

A Case of a Left Atrial Mass in an Orthotopic Heart Transplant Recipient



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INTRODUCTION

Identification of intracardiac masses on noninvasive imaging is important clinically because it determines and guides medical treatment for the structure. Examples of intracardiac masses include neoplasms, vegetations, thrombus, and foreign material. The most common primary cardiac neoplasm is cardiac myxoma, representing 20% to 40% of intracardiac masses.^{1,2} Patients with cardiac myxomas may present with breathlessness, syncope, weight loss, fever, embolization, and even sudden cardiac death.³ The most common location for myxoma is the left atrium, attached to the atrial septum at the fossa ovalis; however, they may be found in all four chambers of the heart. Cardiac myxomas have smooth surfaces, are irregularly shaped, are typically nonhomogenous, and are connected by a stalk.^{1,4} Atrial myxomas can produce obstruction to flow if they occlude the mitral or tricuspid inflow tract.³ A characteristic “tumor plop” may be auscultated after the second heart sound on physical examination if these tumors obstruct inflow tracts.³ Given the characteristic appearance and location of the tumor, echocardiography is typically diagnostic for myxoma.¹

Another important cardiac mass is intracardiac thrombus. Thrombus is typically associated with intracardiac devices, the left atrial appendage in atrial fibrillation, and areas of poor contractility such as the left ventricular after a myocardial infarction and less commonly involves the atrial septum. Left atrial thrombus is most common in patients with mitral stenosis, atrial fibrillation, or amyloidosis.² In rare situations, a thrombus may organize and attach to the atrial septum and have a similar appearance as other cardiac masses. Intracardiac thrombus on echocardiography typically has a popcorn- or snake-like appearance with shaggy edges.² When it involves the atrial septum, it usually is associated with a patent foramen ovale or atrial septum aneurysm with redundant tissue.² Embolization of thrombus may cause devastating strokes and intestinal or limb ischemia and require several months of oral anticoagulation to prevent this occurrence.^{5,6} Intracardiac thrombus is usually silent on cardiac auscultation and is typically present only on physical examination if distal embolization has occurred. The ability to differentiate myxoma from thrombus by echocardiography enables decisions about treatment, as most myxomas are removed surgically to prevent embolization, and most thrombus is treated with oral anticoagulation.^{3,4,7,8}

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VIDEO HIGHLIGHTS

Video 1: Two-dimensional echocardiographic video clip, four-chamber view, of large left atrial mass adherent to the interatrial septum. The mass appears to have smooth borders and an irregular shape and appears echogenically heterogenous in composition.

Video 2: Two-dimensional echocardiographic video clip, zoomed four-chamber view, of large left atrial mass adherent to the interatrial septum. The mass appears to have smooth borders and an irregular shape and appears echogenically heterogenous in composition.

Video 3: Two-dimensional echocardiographic video clip, three-chamber view, of large left atrial mass adherent to the interatrial septum. The mass appears to have smooth borders and an irregular shape and appears echogenically heterogenous in composition.

Video 4: Two-dimensional echocardiographic video clip, subcostal four-chamber view, of large left atrial mass adherent to the interatrial septum. The mass appears to have smooth borders and an irregular shape and appears echogenically heterogenous in composition.

Video 5: Two-dimensional echocardiographic video clip showing a four-chamber view with contrast enhancement of large left atrial mass. On contrast study, the mass appears homogeneously absent of contrast uptake.

Video 6: Transesophageal echocardiographic video clip of a large homogenous well-circumscribed left atrial mass: mid-esophageal biatrial view of left atrial mass.

Video 7: Transesophageal echocardiographic video clip of a large homogenous well-circumscribed left atrial mass: mid-esophageal biatrial X-plane view of left atrial mass appears attached to atrial septum.

Video 8: Transesophageal echocardiographic video clip of a large homogenous well-circumscribed left atrial mass: three-dimensional midesophageal echocardiographic view showing large left atrial mass with thin stalk projecting from atrial septum.

Video 9: Two-dimensional echocardiographic video clip, basal short-axis view, of ill-defined left atrial mass after 3 months on oral anticoagulation with apixaban 5 mg twice daily.

Video 10: Two-dimensional echocardiographic video clip, subcostal view, of ill-defined left atrial mass after 3 months on oral anticoagulation with apixaban 5 mg twice daily.

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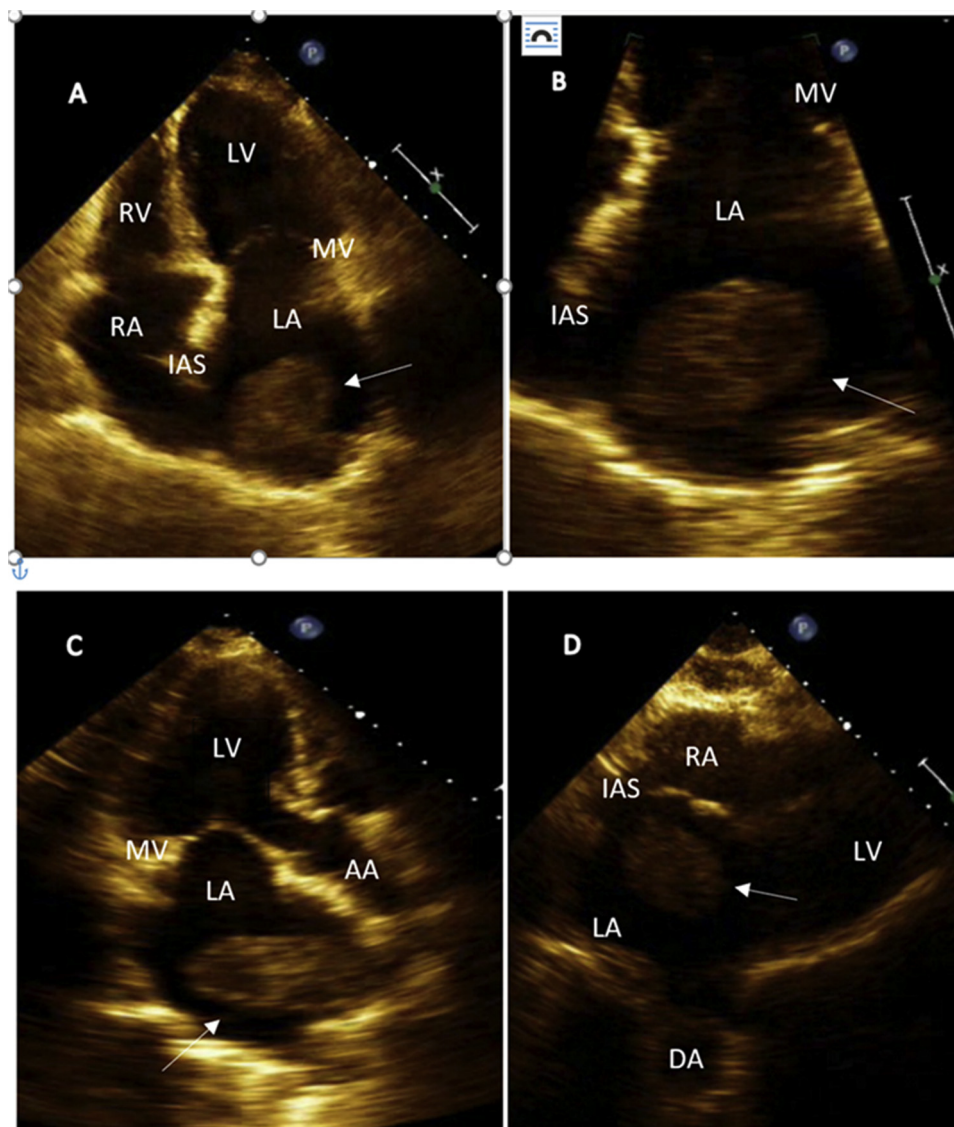


Figure 1 Two-dimensional echocardiographic images including (A) four-chamber view, (B) zoomed four-chamber view of left atrium, (C) three-chamber view, and (D) subcostal four-chamber view of large left atrial mass adherent to the interatrial septum (white arrow). The mass appears to have smooth borders and an irregular shape and appears echogenically heterogenous in composition. AA, Ascending aorta; DA, descending aorta; IAS, intra-atrial septum; LA, left atrium; LV, left ventricle; MV, mitral valve; RA, right atrium; RV, right ventricle.

CASE PRESENTATION

A 73-year-old woman who underwent orthotopic heart transplantation in 1999 for nonischemic cardiomyopathy presented 19 years after transplantation for symptoms of lightheadedness. She had been maintained on a corticosteroid-free regimen of mycophenolate and cyclosporine and had never had an infection related to immunosuppression. In addition to heart transplantation, she had a history of gastroesophageal reflux, hypertension, hyperlipidemia, osteoporosis, and remote cerebrovascular accident with no residual motor or sensory deficits. She underwent routine surveillance coronary angiography in 2016 that showed no significant allograft coronary artery disease. Routine echocardiography performed October 2018 showed a normal ejection fraction of 65% with enlarged left and right atria, consistent with previous transplantation, and a large left atrial mass,

new since March 2018 (Figure 1, Videos 1-4). Color Doppler imaging did not show flow within the mass, and contrast administration showed absent homogenous uptake (Figure 2, Video 5).

After echocardiography was performed, the patient was seen in the clinic for urgent follow-up and at that time had stable vital signs, no neurologic changes, and no remarkable cardiac auscultatory findings, and electrocardiography showed normal sinus rhythm with lateral nonspecific ST-wave flattening. The only symptom reported was lightheadedness. Because of the location and appearance of the mass, initial concern was that the mass likely represented a myxoma rather than a thrombus. To further evaluate the mass, transesophageal echocardiography was performed in October 2018 with visualization of a $5.2 \times 5.1 \times 3.1$ cm mass in the left atrium near the anterosuperior portion of the interatrial septum with a stalk projecting from the fossa ovalis (Figure 3, Videos 6-8).

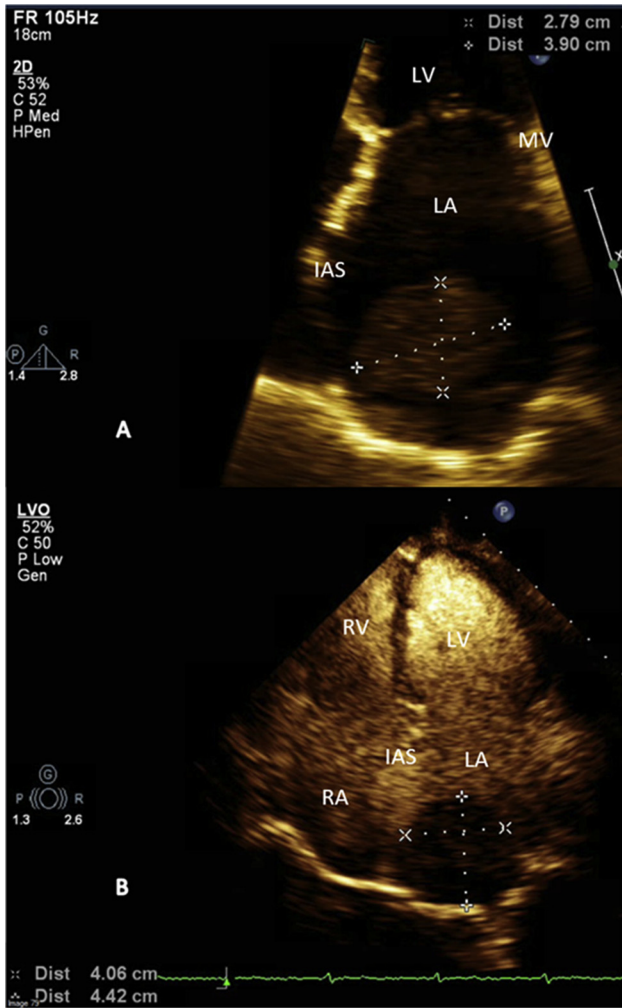


Figure 2 Two-dimensional echocardiographic images of large left atrial mass (dashed white lines). **(A)** Zoomed four-chamber view of left atrium with large left atrial mass extending out from the interatrial septum measuring 2.79 × 3.90 cm. **(B)** Contrast-enhanced four-chamber view of large left atrial mass measuring 4.06 × 4.42 cm. On contrast study, the mass appears homogeneously absent of contrast uptake. IAS, Intraatrial; LA, left atrium; LV, left ventricle; MV, mitral valve; RA, right atrium; RV, right ventricle.

The patient was referred to a cardiothoracic surgeon, and at the time of this consultation observation of the mass was recommended because of the rapid development of the mass and concern that it might be a thrombus. The patient was initiated on apixaban 5 mg twice daily in October 2018 for anticoagulation, and because of claustrophobia, cardiac computed tomographic angiography was ordered rather than cardiac magnetic resonance imaging. Angiography demonstrated a large homogenous mass in the left atrium measuring 3.9 × 5.2 cm (Figures 4 and 5). The mass appeared to arise from the interatrial septum at the fossa ovalis and was thought to more likely represent a left atrial myxoma. No other coronary or cardiac abnormality was appreciated on the study.

Results of noninvasive imaging were sent to a second cardiothoracic surgeon in November 2018. The consensus among the providers was that the mass likely represented a myxoma rather than a thrombus; however, given such rapid development, a thrombus could

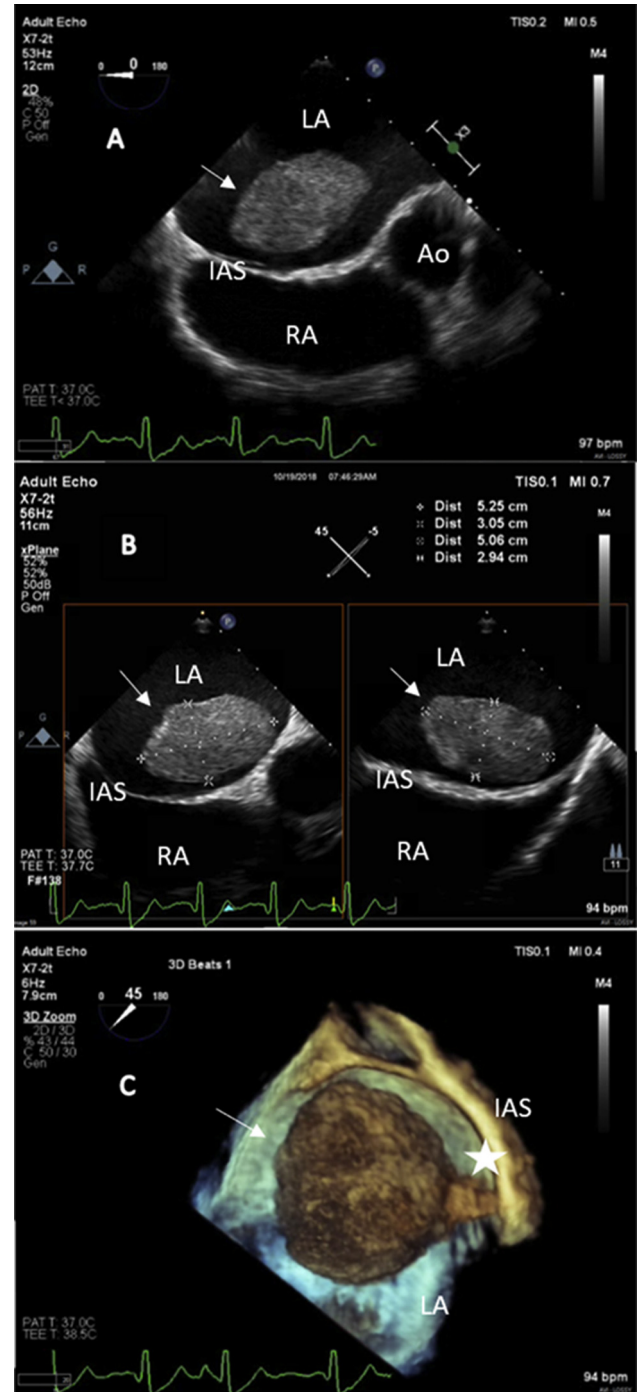


Figure 3 Transesophageal echocardiographic images of a large homogenous well-circumscribed left atrial mass (white arrow). **(A)** Midesophageal biatrial view of left atrial mass. **(B)** Midesophageal biatrial X-plane view of left atrial mass appears attached to atrial septum measuring 5.25 × 3.05 cm at the largest point. **(C)** Three-dimensional midesophageal echocardiographic view showing large left atrial mass with thin stalk (white star) projecting from atrial septum. Ao, Aorta; IAS, intra-atrial septum; LA, left atrium; RA, right atrium.

not be safely excluded. The patient was recommended to continue oral anticoagulation until surgical excision of the mass could be performed. Perioperative echocardiography was ordered in January

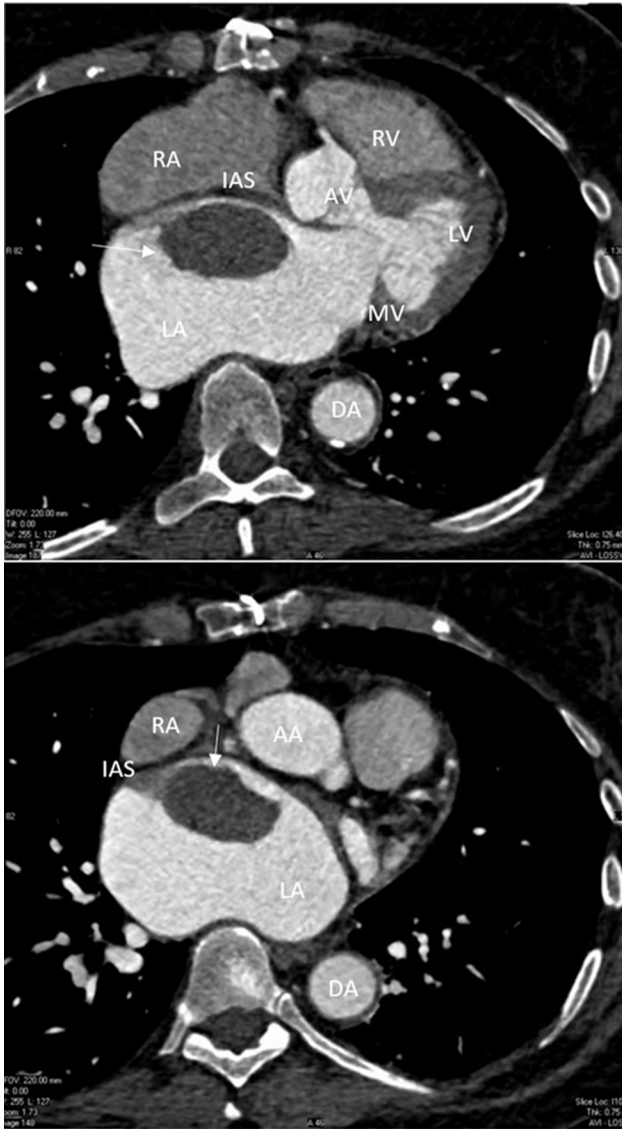


Figure 4 Cardiac computed tomographic angiographic contrast-enhanced images of large homogenous left atrial mass (*white arrow*) that appears adherent to the atrial septum by stalk. AA, Ascending aorta; AV, aortic valve; DA, descending aorta; IAS, intra-atrial septum; LA, left atrium; LV, left ventricle; MV, mitral valve; RA, right atrium; RV, right ventricle.

2019 that showed the left atrial mass, less well defined than on previous echocardiography; contrast was not used for this study (Figure 6, Videos 9 and 10). With plans for surgical excision of the mass, the patient requested a second opinion from a different cardiologist, who also recommended to proceed with surgical excision of the mass given suspicion that it was a myxoma.

The patient underwent redo sternotomy with excision of the left atrial mass in March 2019, with no acute perioperative complications. The specimen was sent for pathologic assessment, which showed a $3.6 \times 3.4 \times 2.2$ cm mass of red-brown tissue that contained red clot material determined to be a thrombus. The patient had an unremarkable recovery from surgery and was discharged from the hospital to a skilled nursing facility for acute postoperative recovery. She was followed up by cardiology and cardiothoracic surgery in April 2019

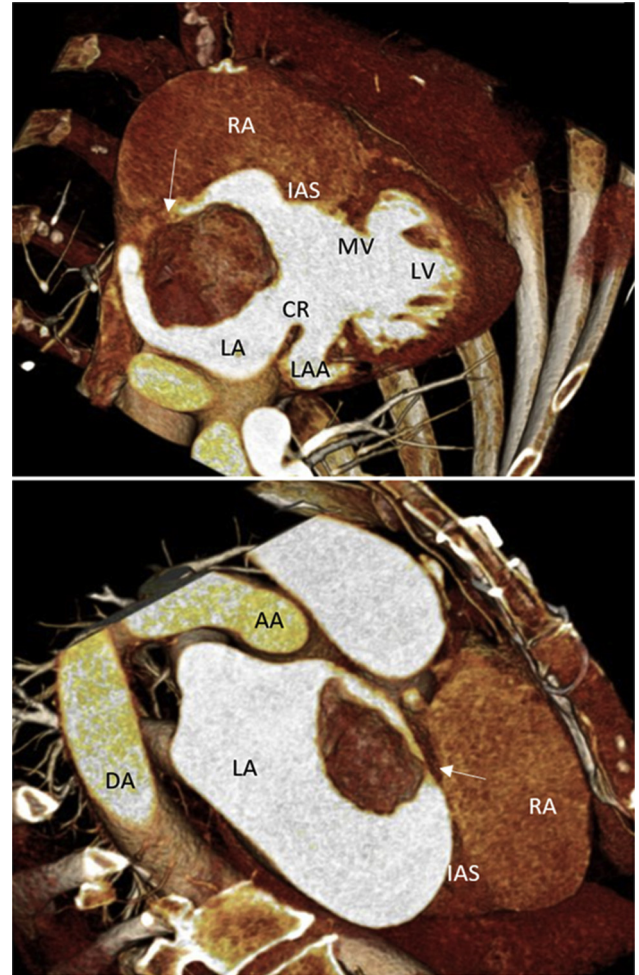


Figure 5 Cardiac computed tomographic angiographic three-dimensional rendering showing a large homogenous left atrial mass (*white arrow*) that appears adherent to the atrial septum by stalk. AA, Ascending aorta; CR, Coumadin ridge; DA, descending aorta; IAS, intra-atrial septum; LA, left atrium; LAA, left atrial appendage; LV, left ventricle; MV, mitral valve; RA, right atrium.

and was doing well postoperatively, with no major symptoms. The patient had been maintained on apixaban 5 mg twice daily for 6 months at the time of her postoperative follow-up in April 2019, and it was recommended that she continue lifelong anticoagulation with apixaban 5 mg twice daily because of the finding of a spontaneous left atrial thrombus. Repeat echocardiography was completed after her follow-up with cardiology in May 2019, which confirmed absence of the left atrial mass after surgical excision.

DISCUSSION

This case demonstrates a unique finding of a left atrial thrombus with features highly suggestive of an atrial myxoma. This left atrial mass had echocardiographic features very similar to a myxoma, including a well-organized structure, a smooth surface, and a stalk that appeared to arise from the fossa ovalis on several different imaging modalities.⁸ With such a classical appearance and location of a mass, myxoma was the agreed-upon diagnosis among several different cardiologists and

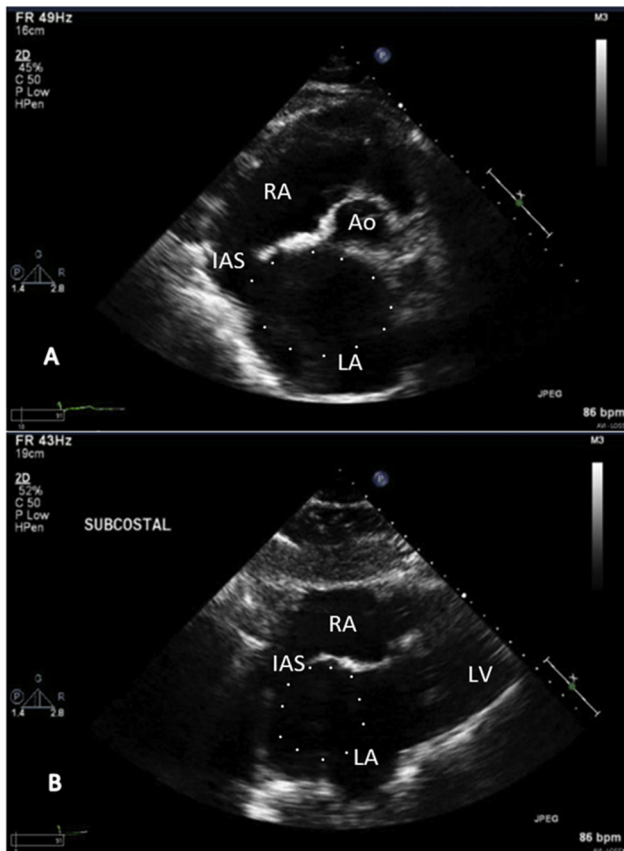


Figure 6 Two-dimensional echocardiographic images (A) basal short-axis and (B) subcostal view of ill-defined left atrial mass (outlined in white dots) after 3 months on oral anticoagulation with apixaban 5 mg twice daily. Ao, Aorta; IAS, intra-atrial septum; LA, left atrium; LV, left ventricle; RA, right atrium.

cardiothoracic surgeons. Typical treatment for myxomas includes surgical excision to prevent embolization and inflow obstructions, which can result in syncope and death.³ Even with thorough excision of the tumor, myxomas may reoccur, and annual follow-up is required for several years.¹ A thrombus attached to a previous suture site or patent foramen ovale patch repair has been described previously.⁹ In our patient's case, however, the heart transplantation had taken place 19 years before this thrombus formation, making exposed suture site attachment unlikely, and no patent foramen ovale or atrial septal aneurysm was appreciated on noninvasive imaging.

Some features of this case may have favored the diagnosis of thrombus over myxoma on noninvasive imaging. First, there was rapid appearance of a large mass in 7 months. Rapid growth is more commonly associated with thrombus but has been seen in myxomas; growth rates can range from 1.3 to 6.9 mm/mo.^{10,11} This mass was initially imaged at approximately 5 cm. At the highest growth rate of 7 mm/mo and over a 7-month interval between echocardiographic examinations, it is plausible that this mass could have represented a rapidly growing myxoma. Second, the mass was poorly visualized on repeat transthoracic echocardiography performed 2 to 3 months after oral anticoagulation; however, this study did not involve the use of contrast, which would have made for a better comparison. Third, the initial contrast transthoracic echocardiographic study showed the mass to be homogeneously dark and absent of contrast uptake. Because myxomas are partly vascularized, they take up contrast

on microbubble contrast echocardiography.⁴ This gives myxomas a mixed bright and dark heterogenous appearance on contrast echocardiography, as opposed to the dark absent homogenous appearance more common with thrombus.⁴

There have been several case reports of isolated thrombus formation with attachment sites at the atrial septum. Several of these patients were in atrial fibrillation at the time of diagnosis,^{12,13} but our patient did not have a documented history of this diagnosis. One case report comparable with ours involved a bicaval transplant recipient approximately 1 year after surgery. Surgical excision was required to identify a left atrial mass thought to be a myxoma, which was in fact identified as a thrombus after pathologic examination.⁷ In this case, no other identifiable cause for thrombus formation was found. Several imaging modalities, including three-dimensional transesophageal echocardiography, have increased sensitivity to detect atrial myxomas and aid in diagnosis.⁸ Cardiac computed tomography and magnetic resonance imaging are other imaging modalities to further characterize anatomic features of cardiac masses.^{14,15} Both three-dimensional transesophageal echocardiography and cardiac computed tomography were used in our patient's care and provided very useful anatomic information, but even in a case with compelling imaging and high clinical suspicion, the correct diagnosis may be difficult to make with such a typical presentation, highlighting the importance of this case.

CONCLUSION

Despite the sensitivity of the different imaging modalities used in this case, there was still difficulty in confirming the diagnosis before surgery. This case demonstrates some of the limitations of noninvasive imaging. Hopefully, with time these advanced imaging modalities will improve not only in their identification of cardiac masses but also in their specificity for differentiating cardiac masses.

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SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2019.10.011>.

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