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CASE REPORT

An unusual polynodular liver disease: Multiple biliary hamartoma

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Summary We report a 73-year-old male patient who suffered from right upper abdominal dull pain, fever, and chills for 2 days. Gall bladder stones and acute cholecystitis were diagnosed, and parenteral antibiotics were used. The abdominal ultrasonography showed multiple tiny hyperechoic nodules over both lobes of the liver. His computed tomography scan revealed low-density lesions without contrast enhancement. Magnetic resonance imaging and magnetic resonance cholangiopancreatography further revealed lesions that were hypointense on T1-weighted images and hyperintense on T2-weighted images. Laparoscopic cholecystectomy and liver biopsy were performed, and biliary hamartoma was diagnosed based on the pathology. This report describes an unusual case of multiple biliary hamartomas and reviews the literature regarding the incidence, pathogenesis, and diagnosis of this disease.

Introduction

Multiple biliary hamartomas, also known as von Meyenburg complexes, were first described by von Meyenburg in 1918. They are rare, benign malformations consisting of multiple well-circumscribed collections of duct-like structures lined by the biliary epithelium and surrounded by the fibrous stroma [1]. Multiple biliary hamartomas are asymptomatic and usually found incidentally. Their clinical significance is that they may be easily confused with liver metastasis, microabscess, and other cystic liver diseases [2]. In this report, we present a patient with this unusual intrahepatic bile duct malformation.

Case report

A 73-year-old male patient was admitted to the Gastroenterology Department of Cheng-Ching General Hospital,

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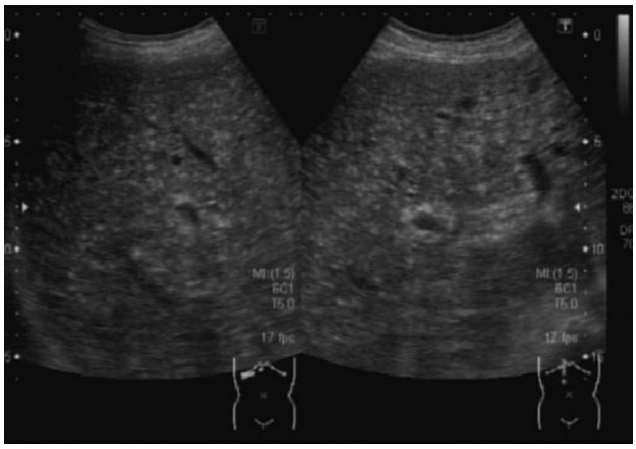

Figure 1 Abdominal ultrasonography showing multiple tiny hyperechoic nodules, 0.1—0.5 cm in diameter, distributed uniformly throughout the whole liver.

Taichung, Taiwan with fever and abdominal pain for > 2 days. The patient had a medical history of hypertension that was controlled by amlodipine (5 mg/d for > 20 years). This time, he suffered from right upper abdominal dull pain for 2 days prior to his admission. The painful sensation occurred nearly all day, and neither exacerbated nor relieved factors could be traced. There was no nausea, vomiting, or diarrhea but fever up to 39.2°C with chills occurred.

Upon admission, results of his physical examination revealed tenderness over the right upper quarter of the abdomen and positive finding of Murphy's sign. No jaundice was noted. His laboratory data showed leukocytosis with left shift (his white blood cell count was 18,500/mm^3, with segment 94.3%, lymphocyte 1.23%) and elevated C-reactive protein level (10.2 mg/dL). All other aspects of the complete blood count as well as liver function tests, bilirubin level, alkaline phosphatase, blood urea nitrogen, creatinine, electrolytes, and blood coagulation tests were within normal ranges.

An abdominal sonography was performed and revealed multiple small hyperechoic nodules, 1—5 mm in diameter, distributed uniformly throughout the liver (Fig. 1), multiple gall bladder stones, and thickness of gall bladder wall.

Further investigation with computerized tomography revealed multiple small low-density lesions over both lobes without contrast enhancement (Fig. 2) and suspicion of cholecystitis.

Then antibiotics including cephalosporin and aminoglycoside were administered. Magnetic resonance imaging (MRI) and magnetic retrograde cholangiopancreatography showed numerous tiny lesions ranging from 1 mm to 10 mm diffusely distributed in both lobes. These lesions had low signal intensity on T1-weighted images and increased signal intensity on T2-weighted images. The signal intensity on T2-weighted images was slightly less than that of simple fluid (Fig. 3). No communication of these lesions and biliary tree was found by MRCP (Magnetic Resonance Cholangiopancreatography).

A surgeon was consulted for laparoscopic cholecystectomy and liver tumor biopsy. During the operation, multiple whitish lesions, 2—3 mm, scattered diffusely over the liver surface was noted, and biopsy and cholecystectomy were performed. The pathological report showed multiple biliary channels lined by the regular cuboidal epithelium with dense fibrous stroma (Fig. 4). A diagnosis of biliary hamartoma was made.

The patient was discharged 5 days after the surgery. A follow-up abdominal ultrasonography done 6 months later showed no significant changes in the biliary tree.

Discussion

Biliary hamartomas are uncommon benign biliary malformations. The prevalence rates of biliary hamartomas are 0.69% [1] and 5.6% [3] in two large autopsy series. They are typically grayish-white nodules, 0.1—0.5 cm in diameter, and are well circumscribed, unencapsulated lesions usually scattered throughout both lobes of the liver, although they may aggregate [1,3]. Histologically, they are multiple collections of disorganized or dilated ducts or ductules with bile or not, embedded in fibrous stroma, without cellular atypia [3]. There is a classification of biliary hamartomas based on the degree of hamartoma consistency/biliary dilatation, as follows: Class 1, predominantly solid pattern with narrow bile channels; Class 2, intermediate pattern; and Class 3, marked cystic dilatation of bile ducts within the lesions [4]. With the different pattern of histology, biliary hamartomas may exhibit different echogenecity on ultrasonography. For example, Class 1 biliary hamartomas may be hyperechoic lesions by ultrasonography.

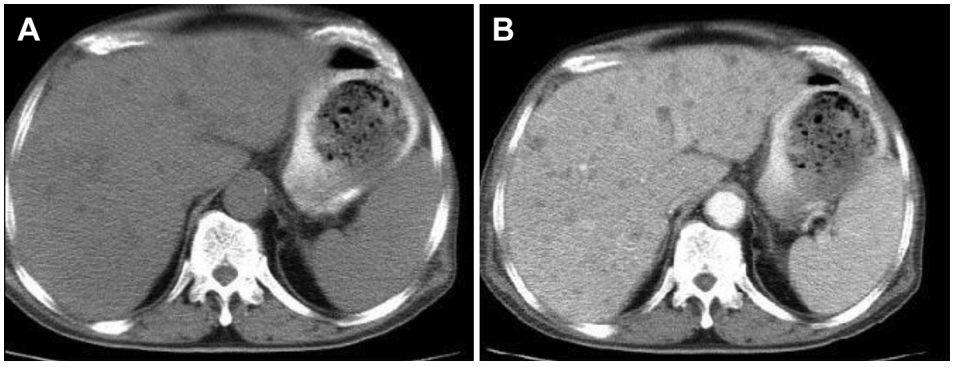

Figure 2 Abdominal computed tomography reveals (A) multiple low density lesions (B) without contrast enhancement.