BILIARY HAMARTOMA

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Abstract

Biliary hamartoma or von Mayenburg complex (VMCs) is a rare benign congenital malformation of the biliary duct. Patients are usually asymptomatic. Hepatic lesions are incidentally discovered on ultrasonography (US) and subsequent radiological methods are necessary for confirmation. A correct diagnosis is established when typical imaging findings are present, otherwise histological confirmation might be needed.

Keywords: Biliary hamartoma; von Mayenburg complex; Ultrasonography; MRI; CT.

Case presentation

A 71-year old Caucasian male with dilative cardiomiopathy and irrelevant family history was admitted to our hospital for the evaluation of a suspected liver disease. The physical examination was within the normal limits, except for hepatomegaly. Laboratory findings revealed a slight anemia (hemoglobin level of 12,5 g/dl, hematocrit of 38%, red blood cells count of 3,9 mil/mm³). White blood cells count was 8730/mm³ and platelet count was 258000/mm³. Biochemical test showed normal serum levels of alanine aminotransferase (ALT=20 IU/L), aspartate aminotransferase (AST=15 IU/L) and alkaline phosphatase (ALP=120 IU/L) and elevated gammaglutamyltranspeptidase (GGT=230 IU/L). Alpha-fetoprotein and carcinoembryonic antigen were within normal limits. Ultrasound showed multiple small hypo- and hyperechoic lesion foci, some cystic lesions with comet-tail echoes, the biggest cyst measured 8 mm in the VII[th] segment (Fig.1). CT scan revealed multiple small cystic lesions; the largest hypodense nodule was in the VII[th] hepatic segment with no peripheral or central enhancement (Fig. 2). The CT examination was not conclusive and a MRI with MRCP was performed next. Multiple small cystic lesions were detected with T1 hyposignal and T2 hypersignal, the largest being in segment VII (Fig. 3). Corroborating data from these imaging techniques with 6 month follow up, the final diagnosis was biliary hamartoma (complex von Meyenburg).

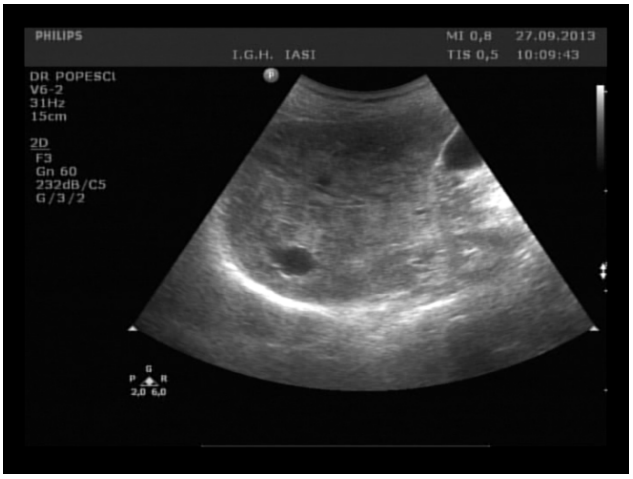

Figure 1. B-mode US scan showed some cystic lesions, the biggest in with 8-mm in the VII[th] segment.

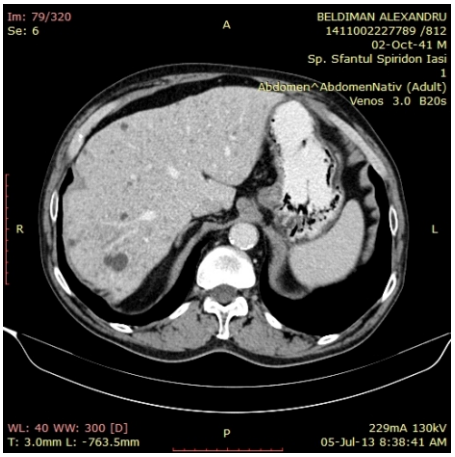

Figure 2. CT scan portal phase. After contrast enhancement, there was no enhancing portion of the lesion.

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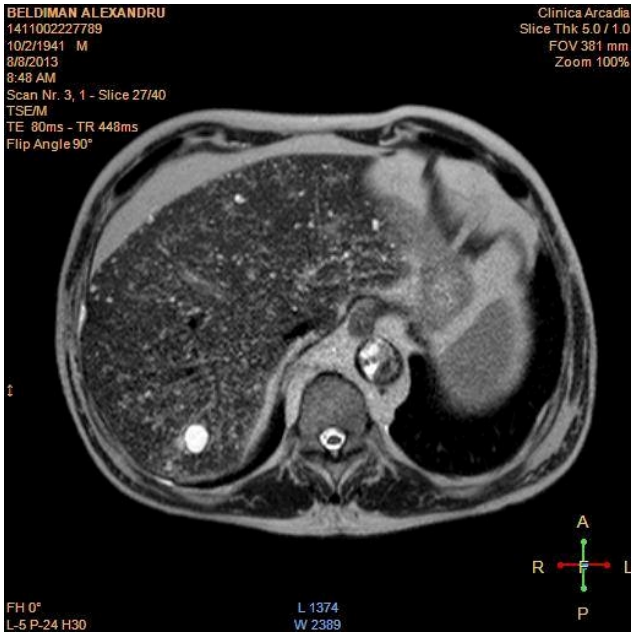

Figure 3. MRI scan. T2 hypersignal cystic lesion.

Discussion

Biliary hamartoma is a benign congenital malformation of the biliary duct which was described for the first time in 1918 by von Mayenburg [1], therefore it is also called von Mayenburg complex (VMCs). Although jaundice and portal hypertension may arise as a result of mass effect, patients are usually asymptomatic [2]. The biliary hamartomas may be single or multiple, with size ranging between 1 and 15 mm [3]. Due to the small size of the lesions, final description is difficult. The autopsy prevalence ranges from 0.6 to 2.8% [4]. Histologically, it consists of disorganized and dilated bile ducts and ductules surrounded by fibrous stroma [5]. US imaging shows hypoechoic, hyperechoic or mixed heterogenic echoic structures [1,3,4]. The multiple comet-tail sign is thought to be the specific US finding of VMCs (6). It has been suggested that lesional echogenicity may depends on the number and size of the dilated bile ducts and on the degree of fibrosis [5]. On contrast enhanced CT, biliary hamartomas are usualy of low attenuation and may have irregular margins. The majority of cases reported suggest that VMC does not show contrast enhancement [5,6]. On MRI, VMCs are described as hypointense on T1 and hyperintense on T2 in comparison with surrounding liver parenchyma [5,7]. Although biliary hamartoma is a benign condition, there are some isolated reports of hepatic malignancies on a background of VMC, including hepatocellular carcinoma and cholangiocarcinoma [8,9].

Biliary hamartomas usually presents as multiple small nodules and despite the fact that they are rare, they may be confused with liver metastatic disease, microabscesses, diffuse primary hepatocellular carcinoma, biliary cysts or Caroli's disease [1,3,4].

In conclusion, associating different imaging modalities with the follow-up are very useful in the diagnosis of biliary hamartoma. A correct diagnosis is established when typical imaging findings are present, otherwise histological confirmation might be needed.

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