

CARDIOVASCULAR IMAGES

Atrial Septal Defect Mimicking Right Atrial Myxoma

Multimodality Imaging of Cardiac Mass

Primary cardiac tumors are extremely rare: in a series of over 12 000 autopsies, only 7 were identified, leading to an estimated incidence of <0.1%.¹ Myxomas are the most common primary cardiac neoplasm, followed by fibroelastomas and lipomas.² As illustrated in this case report, there may be conditions that mimic the presence of a cardiac tumor.

CASE PRESENTATION

A 54-year-old woman was referred to the Internal Medicine Department for analysis of chronic abdominal pain. Apart from ovarian resection because of cysts 20 years earlier, her medical history was unremarkable. Workup included a computed tomographic scan of the abdomen that did not reveal an explanation for the complaints, but accidentally, a hypodense mass was found in the right atrium. She was sent to the cardiologist for further analysis.

Dedicated cardiac imaging techniques were used to better visualize the cardiac tumor. Transthoracic echocardiographic image quality turned out to be suboptimal, and only a relatively poorly visible, not-very-mobile mass of 20 by 24 mm in the right atrium adjacent to the interatrial septum could be identified (Figure 1; Movie I in the [Data Supplement](#)). The size and function of the right atrium and ventricle were normal. More detailed transesophageal echocardiography was performed and identified a roundish tumor with both echogenic and echo lucent areas, again adjacent to the interatrial septum (Figure 2; Movie II in the [Data Supplement](#)). There was no clear shunt visible through the interatrial septum although there seemed to be a patent foramen ovale. After these investigations, the patient was discussed in the heart team and accepted for surgical excision of the mass that was considered to be a myxoma. It was suggested, however, to perform a cardiac magnetic resonance imaging study as well to gather additional information on the mass. Cine imaging showed a round mobile structure in the right atrium with a low signal intensity attached to the interatrial septum (Figure 3; Movie III in the [Data Supplement](#)). The combination of findings from T1- and T2-weighted perfusion and delayed enhancement imaging revealed characteristics of a thrombus and not of a myxoma.

Surgery was performed and confirmed a fenestrated atrial septal defect (ASD) filled with thrombus. After removal of the mass, the ASD was closed primarily. No rest defect could be identified afterward. Microscopy revealed organized thrombus with chronic inflammation and irregular fragments of collagenous connective tissue with endothelium on the surface, with transitions to extensive areas of fibrin deposition (Figure 4). Our patient recovered well from surgery.

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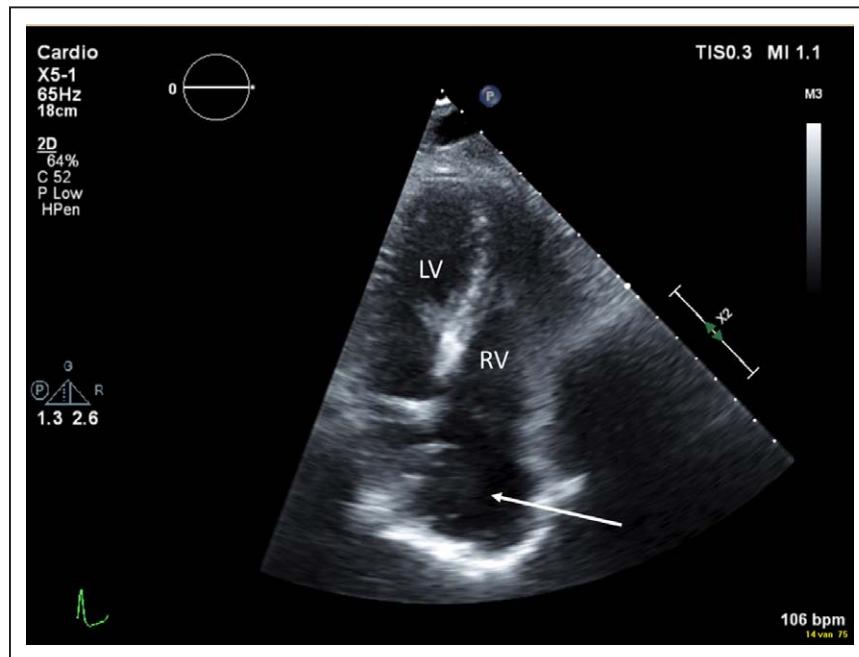
DISCUSSION

An ASD is the most common congenital heart disease at adult age.³ Patients with an ASD are often asymptomatic until adulthood, and, therefore, an ASD may be found during routine cardiac screening. In case of symptoms, dyspnea on exertion and palpitations are most often identified. The presentation of an ASD may vary, however, and fewer patients present with paradoxical emboli or angina complaints because of right ventricular ischemia. Stroke caused by so-called thrombus in transit through a patent foramen ovale has also been described.

A cardiac tumor mimicking a myxoma is a rare initial manifestation of an ASD. Our patient had no cardiac symptoms or signs of right heart overload, so the defect was not hemodynamically important. There was a fenestrated ASD with multiple perforations spread irregularly over the septum. According to the classification of such defects, this ASD was a type 4 perforated atrial septum, for which surgical closure or conservative treatment with anticoagulants should be considered.⁴ Because of the accumulated thrombus in the fenestrated septum, protruding into the right atrium, the ASD mimicked a myxoma. By using cardiac magnetic resonance tissue characterization, this disguise could be uncovered before cardiac surgery.

ARTICLE INFORMATION

The Data Supplement is available at <https://www.ahajournals.org/doi/suppl/10.1161/CIRCIMAGING.119.010004>.



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Disclosures

None.

REFERENCES

1. Lam KY, Dickens P, Chan AC. Tumors of the heart. A 20-year experience with a review of 12,485 consecutive autopsies. *Arch Pathol Lab Med*. 1993;117:1027-1031.
2. Cresti A, Chiavarelli M, Glauber M, Tanganeli P, Scalese M, Cesareo F, Guerrini F, Capati E, Focardi M, Severi S. Incidence rate of primary cardiac tumors: a 14-year population study. *J Cardiovasc Med (Hagerstown)*. 2016;17:37-43. doi: 10.2459/JCM.0000000000000059
3. Yared K, Baggish AL, Solis J, Durst R, Passeri JJ, Palacios IF, Picard MH. Echocardiographic assessment of percutaneous patent foramen ovale and atrial septal defect closure complications. *Circ Cardiovasc Imaging*. 2009;2:141-149. doi: 10.1161/CIRCIMAGING.108.832436
4. Ewerth P, Bergera F, Vogela M, Dähnerta I, Alexi-Meskishvili V, Langea PE. Morphology of perforated atrial septal aneurysm suitable for closure by transcatheter device placement. *Heart*. 2000;84:327-3231.

Figure 1. Transthoracic echocardiography-derived image focusing on the mass in the right atrium (arrow).

LV indicates left ventricle; and RV, right ventricle.

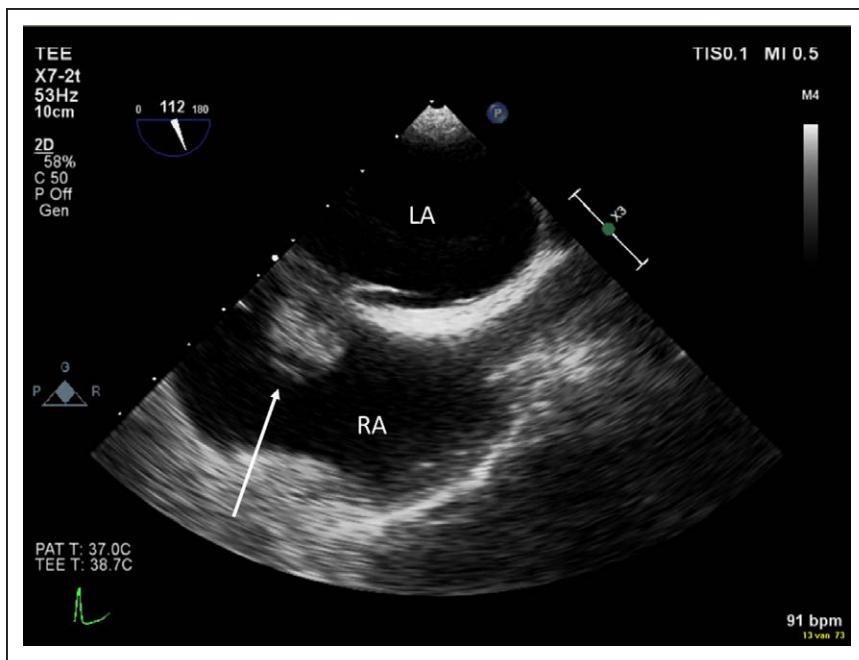


Figure 2. Transesophageal echocardiography (TEE)-derived image focusing on the mass in the right atrium (arrow). LA indicates left atrium; and RA, right atrium.

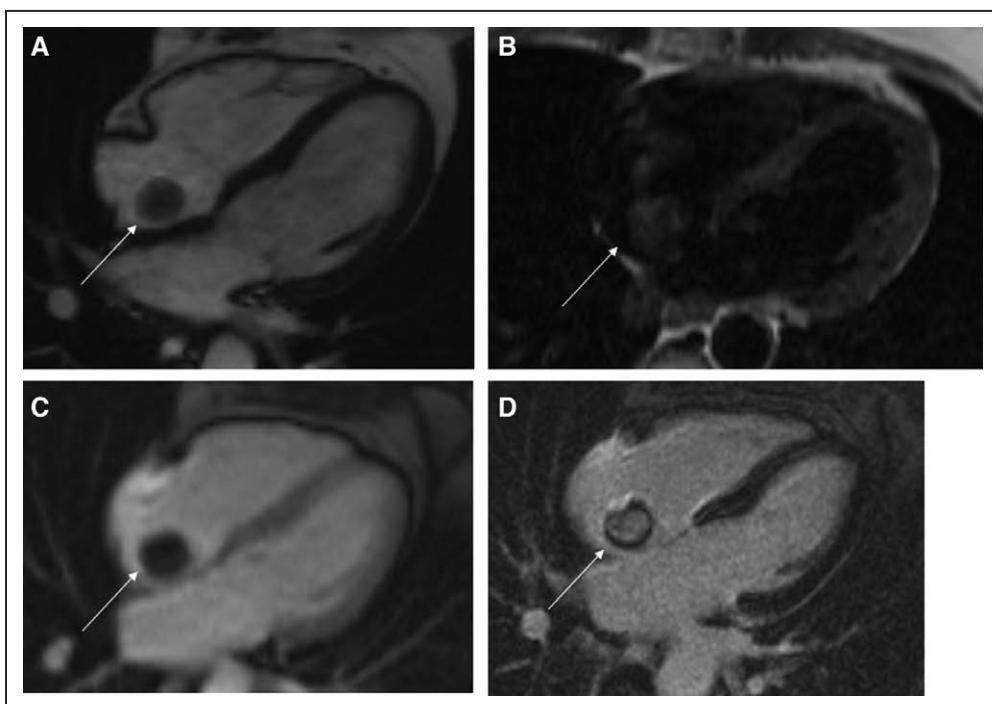


Figure 3. Cardiac magnetic resonance imaging.

A, Cine imaging showing a round mobile structure with low signal intensity in the right atrium attached to the interatrial septum. **B**, On T1- (shown here) and T2-weighted imaging, the mass had a medium-to-high signal intensity, and no fat suppression on additional imaging was seen. **C**, First-pass perfusion using a T1-weighted fast gradient echo sequence. A vascularized tumor seemed unlikely as there was no perfusion of the structure on perfusion imaging. **D**, On conventional delayed enhancement imaging (T1 inversion recovery), a thrombus appears gray or bright with a dark rim surrounding it as in our case. To selectively null avascular tissue, such as a thrombus, the inversion time can be prolonged to, for example, 600 ms, leading to a black appearance of thrombus, whereas the surrounding myocardium will be white (this is not shown here).

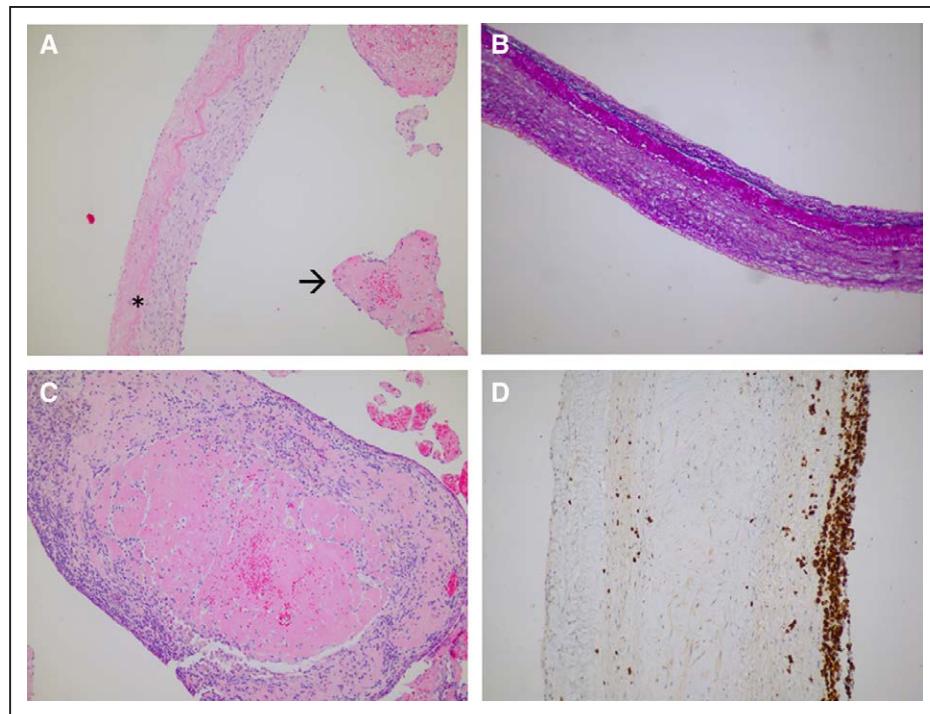


Figure 4. Microscopic images obtained from the pathology specimen.

A, Membrane (*) and thrombus (arrow). **B**, Elastica van Gieson staining solution, showing connective tissue in red and nuclei in blue. **C**, Organized thrombus with chronic inflammation. **D**, CD 20 coloring; CD 3 coloring revealed a comparable pattern, indicating a reactive infiltrate.