Biliary duct hamartomas in polycystic liver disease

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DESCRIPTION

A 45-year-old Caucasian woman, with no known liver disease, was referred to hepatology clinic for liver biopsy after abnormal image findings during a recent adult chicken pox infection. She presented with mildly elevated alkaline phosphatase of 170 U/L and γ -glutamyltransferase of 267 U/L. In clinic, she was asymptomatic with a negative review of systems. Initial laboratory evaluation, including a basic metabolic panel, complete blood count, aspartate aminotransferase, alanine aminotransferase and direct/indirect bilirubin levels were normal. Liver testing for viral hepatitis, primary biliary cirrhosis and haemochromatosis were negative. An abdominal ultrasound was significant for hepatomegaly with diffusely non-homogenous liver density and no gallstones or pericholecystic fluid. Follow-up MRI (figure 1) revealed extensive micronodularity. Subsequently, a liver biopsy was performed, revealing liver parenchyma with bile duct hamartomas and features of ductal plate malformation. No diagnostic intracytoplasmic inclusions,

iron deposition or significant fibrosis were seen with Periodic Acid-Schiff, iron and trichrome staining.

Biliary duct hamartomas, also known as von Meyenburg complexes, are benign lesions of the liver.¹ These lesions arise from a dilation of small intrahepatic bile ducts at ductal plate malformations within a biliary hamartoma. These changes result in a polycystic phenotype on imaging.² This is evident in our patient's imaging, which displayed extensive micronodularity on MRI. These lesions are often first identified incidentally on imaging or at autopsy as they are asymptomatic in most patients.³ An elevated ALP and GGT in our patient are an atypical finding for this condition,¹ but no other causes of cholestasis were identified.

Learning points

- ▶ Polycystic liver disease can be present in patients who are clinically asymptomatic, with minimal or no laboratory abnormalities.
- ▶ Ductal plate malformations are indicative of underlying polycystic disease.

Contributors AS, JH and RF contributed to this project as a team; portions were written and edited by each member of the team multiple times. RF, the attending physician, gave the final approval to this work.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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Figure 1 Markedly unusual appearance of the liver with diffuse micronodularity, as well as confluent areas of abnormal signal, an atypical presentation of multiple biliary hamartomas.

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