A 29-year-old white man was brought to our department with a presumptive diagnosis of diffuse hepatic metastasis. He was complaining of anorexia and abdominal right upper quadrant discomfort over the last 2 months. On physical examination he had a nodular liver border, 2-cm below the right costal margin. Abdominal ultrasonography and computed tomography were performed, revealing the presence of multiple nodules diffusely distributed throughout the liver parenchyma. The patient referred a history of asthma, recently medicated with montelukast, and denied alcohol intake or hepatotoxic drug exposure. Liver tests demonstrated slightly elevated aspartate aminotransferase (64 U/L), alanine aminotransferase (98 U/L), and \( \gamma \)-glutamyltransferase (270 U/L) levels. The remaining blood tests (tumor markers—\( \alpha \)-fetoprotein, carcinoembryonic antigen, and CA19.9; viral serologies; autoimmunity; ceruloplasmin; HFE genotype), as well as upper GI endoscopy and ileocolonoscopy, were normal. The subsequent investigation with abdominal magnetic resonance imaging and magnetic resonance angiography (Figure A, B) revealed an abnormal vascular structure (arrows) and elucidated this clinical situation.

What is the most likely diagnosis?

Look on page 000 for the answer and see the GASTROENTEROLOGY web site (www.gastrojournal.org) for more information on submitting your favorite image to Clinical Challenges and Images in GI.

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Answer to the Clinical Challenges and Images in GI Question: Image 1 (page 126●●): Abernethy Malformation Type 2 With Hepatic Nodular Regenerative Hyperplasia

Magnetic resonance angiography confirmed an extrahepatic porto-systemic shunt. Schematically (Figure C), there is an aneurysmatic dilation with a lateral communication between the portal vein and the inferior vena cava—an Abernethy malformation type 2; notice that there is still blood flow through the portal branches.

Venovenous portosystemic malformations are rare and underdiagnosed. They are divided in Abernethy (extrahepatic, types 1a, 1b, and 2) and Park (intrahepatic, types 1–4), and can be diagnosed incidentally, after liver tests changes, or by clinical signs of hepatic shunting (encephalopathy, hepatopulmonary syndrome, or hypoglycemia).1 Congenital extrahepatic portosystemic shunt was first described by Abernethy in 1793, and later classified by Morgan as type 1, if the entire portal blood drains to the vena cava, and type 2 when there is a partial side-to-side anastomosis with a patent intrahepatic portal vein. To our knowledge, there are only 23 cases reported with the type 2 malformation. The malformation type is important for approach decision, even though there is no standard treatment. Asymptomatic patients should be maintained under surveillance; for symptomatic cases, some authors manage conservatively, whereas others prefer shunt correction with surgery or endovascular methods.2

Extrahepatic shunts are frequently associated with other anomalies (heart, spleen, kidney, or biliary) that were not present in our patient. They are also associated with an increased frequency of hepatic neoplasms, either benign (focal nodular hyperplasia, adenoma, nodular regenerative hyperplasia) or malignant (hepatocellular carcinoma or hepatoblastoma). It has been proposed that the diversion of hepatotrophic substances away from the liver, along with increased arterial hepatic flow, results in alterations of development, function, and regenerative capacity of the liver. Malignant transformation of benign neoplasms has also been reported in the setting of Abernethy shunts; therefore, long-term follow-up is recommended for these patients. The histopathology of our patient’s ultrasound-guided liver biopsy presented an apparent division of the liver specimen in small nodules without fibrous septation and also dilation of the center-lobular veins with focal perivenular fibrosis, all consistent with nodular regenerative hyperplasia (Figure D). This is also a rare disorder and literature on this subject is scarce. In fact, it seems to result from focal alterations in blood flow, and in addition to vascular disorders (Budd–Chiari syndrome, congenital agenesis or shunts of the portal vein), it can be associated with the exposure to certain drugs and with rheumatic or myeloproliferative diseases.3 Since diagnosis, our patient is being maintained under surveillance (clinical, biochemical and imaging at 6-month intervals), avoiding alcohol intake and hepatotoxic drugs. He has experienced no further complications at present.

References

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