Ectopic Liver Tissue Presenting as a Right Atrial Mass on Echocardiography

Brett A. Izzo, MD, Charles J. Stoudenmire, MD, and Benjamin F. Byrd, III, MD, Nashville, Tennessee

INTRODUCTION

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

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 cm², 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). 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 cm²) 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 static 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%. 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. 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. However, there is at least one reported case of the de novo development of an intracardiac ectopic liver mass. 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. Our case, along with the other reported cases with discrete intracardiac heterotopic masses without connecting stalks, 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

From the Departments of Medicine (B.A.I., B.F.B.) and Pathology (C.J.S.), Vanderbilt University, Nashville, Tennessee.

Keywords: Ectopic, Heterotopic, Liver, Right atrium

Conflicts of interest: The authors reported no actual or potential conflicts of interest relative to this document.

Copyright 2017 by the American Society of Echocardiography. Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

2488-6441
http://dx.doi.org/10.1016/j.case.2017.05.007

http://creativecommons.org/licenses/by-nc-nd/4.0/
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...
the right atrial mass explained the patient’s episode of syncope, given its location at the RA-IVC junction. Chapman-Fredrick et al. reported a case of an ectopic hepatic mass causing IVC obstruction. In addition, downstream embolization leading to pulmonary embolism is also a possible complication of these right atrial masses.

Other previously noted clinical implications include increased risk for carcinogenesis in the heterotopic tissue. In fact, in a review of 100 cases of aberrant hepatic tissue, 28 patients had hepatocellular carcinoma found on pathologic review of the tissue. Although such was not the case in our patient, the potential for malignant transformation increases the urgency for surgical removal, ensuring negative margins during surgical excision, and careful pathologic review of the ectopic tissue.

Our patient’s mass and all of the previously reported cases of right atrial ectopic hepatic tissue were diagnosed by transthoracic echocardiography. Although echocardiography may not be able to differentiate these masses from an atrial myxoma, primary and secondary malignant masses, and in some cases thrombus or vegetations, important clinical information from the study can be gleaned. Although the location of the mass does not confirm the diagnosis, on the basis of the current case literature, right-sided masses should raise suspicion of ectopic hepatic tissue. This is particularly true of masses at the RA-IVC junction. In the subcostal view, the IVC can be followed for possible evidence of a connection to the liver. Transesophageal echocardiography can also be considered for further evaluation of the RA-IVC junction and characterization of the mass.

Cardiac MRI can narrow the differential diagnosis in patients with intracardiac masses. Interestingly, in our case, MRI demonstrated a mass that could not be fully characterized, as it did not meet MRI criteria for diagnosis of atrial myxoma, papillary fibroelastoma, thrombus, fat, angiosarcoma, or a malignant or metastatic tumor and was isointense on T1 and T2 imaging. When an intracardiac mass cannot be characterized into one of the above diagnoses by cardiac MRI, heterotopic hepatic tissue should be considered.

CONCLUSION

Intracardiac heterotopic hepatic tissue is a very rare finding, with all reported cases detected by echocardiography showing right atrial masses. Although echocardiography alone may be unable to differentiate these masses from atrial myxomas or other intracardiac tumors, the RA-IVC location and subsequent cardiac MRI can help increase suspicion. Connection of the mass to the liver should be sought on echocardiography.

Ultimately, surgical pathology is necessary for a definitive diagnosis.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at http://dx.doi.org/10.1016/j.case.2017.05.007.

REFERENCES

1. Sarsam S, Arsene C, Abu Rashed K, Liu Q, Rajagopal R. Heterotopic hepatic polyp identified in the right atrium. J Med Cases 2012;3:274-6.
2. Matsuyama M, Sugiura S, Kakita A, Sato Y, Kuroda M. Hepatocellular carcinoma arising from ectopic tissue in the spleen producing insulin-like growth factor II. Pathol Res Pract 2011;207:124-6.
3. Brustmann H. Heterotopic liver in the right cardiac auricle. Ann Diagn Pathol 2002;6:248-9.
4. Moody W, Hubscher S, Rooney S, Doshi S. Intracardiac ectopic liver mimicking atrial myxoma—an unusual cause for a right atrial mass. Int J Cardiol 2016;209:210-2.
5. Trocciola S, Balsam L, Yee H, Gianos E, Srichai M, DeAnda A Jr. Ectopic liver: unexpected finding in a right atrial mass. Ann Thorac Surg 2011;92:715-8.
6. Xu L, Jeudy J, Burke A. Ectopic hepatic tissue presenting as right atrial mass. Human Pathol 2012;43:958-60.
7. Collan Y, Hakkiluoto A, Hastbacka J. Ectopic liver. Ann Chirg Gynaecol 1978;67:27-9.
8. Champman-Fredricks J, Birusingh R, Ricci M, Rodriguez M. Intracaval liver with cardiac extension. A new development anomaly? Fetal Pediatr Pathol 2010;29:401-6.