A Left Ventricular Papillary Fibroelastoma Presenting as an Acute Coronary Syndrome

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INTRODUCTION

Primary cardiac neoplasms are rare, with a prevalence of 0.001%–0.03% according to echocardiographic and autopsy studies.[1,2] Approximately three-quarters of these tumors are benign in nature. Aortic valve tumors are four times and left atrial tumors 1.9 times more likely to result in embolic events compared with other cardiac tumors.[3] We describe a case of a left ventricular (LV) papillary fibroelastoma (PFE) that manifested as an acute coronary syndrome.

CASE DISCUSSION

A 63-year-old woman with a medical history of obstructive sleep apnea, obesity, hypothyroidism, and pseudotumor cerebri presented to the emergency department with left-sided chest pain. The pain was described as a "heavy weight" that gradually progressed to five out of 10 in severity and occurred when the patient was going to bed. The pain worsened with deep inspiration and was not radiating in nature. The patient endorsed recurrence of chest pain in the emergency department. She denied exertional chest pain at baseline but had chronic exertional dyspnea. She had returned from Italy 1 week previously. Review of systems was negative for fever or swelling of the lower extremities. Vital signs were significant for a blood pressure of 190/88 mm Hg, with a heart rate of 72 beats/min. Physical examination demonstrated no signs of respiratory distress, with regular but distant heart sounds. The initial electrocardiogram revealed normal sinus rhythm with limb lead criteria for LV hypertrophy satisfied and no significant ST-T-wave changes.

Initial cardiac biomarkers were negative. D-dimer was positive, and computed tomography of the chest ruled out pulmonary embolism. Subsequent high-sensitivity troponin I levels were 0.1, 0.34, and 1.13 with negative creatine kinase MB levels. The patient was initiated on the standard upstream therapy for non–ST-segment elevation myocardial infarction with low–molecular weight heparin, dual-antiplatelet therapy with aspirin and ticagrelor, a β-blocker, and a statin. No subsequent electrocardiographic changes were seen. The patient had experienced transient monocular (left-sided) visual loss a few weeks previously. Bedside transthoracic echocardiography (TTE) done as part of risk stratification for acute coronary syndrome revealed normal LV function with no regional wall motion abnormality. A 1 × 1.6 cm contrast-enhancing mass was seen in the LV cavity with a stalk attached to the interventricular septum (Figures 1 and 2, Videos 1 and 2). On transesophageal echocardiography, the mass was noted to be partially contrast enhancing, with a false tendon attached to it (Figures 3 and 4, Videos 3 and 4). Cardiac catheterization revealed normal coronary arteries. Therefore, the mass visualized on echocardiography was postulated to be the source of the coronary and possibly the cerebrovascular embolic event. Cardiac magnetic resonance imaging (CMR) done to further delineate tissue characteristics failed to visualize the mass (Figure 5).

Given the clinical presentation, surgical exploration and excision of the mass was advocated. The patient underwent successful surgical resection of the LV mass with an uneventful postoperative course. Surgical pathology revealed a 1.0 × 1.0 × 0.4 cm, soft, gelatinous, ovoid, tan-colored, and focally hemorrhagic mass consistent with a PFE (Figure 6). It was attached to the lateral wall of the left ventricle by thin chordlike filaments. Light microscopy revealed multiple frondlike structures consisting of a central core of dense fibroelastic tissue surrounded by loose connective tissue (Figure 7).

DISCUSSION

PFEs are the second most common benign cardiac tumors and are located predominantly on the cardiac valves. PFEs have dense central stalks with frondlike extensions and are characterized as mobile and pedunculated masses that appear speckled when imaged by TTE.[4]

PFEs are attached to the cardiac valves in about 83% of cases, with the aortic valve implicated in fewer than half of these.[5,6] Rarely, these masses are seen attached to the endocardium or papillary muscles in the left ventricle. Despite their benign nature, embolism of cerebrovascular and coronary circulations has been observed in about 30% of cases. Sudden cardiac death is a rare presentation. Pulmonary, mesenteric, retinal, and peripheral embolism has been reported in a minority of cases.[7] There is a strong association between left-sided mobile PFEs with stalks and future embolic phenomena.[5]

The real-time approach and temporal and spatial resolution of TTE make it the initial modality of choice in the diagnosis of PFE. The use of contrast in echocardiography may help differentiate highly vascular malignant tumors from avascular masses such as thrombi or vegetations.[8] On contrast injection, malignant tumors hyperenhance, stromal tumors partially enhance, and thrombi do not enhance.[9] CMR

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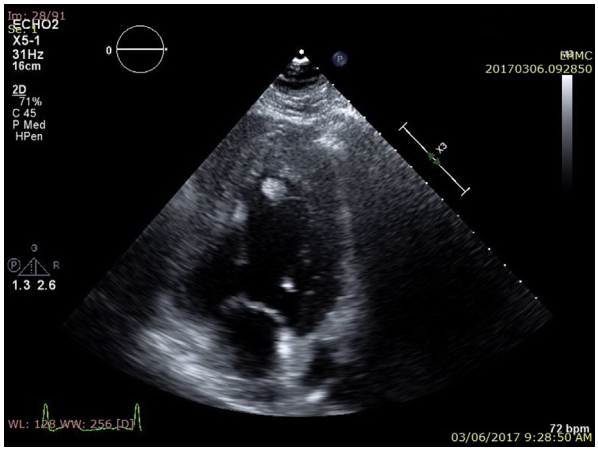

Figure 1 Transthoracic apical two-chamber view depicting a highly mobile rounded mass attached to a thin linear structure within the LV cavity.

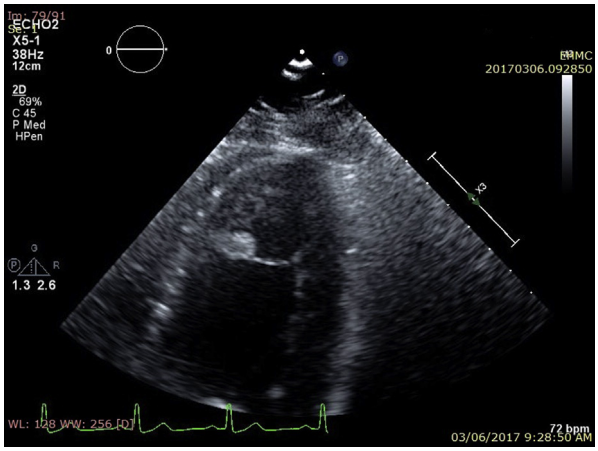

Figure 2 Transthoracic zoomed apical four-chamber view depicting a round mobile mass attached to the lateral wall of the LV via a thin chord-like filament.

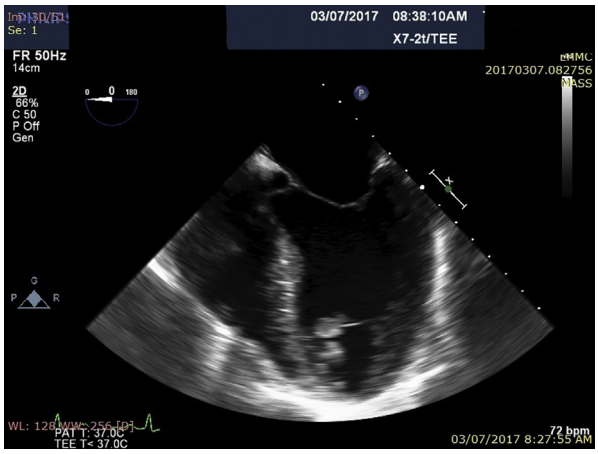

Figure 3 Midesophageal four-chamber view at 0 degrees demonstrating a round mass attached to the lateral wall of the LV.

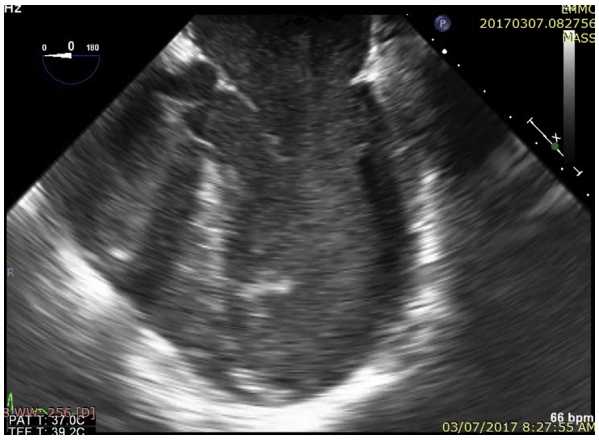

Figure 4 Midesophageal four-chamber view using intravenous contrast. The partially contrast enhancing of the mass is revealed.

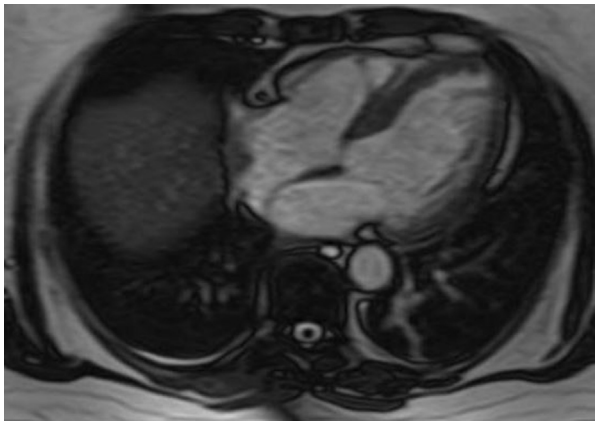

Figure 5 Cardiac MRI four-chamber view (bright blood) with no evidence of LV soft tissue mass.

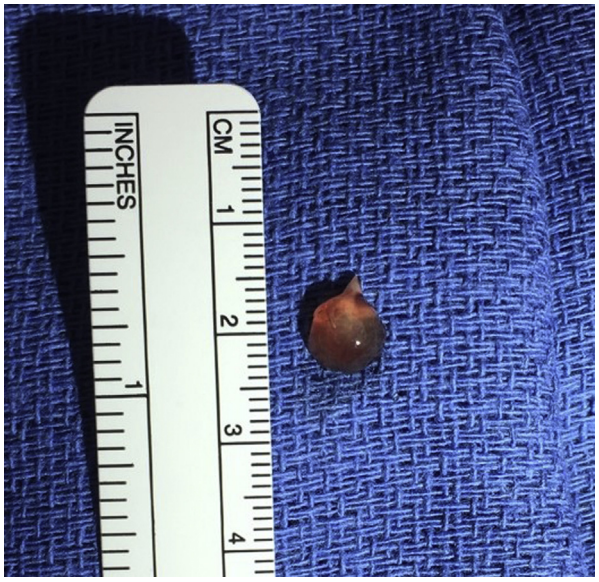

Figure 6 Gross pathology revealing a soft, gelatinous, ovoid, tan-colored focally hemorrhagic mass measuring 1.0 x 1.0 x 0.4 cm.

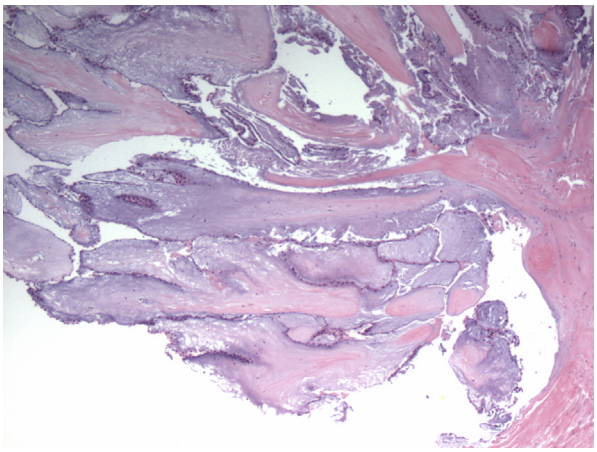

Figure 7 Light microscopy of PFE revealing multiple frondlike structures consisting of a central core of dense fibroelastic tissue surrounded by loose connective tissue.

and computed tomography of the chest may be used to differentiate tumor from thrombus, especially by evaluating for contrast enhancement.[9,10] On CMR, PFE appears as a small, highly mobile homogeneous mass that appears hypointense on cine, isointense on T1-weighted, and hyperintense on T2-weighted images with late gadolinium enhancement.[10-12] One case report revealed gradual centripetal enhancement of a PFE on serial CMR, which helped differentiate it from a thrombus.[13]

CONCLUSION

We present an atypical case of PFE located in the LV cavity that presented with two embolic events. The mass was attached to the lateral wall of the left ventricle by a false tendon. The tissue characteristics and incomplete contrast-enhancing nature of the mass on TTE and transesophageal echocardiography helped differentiate it from a thrombus. We hypothesize that the inadequate temporal resolution and absence of serial postcontrast imaging on CMR resulted in failure to visualize the mass. The consensus is that symptomatic PFEs warrant curative surgical excision, as in our case. In addition, asymptomatic, large (>1 cm), mobile, left-sided PFEs may benefit from valve-sparing surgery given the substantial risk for embolization.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at http://dx.doi.org/10.1016/j.case.2017.08.002.

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