ABSTRACT

Multiple biliary hamartomas (MBHs) are a rare source of cystic lesions of the liver that radiologic detection is difficult, because of its tiny size. We report the case of a 64-year-old woman, with recurrent abdominal pain. The ultrasonographic configuration in this patient showed multiple cystic hepatic lesions. However, the diagnosis of MBH was made by magnetic resonance imaging (MRI) and magnetic resonance cholangiopancreatography (MRCP). A regular follow-up of patient presenting this benign disease is recommended because of its potential for malignant conversion.

Keywords: Hamartoma, Magnetic resonance imaging, Magnetic resonance cholangiopancreatography, Multicystic biliary

INTRODUCTION

Biliary hamartoma (BH), also known as Von Meyenburg complex, is a rare benign hepatic tumor. It is characterized by cystic dilatation of the bile duct surrounded by fibrous stromal elements. Its diagnosis is usually difficult because it is found incidentally and presented as small multiple nodules. It can be confused with other liver diseases like malignant tumors. Therefore, the development of imaging modalities has eased its diagnosis and decreased the need for invasive methods like biopsy. Several case reports have identified BHs.

CASE REPORT

The responsible service received a 64-year-old woman with three months of acute epigastric pain. The initial examination was normal. Laboratory analysis found a slight elevation of gamma-glutamyl transferase (133 IU/L) for reference range understood between 10 and 40 IU/L. However, other biologic values were normal. An abdominal ultrasonography (US) was performed by a generalist showing multiple small cystic hepatic lesions. In our service, subsequent MRIs revealed multiple small liver lesions that were hyperintense on T2-weighted images (Figure 1) and hypointense on T1-weighted images without enhancement after administration of gadolinium (Figure 2). Magnetic resonance cholangiography showed multiple small cystic liver lesions distributed through right and left lobes of the liver, without communication with the draining bile ducts which had normal appearances. That created a “Starry sky” appearance (Figure 3). Depending on US and MRI typical results, the patient was diagnosed with BH, without needing of histological confirmation.

DISCUSSION

Various sorts of cystic lesion could be found in the liver, like BH. This rare disease, which was defined by Von Meyenburg in 1918 [1], is a benign congenital condition,
without clinical significance and with usually incidental diagnosis. Biliary hamartoma arises as a consequence of ductal plate malformations, including interlobular bile ducts. It is consisted of disorganized clusters of dilated bile ducts, which are ceiled with cuboidal epithelium and bordered by fibrosis stroma [2–4], that conduct to cystic dilatations of duct-like formations, uniformly distributed in the liver.

Biliary hamartoma could be isolated, but it is recognized by its association with Caroli syndrome, autosomal dominant polycystic kidney and liver diseases, congenital hepatic fibrosis. It constitutes a risk factor for hepatocellular carcinoma and cholangiocarcinoma [5–7].

The US represents usually the first line imaging technique used in patient with BH, but it is considered as insufficient, because lesions could appear hypoechoic, hyperechoic, or even heteroechoic lesions with variable size and number and with comet-tail sign. This aspect could mimic biliary stones, microabscesses, or metastases [4, 8, 9]. Computed tomography (CT)-scan configuration of BH could be also ambiguous because of the tiny size of lesions, with irregular circumferences and humble attenuation [8, 10].

Magnetic resonance imaging and MRCP are esteemed as the cornerstone imaging modality to determine the diagnosis of BH. They showed cystic lesions hyperintense on T2, without enhancement after gadolinium. Those hamartomas do not communicate with the bile ducts and had a scattered distribution in the liver, realizing the aspect of “starry sky” [11].

Considering the risk of cholangiocarcinoma development, portal hypertension, and jaundice in a patient with BH, a regular follow-up is necessary and liver biopsy is required if a neoplastic process is suspected [12].

CONCLUSION

Biliary hamartoma is a rare asymptomatic disease. Thanks to MRI and MRCP. The recognition from other liver lesions like metastasis and Caroli syndrome has become less difficult. This benign condition has a potential for malignant conversion, where the need of a long-term follow-up is required.

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Authors declare no conflict of interest.

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All relevant data are within the paper and its Supporting Information files.

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