Ectopic liver masquerading as a floating intracaval mass

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Ectopic liver is defined as liver parenchyma situated outside the liver proper with no connection to native hepatic tissue. This rare developmental anomaly is most commonly described as an attachment to the gallbladder with an incidence <0.3%, but it has been reported in other locations within the abdomen and thorax. Most cases are found incidentally in asymptomatic patients, but ectopic liver has been known to cause visceral or vascular obstruction. Herein we present a unique case of ectopic liver attached by a thin stalk seemingly floating in the suprahepatic inferior vena cava. (J Vasc Surg 2012;55:1759-61.)

A 36-year-old female presented with right upper quadrant and flank pain 2 months after a laparoscopic cholecystectomy for symptomatic gallstones. Evaluation demonstrated a retained common duct stone and intracaval mass. The patient underwent successful endoscopic retrograde cholangiopancreatography, which alleviated her symptoms, and was then referred for evaluation of her caval mass. Medical history was significant for two vaginal births, but no prior history of liver disease nor oral contraceptive use. Physical examination and liver tests were unremarkable except for a body mass index of 41. A computed tomography scan demonstrated a 4-cm intracaval mass just cephalad to the origin of the middle hepatic vein in the suprahepatic inferior vena cava (IVC), which seemed to produce partial outflow obstruction suggested by dilatation of the middle hepatic vein (Fig 1). There was no ascites or hepatomegaly. Magnetic resonance imaging demonstrated a soft tissue mass, which exactly paralleled the signal intensity of adjacent liver parenchyma on both T1 and T2 weighted images (Fig 2). Magnetic resonance imaging cine loop images demonstrated the mass floating to-and-fro within the IVC, attached by a thin stalk and appearing to originate from the suprahepatic IVC at the junction with the middle hepatic vein. Because of the precarious location of this floating mass, the observed partial outflow obstruction from the middle hepatic vein, and theoretic possibility of detachment, becoming a probable fatal pulmonary embolus, the decision was made to resect the mass.

A right subcostal incision was extended superiorly to allow for a median sternotomy. The liver was mobilized to expose the retrohepatic vena cava. The infrahepatic and suprahepatic vena cava were isolated, and venovenous bypass initiated from the infrahepatic vena cava to the right atrium. Temporary hepatic inflow occlusion was achieved via a vascular clamp across the porta hepatis with total clamp time of 17 minutes. An oblique incision was made on the anterior surface of the suprahepatic vena cava just cephalad to the origin of the middle hepatic vein. Upon opening the vena cava, a pedunculated lesion attached by a thin stalk was visualized (Fig 3, A). The lesion was removed by shaving the stalk base flush with the intima of the vena cava. The stalk was not ligated. The encapsulated mass measured 17 mm × 25 mm × 10 mm (Fig 3, B). Histopathology demonstrated benign liver tissue with normal-appearing sinuses and portal triads with chronic inflammatory changes. The patient made an uneventful recovery and was discharged home within 7 days. Follow-up liver function studies were normal.

DISCUSSION

Collan initially described abnormal liver tissue as accessory or ectopic. An accessory liver lobe is situated outside the liver proper without direct connection, while ectopic liver is situated outside the liver proper without direct connection to hepatic parenchyma.1 The incidence of ectopic liver is quite low.2-5 Gallbladder-associated liver is most common, but ectopic liver has been reported within thorax, lung, spleen, pancreas, umbilicus, adrenals, pylorus, and IVC.4-11 Ectopic liver is generally an incidental finding discovered during surgery or autopsy.5,8 Visceral and portal obstruction with associated symptoms has been reported; however, ectopic liver is generally asymptomatic. This case

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represents an example of ectopic liver, as there was no macroscopic or microscopic connection to the liver proper, save its proximity.

Ectopic tissue does not receive arterial supply from the hepatic artery and, furthermore, microscopic evaluation may demonstrate absent or incomplete portal and biliary systems. Biliary drainage varies by location of ectopic liver with drainage into the normal biliary system, other organ, or no drainage. Hepatocytes in an ectopic liver behave like normal hepatocytes and may demonstrate an increased risk of carcinogenesis likely secondary to incomplete anatomic architecture and metabolic derangement.

Theories to explain intra-abdominal ectopic liver include formation of accessory lobe with atrophy of connection to the liver proper or abnormal migration of liver buds during organogenesis. In this case, one can surmise that the ectopic liver development stemmed from the embryologic assembly of the IVC and the intimate association of the maturing liver bud, septum transversum, and IVC. Ordinarily, the IVC derives its origin from three different sources: the posterior cardinal, subcardinal, and supracardinal venous systems. The retrohepatic IVC and hepatic veins are thought to derive their origin from a coalescence of intrahepatic sinusoids fusing with a “postero-caudal” vein originating in or around the right adrenal gland. This portion of the IVC develops during days 54 to 56 of embryogenesis. The fact that hepatoblasts migrate from the endoderm of the primitive foregut into the septum transversum mesenchyme to form the nascent liver bud attests to the intimate relationship of maturing liver epithelium and mesenchymal sinusoidal structures. In fact, it appears that signals for hepatoblast migration and proliferation within the stroma of the septum transversum originate in a paracrine fashion from neighboring endothelial cells. It is understandable that a developing focus of liver tissue may be trapped within this mesenchymal milieu and matures as intravascular ectopic liver. We would postulate that this, indeed, is the explanation, as the ectopic liver removed had all the elements of normal hepatic parenchyma.
There have been two prior reports of intracaval ectopic liver tissue occurring in a 12-year-old child with Arnold Chiari malformation and an obese 42-year-old woman with chronic anemia. In the first case, the lesion was situated at the IVC–right atrial junction; however, in the second case, the lesion originated from a hepatic vein and extended into the right atrium. These two tumors consisted of benign hepatic tissue much like the specimen in this case.

Whether the present case could have been treated non-operatively or with endovascular approaches is a matter of debate. Certainly, the thin stalk to which this liver tissue was attached appeared tenuous and capable of rupturing. The consequences would have been disastrous, with a massive pulmonary embolus likely. It is unlikely that the symptoms of right upper quadrant pain in this case were related to her lesion. The ectopic liver had probably been present since birth. It is possible that progressive dilatation of the middle hepatic vein from partial obstruction of flow could have produced some stretching of Glisson's capsule and subsequent pain. There was no radiologic evidence of a Budd-Chiari syndrome. More probable, this was a serendipitous finding during the course of evaluating postcholecystectomy pain.

An open approach was favored over endovascular therapy as we were uncertain of the etiology of this tumor. The surgical approach was designed to minimize blood loss and provide a bloodless surgical field for removal of this tumor. While venovenous bypass might have been possible without a chest incision, the lesion was near the diaphragm and control of the IVC above the diaphragm was necessary. The use of venovenous bypass and complete vascular isolation of the liver allowed resection of this unusual retrohepatic intracaval lesion in a bloodless surgical field and allowed complete transection of the stalk base at the level of the intima for complete removal of the tumor and stalk.

REFERENCES


