Multiple Biliary Hamartomas (von Meyenburg complexes) – A Case Report

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Abstract

Multiple biliary hamartoma (MBH) is a rare cause of multiple benign hepatic lesions. The condition is also known as von Meyenburg complexes, multiple bile duct hamartomas and biliary microhamartomas. MBH is asymptomatic and usually found incidentally where it is important to differentiate from other causes of multiple liver lesions particularly liver metastases. Histologically, they consist of cystic dilatation of the bile duct, encompassed by fibrous stroma. We reported a 74 year old male patient with multiple hepatic lesions which ultrasonographic appearance suggested multiple hepatic cysts. The diagnosis of MBH was made by MRCP of whole abdomen. Recognition of this unusual lesion is essential to avoid confusion with other cystic tumors of the liver specially liver metastases and to learn more about its natural history and response to treatment.

Keywords

Hamartoma, Liver cysts, Von Meyenburg complexes.

Introduction

Multiple biliary hamartomas (MBH) are a rare cause of multiple benign hepatic lesions. The condition is also known as von Meyenburg complexes, multiple bile duct hamartomas and biliary micro-hamartomas. MBH is asymptomatic and usually found incidentally, where it is important to differentiate from other causes of multiple liver lesions, particularly metastases. MBH is known to be associated with autosomal dominant polycystic kidney disease (ADPKD) and polycystic liver disease. Several case reports have identified possible malignant transformation of biliary hamartomas into cholangiocarcinoma and hepatocellular carcinoma.

Case Report

A 74 year old gentleman came to Gastroenterology OPD of Apollo Hospitals Dhaka with the complaints of abdominal pain associated with vomiting for 10-12 days. On admission, he was conscious, oriented, icteric, his vitals were stable, abdomen was soft and mildly tender. His biochemical investigations reaveled elevated ALT (130 IU/L), Alkaline Phosphatase (207 U/L), GGT (727 IU/L), serum amylase (529 U/L) and serum creatinine (1.65 mg/dl). CA-19.9 and CEA were within normal limits. He also underwent endoscopy which revealed mild

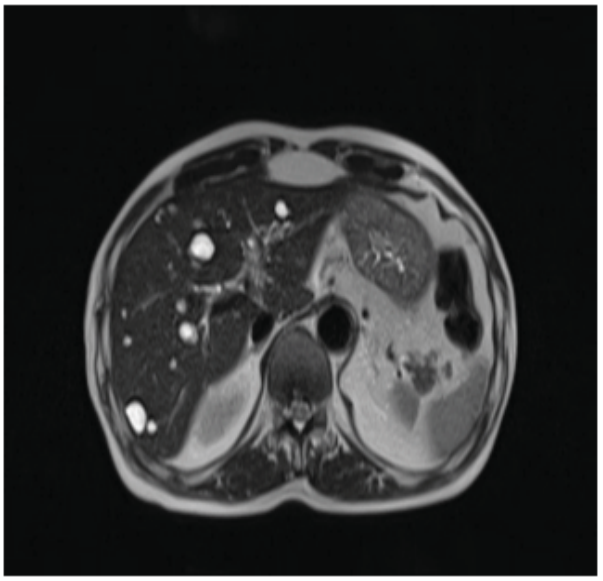

Fig. 1: T2W axial scan

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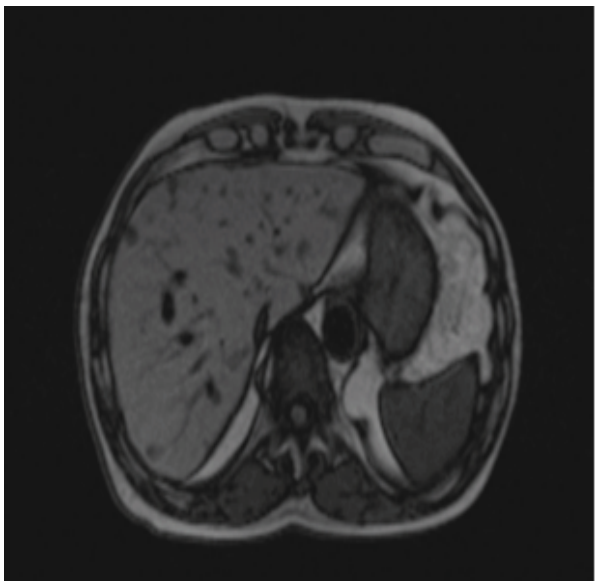

Fig. 2: T1W axial scan

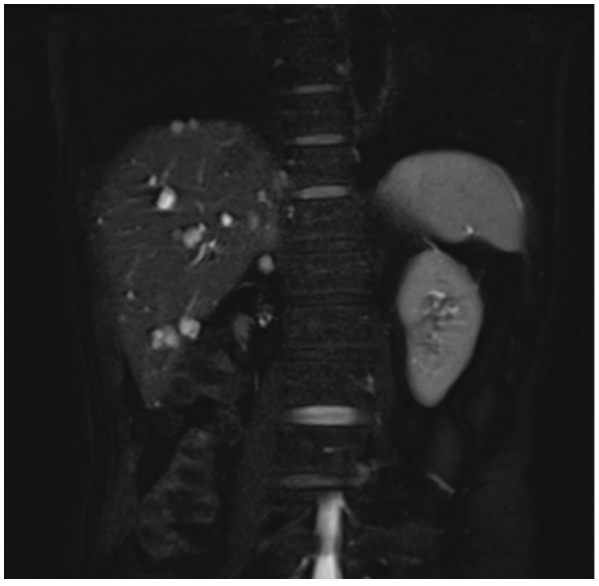

Fig. 3: T2W coronal scan

erosive gastritis with coarse papilla, colonoscopy revealed no polyp, no ulcer or growth. He had done ultrasonography of whole abdomen outside AHD which was reported as ? multiple cysts in both lobes of liver and bilateral renal cortical cysts. From Gastroenterology department he was sent to Radiology department to undergo MRCP which revealed multiple biliary hamartomas with mildly dilated CBD, sludge in

Fig. 4: MIP image

the lumen of Gallbladder, bilateral renal cortical cysts (Fig. 1- 4). ERCP was done which revealed mildly dilated CBD with sludge, papillotomy was done without any complication. Histological examination could not be done as the patient could not afford to undergo surgery. The patient was treated conservatively and discharged after he was haemodynamically stable. Fig (1-4) showing numerous cysts of variable sizes within hepatic parenchyma diffusely distributed throughout both lobes along the intrahepatic biliary channels resembling "starry sky"configuration with mildly dilated CBD.

Discussion

Multiple biliary hamartomas (MBH) is a rare cystic disease of the liver[1]. The condition is also known as Von Meyenburg complexes and biliary microhamartomas. MBH is asymptomatic and usually found incidentally, where it is important to differentiate from other causes of multiple liver lesions, particularly metastases. The prevalence of MBH is about 3% at autopsy, however prevalence on imaging is <1% as most

hamartomas are <5mm in size and often not detected. MBH is reportedly three times more common in women than men.[2] Biliary hamartomas are comhamartomas are composed of small disorganised clusters of dilated cystic bile ducts lined by a single layer of cuboidal cells and surrounded by an abundant fibrocartillageneous stroma.[1,6,7] Although they may communicate with the biliary tree, they generally do not. They are thought to arise from embryonic bile duct remnants that have failed to involute. MBH is known to be associated with autosomal dominant polycystic kidney disease (ADPKD)[3], polycystic liver disease[3], Caroli's disease, congenital hepatic fibrosis, cholangiocarcinomas and cholangitis.[10,11] VMCs can be misdiagnosed on ultrasonography with interpretation as metastatic hepatic disease.[12] In ultrasonography the lesions may appear hyperechoic or hypoechoic nodules with poorly or well limited margins.[11,13-15] Biliary hamartomas are hypoattenuating and often show no enhancement.[6] On MRI most biliary hamartomas are hypointense compared to liver parenchyma on T1 and hyperintense on T2. Usually no enhancement or thin peripheral enhancement after giving contrast.[13,14] In angiography, they are seen as grape like clusters of abnormal vascularity with contrast persisting into the venous phase.[9] Nuclear scan shows delayed uptake and delayed emptying of tracer within larger biliary hamartomas.[11] The definitive diagnosis of these lesions can be confirmed by liver biopsy. Histologically VMCs include dilated small bile ducts, surrounded by fibrous stroma. Microscopically, they are characterized by abundant fibrous stroma with cystic dilatations of the bile duct and sometimes with associated periductal glands.[17] VMCs are usually found along portal tract.

Conclusion

Aside from possible risk of malignant transformation, multiple biliary hamartomas are benign asymptomatic condition with no long term consequences and no treatment is required. Differentiation from liver metastases may require resection by wedge biopsy. However, MRI helps differentiate them from metastases. Bile duct hamartomas do not enhance after intravenous contrast but metastatic lesions do. Bile duct hamartomas can also be differentiated from Caroli's disease by their lack of communication with the biliary tree. It has been suggested that similar multiple, small, non enhancing cystic lesions in the liver without renal involvement on CT and MRI are highly suggestive of biliary hamartomas, thus avoiding the need of risk of biopsy procedure.[5]

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