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IMAGE | LIVER

Starry Liver: An Unexpected Diagnosis

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Case Report

A 65-year-old man presented with mildly elevated gamma-glutamyltranspeptidase (GGT) serum level on blood tests done for general check-up, and the finding of hepatomegaly and diffuse heterogeneity of the hepatic parenchyma on abdominal ultrasound. The patient's history included essential hypertension treated with angiotensin converting enzyme-inhibitors. The patient had no other features of the metabolic syndrome and did not drink alcohol. Physical examination revealed a mildly enlarged liver with a smooth, non-tender margin and a regular surface. Serum GGT was 75 IU/L (normal: 11-50 IU/L); liver enzymes and testing for viral and autoimmune hepatitis were normal. Abdominal ultrasound showed diffuse hyperechoic liver echotexture with multiple, small hypoechoic lesions in both hepatic lobes, particularly evident in the hepatic dome. It also showed one 8-mm gallbladder stone with no intrahepatic and main bile duct dilation, and no signs of portal hypertension. Contrast-enhanced computed tomography showed small, multiple, round and irregular hypodense lesions in both hepatic lobes that did not enhance with contrast. Magnetic resonance imaging (MRI) and MR-cholangiography showed the presence of numerous small (<1.0–1.5-cm diameter) liver lesions hypointense on T1-weighted scans and hyperintense on heavily T2-weighted scans, not communicating with the bile ducts, giving the liver a "starry sky" appearance (Figure 1). Findings were consistent with the diagnosis of bile duct hamartomas (Von Meyenburg complexes; VMC).

VMC are non-hereditary bile duct malformations that originate in the peripheral biliary tree, and may occur within the context of polycystic liver disease.[1] These interlobular benign cystic nodules are characteristically distributed throughout the liver at peripheral bile ducts below the liver capsule.[1] VMCs are usually asymptomatic, and may be incidentally diagnosed at liver biopsy, surgery or at autopsy with an incidence ranging between 0.6% and 2.8%.[1] Altered serum GGT can be found in these patients, although it is not diagnostic.[2] Abdominal ultrasound and computed tomography findings are non-specific, and although liver ultrasound imaging may show the "comet tail" sign, the findings at these imaging techniques are non-diagnostic and may suggest hepatic micro-abscesses, granulomas, or diffuse metastatic cancer.[3] Differentiation of VMC from these lesions may require resection or biopsy, although MRI and MR-cholangiography findings are useful to reach a definite diagnosis without invasive procedures. VMCs are hypointense on T1-weighted scans and hyperintense relative to liver parenchyma on T2-weighted scans, which differentiates these lesions from metastases. They lack communication with the biliary tree, which differentiates them from Caroli's disease.[3] VMC may rarely progress to cholangiocarcinoma, but, in the absence of complications, do not require any follow-up evaluation.[1,2]

Figure 1. T2-weighted, fat-saturated, coronal MR-cholangiography showing numerous small (<1.0–1.5 cm diameter) hyperintense liver lesions not communicating with the bile ducts, giving the liver a "starry sky" appearance.

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Disclosures

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