Ectopic Liver Tissue Presenting as a Right Atrial Mass on Echocardiography

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INTRODUCTION

Ectopic liver tissue is a rare phenomenon that may have clinical implications ranging from local mechanical complications to malignant transformation.[1] The vast majority of reported cases of ectopic liver are intra-abdominal,[2] while most intrathoracic cases are extracardiac. Only five cases of intracardiac involvement in adults have been reported to our knowledge.[1,3-6] One of the five cases was discovered incidentally postmortem,[3] and four of the five described a discrete mass without an anatomic connection to the liver.[1,4-6]

CASE PRESENTATION

A 43-year-old African American woman with a history of prediabetes and congenital deafness presented to the emergency department after an episode of syncope. She was at work when she developed sudden onset of lightheadedness and diaphoresis followed by syncope. She denied other concomitant symptoms. On arrival she was afebrile but hypertensive to 161/101 mm Hg. Physical examination was notable for a systolic murmur. Serial troponin testing was negative, and electrocardiography showed normal sinus rhythm without evidence of ischemia or infarction. Chest radiography and head computed tomography did not demonstrate any pathology. She was admitted for initiation of antihypertensive medications and further evaluation.

Transthoracic echocardiography was performed. It was notable for normal biventricular size and function, a poorly visualized aortic valve with moderate aortic stenosis on the basis of a calculated valve area of 1.4 cm^2, a moderately dilated ascending aorta with a diameter of 4.2 cm, and a 1.3- × 1.5-cm globular, immobile echo density that appeared adherent to the posterolateral aspect of the right atrium (Figures 1 and 2, Videos 1 and 2). Cardiac magnetic resonance imaging (MRI) was recommended to better characterize the mass.

Cardiac MRI demonstrated a bicuspid aortic valve with moderate aortic stenosis (calculated valve area 1.2 cm^2) and moderate aortic insufficiency, moderate fusiform aneurysmal dilation of the ascending aorta (5 cm in diameter), and a round mass at the inferior vena cava (IVC)–right atrium (RA) junction (Figures 3 and 4, Video 3). The mass was thought to likely be a benign tumor, given that it was isointense on T1 and T2 imaging, was not hypervascular, did not exhibit late gadolinium enhancement, and did not show signal characteristics consistent with thrombus, fat, angiosarcoma, or a malignant or metastatic tumor. Hence, the differential diagnosis was narrowed to an atrial myxoma or a papillary fibroelastoma, although it was noted that the MRI appearance was not classic for either.

Given the MRI findings, the patient was referred for a cardiothoracic surgical evaluation. With a congenitally bicuspid valve and a 5-cm ascending aorta, moderate aortic stenosis, and her right atrial mass, she was scheduled for aortic valve replacement, ascending aortic aneurysm repair, and excision of her right atrial mass. Preoperatively, she underwent left heart catheterization and coronary angiography, which demonstrated no coronary artery disease. Intraoperative transesophageal echocardiography confirmed the above findings with better localization of the mass at the RA-IVC junction (Figures 5 and 6, Videos 4 and 5). Aortic valve replacement was accomplished with a 19-mm St. Jude Regent mechanical valve prosthesis, and the ascending aorta was replaced with a 22-mm Gelweave graft. The right atrial mass was located immediately anterior to the IVC, without evidence of IVC obstruction or a connecting stalk, and was successfully excised. Surgical pathology was notable for heterotopic hepatic parenchyma without evidence of hemorrhage, necrosis, or malignancy (Figures 7–9). The postoperative course was uneventful, and the patient was successfully discharged on postoperative day eight. Follow-up echocardiography has shown no recurrence of the right atrial mass.

DISCUSSION

Heterotopic liver tissue remains an infrequently described phenomenon, with a reported incidence of <0.5%.[5] Cases of ectopic liver have been classified into four types: (1) an accessory liver lobe attached to the liver, (2) a large accessory liver lobe with a connecting stalk to the liver, (3) ectopic liver without connection to the liver, and (4) microscopic ectopic liver tissue.[7] The present case is consistent with the third type of aberrant liver tissue.

The pathogenesis of heterotopic liver tissue in the absence of trauma is unknown. It is hypothesized that it arises from a congenital defect of the septum transversum, embryonic tissue that differentiates into both the diaphragm and ventral mesentery of the foregut.[1] However, there is at least one reported case of the de novo development of an intracardiac ectopic liver mass.[4] The patient in that case had no evidence of the mass on surface echocardiography done 18 months before her presentation. Therefore, the authors of that case report proposed a theory that there can be hematogenous migration of hepatic cells with regenerative abilities even after intrauterine development.[6] Our case, along with the other reported cases with discrete intracardiac heterotopic masses without connecting stalks,[1,4-6] could be consistent with either premise, as all these reports noted right atrial masses. To our knowledge, there are no reports of intracardiac ectopic hepatic tissue in the left heart chambers. Given the hepatic venous drainage system, such a mass would make the hematogenous migration theory less likely, although not impossible in a

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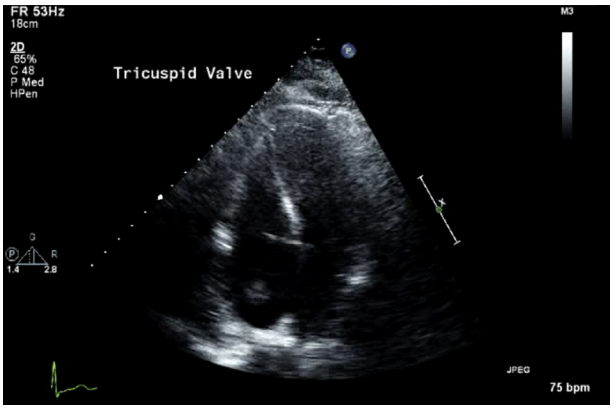

Figure 1 Transthoracic echocardiogram 1. Well-circumscribed, nonmobile, 1.3- × 1.5-cm mass in the body of the RA.

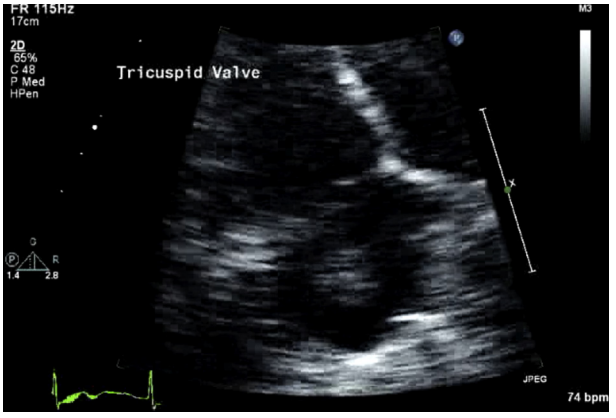

Figure 2 Transthoracic echocardiogram 2. Focused view of the RA, right ventricle, and right atrial mass.

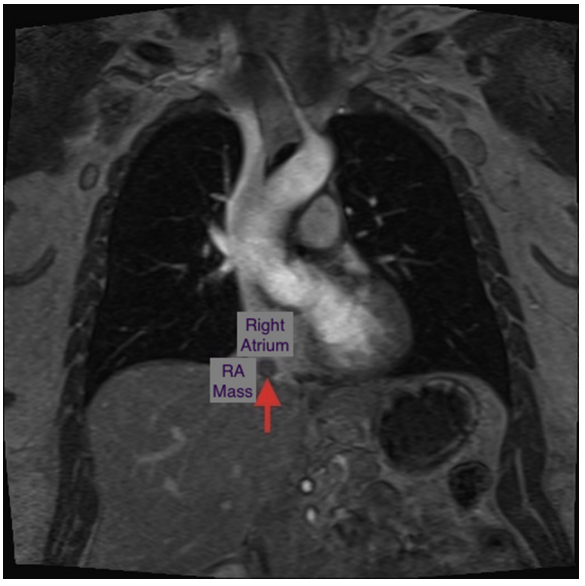

Figure 3 MRI 1. Contrasted magnetic resonance image demonstrating no early uptake of contrast by the right atrial mass (*red arrow*).

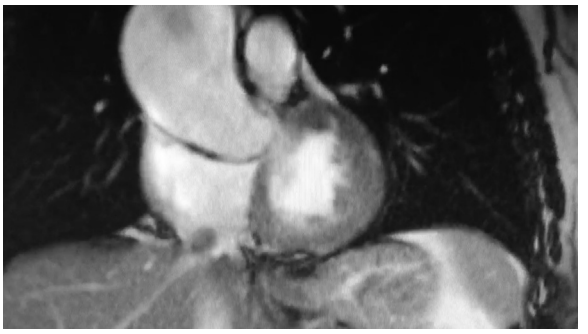

Figure 4 MRI 2. Cine video of contrasted magnetic resonance image of right atrial mass.

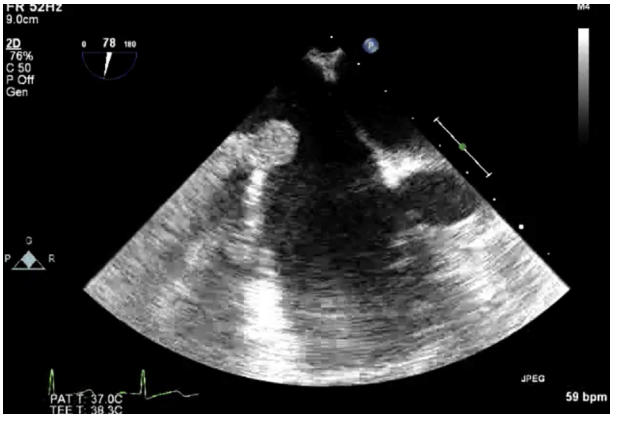

Figure 5 Transesophageal echocardiogram 1. Mass well visualized at the posterolateral aspect of the RA at the junction of the inferior vena cava.

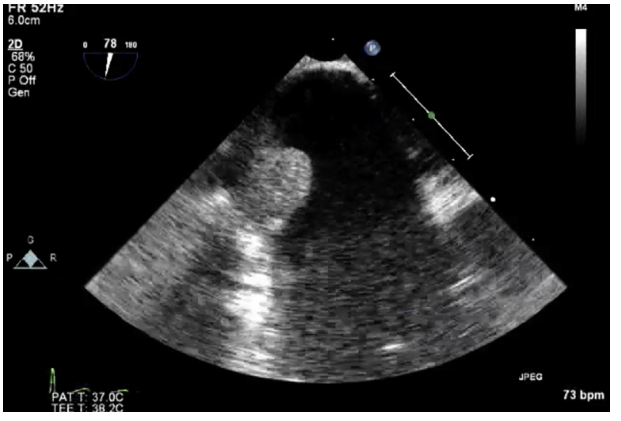

Figure 6 Transesophageal echocardiogram 2. Focused view of RA and right atrial mass. The mobile Eustachian valve is also seen.

patient with a patent foramen ovale or other right-to-left intracardiac or intrapulmonary shunting.

In addition to being considered in the differential diagnosis of right atrial masses found during cardiac imaging, this often incidental finding may have clinical implications. In our case it is possible that