

A diagnostic dilemma upon discovery of a left atrial mass using an intracardiac echocardiogram



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Introduction

It is important to discern the nature of any mass that may appear on the left atrial (LA) ridge, or “coumadin ridge,” which is situated between the LA appendage and the left superior pulmonary vein prior to any procedures involving the left atrium. Despite the rarity of the appearance of LA ridge mass, the shape and location are important in distinguishing a cardiac tumor or thrombus.¹ In this case report, a mass on the LA ridge was discovered during an intracardiac echocardiogram prior to transseptal puncture for ablation of atrial fibrillation, posing a diagnostic dilemma and leading to a cascade of imaging studies.

Case report

A 59-year-old man was admitted for ablation for his symptomatic and rapid atrial fibrillation with near syncope. The patient had been complaining of episodic lightheadedness and near syncope after being on beta blockers and rivaroxaban for 6 months after diagnosis with atrial fibrillation. He had an event monitor 2 months prior to the procedure that revealed paroxysmal atrial fibrillation with multiple conversion pauses up to 4 seconds, which correlated with near syncope, leading to his decision to move forward with atrial fibrillation ablation. A transthoracic echocardiogram showed cardiomyopathy with a left ventricular ejection fraction of 25% without any appreciable masses or other abnormalities.

Since the patient had been on rivaroxaban for 6 months without interruption prior to ablation, there was no transesophageal echocardiogram (TEE) performed preprocedurally. During the procedure, after gaining femoral access using ultrasound guidance, an intracardiac echocardiography catheter (ACUSON AcuNav V ultrasound catheter and SC2000

imaging platform; Siemens Healthcare, Mountain View, CA) was advanced to the right atrium to create an ultrasound-guided map of the left atrium. Incidentally, a mobile mass attached to the LA ridge was visualized (Figure 1). A TEE was then performed, revealing an oval and pedunculated mass with fronds measuring 6 × 8 × 11 mm with a 4 mm pedicle adhering to the LA ridge edge (Figure 2). The mass was mobile and protruding into the appendage during atrial systole (Supplemental Videos 1–3). There was no mass or thrombus in the appendage itself. The atrial fibrillation ablation was aborted to avoid the risk of embolization with manipulation of catheters.

At this point, the differential diagnoses for the LA mass included vegetation, thrombus, or a tumor. There was no history of fever, chills, or recent infection. Blood and urine cultures were negative. Therefore, infectious etiology was more unlikely, as criteria for endocarditis were not met. There was no personal history or symptoms of malignancy, with routine screening examinations being negative to date. Given the history of atrial fibrillation, a presumptive diagnosis of thrombus was made, and the patient was maintained on uninterrupted oral anticoagulation followed up with interval imaging. The anticoagulation regimen was switched to warfarin with a goal international normalized ratio of 2.5–3 with weekly checks.

On repeat TEE after 6 weeks with uninterrupted anticoagulation, the mass was unaltered in size and shape (Supplemental Video 4). The patient then underwent a cardiovascular magnetic resonance to better characterize the mass. The patient was in atrial fibrillation with a rapid ventricular response during the cardiovascular magnetic resonance, not allowing for optimal gating the study with inability to distinguish in features of the LA mass being elicited except gadolinium uptake. This characteristic made a thrombus significantly less likely, with the mass being a likely papillary fibroelastoma or a myxoma.

KEYWORDS Atrial fibrillation ablation; Conversion pauses; Intracardiac echocardiogram; Left atrial mass; Papillary fibroelastoma (Heart Rhythm Case Reports 2021;7:507–509)

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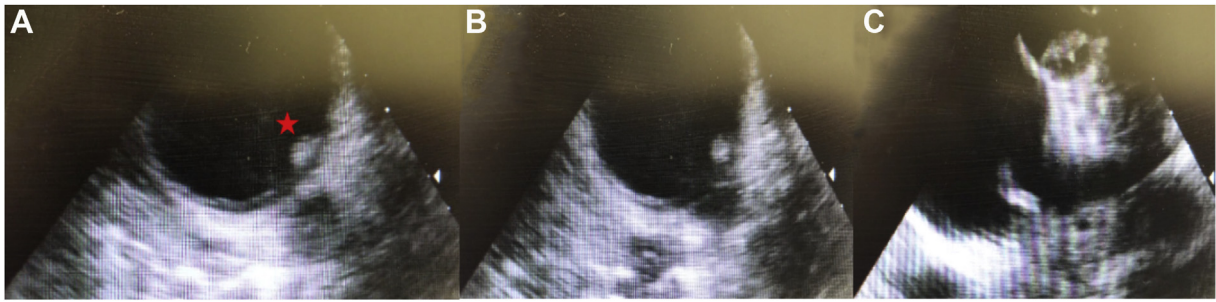


Figure 1 A–C: Intracardiac echocardiography showing a mobile mass attached to the left atrial ridge (*). Images A and B were obtained with intracardiac echocardiogram in the right atrium and C showed the mass on the ridge from the right ventricular outflow tract.

Since there was an inherent risk of potential embolization associated with the persistent mobile pedunculated myxoma or fibroelastoma, surgical removal of the mass became necessary, as the patient had recurrent admissions for persistent and symptomatic atrial fibrillation with exacerbation of his systolic heart failure. The patient underwent removal of the LA mass, modified maze, and LA appendage excision. The excised mass had the consistency of white soft mucinous material with a scant amount of lobular adipose tissue. Histopathology findings were significant for acellular papillary fronds consistent with papillary fibroelastoma (Figure 3).

The patient had a recurrence of persistent atrial fibrillation and developed atrial flutter. He continued to have multiple conversion pauses that worsened, leading to syncope and a fall leading to intracranial hemorrhage after his surgery. After recovery of his subdural hematoma and reinitiation of his anticoagulation, he underwent atrial fibrillation and cavotricuspid isthmus ablation. He has had no further arrhythmias in over 12 months, with recovery to his ejection fraction.

Discussion

The current case report highlights the importance of intracardiac echocardiogram use during ablation of LA structures to not only aid in safety with anatomic visualization and assist with transeptal puncture, but also assist with detection of unusual masses found in unexpected positions in the left atrium. The fibroelastoma in this case arose from the LA ridge, or “coumadin ridge,” an embryological remnant in the left atrium between the left atrium

appendage and the left superior pulmonary vein that can be mistaken for a primary cardiac tumor or thrombus itself.¹ Posterior LA structures cannot be visualized well on transthoracic echocardiogram, and with adequate anticoagulation, intracardiac echocardiogram is often the subsequent modality of anatomic delineation during ablation in the left atrium. Though the LA ridge can mimic a thrombus or primary cardiac tumor, the presence of a mass in an unusual position should initiate further investigation.² The fixed location, the lack of mobility, and the unique linear structure usually help distinguish the LA ridge from other masses or contiguous structures.³

LA masses have a varied differential including tumors, vegetation, or thrombus. Cardiac tumors are rare, with an incidence of 0.05%–1%. Most of the cardiac tumors are primary in nature, with the most common ones being myxoma (45%), lipoma (20%), and papillary fibroelastoma (15%).⁴ Past medical history, location, size, shape, symptoms, and imaging modalities are important to help guide in the differentiation between various LA masses.⁵

Transthoracic echocardiogram helps define the location, size, shape, attachment, and mobility of masses, but posterior LA structures are better visualized with the use of TEE. Contrast echocardiographic imaging helps recognize perfusion patterns between malignancies (hyper-enhancing pattern), benign stromal tumors (hypo-enhancing pattern), and thrombus (nonenhancing pattern). Three-dimensional TEE can further elucidate shape, site of attachment, and surface characteristics.⁶ Fibroelastomas commonly originate from cardiac valves on transthoracic echocardiogram and

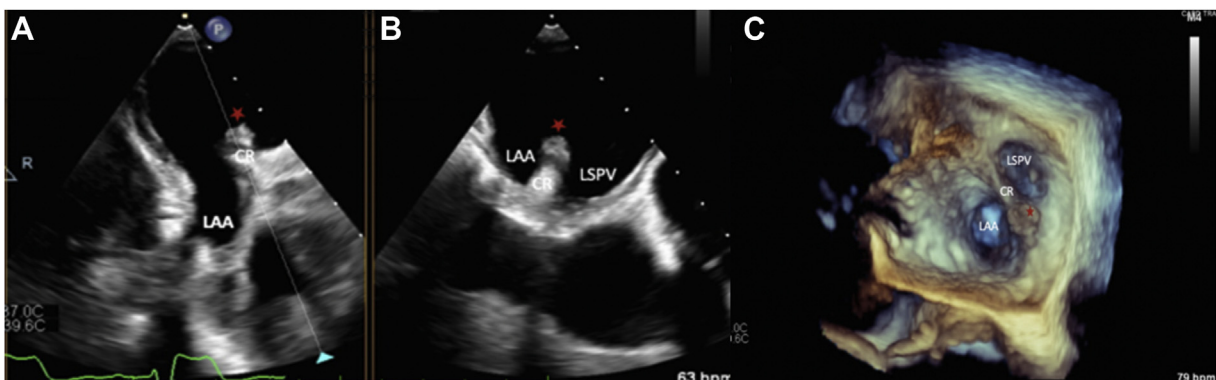


Figure 2 Transesophageal echocardiogram (TEE) images. **A:** TEE showing a mass arising from the left atrial ridge (*). **B:** TEE of the left atrium in orthogonal views showing the left atrial mass arising from the left atrial ridge (*). **C:** A 3D TEE of the left atrium showing the mass arising from the left atrial ridge (*). CR = left atrial or “coumadin” ridge; LAA = left atrial appendage; LSPV = left superior pulmonary vein.

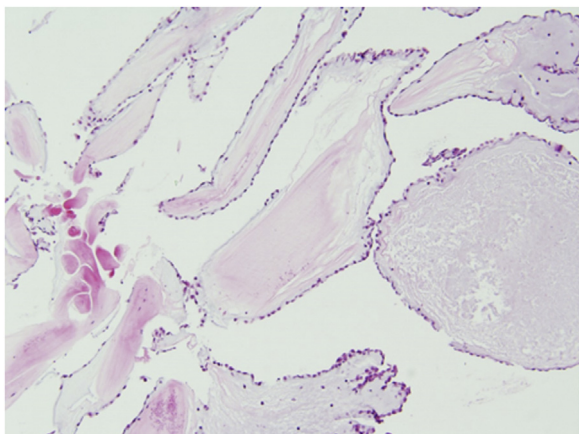


Figure 3 Histopathology showing acellular papillary fronds consistent with papillary fibroelastoma.

can present with thromboembolic events. Paroxysmal atrial fibrillation is a rare presentation reported when fibroelastoma arises from the LA ridge.⁷

Cardiac magnetic resonance imaging with gadolinium contrast with a focus on early vs late enhancement can provide additional insight into cardiac mass. LA ridge, being a normal cardiac tissue, should have the same signal intensity as the surrounding myocardium on both T1- and T2-weighted imaging and should not show late enhancement with the use of gadolinium contrast. A thrombus is usually a low-signal-intensity mass surrounded by high-intensity structures on late gadolinium enhancement images. With the use of delayed gadolinium imaging with a long inversion time of 600 ms, avascular tissue such as thrombus looks homogeneously dark as compared with the myocardium, which shows intermediate signal intensity.⁸ Computed tomography has a limited role in the distinction between intracardiac masses owing to the lack of dynamic imaging of cardiac structures. Atrial arrhythmias might be seen with fibroelastoma, although there are no specific electrocardiogram abnormalities associated with fibroelastoma.⁷

Fibroelastoma shows an increased signal intensity on late gadolinium imaging.⁹ A limitation the cardiac magnetic resonance highlighted is the presence of rapid rates that can cause poor cardiac gating. Ultimately, histopathology confirmed the diagnosis of fibroelastoma with the presence of alveolar papillary collagen fronds with elastin fibers.¹⁰ Fibroelastomas usually arise from endocardial surfaces attached to the ventricular surface of semilunar valves and atrial structures of atrioventricular valves. They have a frondlike appearance with filiform projections resembling sea anemone. Since fibroelastomas can cause embolization to cerebral and coronary arteries, surgical resection should be considered for larger masses.^{5,11} Rare complications such as pulmonary embolism, congestive heart failure, near syncope, ventricular arrhythmias, and sudden death have also been reported.¹¹ Treatment of acute embolic events is controversial, since it is unknown whether the embolic component of papillary fibroelastoma is a portion of the tumor itself, or thrombus or thrombotic material adherent to the fronds. Generally, anticoagulation and simple surgical excision with valve-sparing

approach are used if there is a thromboembolic incident.¹² The preferred treatment with a good prognosis is simple surgical excision for fibroelastoma.¹³ Since there can be difficulty in distinguishing between fibroelastoma and thrombus on echocardiography, cardiac magnetic resonance can help with the distinction, which can save patients from risks of lifelong anticoagulation and possible surgical excision to prevent thromboembolic complications.¹⁴

Conclusion

LA masses discovered on intracardiac echocardiogram prior to ablation can be challenging to diagnose and treat. This case highlights how an LA mass in proximity to the LA ridge can easily be mistaken for a thrombus. We described a rare case of a papillary fibroelastoma incidentally discovered at this site that needed excision after persistent appearance on various imaging modalities despite anticoagulation changes over a protracted period. Contemporary multimodality noninvasive imaging tools are of utmost importance in the work-up of patients with LA masses. Further delineation and potential treatment of the intracardiac mass is necessary to prevent thromboembolic events prior to ablation in the left atrium.

Appendix Supplementary data

Supplementary data associated with this article can be found in the online version at <https://doi.org/10.1016/j.hrcr.2021.06.004>.

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