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Case Report

Ciliated Hepatic Cyst: Report of a Case and Review of the Literature

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Keywords: Ciliated hepatic cyst (QCH); Intraabdominal abscess; Hepatic lesion

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Abstract

The ciliated hepatic cyst of the anterior intestine is a less frequent benign entity that arises from the alteration in the migration of embryological remains. Most of them are found in the left lobe of the liver, especially in segment IV. Its wall is covered by a pseudostratified ciliated columnar epithelium, a layer of connective tissue, smooth muscle, and a surrounding fibrous outer layer. We present the case of a 61-year-old man who, in the context of a scheduled admission for drainage of an intraabdominal abscess, was incidentally discovered to have a hepatic lesion of cystic aspect. The anatomopathological diagnosis was that of a ciliated hepatic cyst. Due to its low frequency in clinical practice (in part due to its incidental character), a review of the case and a review in the literature of the peculiarities of said entity are proposed.

Introduction

The ciliated hepatic cyst (QCH) constitutes a less frequent entity, although its diagnosis and detection have increased due to the greater use of imaging tests, having reported the majority of cases in the last 20 years [1]. It is usually solitary, unilocular [2] less than 30mm, and in the middle of the liver location, especially in segment IV. Due to its incidental nature and increased diagnostic frequency, the aim is to analyze the histopathological characteristics of the entity and review the literature [3].

Presentation of the case

We present the case of a 61-year-old man with cardiovascular risk factors (hypertension, dyslipidemia, obesity, occasional smoking). In 2013, in a TAC performed during a scheduled admission to drain an intraabdominal abscess in the context of a cholecystectomy for a cholecystoduodenal fistula, a hypodense lesion of 12 mm in segment IVa was described for the first time.

In March 2020, he was admitted to the Internal Medicine Department for bilateral interstitial pneumonia caused by SARS-CoV2, during which mild alterations of transaminases were presented without data of hepatocellular dysfunction. It performs TAC abdominal (Image 1), in which it continues to visualize the lesion of the cystic aspect, without changes with respect to the last years.

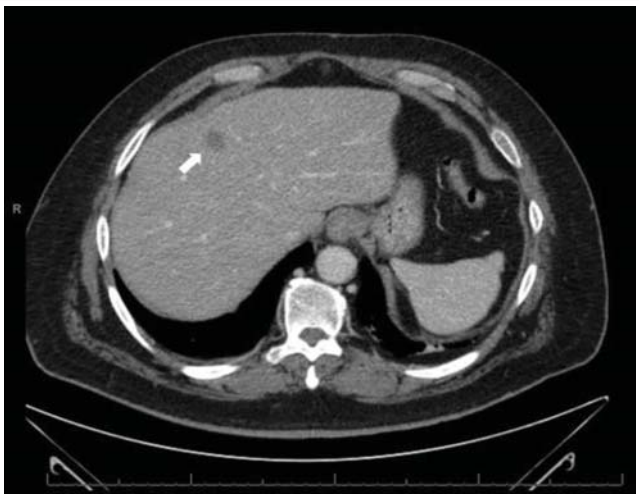

Image 1: TAC abdominal with contrast. Hypodense indeterminate lesion in segment IVa hepatica (arrow). 12 de Octubre University Hospital, Madrid.

The patient is referred to the Digestive Apparatus Service, where an RMN hepatic (Image 2) is requested, in which the lesion in segment IVA is described as hyperintense in T2 and with liquid-liquid levels in T1, with a thin wall in the Late phase of realisation, without septa or solid areas. These findings suggest as the first possibility, and given the location, a QCH.

Echography (Image 3) is carried out, in which the lesion is objective, which is anechogenic, and measuring 15 mm in its maximum axis, with posterior reinforcement; presenting

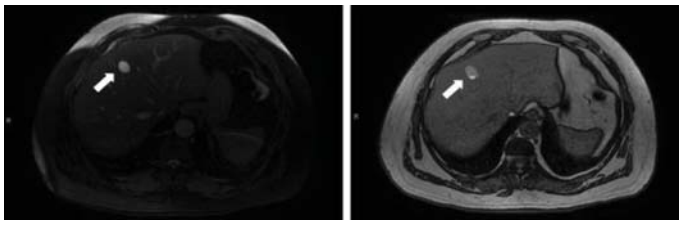

Image 2: RMN abdominal. The lesion is hyperintense in T2 (arrow) and with liquid-liquid levels in T1 (arrow), with a thin wall in the late phase of realisation, without layers or solids. 12 de Octubre University Hospital, Madrid.

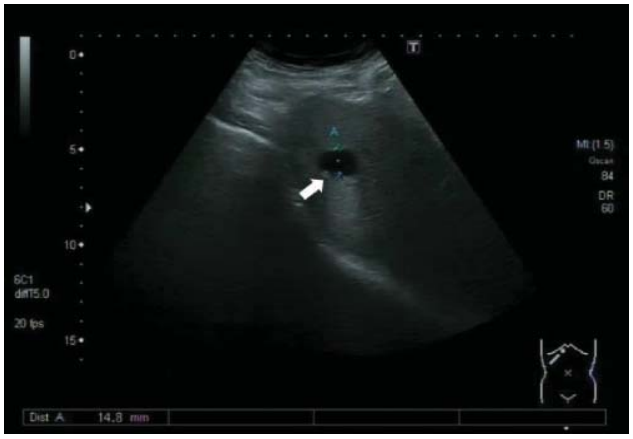

Image 3: Echography abdominal. Anechoic lesion of 15 mm, with posterior reinforcement (arrow). 12 de Octubre University Hospital, Madrid.

in one of its extremes a focal millimetric hyperechogenic thickening with posterior acoustic shadow, suggesting calcification.

Since the Service of Digestive Apparatus requests an evaluation by Surgery General. Given the history of the patient with two previous laparotomies, it was decided to perform surgical resection with an open approach, which was completed without significant complications.

In our service, we received the postoperative gross specimen, and it was a piece of subcapsular hepatic resection of 3.6 x 3.4 x 3 cm. In the serial sections of the same, a cystic lesion of 1.5 x 1.5 x 1.5 cm was identified, with a fibrous wall of whitish colour, of maximum thickness of 0.4 cm, with clear mucoid content in the interior.

At a microscopic level, a cystic lesion is observed (Images 4, 5), covered by a pseudostratified columnar epithelium with the presence of patched cilia, accompanied by a wall composed of connective tissue, a layer of thickened smooth muscle and externally fibrous tissue with areas of calcification and cholesterol crystals. These four described layers correspond to the classic histology of QCH [4].

The epithelium lining the cyst was positive for cytokeratin 7 (Image 6), cytokeratin 8, cytokeratin 19, and BerEP4; and negative for cytokeratin 20 and TTF1. In some reviews [5], it is recommended to carry out an immunohistochemical study with cytokeratin in case the epithelium has been flattened,

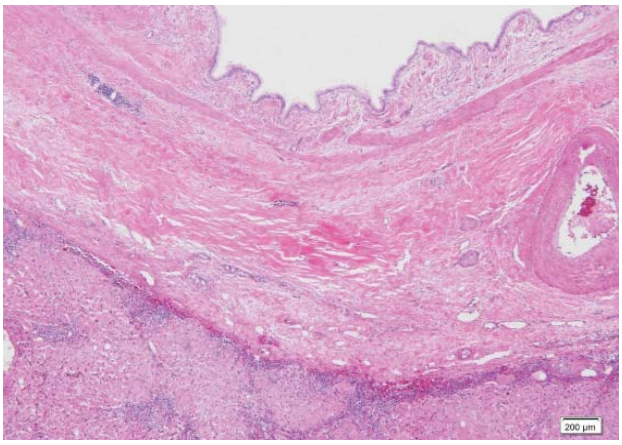

Image 4: View of the QCH, observing all four layers, in continuity with the hepatic parenchyma (hematoxylin-eosin, 4X). 12 de Octubre University Hospital, Madrid.

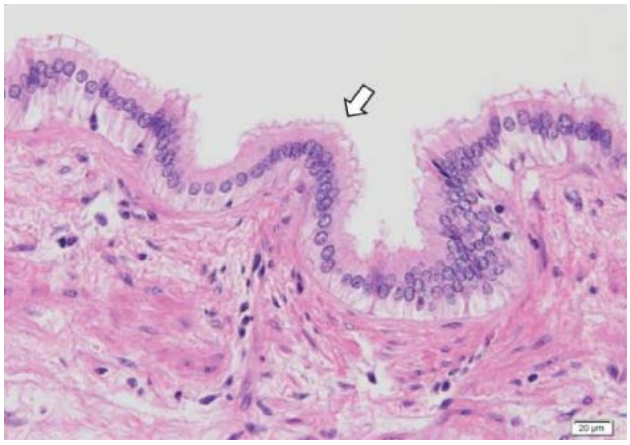

Image 5: Detail of pseudostratified ciliated epithelium (arrow) (hematoxylin-eosin, 40X). 12 de Octubre University Hospital, Madrid.

Image 6: The epithelium shows positivity for cytokeratin 7 (20X). 12 de Octubre University Hospital, Madrid.

which occurs especially in the case of biliary communication, due to the action of bile.

The remainder of the hepatic parenchyma presents with mild architectural distortion, with periportal fibrosis and incomplete septa. Moderate macrovacuolar steatosis is observed, affecting 45% of the hepatocytes. In the hepatic lobule, at least two foci of infiltrating inflammatory mixed

were identified by fields of great increase, and hepatocytes ballooned with Mallory's hyaline.

The patient has undergone imaging tests (CT and ultrasound) for follow-up without recurrence and is currently being monitored for steatohepatitis by the digestive service.

Discussion

The QCH is an extremely infrequent entity, which tends to be solitary, unilocular and the clinical course is essentially benign. The first description dates back to 1857 and was provided by Friedreich, who postulated that it was dealing with an embryological malformation [6]. It was not until 1984 when Wheeler and Edmondson used for the first time the term "ciliated hepatic cyst of the anterior intestine" [7]. Proposed that the ciliated hepatic cyst and the bronchogenic and esophágic cysts share a common embryological origin, which could be in the migration of yolks of the bronchial and oesophageal regions from the anterior primitive intestine and its posterior entrapment by the stomach.

The most common location of the QCH is in segment IV of the liver, which may explain why the left lobe, in particular that segment, constitutes the majority of the hepatic volume during the fourth to sixth weeks of development. Subsequently, this is followed by a period of remodeling, during which the left lobule undergoes peripheral degeneration and the right lobule increases in size [8].

The ciliated cysts of the anterior intestine can originate, in addition to the liver, in relation to the tracheobronchial tree, mediastinum, pancreas [9], tongue, biliary vesicle [10], and superior digestive tract. These have been described cases in the common hepatic duct [11] and in the triangle of Calot [12].

Although QCH exhibits some typical symptoms in imaging tests, these do not come close to providing a definitive diagnosis, and none of them have sufficient sensitivity or specificity to confirm or rule out this condition. Echographically, while presenting a solid appearance, they are usually described as unilocular hypoechoic cysts, sometimes with a solid appearance or with echogenic material in their light [13]. In computerized tomography without contrast, these lesions are also usually hypodense. The magnetic resonance imaging in T1 shows an image with traces of variable densities. However, in T2 the images are characteristically hyperintense.

The differential diagnosis should include hepatocellular carcinoma as well as other benign hepatic tumors, such as the biliary cystoadenoma that tends to be multilocular and is covered by columnar non-ciliated epithelium.

Fine needle aspiration (PAAF) can be used as the next step if the information generated by imaging tests continues to be incomplete, giving a positive predictive value of 76% [1].

In PAAF, the QCH results in the presentation of ciliated columnar epithelial cells of benign appearance over a mucoid background [14]. Caliciform cells, macrophages, and fragments of smooth muscle can also be seen. The absence of these characteristics does not completely rule out the diagnosis, because it may occur that the material aspirated is not sufficiently cellular. It has also been said that establishing the differential diagnosis with bronchogenic cysts is complicated, as both have the same type of epithelium.

QCH is a fundamentally benign entity, however, 5 cases have been described in the literature to date that developed squamous cell carcinoma [3,15-20].

The malignant transformation of QCH adds further difficulty to diagnosis, given the limitations of imaging tests and the possible sampling errors made during the macroscopic processing of surgical specimens. Serological markers such as CA 19-9 appear to be of some efficacy, as it has been observed that it is elevated in benign QCH and can be normal in case of malignant transformation. The size may help to suspect a possible malignancy, as QCH rarely exceeds 4 cm in diameter, and in the 5 cases described to date, they all averaged more than 5 cm in diameter, with the exception of the case reported by Zhang, et al. which presented multiple small cysts (3-5 mm) [17].

While it was thought that pain in the right quadrant was typically most suggestive of malignancy of the QCH, Wilson, et al. presenting a case in which the patient was found to be completely asymptomatic [19]. Nor does age seem to help discern benignity from malignancy, because curiously, one of the 5 patients who developed squamous cell carcinoma was only 21 years old, while the median age at presentation of ciliated ciliated cysts was 48 years [16].

It has been suggested that there exists a metaplasia-carcinoma sequence and that the source of the irritation may be related to the cystic content. The development of squamous cell carcinoma is associated with an aggressive clinical course, with a survival of 2 and 9 months in two of those cases, and pulmonary or abdominal recurrence in others [15,17].

The ciliated hepatic cyst in children is a less frequent entity, with fewer than one in 10 cases reported [4]. It affects both sexes equally, and most of them are diagnosed before the age of 5 years (including prenatal diagnoses). More than half of the children were found asymptomatic at the time of diagnosis, while another subgroup presented with abdominal pain and swelling. Data indicate that these infantile cysts tend to have biliary communication and that their continuous filling by bile makes them large at the time of diagnosis, with cases reported as large as 11 cm.

Macroscopically, the cysts are usually pinkish and unilocular. The internal surface tends to have smooth walls, sometimes being fibrous and irregular or with verrucous plates [21]. The content of QCH tends to be mucoid, clear, or viscous,

although others have described a gelatinous consistency, and a wide range of colorations, greenish, yellowish, or whitish [22]. A case has been reported in which the content was biliary, in which the QCH was compressing the portal vein and causing portal hypertension in a young woman of 17 years [23].

Regarding histological peculiarities, cases of QCH with squamous metaplasia have been described, the majority of them associated with squamous carcinoma [24]. Also observed are epidermoid hepatic cysts without other features of specificity [25], which could correspond to QCH or biliary cysts with extensive metaplasia squamous. There exist ciliated cysts without a layer of muscle tissue, as this shows the absence of staining for desmina and actin of muscle tissue [26]. In some, caliciform cells have been observed [5,22].

Within the differential diagnosis, we must include the simple hepatic cyst, the hydatid cyst [27], the pyogenic abscess, the mucinous cystic neoplasia (formerly called cystoadenoma biliary), the cystadenocarcinoma, the bronchogenic cyst, solid hypovascular tumors, and the cystic looking hepatic metastases. Observing histologically the four layers of the cyst wall allows us to reach the diagnosis.

The main differential diagnosis is established with the bronchogenic quest, both sharing an embryonic origin. In this case, we observe cartilage and seromucous glands accompanying a pseudostratified columnar or cuboidal epithelium [8].

In children, to the previous differential diagnosis, we must add the mesenchymal hamartoma, the epidermoid cyst, and the choledochal cyst [28].

Conclusion

The QCH is a less frequent entity, although, in the last few years, the reported cases have increased. Typically the course is asymptomatic, although on some occasions patients experience abdominal pain or ictericia. The diagnosis may be incidental to performing some imaging test, usually localizing in the medial segments of the left hepatic lobule at the subcapsular level. They usually present as solitary, unilocular lesions or less than 3 centimeters in diameter.

Surgical treatment is recommended, as cases of malignant transformation have been described.

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