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CASE REPORT

ADVANCED

CLINICAL CASE

# An Anatomic Curiosity

## A Sliver of a Liver in an Unusual Location

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**ABSTRACT**

A 48-year-old woman underwent preoperative cardiac testing prior to gastric bypass. She was incidentally found to have a right atrial mass on transthoracic echocardiography. Subsequent cardiac magnetic resonance confirmed this finding. She underwent excision of the mass. Tissue pathology revealed ectopic hepatic tissue. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2021;3:1541–1544) © 2021 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

### HISTORY OF PRESENT ILLNESS

A 48-year-old woman underwent preoperative cardiac testing prior to gastric bypass surgery.

### PAST MEDICAL HISTORY

The patient's history was significant for hypertension and morbid obesity.

### DIFFERENTIAL DIAGNOSIS

The differential diagnosis for the patient's right atrial mass included thrombus, lipoma, myxoma, fibroma, metastatic renal cell carcinoma, and primary cardiac malignant tumors.

**LEARNING OBJECTIVES**

- To be able to create a differential diagnosis of a right atrial cardiac mass.
- To understand the role of multimodality imaging in the evaluation of cardiac masses.

### INVESTIGATIONS

A transthoracic echocardiogram (TTE) was obtained as part of the patient's preoperative cardiac testing for long standing dyspnea. The TTE demonstrated normal biventricular function, and no valvular disease. However, a large, ellipsoid echogenic mass measuring 29 × 29 mm was visualized. The patient then underwent transesophageal echocardiography (TEE), including 3-dimensional imaging that demonstrated the presence of a medium-sized, mobile echodensity, attached to the lateral wall of the right atrium. Subsequent cardiac magnetic resonance (CMR) was significant for a well-circumscribed mass originating in the inferior vena cava extending into the posterior right atrium (**Figure 1**).

CMR with steady-state free precession imaging demonstrated a well-circumscribed, mobile mass

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**ABBREVIATIONS AND ACRONYMS**

**CMR** = cardiac magnetic resonance

**TEE** = transesophageal echocardiography

**TTE** = transthoracic echocardiography

most prominent in the posterior right atrium with its attachment beginning in the distal inferior cava. Native T1 map demonstrated qualitatively low relaxation time relative to myocardium (Figure 2). There was no increased signal intensity on T2-weighted black blood imaging. There was no change in signal intensity with the addition of fat saturation. First-pass perfusion imaging demonstrated late, subtle perfusion without hyperemia. There was no central or peripheral enhancement of the mass on late gadolinium enhancement imaging with a standard or long (600 ms) inversion time. The low native T1 values made fibroma or acute thrombus unlikely. The lack of increased signal intensity of black blood imaging with or without fat saturation argue against myxoma or lipoma. Late, subtle perfusion is not typical for thrombus or a malignant tumor (primary or secondary). A benign cardiac tumor of different tissue characterization from myocardium was the most likely diagnosis.

## MANAGEMENT

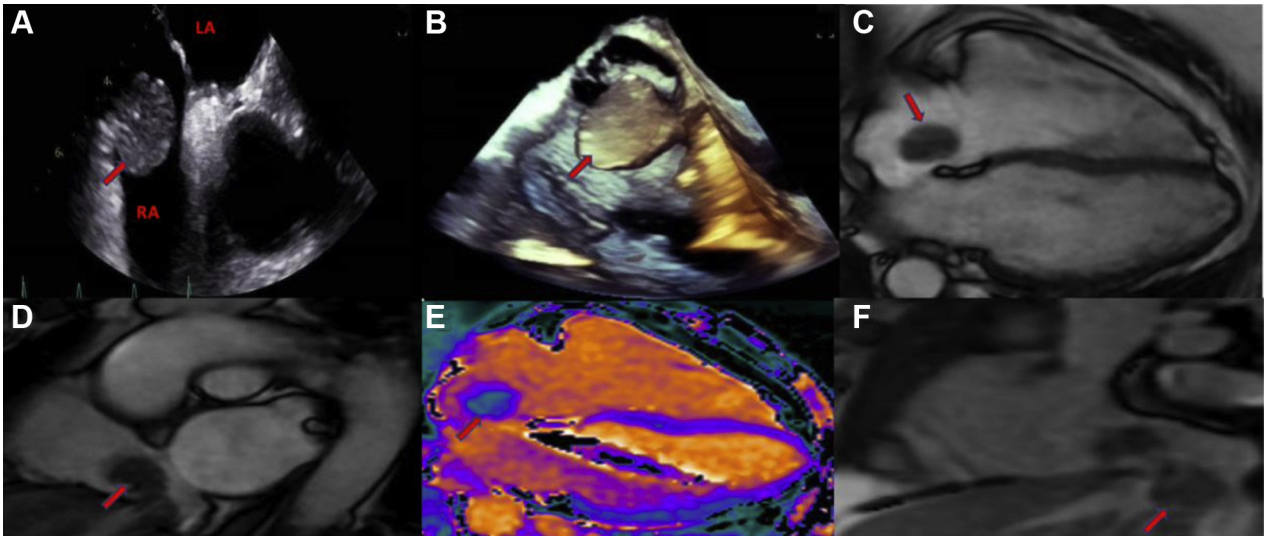
The patient underwent open heart surgery and successful excision of the cardiac mass. Gross pathology demonstrated a well-circumscribed, round mass measuring 2.5 cm in diameter with a 1-cm stalk. Tissue pathology revealed ectopic hepatic tissue with normal hepatic architecture, and was not consistent with metastatic disease (Figure 3).

## DISCUSSION

We illustrate the diagnostic challenges of a right atrial mass and the presentation of intracardiac ectopic liver tissue, an exceedingly rare phenomenon. To our knowledge, there are only 4 cases of intracardiac ectopic liver tissue reported in the literature, with most of these cases presenting incidentally (1-4). The most common causes of a right atrial mass are myxoma, thrombus, metastases, and primary malignant cardiac tumor, namely angiosarcoma. Given the wide array of possibilities, each with their own clinical implications, accurate diagnosis of a right atrial mass requires multimodality imaging in its evaluation.

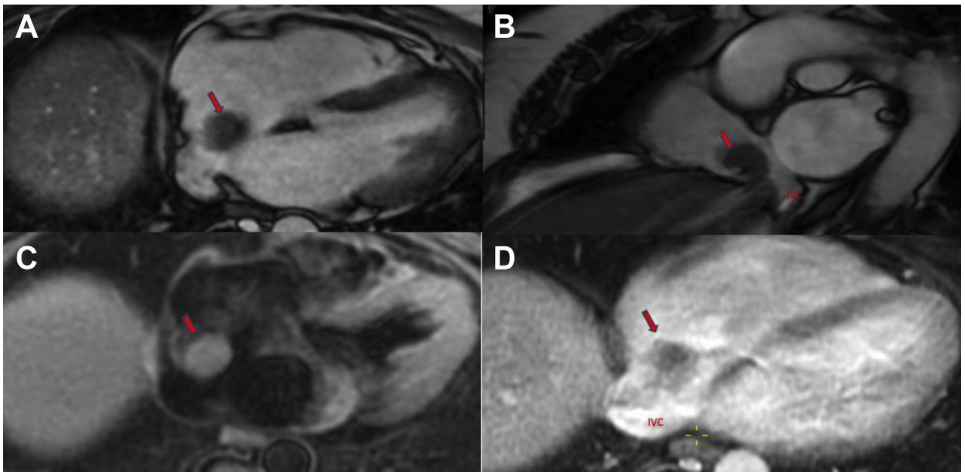
Echocardiography serves as an accessible and affordable initial imaging modality of an intracardiac mass. Beyond TTE, TEE is often subsequently used to supplement information gathered from a TTE, and can more accurately characterize a mass's size, shape, and location with high-resolution images. Additionally, TEE provides insight into surrounding structures, such as the right atrial appendage and inferior vena cava, as they may spatially relate to the intracardiac mass. However, both TTE and TEE do carry

**FIGURE 1 Multimodality Evaluation of Right Atrial Mass**




**(A)** Two-dimensional and **(B)** 3-dimensional transesophageal echocardiogram images at 70°, showing a mass that appears to originate from the lateral wall of the right atrium (RA). **(C)** Steady-state free precession imaging demonstrating mass in the RA **(D)** with extension into the inferior vena cava **(arrow)**. **(E)** T1 mapping has qualitatively low relaxation time. **(F)** There was no central or peripheral enhancement of the mass on phase sensitive inversion recovery late gadolinium enhancement imaging. LA = left atrium.

**FIGURE 2** Cardiac Magnetic Resonance Perfusion Characteristics




**(A)** Cardiac magnetic resonance steady-state free precession demonstrates a well-circumscribed lobulated mass within the posterior inferior right atrium. **(B)** The mass's attachment site begins inferiorly within the inferior vena cava (IVC) and appears confluent with the adjacent hepatic parenchyma in vertical long-axis view. The mass does not definitively perfuse on first-pass perfusion imaging on T1 **(C)** pre-contrast and **(D)** post-contrast but appears isointense to adjacent hepatic parenchyma on both sequences.

limitations, which include limited acoustic windows often related to patient's body habitus as well as variation in operator experience. TEE requires an invasive procedure with inherent risk, albeit low in selected patients. Alternatively, cardiac computed tomography is an imaging modality that can be used to assess cardiac masses in patients with known contraindications to CMR. Computed tomography imaging can uniquely offer optimal evaluation of calcified masses. (5).

Myxomas are the most common intracardiac primary tumor, with 75% occurring in the left atrium and 20% in the right atrium (6). On echocardiography, characteristics of a myxoma include smooth contours and association with a stalk. CMR is often revealing of heterogeneous signal intensity with an increased signal intensity on the fat-saturated sequence. Alternatively, a lipoma would have decreased signal intensity with fat suppression and increased signal intensity on T2-weighted images. Cardiac thrombi represent the majority of cardiac masses in the right atrium. However, thrombi located in the right atrium are relatively uncommon, and most often are found in patients with a central venous line or catheter, which was not the case for our patient. On magnetic resonance imaging, we would expect the relaxation time of the mass to be similar to myocardium on native T1 mapping (7). Last, malignant tumors are hypervascular with areas of necrosis demonstrating increased signal intensity on first-pass perfusion and late gadolinium enhancement. In this case, the mass demonstrated none of these characteristics.

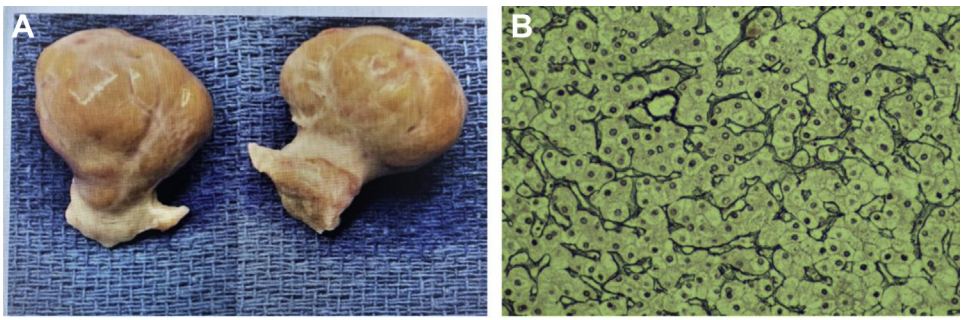
With our patient, the use of multimodality imaging was able to effectively rule out myxoma and thrombus, the 2 most common types of cardiac masses. With a high suspicion that the mass was a chronic thrombus or benign tumor, the decision was made to pursue surgical excision.

Although intracardiac ectopic liver tissue is an incredibly rare entity, it does come with important clinical implications. There is risk for carcinogenesis and transformation into hepatocellular carcinoma, thereby understating the necessity of surgical removal (1). There are no reports in the literature of ectopic liver tissue arising in the left sided cardiac chambers. Therefore, this speaks to the likely role of hepatic venous drainage and hematogeneous spread leading to the development of hepatic tissue in the right atrium. In addition to being on the right, specifically being located at the right atrial-inferior vena cava junction, as with our patient, may raise suspicion for a possible connection to the liver.

## FOLLOW-UP

Postoperatively, the patient did well without complications. She followed up in clinic with

**FIGURE 3 Pathologic Findings of Right Atrial Mass**




Gross specimen of the excised cardiac mass with attached stalk. **(A)** The mass was approximately 2.5 cm in diameter with a 1-cm stalk. **(B)** Reticulin (20×) stain depicting the architectural pattern of benign liver parenchyma.

plans to proceed with bariatric surgery in the near future.

## CONCLUSIONS

This case illustrates the diagnostic challenge that is often presented with a right atrial mass, while highlighting the utility of multimodality imaging in its evaluation. While CMR sequencing did not clinch the final diagnosis, it was crucial in excluding a number of the potential pathologies, such as acute thrombus, myxoma, and malignant tumor.

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## REFERENCES

**1.** Doshi A, Shah D, Gupta S, et al. A rare case of an ectopic liver presenting as right atrial mass. *Case Rep Cardiol*. 2019;2019:4103827.

**2.** Moody WE, Hubscher SG, Rooney SJ, Doshi SN. Intracardiac ectopic liver mimicking atrial myxoma–An unusual cause for a right atrial mass. *Int J Cardiol*. 2016;209:210–212.

**3.** Trocciola SM, Balsam LB, Yee H, Gianos E, Srichai MB, DeAnda A Jr. Ectopic liver: an unexpected finding in a right atrial mass. *Ann Thorac Surg*. 2011;92:715–718.

**4.** Xu L, Jeudy J, Burke AP. Ectopic hepatic tissue presenting as right atrial mass. *Hum Pathol*. 2012;43:958–960.

**5.** Auger D, Pressacco J, Marcotte F, Tremblay A, Dore A, Ducharme A. Cardiac masses: an integrative approach using echocardiography and other imaging modalities. *Heart*. 2011;97:1101–1109.

**6.** Motwani M, Kidambi A, Herzog BA, Uddin A, Greenwood JP, Plein S. MR imaging of cardiac tumors and masses: a review of methods and clinical applications. *Radiology*. 2013;268:26–43.

**7.** Saba SG, Bandettini PW, Shanbhag SM, Spottiswoode BS, Kellman P, Arai AE. Characterization of cardiac masses with T1 mapping. Abstracts of the 18th Annual SCMR Scientific Sessions. *J Cardiovasc Magn Reson*. 2015;17:Q32.

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