CASE REPORTS

LEFT VENTRICULAR PAPILLARY FIBROELASTOMA

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Abstract

Introduction: *Cardiac papillary fibroelastoma is a rare benign primary cardiac tumor typically found on the valvular structures of the heart. Nonvalvular papillary fibroelastomas are exceedingly rare. We report the case of a 66-year-old male who presented with cerebellar infarctions and was found to have a mobile mass attached to the endocardial surface of the anteroseptal wall of the left ventricle. Cardiac magnetic resonance imaging demonstrated late gadolinium enhancement. Surgical excision of the cardiac mass was performed via the transaortic approach without intra-operative complications. Histopathologic examination confirmed the diagnosis of a papillary fibroelastoma. Some aspects related to the etiology, diagnosis and management of this entity are discussed.*

Keywords: *papillary fibroelastoma, heart neoplasms, echocardiography, cardiac magnetic resonance imaging.*

INTRODUCTION

Primary cardiac tumors are rare and the majority are benign. Estimated incidence ranges between 0.02% in autopsy series and 0.089% in echocardiographic studies[1-3]. Papillary fibroelastoma (PFE) is a benign tumor of the endocardium. Until the introduction of echocardiography, most PFEs were incidental findings in autopsy studies or during cardiac surgical procedures. However, with the widespread application of ultrasonic studies as well as computed tomography scans (CT-Scans) and magnetic resonance imaging (MRI), the reported numbers of diagnosed cases of PFE have notably increased[1-3]. Traditionally, PFE has been considered as the third most frequent cardiac tumor after cardiac myxoma and cardiac lipoma 1. However, based on more recent echocardiographic studies in selected populations, some authors reported that PFE was the most common primary cardiac tumor[2, 3]. In more than 80% of the reported cases, PFEs are attached to the valvular endocardium, with the aortic valve being the most frequently involved. However, they can also be related to the non-valvular endocardium of any cardiac chamber [1-4]. Left ventricular PFE is very rare, usually presenting as a mobile mass anchored to the endocardial surface of the left ventricle (LV) and whose differential diagnosis includes mural thrombi, other cardiac tumors, and vegetations [5-7]. In this report, we describe a rare case of a 66-year-old patient who presented with cerebellar stroke secondary to emboli from left ventricular PFE.

CLINICAL CASE

A 66-year-old man with a history of arterial hypertension, dyslipidemia, and prostate hyperplasia presented to our hospital with a 48-hour history of headache, vertigo, nausea and vomiting. Physical examination was unremarkable except

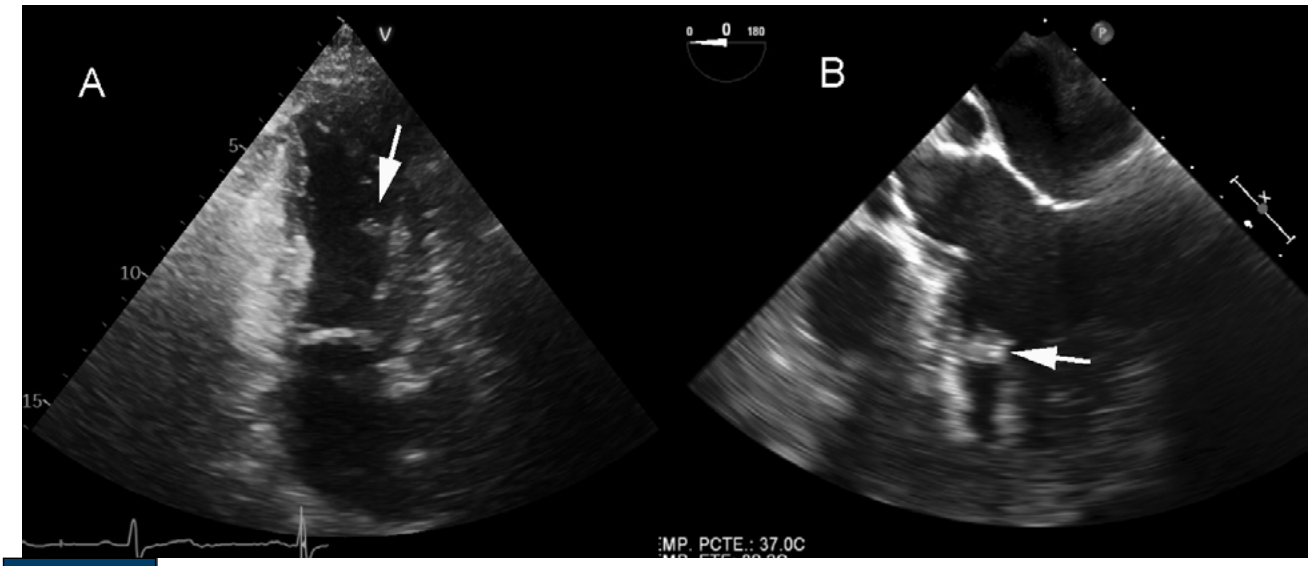

Figure 1

Transthoracic and transesophageal echocardiography. A. Apical two-chamber view on transthoracic echocardiogram showing a 1x1 cm mass (arrow) arising from the anteroseptal wall of the left ventricle. B. The same mass is seen in apical five-chamber view on transesophageal echocardiogram (arrow).

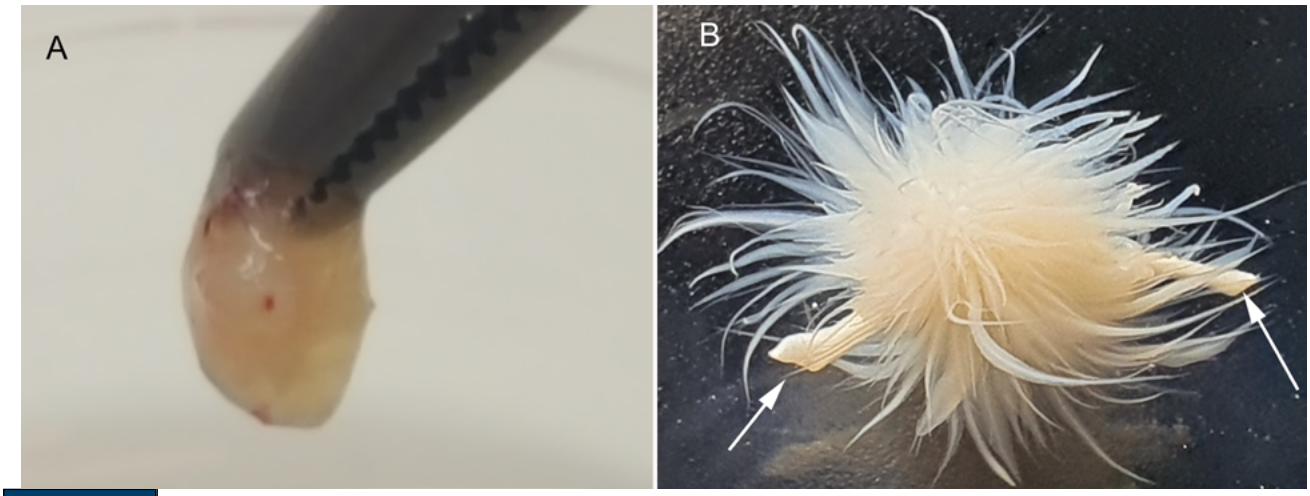

Figure 2

Cardiac magnetic resonance imaging. A round 10 mm diameter mobile mass is attached to anteroseptal left ventricular wall that shows enhancement 10 minutes after gadolinium administration (arrow).

for gait instability. His laboratory tests and electrocardiogram did not show any abnormal findings. CT scan of brain demonstrated a hypodense lesion in the left posteroinferior cerebellar territory compatible with subacute infarction.

To exclude a cardiac source for emboli, a transthoracic followed by transesophageal echocardiograms were performed. These showed a 10x8 mm mobile intraventricular mass of intermediate echogenicity that was anchored at the level of the anteroseptal wall of the LV close to the base of papillary muscle (figure 1). Cardiac MRI confirmed these findings and showed a late gadolinium enhancement consistent with a primary cardiac tumor (figure 2).

With the presumed diagnosis of a possible cardiac tumor, the patient was referred to our cardiac surgery unit for surgical resection of the mass four weeks after his stroke. The procedure was performed under cardiopulmonary bypass and using the transaortic approach. Intraoperatively, there was a gelatinous mass of approximately 1 cm in diameter, anchored by a thin pedicle similar to a tendinous chord to the ventricular wall in proximity to the anterior papillary muscle, which was ressected with a small strip of endocardium to obtain a tumor free margin. Macroscopic features and histopathological examination were consistent with PFE (figure 3, and figure 4). Postoperative course was uneventful, and the patient was discharged home on postoperative day 12th after recovering gait stability.

DISCUSSION

PFE can affect patients of any age, however the peak incidence occurs around 60 years of age [1], and it does not seem to have sex predilection. It usually presents as a solitary mass

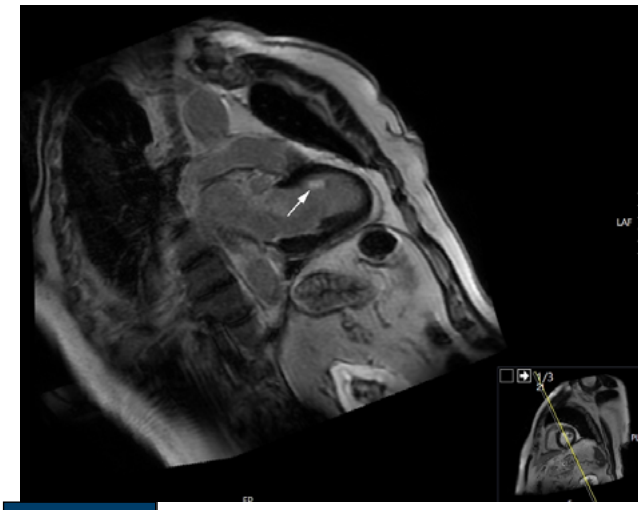

Figure 3

A mucinous friable mass attached to the left ventricular wall was excised A. Out of water the tumor looked like a solid mass covered with gelatinous material B. In water the tumor had the appearance of a sea anemone. The tumor presented two stalks mimicking small chordae tendineae (arrows).

but cases of multiple PFEs have also been described[8]. The most frequent location is the endocardium of the non-coronary leaflet of the aortic valve, although it can affect any valve or cardiac chamber[1-3].

Macroscopically, PFE is a small, cream-coloured, soft gelatinous mass of friable fronds[1, 5,6]. Thrombi can frequently be found adhered to its surface, hence the high risk for systemic embolization [1-3]. When immersed into a liquid medium, they resemble the appearance of a sea anemone [1-4]. Microscopically, avascular papillary fronds consisting of collagen and elastin lined by endothelial cells can be observed[1-3].

It has been suggested that PFEs are not true tumors but lesions that are reactive to mechanical trauma, radiotherapy, or inflammatory processes [1-3,9]. The presence of small chordae tendinae at the base of many PFEs, as we were able to observe in our patient, supports the hypothesis that it could be a hamartoma [1-3]. However, rKRAS gene mutations have been identified in up to 8% of PFEs, which may suggest the neoplastic origin of this tumour in this subtype [2].

Although PFEs are benign tumors, most are clinically silent and very often incidental findings in echocardiographic and radiological studies [1, 2].The most common presenting scenarios are cerebrovascular ischemia, followed by myocardial ischemia in the form of infarction or angina[1-3]. Other forms of presentation include dyspnea, syncope, sudden death, renal and mesenteric embolism, fever, and thrombocytopenia [1, 6]. Right-sided valvular or intra-cavitary PFEs can cause pulmonary embolism and pulmonary hypertension [1, 2].

The echocardiographic appearance of PFE is very characteristic with a specked appearance and stippling along the edges. Transesophageal echocardiography is more sensitive than transthoracic echocardiography for PFE [1, 2]. Cardiac MRI is also useful for the diagnosis of PFE due to its greater capacity for myocardial tissue characterization as compared to CT-scan [2, 3, 7].

Some authors suggest a conservative non-surgical approach and the administration of anticoagulant/antiplatelet therapy in small and slightly mobile PFEs due to the low risk of complications[1]. However, most authors recommend surgical resection as soon as possible, regardless of the size, due to the potential for systematic embolism [2-3]. In our case, the neurologists decided to delay the surgical procedure by four weeks because of the persistence of vertigo, vomiting and gait instability.In terms of surgical approach, valvular PFEs can be excised without the need for prosthetic valve replacement [1-3,6]. During the intervention, careful manipulation of the tumor is required to avoid its fragmentation and embolization [2, 3, 6]. There are some reports of recurrent PFEs after successful surgical resection, therefore, long-term follow-up with periodic echocardiographic assessment is advised [4, 10].

CONFLICTS OF INTEREST

The authors declare no conflicts of interest.

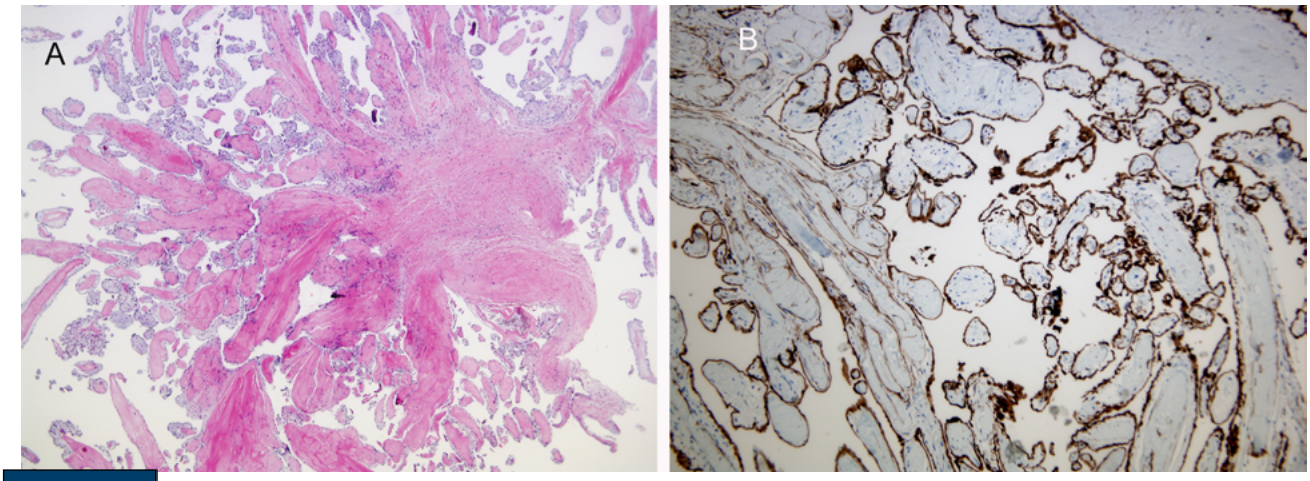

Figure 4

Microscopic images. A. Elongated and branching papillary fronds consisting of hypocellular collagenous cores lined by a single layer of endothelial cells (hematoxilyn-eosin). B. Immunohistochemistry stain for CD34 highlights the endothelial lining.

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