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Case Report

Ciliated Hepatic Foregut Cysts: About A Case and Review of the Literature

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Abstract

Ciliated hepatic foregut cysts (CHFCs) are a benign cyst of the liver with a ciliated coating. It is an extremely rare solitary cystic lesion. In this paper, we present a case of CHFCs in a 41-year-old woman. Our goal was to insist by the report of this clinical observation and the review of the literature on the main issue of this pathology which is to ensure a regular and rigorous monitoring of patients with CHFCs or a radical surgical treatment to anticipate or avoid malignant complications.

Keywords: Ciliated hepatic foregut cysts; Ultrasound; CT; Magnetic resonance imaging.

INTRODUCTION

Ciliated hepatic foregut cysts (CHFCs) is a benign cyst of the liver with a ciliated coating. It is an extremely rare solitary cystic lesion with only about 100 cases reported in the literature since 1857. However, the detection rate of this tumor has increased over the last decades, due to the increasing use and progress of radiological complementary examinations. CHFCs is due to a migration of cells of the foregut from the ventral bud (origin of the trachea and esophagus) to the dorsal bud (origin of the liver) during embryonic life. The long-term evolution of these cysts is still poorly understood with four reported cases of malignant degeneration. The challenge is therefore to ensure regular and rigorous monitoring of patients with CHFCs or radical surgical treatment to anticipate or avoid malignant complications. In this article, we present a case of CHFCs in a 41-year-old woman.

OBSERVATION

In March-July 2020, a 41-year-old woman, married and mother of two children, housewife, was admitted to our hepato gastroenterology department, for diagnostic and therapeutic management of abdominal pain in the right hypochondrium with discovery of a hepatic lesion on ultrasound done in ambulatory.

Our patient had no notable personal or family history.

The history of his illness goes back to one year after his admission with the progressive installation of an abdominal pain in the right hypochondrium in hemi-girdle towards the right scapula, cramp-like, of minimal to moderate intensity rated at 4/10 on the pain scale, without aggravating or calming factors and without associated signs. Thus, it was an isolated hepatic colic type pain evolving in a context of apyrexia and conservation of the general state.

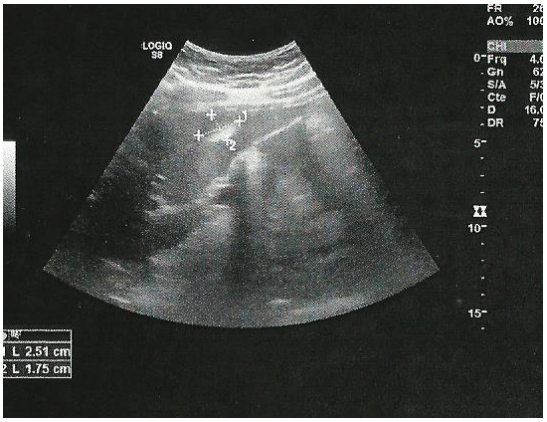
The physical examination found a conscious patient well oriented in time and space with a Glasgow Coma Score (GCS) equal to 15 out of 15, hemodynamically and respiratory stable (eupneic, normocardial and normotensive), The body mass index (BMI) was equal to 34 kg/m2 which reflected an obesity and waist circumference was increased with a value equal to 115 cm, the temperature was 37°C, the conjunctiva were normally colored with absence of signs of hepatocellular insufficiency. The abdominal examination showed abdominal distension without CVC or abnormal dullness or abdominal tenderness, liver and were clinically normal. Examination of the lymph node areas showed no palpable adenopathy. Clinical examination of the other systems was unremarkable.

Biologically, laboratory blood tests on admission revealed a hemoglobin level of 14.1 g/dL, hematocrit of 45%, platelet count of 331000/µl, and white blood cell count of 7450/µl. The nonspecific liver workup showed absence of cytolysis with aspartate aminotransferase, alanine aminotransferase were 28, 22

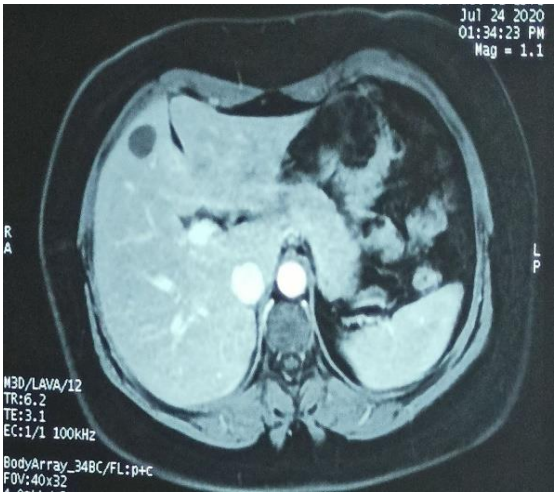
IU/L respectively and absence of cholestasis with alkaline phosphatase, gamma-glutamyltransferase and total bilirubin were 75 and 28 IU/L and 0.7 mg/dL, respectively, absence of biological stigma of hepatocellular failure with a coagulation profile which was normal and albumin at 42 g/L. Other laboratory data showed an AFP (Alpha Fetoprotein) level of 5.57 ng/ml and an ACE (Embryonic Carcino Antigen) level of 1.36 ng/ml.

Radiologically, the ultrasound showed a liver of normal size and regular contours with a cystic formation with a hyperechoic component. This formation was measured at 25/18 mm and localized at the level of segment IV with absence of other similar images visible at the level of the hepatic parenchyma, the gallbladder was alithiasic with thin wall. The bile ducts were not dilated, the suprahepatic veins and portal vessels were patent and of normal caliber, pancreas spleen and kidneys were without abnormalities with no intraperitoneal fluid effusion. The scannographic aspect of this cystic image was in the form of a capsular hypodense nodule of segment IV with a calcified sediment, this nodule measuring 18*22mm. This appearance was in favor of a ciliated hepatic foregut cyst, the liver was of normal size and regular contours with no other detectable lesions. On magnetic resonance imaging (MRI), the appearance was that of a focal lesion in segment IV, with a thick wall (2 mm), regular, downstage, with heterogeneous content, in frank T2 hypersignal and diffusion hypersignal with a posterior part in T2 hypointense and in diffusion evoking a hepatic cyst with a ciliated coating.

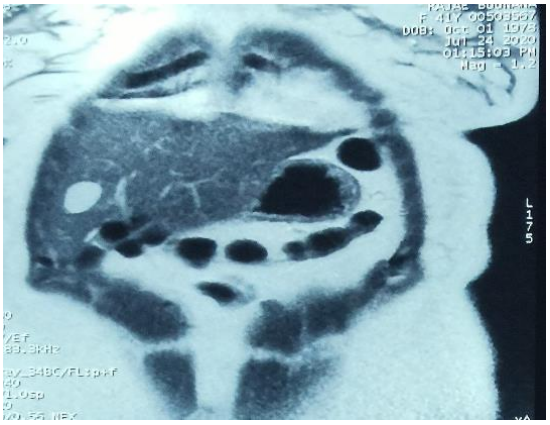
These radiological findings, combined with the clinical and biological data were consistent with the diagnosis of hepatic ciliated foregut cyst. Our patient was put under a close monitoring protocol with an abdominal ultrasound every 6 months and an annual hepatic MRI. The evolution was marked by a spontaneous decrease of the abdominal pain with a stable lesion on ultrasound.



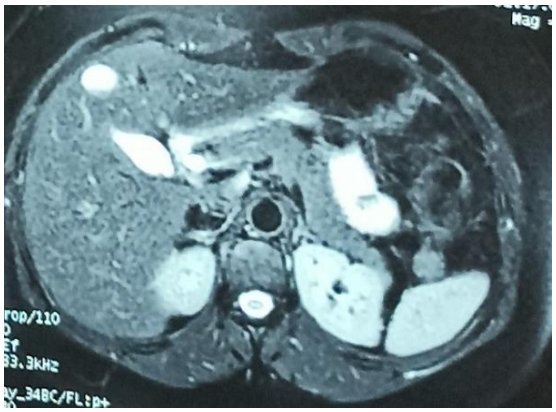
Ultrasound cross-section showing a cystic image with a hyperechoic component of segment IV



Cross-sectional CT scan showing a hypodense nodule in segment IV



Sagittal MRI section showing a nodule in T2 hpersignal



MRI cross-section showing a T2 hypersignal nodule

DISCUSSION

Hepatic ciliated foregut cyst is a benign cystic tumor of the liver. First described as a congenital malformation by Friedrich in 1857. Then, since 1984 this malformation has been called ciliated hepatic cyst [1 .2] It is an extremely rare hepatic lesion. However, its exact prevalence is still difficult to determine, since in most cases, the CHFCs is clinically silent and of

incidental findings. A review of the literature has counted 109 cases reported since 1964 and has shown an increase in the detection rate of this tumor in recent decades with 85% of cases found were reported in the last 20 years, which can be explained by the increasing use and progress of radiological examinations [3 4].

Classically the CHFCs is formed of four layers including a pseudo stratified ciliated columnar epithelium associated with secretory mucus cells, a second layer of sub-epithelial connective tissue, a smooth muscle layer all surrounded by an outer fibrous capsule. The presence of ciliated columnar epithelial cells is pathognomonic of CHFCs [9]. To explain this histological composition, one must understand the histogenesis of this cyst. In fact, during embryonic life there is an abnormal development of the primitive intestine with an abnormal migration of a bronchiolar bud into the peritoneal cavity. This occurs before the closure of the pleuroperitoneal ducts, this bud will be included in the septum transversum surrounded by the cells of the hepatic bud [5-7]. The CHFCs is preferentially located in the left liver at the level of segment IV, which can be explained by the important development of the left liver compared to the right liver at the beginning of embryogenesis (from the fourth to the sixth week) and in particular segment four which is the largest part in size during this period [1, 8].

The average age of diagnosis of CHFCs was 55 years, but this cyst can be diagnosed at any age ranging from 05 months to 82 years depending on the series. Clinically, the majority of patients with CHFCs are asymptomatic at the time of diagnosis. However, when they are symptomatic, the main clinical sign is abdominal pain in the right hypochondrium. This abdominal pain can be explained by the location of the tumor, which can cause distension of Glisson's capsule. Very rare cases have been described of obstructive jaundice by biliary compression and portal hypertension by portal compression [3, 10, 11, 12, 28].

The increasing use of abdominal imaging techniques has improved the detection rate of CHFCs. The suggestive morphological criteria common to all imaging techniques are: the lesion is unique in the vast majority of cases (rare cases reported of multilocular lesion), the diameter most often between 1 and 5 cm, the location of the lesion in the left liver exactly at the level of segment IV in anterior[13] sub capsular.

Abdominal ultrasonography is an innocuous and accessible examination most often performed for another cause, the CHFCs appears on ultrasound as a well-limited, uni-loculated, hypoechoic image (very rare cases have been reported of an echogenic cyst with posterior enhancement), although very rare cases of multi-loculated cysts have been reported, with or without posterior enhancement, avascular, i.e., with no Doppler flow visualized [14, 15].

On CT, the lesion is hypodense with no parietal enhancement after injection of contrast medium. In some cases the lesion may appear spontaneously hyperdense. Mobile calcifications have also been described [8, 16].

In magnetic resonance imaging (MRI), in T2-weighted sequences they are almost exclusively hyperintense, rare cysts had a hypo- or iso-intense aspect, while in T1-weighted sequences they appear in different densities [8, 17, 30].

The sometimes variable appearance on imaging can be explained by the existence of elements of the cyst contents with different characteristics, including a serous material of variable viscosity from one cyst to another. For this reason, it is recommended to perform combined imaging examinations in order to increase diagnostic accuracy [17, 18].

In case of doubt, anatomopathological examination confirms the diagnosis of CHFCs. This is due to the visualization of the pathognomonic four-leaf appearance, this histological appearance resembles the bronchogenic cyst at the thoracic level. This histological similarity also exists in other locations: esophagus as an esophageal cyst, stomach, pancreas, oral cavity, retroperitoneum and oral cavity [9, 19]. Immunohistochemically, the cells express general and specific markers of the foregut which are Cytokeratin 7 or 19. However, the more specific markers of the caudal intestine are usually negative. In the majority of cases, they also express thyroid transcription factor 1 (TTF-1) [20, 21].

In addition to the anatomopathological study of the surgical specimen, the diagnosis can also be affirmed by the demonstration of pathognomonic hair cells thanks to the cytological study of the liquid obtained by percutaneous fine[9] needle aspiration.

Rates of malignant transformation of CHFCs vary from 3% to 5%, with squamous cell carcinoma being the most common malignant transformation. The main risk factor for malignant progression was cyst size [1; 22]. Embryonal carcinoma antigen (ECA) and carbohydrate antigen 19-9 (CA 19-9) bioassays have not been shown to be effective as markers of malignant progression of CHFCs. Therefore, they do not seem to be useful to measure during follow-ups. Some cases of patients followed for CHFCs had high levels of these two tumor markers in the absence of any malignant progression. In addition to the risk of degeneration, two cases of biliary and portal vascular compression have been reported; [2324].

Therapeutic management is still controversial, Some teams recommend that since it is a congenital tumor, regular and rigorous surveillance should be performed using cross-sectional imaging techniques with recourse to either radical surgery if the patient is symptomatic or if the size of the tumor exceeds 5 cm, or during follow-up if the size of the tumor increases significantly or if there are negative signs on imaging such as focal wall abnormalities or thickened septations There is no consensus on the rhythm of surveillance. Other teams opt for a more aggressive strategy with the use of surgery as soon as CHFCs is diagnosed to avoid the risk of degeneration, especially since published cases of degeneration on CHFCs had a fatal evolution in the majority of cases. The recommended route for surgical treatment is the laparoscopic route. Because of the accessible location of the tumor, often subcapsular, and because of the small size of the cyst [16, 25, 26, 27].

CONCLUSION

The ciliated hepatic cyst is a rare benign lesion of the liver. CHFCs is characterized by a nonspecific appearance that is sometimes variable but very suggestive on imaging, especially if multiple techniques are matched. Although in the majority of cases CHFCs were asymptomatic and discovered incidentally on imaging for other reasons, they should require regular and rigorous follow-up or resection because of the rare cases reported of malignant degeneration.

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