was referred to our hospital with a left ventricular (LV) mass and obstructive hypertrophic cardiomyopathy (HOCM). Although he was treated for the LV thrombosis and received anticoagulation therapy, the mass did not decrease in size for three years. His past history was colon cancer which was resected in its early phase. Laboratory studies revealed the absence of any inflammatory and tumor marker symptoms. Transthoracic and transesophageal echocardiography revealed a mass of 24×11 mm in diameter attached to the septal wall of LV and another two or three masses detected in LV wall. He received surgical treatment with complete mass excision with LV dissection and hypertrophied ventricular muscle was resected. Surgical resection of these LV masses and septal myectomy was performed. The histological examination showed that the lesions had a papillary configuration with an avascular connective tissue core lined by a single layer of endothelial cells, which was sufficient for a diagnosis of cardiac papillary fibroelastoma (CPF). The patient recovered without any complications.

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Keywords: Papillary fibroelastoma; Obstructive hypertrophic cardiomyopathy; Cardiac tumor

1. Introduction

The incidence of primary cardiac tumors is low at 0.002–0.33%. Cardiac papillary fibroelastoma (CPF) is a rare primary tumor that accounts for 8% of all cardiac tumors, and is the third most common benign cardiac tumor after myxoma and lipoma [1, 2]. As rare as these tumors are, the incidence of multiple fibroelastoma in left ventricle (LV) is even rarer [3–5].

We describe an unusual case of multiple CPFs of the LV. A 68-year-old man was referred to our hospital from an outside facility with a LV mass in 2006, and he had no symptom. Since 2004, he was treated for the LV thrombosis and received anticoagulation therapy; the mass did not decrease in size for three years. His past history was colon cancer which was resected in its early phase.

By transthoracic and transesophageal echocardiography, a round and cystic mass of 24×11 mm in diameter was found attached to the septal wall of LV and another two or three masses detected in LV wall (Fig. 1). Valve function and LV wall motion was good. LV outflow tract was stenosis by septal wall thickness. A ventricular wall was thickened, accelerated blood flow in LV outflow tract was 4.8 m/s, and it was diagnosed as the obstructive hypertrophic cardiomyopathy (HOCM). Scintigraphy and computed tomography showed no evidence of another tumor and embolism. Laboratory studies revealed the absence of any inflammatory and tumor marker symptoms. Catheter angiography showed that there was no feeding artery for this tumor and coronary artery was intact. Result of physical examination and laboratory profile concluded tumor markers were unremarkable. Surgical excision was recommended in consideration of the known potential for coronary, cerebral, and systemic embolization.

The patient was brought to the operating room. Bicaval and ascending aorta cannulation were accomplished and total cardiopulmonary bypass was initiated. The stenosis of LV outflow tract made the transaortic approach less desirable. Transmitral valve approach was not adequate because there were masses at the back of the mitral valve. A ventricular incision was chosen.

On opening the LV, there were masses attached to the septal wall of LV and another three masses detected in LV wall. A pedicled mass with delicate papillary fronds resembling a sea anemone was seen arising from the endocardial surface at the LV septal wall (Fig. 2). Resection of these masses and septal myectomy was performed. Intraoperative transesophageal echocardiography demonstrated no residual mass and accelerated blood flow in LV outflow tract had decreased to 2.8 m/s.

The histological examination showed that the lesions had a papillary configuration with an avascular connective
tissue core lined by a single layer of endothelial cells, which was sufficient for a diagnosis of CPF.

The postoperative course was uncomplicated.

2. Comment

CPF is a rare primary tumor that accounts for 8% of all cardiac tumors, and is the third most common benign cardiac tumor after myxoma and lipoma [1, 2]. Between 77% and 90% of CPFs appear on cardiac valves, and the remainder have been reported to originate from nearly all portions of the non-valvular endocardium including the LV septum, the LV mural endocardium, the right ventricular outflow tract, the right atrium, the atrial septum, the papillary muscles, the chordae tendinea, the eustachian valve, and the Chiari network. Multiple CPF were detected in 8% [3].

Despite their small size, CPFs have been reported to cause angina and sudden death from either coronary emboli or direct ostial occlusion by prolapsing tumor. Although CPFs are considered benign tumors, their clinical course is deceptive. CPFs should be resected even in the patients who have no symptoms.

It is often avoided by the LV resection with its potential complications; bleeding, arrhythmia and declined LV function. However, a ventricular incision was very useful for this patient complicated with HOCM and multiple masses because all masses could be observed and aberrant cardiac muscle could be resected well.

References


eComment: Cardiac papillary fibroelastoma: a current assessment

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Reading your interesting case report [1], several questions arise. It is notable that the patient was treated for three years with anticoagulants, while the left ventricle mass seems not to have been investigated during the above period. Particularly, symptomatic cardiac papillary fibroelastoma (CPF) should be surgically removed whereas asymptomatic lesions left-sidied or > 1 cm (as in your patient) should be considered for surgical excision according to the current literature [2]. The fact that you did not perform magnetic resonance imaging before the operation is also notable.

Generally, transesophageal echocardiography is the recommended method for diagnosis, with an overall sensitivity of 77% [2, 3]. Computed tomography is inferior to echocardiography in depicting small moving structures, such as on the cardiac valves, because it does not allow true real-time imaging. Magnetic resonance imaging is more valuable than computed tomography as it allows imaging in multiple planes and better soft-tissue characterization of the tumor. The main disadvantage of magnetic resonance imaging is its susceptibility to motion artefact [4]. Generally, cardiac catheterization and selective angiography are not necessary in most patients with CPF, as adequate information can usually be obtained by non-invasive imaging. Especially, cardiac catheterization appears warranted in cases where the other techniques have not fully defined the CPF or when another cardiac condition warranting cardiac catheterization is suspected. In contrast to non-invasive diagnostic techniques, cardiac catheterization can be associated with an added risk to patients because the catheter may dislodge a fragment of the tumor or adhere thrombi, resulting in embolism [3].

CPF is generally slow growing tumors but may serve as a nidus, allowing formation of large superimposed thrombi over a short period of time. About 77% of CPFs originate on the valves, and the other 23% in the endocardial non-valvular surface, whereas > 95% arise in the left heart [3]. Symptomatic CPFs should be surgically resected; the treatment of asymptomatic tumors...
is less clear. Large, left-sided mobile tumors should be excised to prevent sudden death and emboli. Small, non-mobile tumors may be followed with serial echocardiography and removed if they increase in size, or become mobile or symptomatic [2]. CPFs are usually pedunculated and may easily be removed with associated endocardial tissue. The root of the pedicle and the full thickness of endocardium involved are excised. Care should be taken to avoid fragmentation of the tumor to prevent embolization.

Various methods have been proposed for complete excision of left-sided CPFs to avoid a left ventriculotomy with its further complications. When a CPF is located on the aortic valve or on the left ventricular outflow tract, it is best approached via the transaortic route, and when located on the mitral valve, it is best approached via a left atriotomy [5]. For tumors deep within the left ventricular cavity close to the apex, the use of a cardioscope passing through the aortic or mitral valve is recommended to avoid damage to the valvular apparatus. Undoubtedly, surgery is curative with excellent short- and long-term prognosis after surgical removal. Moreover, recurrence after surgical resection has not been reported.

References