Pulmonary Fibroelastoma: A Rare Cardiac Mass Presenting With Dyspnea

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INTRODUCTION

Papillary fibroelastomas (PFEs) are rare primary cardiac tumors that arise from one of the cardiac valves.[1] They comprise approximately 10% of all primary cardiac tumors.[2] While the etiology is unknown, they are believed to occur in areas of endothelial damage.[1] The most affected valves are the aortic (35%-63%) and mitral (9%-55%) valves, with involvement of the pulmonic valve occurring less than 8% of the time.[3] Symptoms are variable and dependent on their location but can be divided into 2 categories: symptoms related to embolization and symptoms related to obstruction. In most cases, embolization from PFEs leads to symptoms related to the downstream sites of embolization. When left-sided heart valves are affected, which they most commonly are, symptoms can present as either stroke or transient ischemic attack. In more rare cases, the symptoms can be obstructive, leading to sudden cardiac death, syncope, and heart failure.

CASE PRESENTATION

A 67-year-old woman with hyperlipidemia and gastroesophageal reflux disease presented to our medical center with complaints of dyspnea on exertion, bilateral lower extremity edema, and intermittent chest discomfort. The patient denied associated palpitations, syncope, or fever. On arrival to the emergency department, the patient was noted to be in no apparent distress. Vital signs were stable with appropriate oxygenation on room air, and cardiopulmonary examination was unremarkable. Initial evaluation in the emergency department included a computed tomography (CT) angiography of the chest, which demonstrated a large filling defect near the pulmonic valve (Figure 1).

An initial transthoracic echocardiogram (TTE) was performed that confirmed the presence of an echodensity on the pulmonic valve (Figure 2, Video 1). The patient had a normal ejection fraction, and the remainder of the cardiac valves were normal.

At this time, the differential diagnosis included infective endocarditis, thrombus, sterile vegetation, and neoplasm. The patient was admitted, and blood cultures were obtained. Empiric antibiotics and therapeutic anticoagulation were started. Transesophageal echocardiogram (TEE) confirmed the presence of a pulmonic valve mass but no stigmata of acute thrombus (Figure 3, Videos 2 and 3).

Thereafter, blood cultures remained negative. It was at this time that in conjunction with the negative microbiological workup it was suspected that the patient could have a primary cardiac tumor versus a sterile vegetation. The empiric antibiotics were discontinued, and the patient was discharged home on oral anticoagulation.

The patient was seen in office several weeks later and underwent further evaluation with outpatient cardiovascular magnetic resonance imaging (CMR). The pulmonic valve was clearly visualized and redemonstrated a mobile mass attached to the ventricular surface of the pulmonic valve (Figure 4, Videos 4 and 5). The mass was of intermediate signal intensity on T1- and T2-weighted imaging sequences (Figures 5 and 6). First-pass perfusion sequences were performed and showed absence of uptake on first-pass perfusion (Figure 7, Videos 6 and 7). Phase-sensitive inversion recovery sequences demonstrated hyperintense signal intensity on late gadolinium enhancement with a central focal hypointense signal in the center of the mass (Figure 8).

The CMR findings were consistent with a benign, poorly vascularized mass given lack of uptake on first-pass perfusion combined with

VIDEO HIGHLIGHTS

Video 1: Two-dimensional TTE of a modified parasternal short-axis view demonstrating a mobile echo density on the pulmonic valve *(arrow)*.

Video 2: Two-dimensional TEE, midesophageal right ventricular inflow-outflow view (87°) demonstrating a mobile mass measuring 1.3 × 1.3 cm attached to the pulmonic valve.

Video 3: Two-dimensional TEE, midesophageal right ventricular inflow-outflow view (87°) with color Doppler demonstrating mild pulmonary regurgitation due to the pulmonic valve mass.

Video 4: CMR cine sequence of the Figure 4A coronal view of the pulmonary artery demonstrating a 1.0 × 1.1 cm mass attached on the ventricular surface of the pulmonic valve.

Video 5: CMR cine sequence of the Figure 4B short-axis view of the pulmonic valve demonstrating a 1.0 × 1.1 cm mass attached on the ventricular surface of the pulmonic valve.

Video 6: CMR first-pass perfusion imaging sequences with injection of gadolinium. Right ventricular outflow tract demonstrating absence of uptake on first-pass perfusion.

Video 7: CMR first-pass perfusion imaging sequences with injection of gadolinium. Short-axis view of pulmonic valve demonstrating absence of uptake on first-pass perfusion.

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https://doi.org/10.1016/j.case.2022.10.003

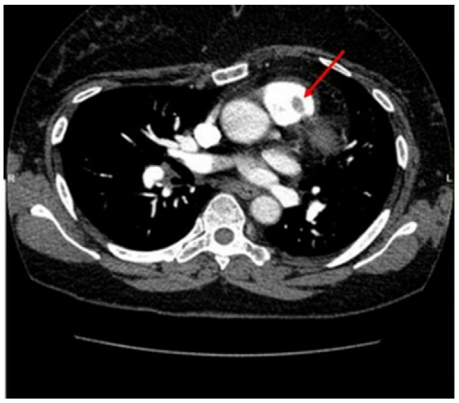

Figure 1 Computed tomography angiogram of the chest demonstrating a 1.2 × 0.7 cm filling defect at the level of the pulmonic valve *(arrow)*.

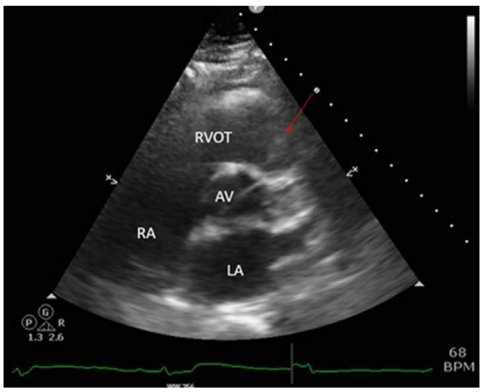

Figure 2 Two-dimensional TTE still frame of a modified parasternal short-axis view at end diastole demonstrating a mobile echo density on the pulmonic valve *(arrow)*. *AV*, Aortic valve; *LA*, left atrium; *RA*, right atrium; *RVOT*, right ventricular outflow tract.

evidence of late gadolinium enhancement on the phase-sensitive inversion recovery sequence. Moreover, the presumptive diagnosis of a PFE was supported by the uniform intermediate signal density on T1- and T2-weighted images, which is reflective of the fibroelastic composition of the mass.[4]

The patient was subsequently referred for evaluation by cardiothoracic surgery, where they successfully underwent resection of the pulmonary valve mass and valve replacement. Pathology obtained from the procedure confirmed the suspected diagnosis of PFE (Figure 9). The patient's postoperative course was uneventful. They were later seen in follow-up and reported improvement in dyspnea.

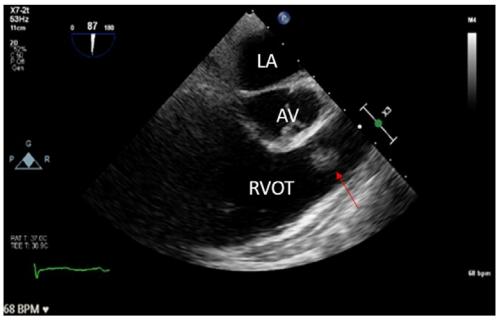

Figure 3 Two-dimensional TEE, midesophageal right ventricular inflow-outflow view (87°) at end diastole, demonstrating a mobile mass measuring 1.3 × 1.3 cm attached to the pulmonic valve *(arrow)*. *AV*, Aortic valve; *LA*, left atrium; *RVOT*, right ventricular outflow tract.

DISCUSSION

Papillary fibroelastoma is a rare primary cardiac tumor. In most instances PFEs are discovered incidentally in asymptomatic patients by direct visualization in autopsy and cardiac surgery.[3] Nevertheless, clinical sequelae and associated symptoms have also been reported, the most common of which is embolization.

Systemic embolization, which occurs more frequently, due to the aortic (prevalence 35%-63%) and mitral valves (9%-35%) being more commonly affected, is often manifested by cerebrovascular accidents.[3,5] Cerebrovascular accident risk has been reported to be increased in patients with PFE who do not undergo surgical removal, occurring in 6% and 13% of these patients at 1 and 5 years, respectively.[5] Other signs of systemic embolization include coronary artery occlusion with related sudden cardiac death and transient ischemic attack.[6] Pulmonary embolization has also been reported in the case of a pulmonary PFE presenting with dyspnea with evidence of bilateral subsegmental perfusion deficits noted on VQ scan.[7]

In our case we suspected the possibility that the presenting complaint of dyspnea could be due to one of the above sequelae, more likely pulmonary embolization, due to location on the pulmonary valve. However, a clear causal link could never be clearly established due to an indeterminate VQ scan and no evidence of flow limitation noted on TTE, TEE, or CMR. This is similar to a previously reported case in which symptoms of dyspnea improved following cardiac surgery, although no real causal link was established as to the etiology of the dyspnea.[8]

In the workup of PFE, a multimodality imaging approach may be the preferred method in establishing a diagnosis. The 3 primary cardiac imaging modalities, echocardiography (both two- and three-dimensional), CT, and CMR, all have distinct strengths and limitations that may be needed in the detection, characterization, and surgical planning needed to treat PFE.

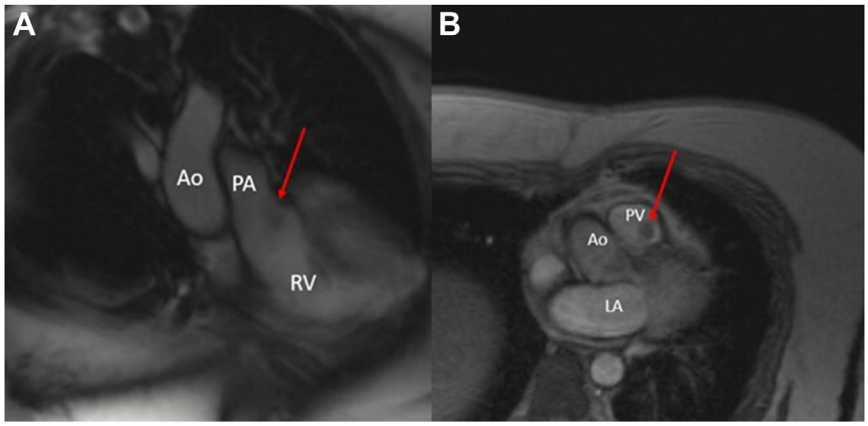

Figure 4 CMR of **(A)** coronal view of the pulmonary artery and **(B)** short-axis view of the pulmonic valve demonstrating a 1.0 × 1.1 cm mass *(arrow)* attached on the ventricular surface of the pulmonic valve. *Ao*, Aorta; *LA*, left atrium; *PA*, pulmonary artery; *PV*, pulmonic valve; *RV*, right ventricle.

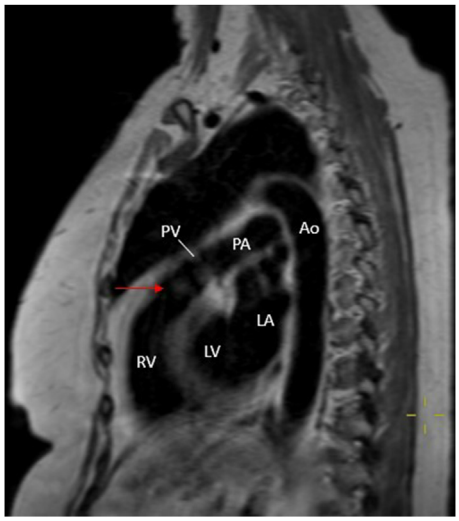

Figure 5 CMR T1-weighted images (dark blood) of the right ventricular outflow tract demonstrating intermediate signal intensity of the pulmonic valve mass *(arrow)*. *Ao*, Aorta; *LA*, left atrium; *LV*, left ventricle; *PA*, pulmonary artery; *PV*, pulmonic valve; *RV*, right ventricle.

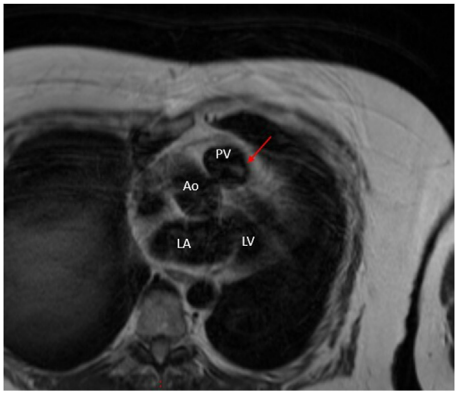

Figure 6 CMR T2-weighted images (dark blood) of the short-axis view of the pulmonic valve demonstrating intermediate signal intensity of the pulmonic valve mass *(arrow)*. *Ao*, Aorta; *LA*, left atrium; *LV*, left ventricle; *PV*, pulmonic valve.

Transthoracic echocardiography is one such modality that has been shown to be useful, but it has limitations. It has the benefit of lower cost and ease of completion, but its constraints are that it is less useful in detecting smaller PFEs and is unable to provide accurate characterization of cardiac masses once they are detected. Sun *et al.*[9] has assigned a sensitivity and specificity of 88.9% and 87.8%, respectively, in the detection of PFEs ≥0.2 cm in size. This number, however, drops significantly, with a sensitivity of 61.9% when PFEs are ≤0.2 cm in size.[9] With a reported wide variation in size from 2 to 70 mm, this limitation may be a reason to augment TTE with further imaging.[3]

Two- and three-dimensional TEE are imaging modalities that are better at detecting smaller lesions and may have a benefit in the anatomical spatial characterization of lesions that can be used in surgical planning.[10-13] Sun *et al.*[9] assigns TEE a sensitivity of 76.6% in detecting PFEs ≤0.2 cm in size.[3]

More advanced imaging including cardiac CT and CMR may be of benefit in the workup of cardiac masses as well. Their limitations are their higher cost, limited availability at various institutions, longer time to completion, and, in the case of cardiac CT, exposure to radiation. Their benefits, however, are that they may be better at characterizing tumors, distinguishing the etiology of various cardiac tumors (malignant, infectious, benign, or thrombus), providing insights into points of attachment, and providing insightful information regarding signal characteristics.

Papillary fibroelastomas can be effectively treated with cardiac surgery. Prognosis is noted to be excellent, with recurrence rates observed to be as low as 1.6%.[5] In Tamin *et al.*,[5] cerebrovascular accident risk in those with histopathologically confirmed PFE removed by cardiac surgery was noted to be 2% and 5% at 1 and 5 years, respectively. In that same study, 98% of those undergoing primary cardiac surgery for removal of PFE had preservation of their native valve.

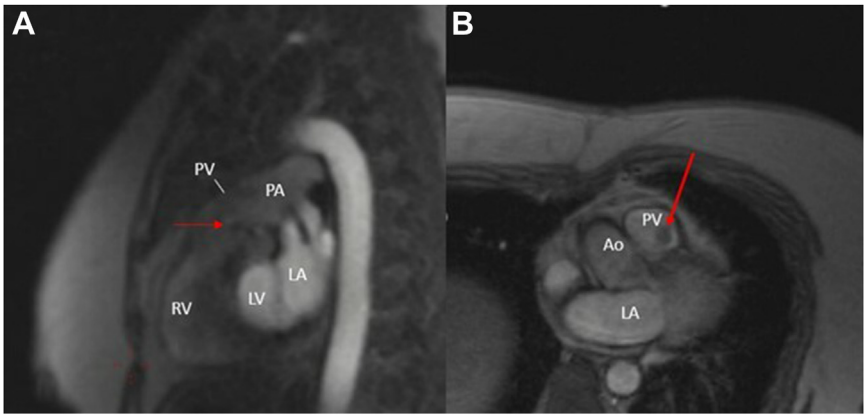

Figure 7 CMR first-pass perfusion imaging sequences with injection of gadolinium. **(A)** Right ventricular outflow tract and **(B)** short-axis view of pulmonic valve demonstrating the pulmonic valve mass *(arrow)* with absence of uptake on first-pass perfusion. *Ao*, Aorta; *LA*, left atrium; *LV*, left ventricle; *PA*, pulmonary artery; *PV*, pulmonic valve; *RV*, right ventricle.

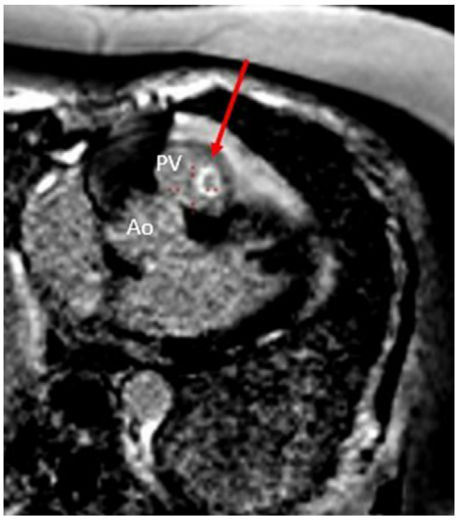

Figure 8 CMR phase-sensitive inversion recovery sequence of a short-axis view of the pulmonic valve demonstrating hyperintense signal intensity on late gadolinium enhancement with central focal hypointense signal in the center of the mass *(arrow)*. *Ao*, Aorta; *PV*, pulmonic valve.

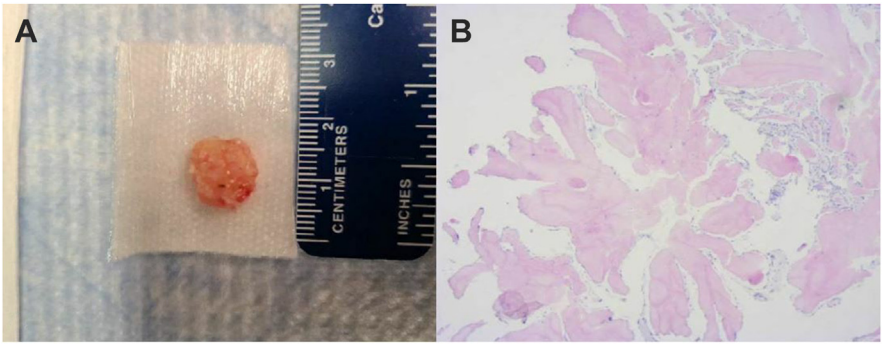

Figure 9 **(A)** Gross specimen of the resected mass attached to the pulmonic valve measuring 1.6 × 1 × 0.3 cm. **(B)** Cytology with bland features.

CONCLUSION

This case highlights the importance of early detection, characterization, diagnosis, and treatment of cardiac masses and more specifically PFE, particularly when involving a valve less commonly affected. It also establishes the benefits of a multimodality imaging approach in the diagnosis and surgical planning of PFE.

ETHICS STATEMENT

The authors declare that the work described has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans.

CONSENT STATEMENT

The authors declare that since this was a non-interventional, retrospective, observational study utilizing de-identified data, informed consent was not required from the patient under an IRB exemption status.

FUNDING STATEMENT

The authors declare that this report did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

DISCLOSURE STATEMENT

The authors report no conflict of interest.

SUPPLEMENTARY DATA

Supplementary data to this article can be found online at https://doi.org/10.1016/j.case.2022.10.003.

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