LETTER / *Gastrointestinal imaging*

Imaging features of ciliated hepatic foregut cyst



Keywords: Ciliated foregut cyst; Liver; Magnetic resonance imaging

Ciliated hepatic foregut cyst (CHFC) is a very rare, benign and solitary cyst [1,2]. To our knowledge, five cases of CHFC were reported in the 19th century and 88 cases in the 20th and 21st centuries. Its medical imaging findings are seldom reported [1,3]. Sometimes it is difficult to differentiate the CHFC from other tumors in particular malignant tumors [1,4,5]. In the literature, the majority of cysts are situated in the left lobe in a subcapsular location. We recently encountered a patient with CHFC. This cyst was located next to the gall bladder which is a situation rarely described in the literature in four cases [6]. Radiology in this case is not specific and posed a problem of differential diagnosis especially gallbladder duplication. The diagnosis was established after laparoscopic resection and an extensive review of the literature on this condition was carried out.

A 19-year-old woman presented with a 9-month history of right upper quadrant pain. There was no associated jaundice, rigors or weight loss. Apart from minimal right upper quadrant tenderness, examination was unremarkable. Full blood count, liver and renal function tests and clotting profile were normal. Sonographic examination revealed a well-delineated oval hypoechoic mass, 55 mm × 38 mm × 34 mm in size. It was located in the medial segment of the left lobe (segment IV). This one was the seat of hyperechoic declive sediment. It was adjacent to the gallbladder without obvious communication (Fig. 1A). Diagnosis

of gallbladder duplication was discussed. So that repeat non-fasting ultrasonographic examination didn't show any changes in shape and volume whereas the gallbladder had collapsed (Fig. 1B). Magnetic resonance (MR) imaging was performed for more detailed examination. The lesion appeared hypointense relative to surrounding liver parenchyma on T1-weighted imaging and markedly homogeneously hyperintense on T2-weighted imaging (Fig. 2A). It was also not enhanced after Gd-DTPA administration (Fig. 2B). This mass was the seat of sediment, which was hyperintense on T1-weighted imaging and hypointense on T2-weighted imaging (Fig. 2A). No direct communication with the biliary ducts was visible on the MR-cholangiography (Fig. 3). As well, no enhancing of the wall of cyst after Gd-DTPA administration or intramural nodules was noted. Otherwise we noted the presence of an angioma of the segment VI and three biliary cysts measuring respectively 12 mm, 9.6 mm and 11 mm on the segments VI, IV and II. Among the differential diagnosis were biliary cystadenoma, lymphangioma and hemorrhagic biliary cyst.

Therefore, in the absence of etiological diagnosis, cholecystectomy with en-bloc excision of the mass was carried. A well-encapsulated mass was identified in the gallbladder fossa and was dissected from the liver bed (Fig. 4). There were no apparent vessels or bile ducts directly communicating with the mass. The patient was discharged on the second postoperative day. Postoperative course was uneventful.

Histological studies of the surgical specimen revealed a fibrous wall lined by ciliated pseudo-stratified columnar epithelial cells (Fig. 5). The final diagnosis was a ciliated hepatic foregut cyst.

The ciliated hepatic foregut cyst consists of a fibrous cyst wall-lined by pseudo-stratified ciliated columnar epithelium

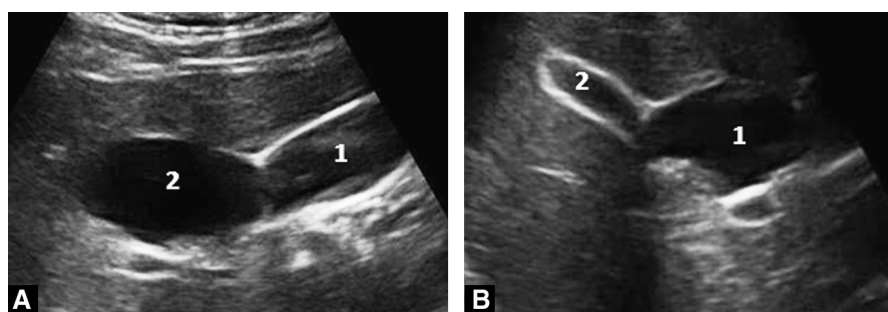


Figure 1. A. Fasting ultrasonography. B. No fasting ultrasonography. Ultra-sonography shows an anechoic well-defined lesion with hyperechoic declive sediment; this lesion did not change in shape and volume on non-fasting ultrasonographic exam. 1: CHFC, 2: gallbladder.

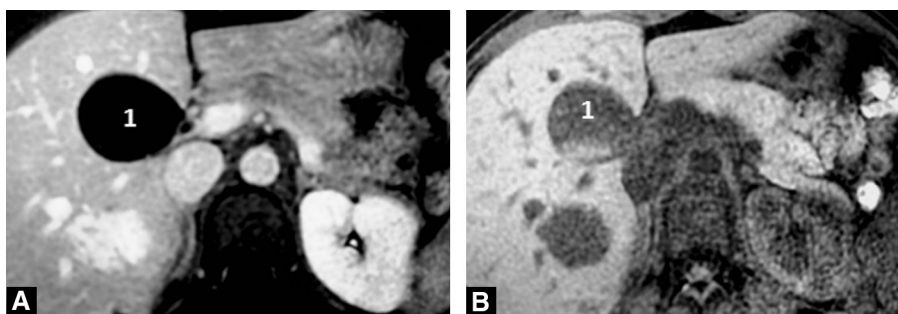


Figure 2. Axial T2 SSFSE TR: 2300 ms TE: 118.27 ms. Axial MR T1 (A) and T2 (B) weighted images shows a well-defined cystic mass which is hypointense on T1 and markedly hyperintense on T2 the seat an hyperintense declive sediment; this mass is adjacent to the gallbladder. 1: CHFC, 2: gallbladder.

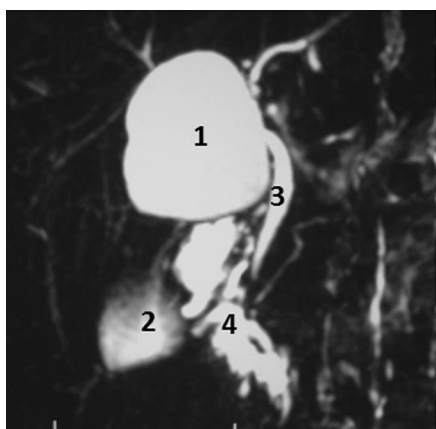


Figure 3. MR-cholangiography (radial 2D-T2 SSFSE- TR: 8000 ms, TE: 1045.82 ms). MR cholangiography shows no communication with the biliary duct. 1: CHFC, 2: gallbladder, 3: biliary duct, 4: duodenum.

and is characterized by the presence of bundles of smooth muscle [2]. It arises from the embryonic foregut in the liver [1]. It is a detached hepatic diverticulum or abnormal trachea-bronchiolar bud that may have migrated caudally to be included with the liver, prior to the closure of the pleuro-peritoneal canals by the end of the eighth week of development [7]. The first case was reported by Friedreich in 1857 [8].

Clinically, the majority of patients were asymptomatic and lesions were found incidentally on radiologic imaging. The most common symptoms include vague right upper quadrant pain as reported in our case [9]. It most commonly

presents in the 4th decade of life, more frequently in men and has a greater occurrence in the right hepatic lobe (with a strong predilection segment IVa).

The preponderance of cases in which CHFC arises in segment IV can be explained by the fact that the left hemiliver, in particular segment IV, accounts for the bulk of the hepatic mass during weeks 4–6 of gestation, during which time these lesions likely arise from detached out-pouchings of the hepatic diverticulum or adjacent foregut during development [10]. In our case, the cyst was developed at the segment IVb next to the gallbladder, this situation is rare and it is reported in 3,6% of cases in the literature [6].

Because of increased availability of radiology, CHFC is increasingly diagnosed. But its diagnosis is often confounded by the radiographic difficulty in distinguishing a benign CHFC from a neoplastic process. Ultrasonography usually shows an anechoic to slightly hypoechoic lesion that may contain spotty hyperechoic areas as reported in our case [5,11]. An unenhanced CT scan also displays these lesions as hypodense in nature in the majority of the patients. A feature that was commonly observed in almost all the reported cases was the absence of enhancement on contrast injection. Although almost all cysts display a high-intensity signal on T2-weighted MRI, T1-weighted imaging has no characteristic signal intensity. The most common combination of imaging features is a unilocular hypoechoic mass on ultrasound that is hypodense on CT without septations or vascular enhancement. Commonly considered diagnoses include other unilocular hepatic lesions, such as simple hepatic cyst, parasitic (echinococcal) cyst, epidermoid (or endometrial) cyst, pyogenic abscess, intrahepatic choledochal cyst, mesenchymal hamartoma, hypovascular solid

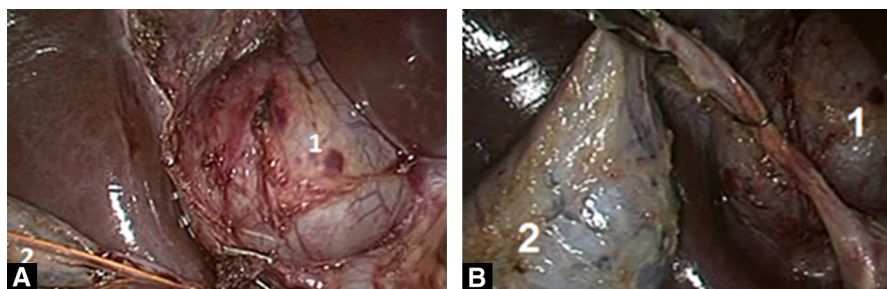


Figure 4. A. Laparoscopic view of CHFC (1) after dissection of the gallbladder (2). B. Intra-operative photograph demonstrating (1) ciliated hepatic foregut cyst, (2) gallbladder dissected from the bed and with accompanying cystic duct.

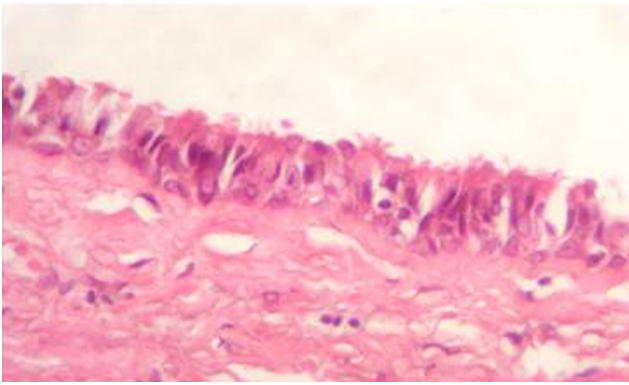


Figure 5. High magnification view ($\times 400$) showing the columnar epithelium with cilia.

tumor, and hepatobiliary cystadenoma or cystadenocarcinoma [12,13]. In this case, cystadenoma may be included in the differential diagnosis because the lesion may be malignant. Differential points are that cystadenomas are usually multilocular and sometimes reveal mural nodules, and these features are well revealed on radiologic imaging but not enough to confirm the diagnosis, which is histological.

CHFC are generally considered benign non-neoplastic processes, however cases showing the development of malignancy and a fatal course have been described; the risk of malignant transformation is about 3% [14]. Thus, hepatic cystic lesions should be removed surgically. The key histological features of CHFC are the presence of a four-layered cyst wall, which consists of a mostly inner lining of pseudostratified columnar epithelium followed by a layer of loose subepithelial connective tissue, smooth muscle layer, and an outer layer of dense fibrous tissue [15].

Conclusion

In conclusion, ciliated hepatic foregut cyst is an increasingly frequently diagnosed condition. Imaging alone is non-diagnostic *per se*. When a well-demarcated lesion in the left hepatic lobe present the radiological aspect described above, the diagnosis of CHFC should be considered among others diagnosis and due to the risk of malignant transformation and potential confusion with other benign and non-benign conditions, surgical resection is warranted.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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