

## Multiple Cardiac Papillary Fibroelastomas



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## INTRODUCTION

Papillary fibroelastoma (PFE) is a rare benign cardiac tumor that classically arises from the valvular endocardium and is usually solitary. We present an interesting case of a patient who presented with multiple cardiac masses consistent with multiple PFEs and tissue in the right atrium that resembled ectopic spleen.

## CASE PRESENTATION

A 76-year-old woman with a history of hypertension, diabetes mellitus, and permanent pacemaker implantation for symptomatic bradycardia two years previously presented with symptoms of fatigue, pedal swelling, intermittent chest discomfort, and exertional shortness of breath. She was hemodynamically stable on admission, saturating 95% on 4 L of oxygen via nasal cannula. Electrocardiography revealed atrial fibrillation with demand ventricular pacing, which was also new for this patient. Because her symptoms were consistent with heart failure, transthoracic echocardiography was performed and showed multiple mobile echodensities on the aortic valve at the left ventricular outflow tract level (Figure 1, Video 1 and Video 2) and on the pacemaker lead (Figure 2) and a large mass in the left ventricle (Figure 3). Transesophageal echocardiography was performed the following day to better visualize these masses and again demonstrated multiple right- and left-sided intracardiac masses, with a large mass on the tricuspid valve (Figure 4, Video 3).

Because this was an unusual case of multiple cardiac masses in an elderly patient, we did a complete malignancy workup. Cardiac computed tomography and positron emission tomography both demonstrated no evidence of malignancy or metastatic disease. Blood cultures were negative. There was no definitive evidence of thrombophilia or connective tissue disease. Cardiothoracic surgery was consulted for evaluation and excision of her masses, and the patient underwent median sternotomy and open heart surgery.

A total of 10 masses were removed during surgery. Pathology was consistent with PFEs for six of the masses, and no malignancy was identified in any of the masses (Figure 5). Movat stain demonstrated elastic tissue in the papillary fronds, a characteristic feature of PFE.

In the right atrial mass, the light microscopic features were consistent with benign splenic tissue (Figure 6). However, CD8 immunoper-

oxidase stain performed on the sinusoidal lining cells was negative for CD8. Splenic tissue is typically positive for CD8 cells.

The patient safely underwent surgery without any complications. Her postoperative echocardiography showed a 1-cm lesion in the left ventricle that was seen previously. Otherwise, she was weaned off oxygen and no longer required diuretics.

## DISCUSSION

Primary cardiac tumors have an incidence of 0.0017% to 0.33%. About 75% of these tumors are benign. Historically, myxomas are the most common, and PFEs are the second most common benign tumors of the heart.<sup>1</sup> However, a Mayo Clinic study published in 2015 reported PFE to be the most common primary cardiac neoplasm.<sup>2</sup>

PFEs are typically described as resembling a sea anemone and have multiple papillary fronds. They are attached to the endocardial surface of the valves by small pedicles. The majority of these lesions are solitary, and multiple PFEs occur in only 7% to 10% of patients. In case studies by Anastacio *et al.*,<sup>3</sup> 82% of lesions were completely solitary, 8% had two distinct masses, and 8% had three distinct masses. To date, the greatest number of PFEs discovered in a single patient was 35 to 40, found in both ventricles in a case report published in 2009.<sup>4</sup> That patient had hypertrophic cardiomyopathy and a history of multiple open heart operations, both of which are risk factors for multiple PFEs. Kumar *et al.*<sup>4</sup> suggested that an aberrant reactive response could have contributed to multiple lesions close to prior surgical sites as well as intrinsic abnormalities caused by hypertrophic cardiomyopathy. Our patient was unique in that she had 9 to 10 distinct masses without a history of cardiac surgery or of anatomic cardiac disease. She did have a pacemaker placed two years previously that was inserted for symptomatic bradycardia.

PFEs generally occur on the valvular endocardium of the left side of the heart. In a retrospective case study by Anastacio *et al.*,<sup>3</sup> among 23 patients who were diagnosed with cardiac PFE between 1996 and 2012 at Washington University, tumors were found primarily on cardiac valves (aortic, 52%; mitral, 33%; pulmonary, 11%; tricuspid, 4%). Only 7% of patients had origins in the endocardial surface of the ventricular and atrial walls. There have been rare cases of PFEs originating within the aorta.<sup>5,6</sup> In our patient, one of the two largest masses was attached to the right atrial lead in the right atrium. The other large mass was in the left ventricle. Both of these locations are unusual for PFEs, as most are located on the valvular endocardium on the left side. In addition, there was no left ventricular outflow tract gradient. Current risk factors associated with the development of PFEs include age, male gender, thoracic irradiation, cardiac valve disease, congenital heart defect repair, open heart surgery, and rheumatic heart disease. Interestingly, the risk factors our patient had were her age and the presence of a pacemaker, without known cardiac disease. PFEs today are classified as neoplasms, but they have also been theorized to be hamartomas, organizing thrombi, and posttraumatic tumors.

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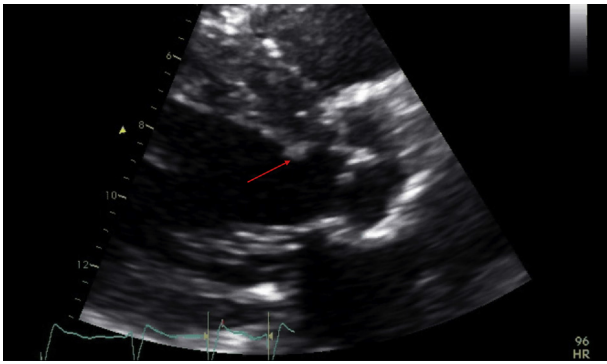
Keywords: Cardiac mass (neoplasm), Papillary fibroelastomas, Ectopic spleen

Conflicts of Interest: The authors reported no actual or potential conflicts of interest relative to this document.

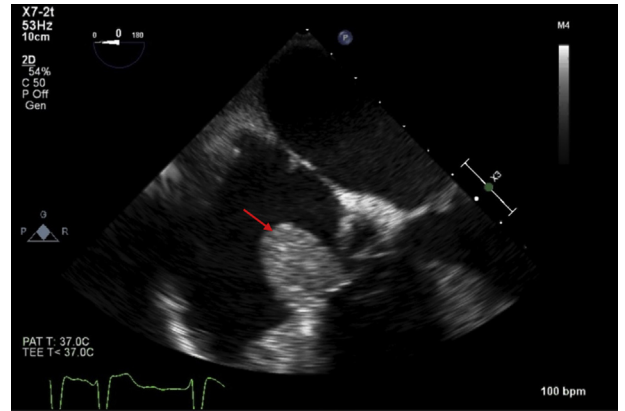
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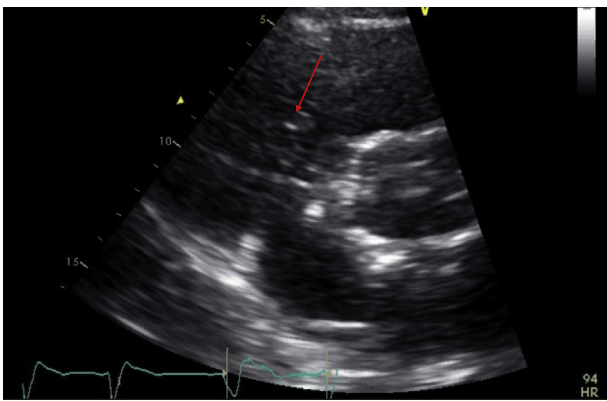
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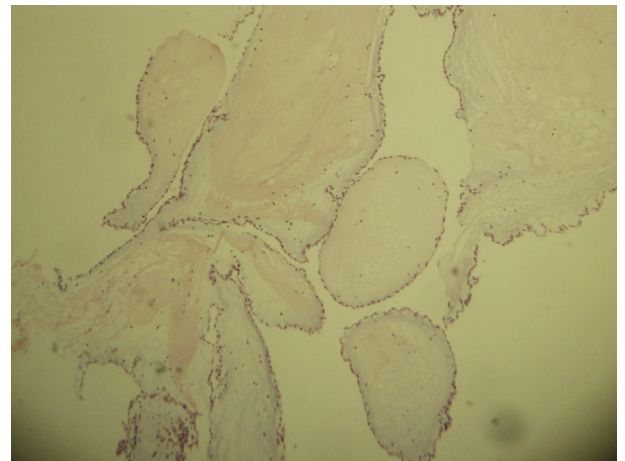
**Figure 1** One of the left ventricular outflow tract masses indicated by the arrow by transthoracic echocardiography.



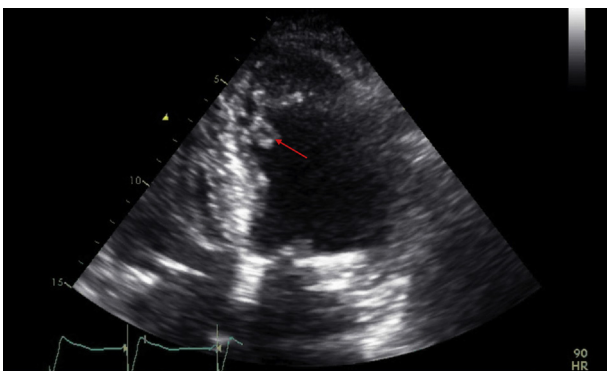
**Figure 4** The arrow points to a large tricuspid valve mass on transesophageal echocardiography.



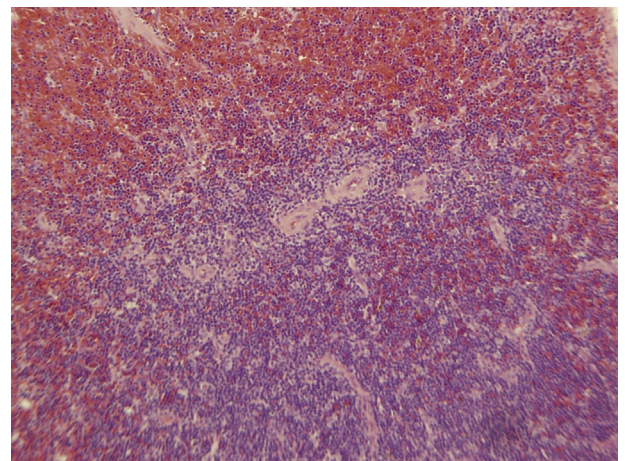
**Figure 2** The arrow points to pacemaker lead masses by transthoracic echocardiography, parasternal short-axis view. Compared with Figure 4, the mass looks smaller, highlighting the advantages of multimodality imaging.



**Figure 5** Magnification of the left ventricular mass showing PFE (Hematoxylin and Eosin stain).



**Figure 3** Left ventricular mass, as indicated by the arrow, seen on four-chamber transthoracic echocardiography.



**Figure 6** Magnification of right atrial mass resembling normal splenic tissue (Hematoxylin and Eosin stain). CD8 stain, which would be positive in splenic tissue, was negative.

PFEs are usually asymptomatic and are incidentally identified. In the case studies by Anastacio *et al.*,<sup>3</sup> 13 patients (57%) were symptomatic, with the most common presentation being a thromboembolic event (33% stroke and 4% pulmonary embolism). About 43% presented with cardiac symptoms, which included angina, unstable angina, or myocardial infarction. Our patient presented with signs of heart failure.

The mass in the right atrium was different from the other masses in that it resembled benign splenic tissue on light microscopy. There

have been few case reports describing ectopic splenic tissue in the heart. Kuijer *et al.*<sup>7</sup> published a case report of right atrial and ventricle masses that represented splenic tissue. When the cardiac tumor was compared with splenic tissue during necropsy, they reported no

differences in terms of histological markers. Our case is unique in that we stained for CD8, and the results were negative. Typically, splenic tissue is CD8 positive.

### CONCLUSION

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Multiple PFEs can also be found on the right side of the heart, not only on the endocardium. Multiple echocardiographic imaging modalities may be required to discover all masses.

PFEs can resemble splenic tissue and still test negative for neoplasm with CD8 staining. PFEs can be successfully resected with surgical intervention to eliminate the risk for embolization.

### SUPPLEMENTARY DATA

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Supplementary data related to this article can be found at <http://dx.doi.org/10.1016/j.case.2017.03.004>.

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