

Cavernous hemangioma with extensive sclerosis masquerading as intrahepatic cholangiocarcinoma — A pathologist's perspective

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A patient presented with an acute episode of bright red blood in her stool. The incidental liver mass seen in segment 4 was suspected to represent a cholangiocarcinoma due to associated mild intrahepatic biliary ductal dilatation and suspicion for capsular retraction. Pathology confirmed that this lesion represented a sclerosing hemangioma. This case report corroborates prior observations that degenerative changes in hemangiomas—sclerosis, narrowing of vascular channels, thrombosis, infarct, hemorrhage—may produce atypical radiographic findings. Since these atypical radiographic features may suggest a primary or metastatic malignancy, the protean appearance of hemangiomas remains an important consideration in the evaluation of hepatic masses.

Case report

A 60-year-old woman presented with an acute onset of bright red blood in her stool. She had no preceding episodes of hematochezia, and the most recent colonoscopy performed within the past 5 years was normal. She had a remote history of a laparoscopic cholecystectomy, but was otherwise healthy and denied recent weight changes, pain, nausea, jaundice, systemic symptoms, or new exposures. Laboratory evaluation revealed a total bilirubin of 0.4 mg/dL, albumin 4.1 g/dL, INR 1.0, platelets 404,000/uL, hematocrit 45%, creatinine 0.8 ml/dL, and liver and pancreatic enzymes within normal limits. Serum 19-9 and carcinoembryonic antigen were not elevated.



Figure 1. 60-year-old woman presented with an acute onset of bright red blood in her stool and an incidental liver mass. Axial CT of the abdomen in the portal-venous phase shows a hypodense peripherally enhancing mass with subtle intrahepatic biliary ductal dilatation in the left lobe.

Initial imaging workup consisted of an ultrasound that showed a heterogeneous 3.5cm mass in segment 4 (images not shown). Minimal intrahepatic biliary ductal dilatation was noted in the left lobe of the liver. Subsequently, a CT

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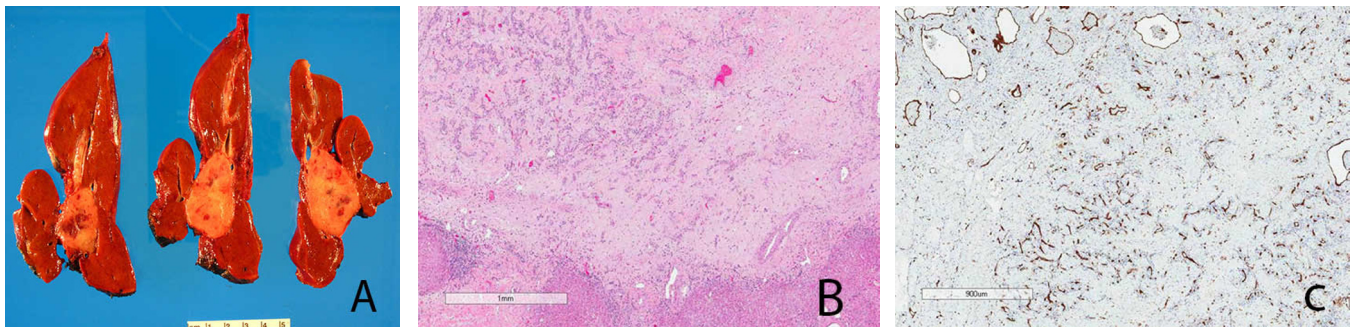
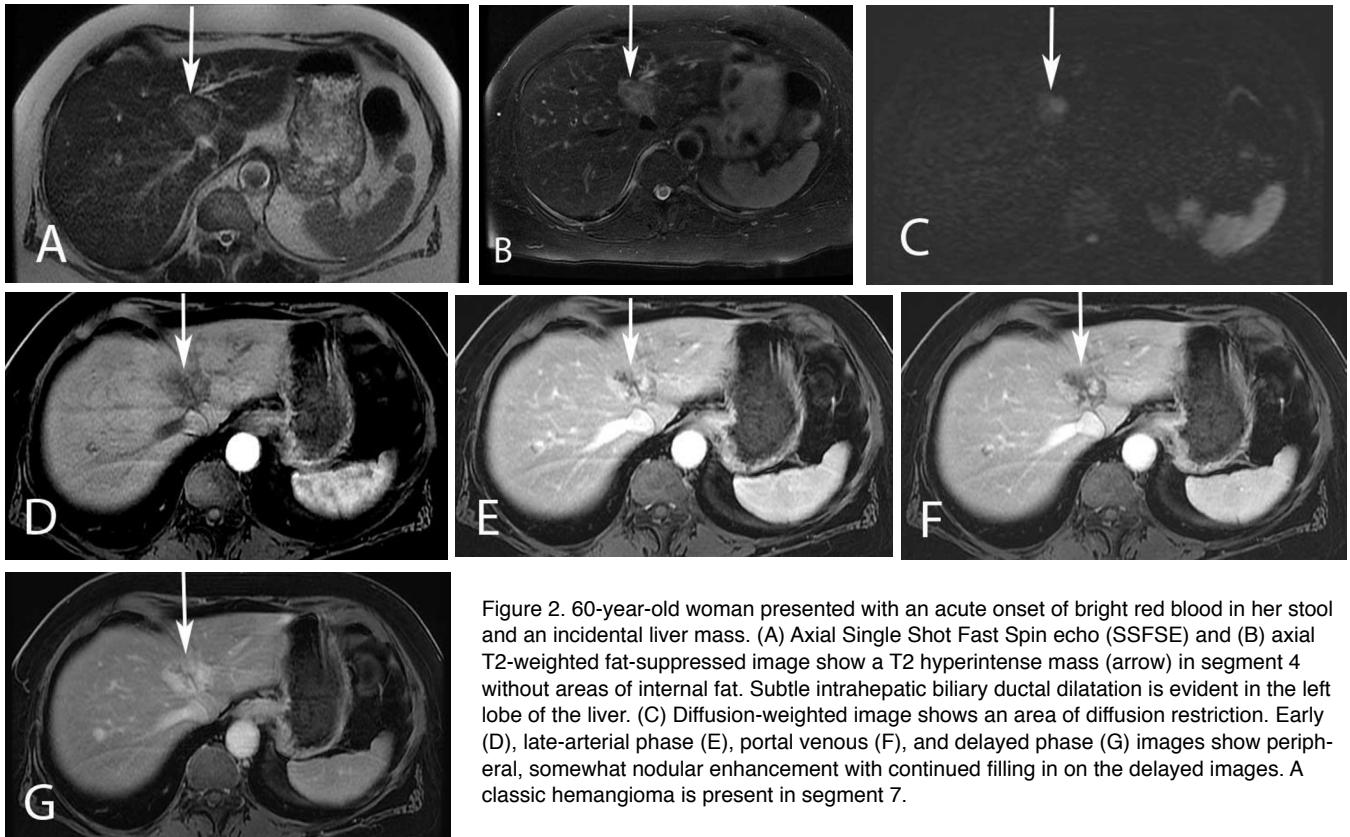


Figure 3. 60-year-old woman presented with an acute onset of bright red blood in her stool and an incidental liver mass. (A) The partial hepatectomy specimen shows a 4.2cm, well-circumscribed, somewhat soft, white, and focally hemorrhagic mass. (B) Microscopy revealed well-demarcated, thin-walled, variably cavernous, and compressed vascular channels lined by endothelial cells without cytologic atypia or mitoses. No bile duct proliferation or carcinoma was present, which was confirmed by a negative CK7 stain (not shown). Extensive zones of sclerosis were present within the hemangioma, more prominent at the periphery. The background liver had mild steatosis. (C) CD31 stain confirmed the vascular nature of the neoplasm.

study confirmed the same findings. The enhancement pattern on CT was nonspecific, and the mass was hypodense (Fig. 1). A 4-phase MRI of the liver for complete characterization showed the 3.5 x 3.1-cm mass as having hypointense T1 signal, mildly hyperintense T2 signal, diffusion restriction, peripheral areas of enhancement, delayed filling in, and associated mild intrahepatic biliary ductal dilatation. Subtle capsular retraction was suspected. A classic hemangioma was seen in segment 8 with peripheral nodu-

lar enhancement on the arterial phase and filling-in on the portal venous and delayed phases (Fig. 2). On further discussion with the tumor board, the mass was referred for surgical resection, as the intrahepatic biliary ductal dilatation was felt not to represent a hemangioma due to associated intrahepatic biliary ductal dilatation. The lesion was surgically removed (partial hepatectomy) without complication. On pathology, the tumor was diagnosed as a sclerosing hemangioma (Fig. 3).

Discussion

In this case of a sclerosing cavernous hemangioma, an unusual MRI appearance lead to radiographic concern for intrahepatic cholangiocarcinoma. Imaging findings correlated pathologically with extensive sclerosis and predominantly compressed vascular channels; no carcinoma was present.

Cavernous hemangiomas are benign vascular proliferations; they are the most common primary tumor in the liver (1). They are usually solitary but may be multiple 10% of the time, and can be associated with hemangiomas in other sites in syndromes such as von Hippel-Lindau and those related to systemic vascular malformations (2). Their pathogenesis is not well understood; some studies suggest a correlation between mast cells and angiogenesis (3). Other investigators have suggested that estrogen therapy may lead to enlargement, though this is not well supported (4). Though usually found incidentally, giant hemangiomas (> 5 cm) may present with nausea, jaundice, consumptive coagulopathy (Kasabach-Merritt syndrome); they may also rarely rupture (5).

Sclerosing cavernous hepatic hemangiomas can present diagnostic radiographic challenges, mimicking primary hepatocellular carcinoma, metastases, abscess (6), or (in this case) cholangiocarcinoma. The typical heterogeneous MRI appearance of a hemangioma is created by sluggish blood flow through large vascular spaces with constant formation and disintegration of thrombi (1). However, atypical radiographic features correlate pathologically with prominent sclerosis, narrowing of vascular channels, and degenerative changes; these alter blood flow and thus the usual finding of peripheral globular enhancement (6). Additionally, although usually well-circumscribed, hemangiomas occasionally contain entrapped bile ducts or extend into the liver parenchyma (1), which may suggest an infiltrative lesion.

In this case, atypical imaging characteristics included the presence of intrahepatic biliary ductal dilatation and a suspicion for capsular retraction. Pathologically, fibrosis usually begins in the center of the lesion and is associated with thrombosis, infarct, and hemorrhage (3); these degenerative changes may cause loss of typically high T2 MRI signal and alter enhancement characteristics (7). This report corroborates prior observations that degenerative changes in hemangiomas—sclerosis, narrowing of vascular channels, thrombosis, infarct, hemorrhage—may produce atypical radiographic findings (6). Since these atypical radiographic features may suggest a primary or metastatic malignancy, the protean appearance of hemangiomas remains an important consideration in the evaluation of hepatic masses.

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