A Large Cardiac Papillary Fibroelastoma Arising from the Coumadin Ridge: Unusual Location and Presentation

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INTRODUCTION

We report the case of a 59-year-old woman presenting with ST-segment elevation myocardial infarction, resulting in cardiogenic shock. Because of advanced cardiomyopathy, she subsequently underwent left ventricular assist device implantation. During her diagnostic workup, a large, mobile, left atrial (LA) mass was diagnosed by transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE), and pathologic examination of the excised mass confirmed the diagnosis of a cardiac papillary fibroelastoma (PFE). This case is unique in that the tumor mass was very large and arose from an unusual location: the coumadin ridge. It is also an illustrative example of the echocardiographic assessment of cardiac PFE.

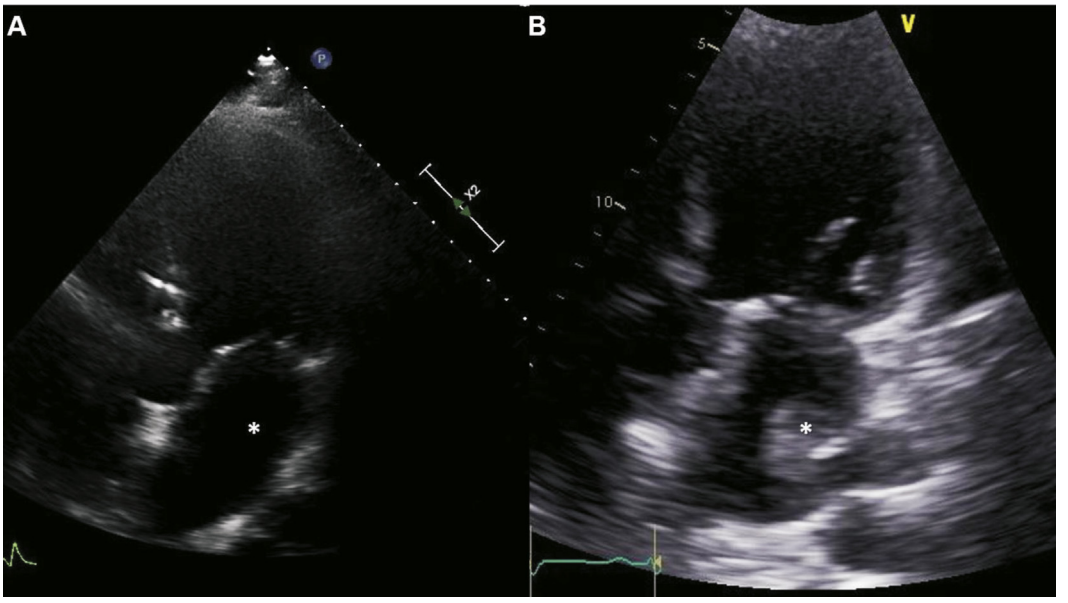

Figure 1 Serial TTE (3 months apart). **(A)** Apical five-chamber view demonstrates the absence of a cardiac mass within the left atrium (*asterisk*). **(B)** Three months later, a similar apical five-chamber view demonstrates the presence of a moderate-sized cardiac mass within the LA chamber (*asterisk*).

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CASE PRESENTATION

A 59-year-old woman presented with left-sided chest pain and anterolateral ST-segment elevation on electrocardiography. Her medical history was notable for type 2 diabetes mellitus and hypertension. Coronary angiography demonstrated thrombotic occlusion of the proximal left anterior descending and proximal left circumflex coronary arteries. The angiographic appearances were consistent with an acute thromboembolic phenomenon. The patient was managed with thrombus aspiration and balloon angioplasty. A detailed workup for a hypercoagulable state (including lupus anticoagulant, β_2-glycoprotein, anticardiolipin antibody, lipoprotein[a], and serum homocysteine level) was unremarkable. The hospital course was complicated by heparin-induced thrombocytopenia

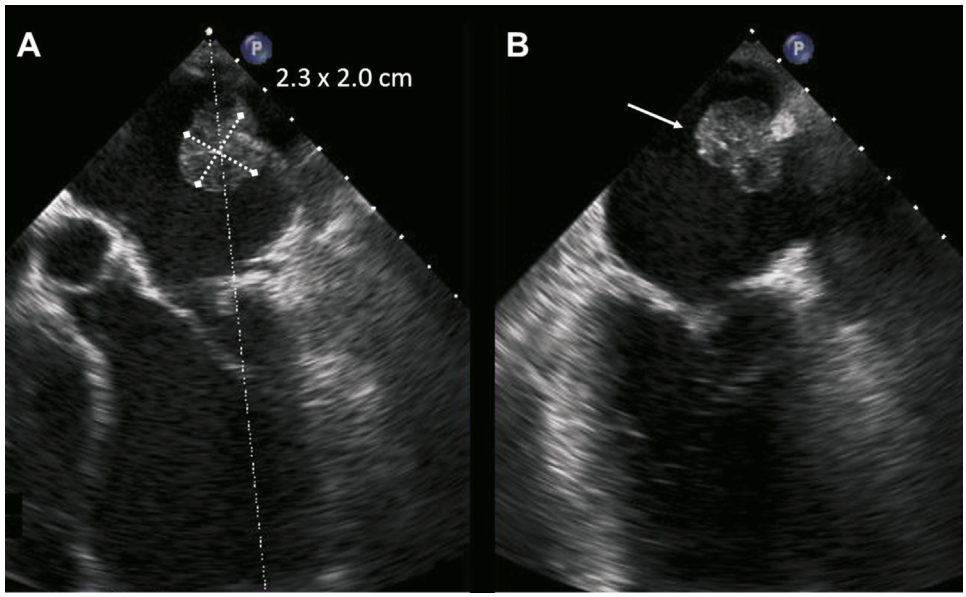

Figure 2 TEE. **(A)** Midesophageal four-chamber view at 0° with biplane orthogonal 90° view **(B)** focusing on a 2.3 × 2.0–cm, globular, heterogeneous, pedunculated mass (*arrow*), which appears attached to the limbus of the LA appendage, also known as the coumadin ridge.

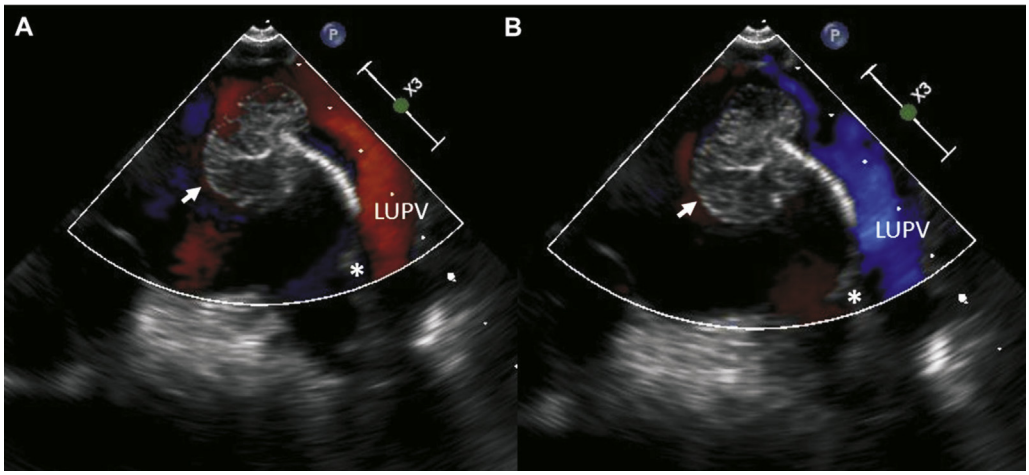

Figure 3 TEE. **(A,B)** Midesophageal view at 0° showing the PFE (*arrow*) within the left atrium attached to the tip of the limbus of the LA appendage, also known as the coumadin ridge, and demonstrating the spatial relationship between the PFE (*arrow*), the limbus (*asterisk*), and the left upper pulmonary vein (LUPV).

(positive platelet factor 4 and serotonin release assays), and the patient was discharged on standard medical therapies, in addition to anticoagulation with warfarin for presumed thromboembolic ST-segment elevation myocardial infarction. Of note, the patient did not have a history of deep vein thrombosis, and there was no family history of thrombophilia. The patient never smoked or used oral contraception. As a result of the myocardial infarction, there was significant myocardial dysfunction, with a severely depressed left ventricular ejection fraction (20%), decreased from a previously normal baseline.

Three months following hospital discharge from the ST-segment elevation myocardial infarction presentation, the patient re-presented with worsening dyspnea on exertion, fatigue, and lower extremity edema. On clinical assessment, she was hypotensive. A positron emission tomographic viability study showed a large region of viable hibernating myocardium in the left anterior descending coronary artery territory, without scar. Repeat coronary angiography interestingly demonstrated absence of significant obstructive coronary artery disease in all three epicardial vessels. TTE confirmed persistently depressed left ventricular systolic function (left ventricular ejection fraction 18%), with akinesis of the apex and extension of severe hypokinesis into the midanterior, midseptal, and midinferior walls. Notably, a new, large mobile echogenic mass was visualized, which appeared to be attached to the lateral wall of the left atrium, likely emanating from the LA appendage (Figure 1B, Video 1). This echogenic mass was not seen when TTE was performed 3 months earlier (Figure 1A, Video 2).

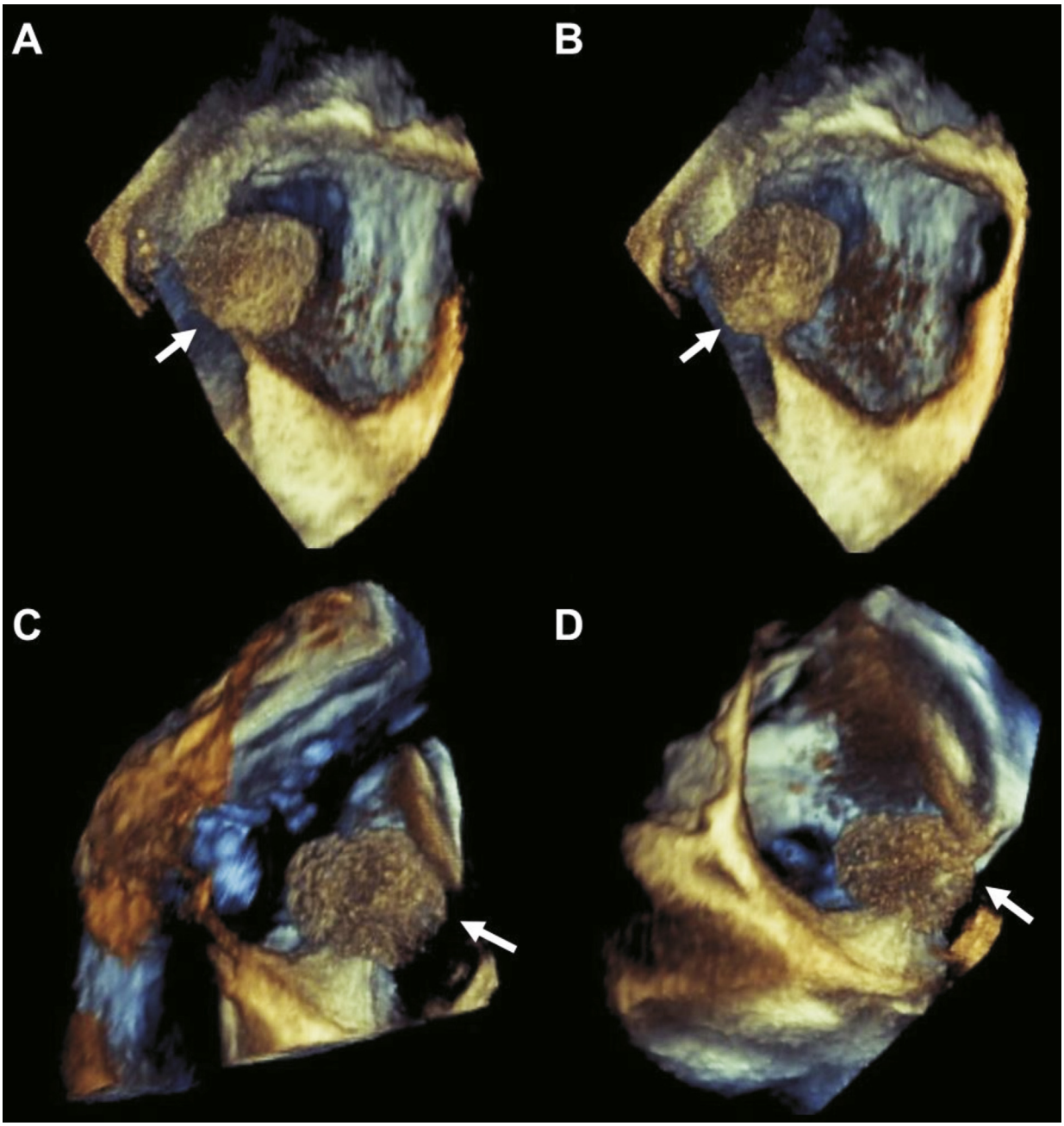

Figure 4 Three-dimensional TEE demonstrating the location of the PFE (*arrow*) as it arises from the coumadin ridge **(A,B)**, with the aortic valve at the top of the image (not visualized) and the LA appendage to the left of the coumadin ridge and the PFE. Note in a more distant plane the anterior leaflet of the mitral valve in **(C)** and the mitral annulus in **(D)**.

The echocardiographic appearance and location of the mass were suspicious for a LA appendage thrombus.

Further characterization of the mass by TEE occurred (Figure 2, Video 3). This demonstrated a large heterogeneous mass (measuring $\geq$2.3 $\times$ 2.0 cm) attached to the coumadin ridge (Figure 3, Videos 4 and 5). On detailed assessment by three-dimensional TEE, tiny fronds were visualized on the surface of this echogenic mass (Figure 4, Videos 6 and 7). The mass did not arise from the LA appendage, and no LA thrombus was visualized. On the basis of these imaging findings, the differential diagnoses considered for the mass included a PFE, an atypical atrial myxoma, or an atypical thrombus, not arising from the LA appendage.

The patient underwent surgical excision of the mass and subsequently underwent implantation of a HeartMate II left ventricular assist device (Thoratec, Pleasanton, CA), given her advanced cardiomyopathy and heart failure (Figure 5, Video 8). Pathologic examination of the mass confirmed a PFE (Figure 6), consistent with findings on TEE. The patient was discharged on standard heart failure therapies to a skilled nursing facility for ongoing rehabilitation. She remains clinically stable.

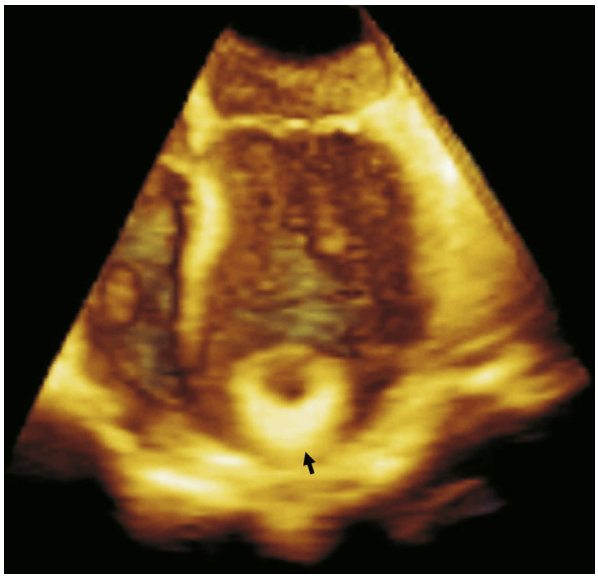

Figure 5 Three-dimensional TEE demonstrating the inflow cannula position (*arrow*) after placement of a HeartMate II left ventricular assist device.

DISCUSSION

Primary cardiac tumors are uncommon, and most are benign in nature.[1] Cardiac myxomas were traditionally thought to be the most common type of primary cardiac tumor, on the basis of surgical series. However, a recent large, single-center series suggested that cardiac PFEs may be more common than cardiac myxomas, with a rate of one PFE per 1,100 echocardiograms from the reporting center's referral base.[2] The differential diagnosis of a PFE is wide and includes thrombus, myxoma, fibroma, cardiac metastases, vegetation, and degenerative or calcific change.

PFEs are benign primary cardiac tumors consisting of small multiple papillary protrusions attached to the endocardium by a short stalk with a "sea anemone" appearance.[3] Although often discovered incidentally, the clinical presentation of PFEs varies widely, and they can present with severe embolic or ischemic events, such as a myocardial infarction.[4] The probability of a PFE resulting in an embolic event likely varies, according to the mobility, friability, size, and location of the tumor. In addition to cerebral and cardiac emboli, PFEs have been reported to cause mesenteric, renal, and limb embolism.[5,6] As a result, surgical resection is generally recommended, especially for those with a high risk for embolization, usually when the size of the tumor is >1 cm and associated with a stalk.[7] Surgical excision of PFEs is generally associated with an excellent long-term prognosis.[8]

Approximately 84% of PFEs arise from the valvular endocardium, favoring the aortic valve in most cases.[9] However, involvement of the pulmonary valve, and even simultaneous involvement of both aortic and pulmonary valves, has been reported.[10] Nonvalvular PFEs represent 15% of all cases reported[8] and can arise from the endocardium of both the left and right ventricles (septum and papillary muscles), atria (septum, appendages, Eustachian valve, and Chiari network), and the intimal surface of coronary artery ostia. Most nonvalvular PFEs originate in the left ventricle, with an incidence of approximately 9%, favoring the interventricular septum.[11] The presence of PFEs in the left atrium is very rare, and its occurrence has been reported to be approximately 1.6%–2%.[8,12] LA PFEs have been previously found to be attached to the free wall,[8,13] interatrial septum, and LA appendage,[8] but to the best of our knowledge, they have not been previously reported to arise from the limbus of the LA appendage, also known as the coumadin ridge. Our case is unique in terms of the unusual location of the PFE.

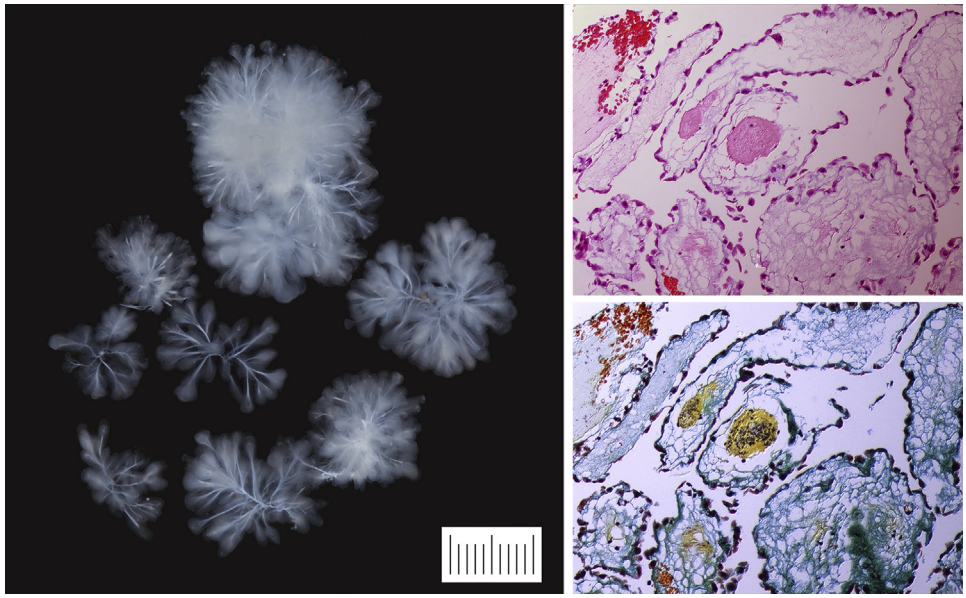

Figure 6 Gross and microphotograph of the excised mass. The gross specimen (*left*) was received fragmented, with the largest piece measuring 3.2 cm, showing pieces of branching papillary tumor with bulbous gelatinous fronds. On light microscopy (*top right*, Hematoxylin and Eosin; *bottom right*, Movat pentachrome, 400× magnification), the fronds showed a central core of fibrous tissue (*yellow* in the Movat stain). Each frond consisted of a mucopolysaccharide-rich stroma (*green*) lined by endothelial cells. The fibrous cores of some of the papillae contained elastic fibers (*black*), as demonstrated in the Movat stain.

Multimodality Cardiovascular Imaging

Before the widespread application of multimodality imaging, PFEs were often incidentally detected during autopsy or cardiac surgery.[3] Multimodality cardiovascular imaging, including echocardiography (TTE, TEE, and three-dimensional echocardiography), cardiac magnetic resonance (CMR), and multidetector computed tomography, has an important role in the diagnosis of PFEs. On echocardiography, PFEs appear as mobile, pedunculated masses with a heterogeneous, speckled appearance. Classically, tiny fronds may be visualized at the surface of the tumors. Three-dimensional echocardiography may provide incremental value in the characterization of PFEs. On multidetector computed tomography, PFEs are generally well-delineated lesions with soft tissue attenuation. CMR can further aid in the tissue characterization of cardiac masses and tumors. On CMR, PFEs are generally homogeneous masses, with an intermediate signal on T1-weighted sequences and a high signal on T2-weighted sequences, which exhibit a lack of suppression with fat saturation and strong delayed enhancement.[14] The differential diagnoses of PFEs can also be effectively assessed by multimodality cardiovascular imaging, with echocardiography being the mainstay initial imaging tool. On echocardiography, cardiac myxomas typically are visualized as mobile, pedunculated masses arising from the endocardial surface. Classically, cardiac myxomas arise from the region of the fossa ovalis. On CMR, atrial myxomas have a heterogeneous appearance, with intermediate signal intensity on T1-weighted sequences and higher signal intensity on T2-weighted sequences.[14] An intracardiac thrombus typically arises from the LA appendage itself, rather than the coumadin ridge, and contrast study may aid in characterizing an intracardiac thrombus by demonstrating lack of contrast uptake. This finding may be further confirmed by delayed enhancement imaging on CMR. An intracardiac thrombus appears as a homogeneously low–signal intensity mass on long–inversion time delayed-enhancement imaging, because of lack of contrast uptake, surrounded by high-signal blood pool.[15] CMR is powerful in its ability to provide detailed tissue characterization of intracardiac masses and can help differentiate PFEs from other intracardiac tumors. Lipomas generally occur as sessile homogeneous masses in the atrium (interatrial septum or atrial wall), which are hyperintense on T1-weighted turbo spin-echo sequences and hypointense on fat-suppressed sequences, as opposed to PFEs, which do not show any signal dropout on fat-suppressed sequences.[14] Fibromas are generally solitary, well-defined intramyocardial masses, which have a smooth morphology, associated with calcification and a broad base. They are inhomogeneous and isointense or slightly hyperintense on T1-weighted sequences and hypointense on T2-weighted sequences, demonstrate no change with fat saturation, and are associated with intense delayed enhancement.[14] Malignant cardiac tumors, including cardiac metastases, may have evidence of tissue invasion morphologically and, for cardiac metastases, may often be associated with a pericardial effusion.[14] Different types of malignant cardiac tumors exhibit varying CMR imaging characteristics, and a detailed discussion is beyond the scope of this article.

CONCLUSION

We describe the comprehensive echocardiographic assessment of a large PFE attached to the coumadin ridge in a 59-year-old woman. TTE provided the initial diagnosis of the large mass in the left atrium, while detailed TEE demonstrated features more suggestive of a PFE than a thrombus. The findings on TEE were confirmed by pathologic examination of the excised mass. This case is unique in that the PFE reported was very large in size and arose from an unusual location, the coumadin ridge. Additionally, this case serves as an illustrative example of the imaging diagnostic pathways involved in the assessment of an intracardiac mass.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at http://dx.doi.org/10.1016/j.case.2017.05.006.

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