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Case Report

Multiple hamartomatosis of the biliary tract: case report

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ABSTRACT

Von Meyenburg complex or multiple biliary hamartomatosis is a rare clinical diagnosis, with a reported prevalence of up to 5.6% in large autopsy series. Since this type of pathology often lacks clinical symptoms, it is usually an incidental finding. We present the case of an 85-year-old female referred to our unit for persistent jaundice, abdominal pain and weight loss. The diagnosis and management of this pathology is often a challenge, the natural history of these cases remains unknown, and neither follow-up nor treatment protocols have been developed in patients with multiple biliary hamartomatosis. Multiple biliary hamartomatosis is a rare pathology which has no specific treatment, however, knowing about it allows a good differential diagnosis with obstructive and parenchymal hepatic pathology.

Keywords: Biliary hamartomatosis, Hepatomegaly, Von Meyenburg, Jaundice

INTRODUCTION

Von Meyenburg complexes or also known as biliary hamartomas, described by Hanns von Meyenburg in 1918, are rare benign malformations of the intrahepatic bile ducts considered part of the spectrum of fibro polycystic liver disease arising from abnormal embryologic development of the ductal plate.[1,2]

Von Meyenburg complex is a rare clinical diagnosis, with a reported prevalence of up to 5.6% in large autopsy series. Since this type of pathology often lacks clinical symptoms, it is usually an incidental finding; on rare occasions it may cause fever, jaundice or abdominal pain and elevations of transaminases, alkaline phosphatase or gamma-glutamyl transferase (GGT) may occur.[3]

The rare nature of this condition, as well as its heterogeneous clinical presentation can make its management difficult, so it is important to recognize biliary hamartomas and distinguish them from other pathologies. In Mexico there is no case reported in the literature, so we present a case of Von Meyenburg complex in a third level hospital in Mexico City.

CASE REPORT

An 83-year-old female patient referred to our unit with the diagnosis of probable liver tumor. She has a history of Systemic Arterial Hypertension of long evolution, denies any history of neoplasia. She started 1 month ago with jaundice, hyporexia and tendency to somnolence, as well as weight loss of 4 kg in 1 month. She went to the doctor where antibiotic therapy was given without improvement, for which reason an imaging study was performed documenting structural alterations related to large cysts that conditioned extrinsic compression of biliary tract. On examination with generalized jaundice, depressible soft abdomen, not painful on palpation, with hepatomegaly, without data of peritoneal irritation. Her laboratories report: glucosa-97, urea-51, BUN-23.8, creatinine-0.62, BT- 26, BD-9.7, BI-6.3, ALT-144, AST-181, FA-1170, GGT-628, DLH-194, ALB-1.5, amylase-79, lipase-43,

CA-8 NA-134.1, K-3.67, CL-99.1, P-2.6, MG-1.9, Hb-8.8, HTO-25.1, LEU-7.15, NEU-68%, PLQ-177,000, TP-15.8, INR-1.51, TTPA-39.4. Magnetic resonance cholangiography is performed to which reports liver with increased size secondary to the presence of multiple lesions of cystic behaviour of diffuse distribution throughout its parenchyma and communicating with the intrahepatic bile duct which is also observed dilated, suggesting that these lesions are dependent on the bile duct, the largest cystic lesion covers the hepatic segment v, presents liquid-liquid level (hypointense in t2) suggesting data of bleeding in chronic phase. Its approximate dimensions are 10.8×8.4×8.2 cm; it displaces the gallbladder to the right and inferiorly, which has heterogeneous content due to multiple lithos in its interior. the extrahepatic biliary tract cannot be visualized due to an important increase of the hepatic size that compresses and displaces the adjacent structures, compatible with multiple biliary hamartomatosis (Figure 1).

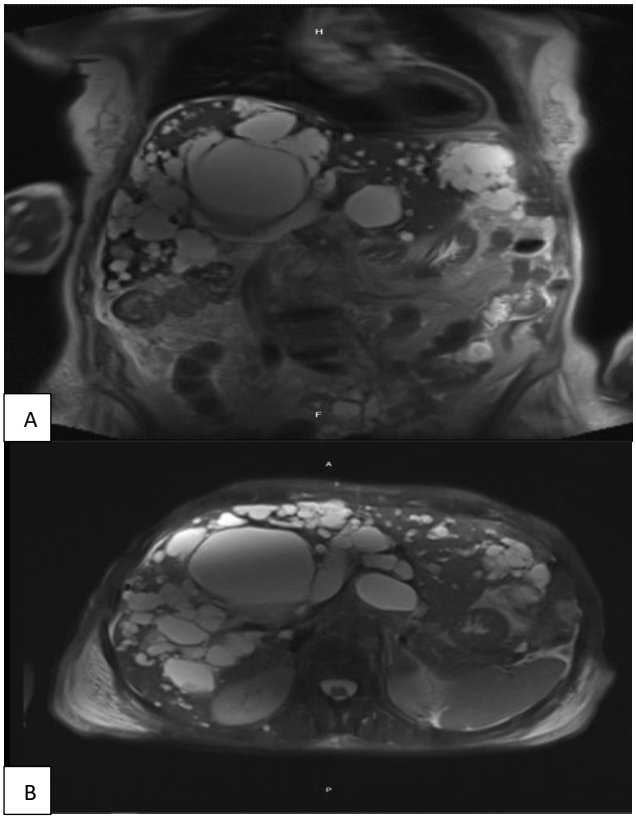

Figure 1 (A and B): Magnetic cholangioresonance en T2 sequence. Liver with multiple hyperintense cystic lesions throughout the parenchyma.

DISCUSSION

This type of pathology represents a rare clinicopathological entity, typically asymptomatic and without alterations in hepatic parameters. Its incidence in autopsies is estimated at around 5.6% in adults and 0.9% in children.[4] Intrahepatic bile ducts begin to form in the first 7 weeks of embryological life. The portal vein grows inward from the hepatic hilum and begins to branch, accompanied by a layer of epithelial cells surrounding each portal venous branch, this is known as the ductal plate, these cells have the ability to differentiate into hepatocytes or cholangiocytes. Von Meyenburg complexes represent malformations of this ductal plaque responsible for the development of most peripheral intrahepatic ducts.[1]

Often, patients do not present with symptoms and the lesion is usually discovered accidentally. A review found that only 36% of patients were symptomatic at presentation, suggesting that the majority (almost 2/3) of patients are asymptomatic Among symptomatic patients, abdominal pain was the most frequent symptom. Other frequently reported symptoms were fever (18.0%), weight loss (14.0%) and jaundice (14.0%). The time from symptom onset to disease presentation varied from study to study and ranged from 1 day to 10 years.[3]

On microscopic histopathology, they appear as minute lesions consisting of malformed bile ducts of variable caliber in a densely collagenized stroma. The ductal structures vary from narrow to markedly dilated, are often irregularly shaped and frequently contain inspired bile. Extreme ductal dilatation may result in a cystic appearance large enough to be observed macroscopically. They are usually lined by a single layer of cuboidal, columnar or flat epithelium that lacks cytologic atypia. The connective tissue stroma in which are immersed in the ducts is denser compared to normal portal tracts and often appears hyalinized. Calcification may be observed.[1]

Von Meyenburg complexes are small lesions, and often go undetected on radiological examinations including ultrasound, computed tomography (CT) and magnetic resonance imaging (MRI).[5] Advances in radiology have made them easily detectable, providing a more accurate diagnosis to avoid biopsy, which must be performed to confirm the diagnosis in case of doubt. Using USG the features of CMV are multiple small hyperechoic lesions associated with small cysts and comet-tailed echoes that are often only visualized after use of a zoom function, with the use of CT multiple, nodular and irregular hepatic cystic lesions, smaller than 15 mm, with low attenuation and no non-enhancement with contrast injection can be detected.[6,7]

MR images of the liver parenchyma are of special importance, most biliary hamartomas are hypointense on T1 compared to the liver parenchyma, if gadolinium is added they usually have no enhancement, sometimes there may be enhancement of the thin peripheral border which may represent compressed normal liver parenchyma, they present hyperintense on T2.[8] The differential diagnosis of MVCs is quite broad due to their non-specific appearance**.**

The natural history of these cases remains unknown, and no follow-up protocols have been developed in patients

with multiple CMVs. At present, CMVs do not require treatment or follow-up imaging tests.[1]

CONCLUSION

Multiple biliary hamartomatosis is a rare pathology which has no specific treatment, however, knowing about it allows a good differential diagnosis with obstructive and parenchymal hepatic pathology.

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