Chordal Papillary Fibroelastoma of the Mitral Valve: A Dramatic Presentation of a Tiny Mass

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Abstract

Papillary fibroelastoma (PFE) is a rare benign endocardial tumor, primarily affecting the cardiac valves. It predominantly affects the aortic valve followed by the mitral valve. It can be silent on presentation or can present with complications like devastating systemic embolization, myocardial infarction, and death. We describe a rare case of a young man hospitalized for the investigation of cerebrovascular events. On transesophageal echocardiography (TEE), a PFE was identified and resected successfully through a minimally invasive surgery. Our case illustrates that the possibility of embolization from a small cardiac tumor as an independent factor for planning the surgical resection.

Keywords

Papillary fibroelastoma, Minimally invasive cardiac surgery, Right anterior thoracotomy, Cardiothoracic surgery, Cardiac tumors

Introduction

Papillary fibroelastoma is the second most common primary cardiac tumor [1]. It primarily involves the cardiac valves and its clinical presentation ranges from asymptomatic patients to those presented with thromboembolic events. Transesophageal echocardiography, along with clinical presentation, is important for its diagnosis [2]. Although guidelines for asymptomatic fibroelastomas are still unclear, both symptomatic and asymptomatic patients are treated with surgical resection of fibroelastomas to avoid thromboembolic complications. It can be treated by conservative procedures but large fibroelastomas and embolic complications associated with them mandate surgical resection for curative and long-term postoperative prognosis [3]. Here we present the case of a 30-year-old male who presented to the emergency department after an episode of ischemic stroke. The stroke manifested as a left-sided weakness that lasted for 30 minutes. Computed tomography (CT) and magnetic resonance imaging (MRI) were done which revealed a right middle cerebral artery infarction. Echocardiography was also done which revealed a mass attached to the anterior mitral leaflet. It was diagnosed as PFE. The mass was surgically removed to avoid cardiovascular and embolic complications.

The patient was then discharged and was doing well on the follow up.

Case

A 30-year-old male presented to emergency department (ED) after an episode of transient ischemic stroke manifested predominantly as left-sided weakness of 30-minute duration before admission with no residual deficits. He had no significant co-morbidities. The patient was a smoker. He underwent a head computed tomography (CT) and brain magnetic resonance imaging (MRI), which revealed right parietal hypo density with right middle cerebral artery infarction with no hemorrhagic changes (Figure 1). Given the concern for potential embolism, a pan-computed tomography...
(CT) was obtained, revealing a wedge shape peripheral hypodensity in the upper pole of the spleen as well as in both of the kidneys. The findings are in keeping with splenic and renal infarctions. Transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE) was performed that revealed a normal ejection fraction with normal mitral valve functions. The sub-valvular mitral apparatus showed a flickering, mobile, echo-dense mass, 5 mm long and 5 mm in diameter, attached to the chordae tendinea of the middle segment of the anterior mitral leaflet A2 (Figure 2 and Figure 3). These characteristics are given the diagnosis of a tumor, a papillary fibroelastoma (PFE) of the mitral valve. Infective endocarditis was ruled out by negative inflammatory blood tests including complete blood count and blood cultures.

After cardiac surgery consultation, a decision of surgical resection was made because of cerebrovascular and cardiovascular events. The patient consented to low operative risk (Euro SCORE II of 0.5%). The surgery was conducted through a small right mini-thoracotomy incision (7 cm). Peripheral cardiopulmonary bypass was installed through the right femoral artery and right internal jugular vein. After cardiac arrest and left atriotomy, we found a gelatinous mass attached to the ventricular aspect of the anterior mitral valve leaflet and sub-valvular apparatus. The mass was shaved carefully without damaging the chordae (Figure 4). Intra-operative TEE showed no residual masses with good mitral valve function. The postoperative period was uneventful, and the patient was discharged on day 6.
During the follow-up period, the patient was doing well. Histopathology of the excised mass confirmed the diagnosis of the papillary mitral valve fibroelastoma (Figure 5).

Discussion

Papillary fibroelastoma (PFE) is a primary cardiac tumor that primarily involves the heart valves. It is the second most common primary cardiac tumor. It is generally benign and asymptomatic but can often lead to fatal complications such as myocardial infarction, stroke, pulmonary embolus, cardiac arrest etc [1]. It can occur anywhere on the endocardium, most commonly arising in the left side of the heart and in the valvular structures. PFEs can arise from aortic valve (29%), mitral valve (25%), tricuspid valve (17%) and pulmonary valve (13%) [2]. In a recent study by Abu Saleh, et al., out of fourteen patients who underwent surgical excision of PFE, 50% originated from the left side of the heart [3].

Clinically these benign tumors are generally small and asymptomatic [4]. However, they may possess life-threatening malignant complications such as stroke, myocardial infarction, acute myocardial infarction, angina pectoris, ventricular arrhythmia, and even sudden death [5].

Echocardiography (transthoracic and transesophageal) is necessary for evaluation of PFE. Laboratory findings, clinical information and blood cultures are also important for differential diagnosis [6].

There is a great deal of debate surrounding the need for surgical treatment of asymptomatic patients, as anticoagulation and antiplatelets can be offered to these patients. However, convincing data from the randomized trial have demonstrated that cerebrovascular accidents were observed in patients suspected to have PFE who did not undergo surgical excision [7].

While the guidelines for asymptomatic patients are still unclear, most of the current evidence strongly recommends surgical resection of in symptomatic patients. The surgical risk is low and repair can be achieved in virtually all patients with excellent post-operative prognosis [8,9]. One study recommends surgical excision for PFEs greater than 10 mm and for masses smaller than 10 mm, serial echocardiographic evaluation rather than surgery was recommended [10].

We present a case of a 30-year-old male who presented to the emergency department after an episode of ischemic stroke. The stroke manifested as a left-sided weakness that lasted for 30 minutes. Computed tomography (CT) and magnetic resonance imaging (MRI) were done which revealed a right middle cerebral artery infarction. Echocardiography was also done which revealed a mass attached to the anterior mitral leaflet. It was diagnosed as PFE. The mass was surgically removed to avoid cardiovascular and embolic complications. The patient was then discharged and was doing well on the follow up. In our experience, regardless of whether the size of the tumor is small or large, it is important to resect it because of the possibility of embolization or valvular obstruction.

Conclusion

Papillary fibroelastoma can cause cerebrovascular events, and surgical excision, regardless of the size of the mass, is curative for long-term management with an excellent prognosis. As compared to the conventional median sternotomy minimally invasive approach through a right mini-thoracotomy for the resection of benign cardiac tumors can be performed safely with improved patient outcome and it should be considered for the patients of papillary fibroelastoma.

Declarations

Conflict of interest

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Availability of data and materials

All data are available and attached.

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References


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