Multimodality Imaging of an Unusual Case of an Obstructive Intracaval Mass by an Aberrant Liver

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A 40-year-old woman presented with recurrent chest pain. Past medical history was positive for schizoaffective disorder, polysubstance abuse, and gestational diabetes mellitus. The physical examination was unremarkable. Results of an ECG and a test for cardiac enzymes, including troponins, were negative. Transthoracic echocardiography showed a well-circumscribed, smooth, homogeneous mass measuring up to 1.5 cm in the distal inferior vena cava (IVC) that was protruding into the right atrium (Figure 1A). Transesophageal echocardiography confirmed the mass and showed it to be arising from the distal IVC near the right hepatic vein entry (Figure 1B).

Magnetic resonance imaging showed a well-circumscribed mass arising just below the junction of the IVC and the right atrium and protruding into the IVC. The mass had signal characteristics similar to those of the liver parenchyma in all sequences, including T1-weighted, T2-weighted, short Tau inversion recovery, half Fourier acquisition single-shot turbo spin echo, volumetric interpolated breath-hold examination, and steady-state free-precession (Figure 2A and 2B) sequences. The mass appeared contiguous with the liver parenchyma and had contrast enhancement similar to that of liver parenchyma in perfusion (Figure 2C; Movie 1 in the online-only Data Supplement) and delayed enhancement images. Positron emission tomography/computed tomography showed the mass within the IVC (Figure 3A) but did not show any uptake (Figure 3B). Cardiac catheterization was negative. Imaging features were suggestive of an intracaval aberrant liver.

Because of IVC obstruction, a decision was made to resect the mass. During the surgery, the mass was identified in the IVC, from the 2-o’clock to 3-o’clock position as visualized from the right atrial side. It arose from a broad stalk that communicated with the liver parenchyma. Pathology showed a pedunculated mass with histopathological characteristics of normal liver parenchyma (Figure 4). This is a very unusual case of an aberrant liver tissue that extended into the IVC to present as an IVC/right atrial mass.

Aberrant hepatic tissue outside the native liver has been described as an accessory hepatic tissue when it retains connection with the native liver by a stalk or an ectopic tissue when there is no such connection.1,2 Abdomen is the most common location of this abnormality (gall bladder, spleen, pancreas),3 and only 4 previous cases of IVC involvement have been reported.2,3

Ectopic/accessory liver is likely caused by aberrant migration of hepatocytes into the IVC lumen during development. There is a close association between the development of liver bud, IVC, and septum transversum. The liver bud is formed in the fourth week of development from hepatoblasts that migrate from the primitive foregut endoderm into the septum transversum mesenchyme. The hepatic portion of the IVC develops between 54 and 56 days of development along the caval fold in the network between the hepatocardinal vein and right subcardinal vein.4 After the IVC is formed, the caval fold mesenchyme to the left of the IVC is invaded by embryonic hepatic tissue and becomes the caudate lobe.4 Aberrant migration and trapping of the hepatic tissue into the IVC results in accessory liver or ectopic liver. Development of a second liver bud and incomplete atrophy or regression of developing liver lobes are other theories.2

The ectopic liver tissue is usually asymptomatic and mostly discovered incidentally in surgery, autopsy, or imaging. However, obstruction of the hepatic veins may cause cirrhosis, portal hypertension, and portal venous thrombus.5 Obstruction of the IVC may result in lower-extremity thrombus. Dislodgement may lead to pulmonary embolus.5

Although very uncommon, this diagnosis should be considered when the signal characteristics and contrast enhancement are similar to those of liver parenchyma. Differential diagnosis of IVC lesions include thrombus, benign neoplasms (leiomyoma, endothehlioma, fibroelastoma), and malignant neoplasms (leiomyosarcoma, metastasis, lymphoma, and tumor extension).

Disclosures

None.

References


Figure 1. A, Parasternal right ventricular inflow view of a transthoracic echocardiogram demonstrates a homogeneous echogenic mass that is extending into the right atrium from the inferior vena cava. B, Transgastric inferior vena cava long-axis view with transesophageal echocardiography demonstrates the mass (white arrow) prolapsing into the right atrium from the inferior vena cava. Note that the mass is contiguous with the liver tissue, and its echogenicity is identical to that of the liver.

Figure 2. A, Axial steady-state free-precession magnetic resonance image shows an isointense mass within the lumen of the inferior vena cava (arrow). B, Coronal steady-state free-precession image shows mass that is extending into the inferior vena cava and the right atrium, with similar intensity as that of the adjacent liver parenchyma, with which it is contiguous. C, Sagittal perfusion image shows the mass within the inferior vena cava (arrow), showing similar contrast enhancement as the liver, with which it is contiguous.

Figure 3. A, Coronal reconstructed computed tomography image shows a mass within the inferior vena cava causing its expansion. The mass has the same attenuation as the adjacent liver parenchyma and is contiguous with it. B, Coronal reconstructed positron emission tomography/computed tomography image shows the mass within the IVC, with no uptake of fluorodeoxyglucose.
Figure 4. Hematoxylin-and-eosin staining of the specimen (magnification ×200) demonstrates normal-appearing nodular hepatic parenchyma with hepatocytes and Kupffer cells.
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Circulation. 2014;129:e310-e312
doi: 10.1161/CIRCULATIONAHA.113.004621
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/129/5/e310

Data Supplement (unedited) at:
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Movie Legend

**Movie 1.** Coronal dynamic perfusion image through the mass and the liver showing the intracava mass having similar signal characteristics and perfusion as that of the adjacent liver. Best viewed with Windows Media Player.