Ciliated Hepatic Foregut Cysts: A Differential Diagnosis in Hepatic Lesions Placed in Segment IV

Abstract
Ciliated hepatic cysts or ciliated hepatic foregut cysts (CHFC) are unusual entities. While their histopathological features are pathognomonic, clinically and radiologically these lesions can be indistinguishable from other liver neoplasms. Its potential malignant transformation renders the surgical treatment mandatory, highlighting the importance of an accurate diagnosis. We report the case of a patient affected with colorectal cancer (CRC) presenting with a presumed liver metastasis which turned to be a CHFC after combined liver and colon laparoscopic resection.

Abbreviations: CHFC: Ciliated Hepatic Foregut Cysts; CRC: Colorectal Cancer; FNA: Fine Needle Aspiration; TTF-1: Thyroid Transcription Factor 1

Introduction
Ciliated hepatic cysts or ciliated hepatic foregut cysts (CHFC) are unusual entities with only approximately one hundred cases published in the Literature.

Originated from the embryological foregut, they are histologically recognized by its ciliated pseudostatified columnar epithelial layer reminiscent of the respiratory epithelium and also found in the bronchial cysts [1]. Due to the variability in appearance on radiographic imaging (often attributed to the elements of the cyst contents), diagnosis can be difficult and differential diagnosis include a large variety of hepatic lesions.

We report the case of a patient affected with colorectal cancer (CRC) presenting with a presumed liver metastasis which turned to be a CHFC after combined liver and colon laparoscopic resection.

Case Report
A 82 year-old male, with no relevant past medical history, presented with changes in bowel habits and a positive faecal blood test. Colonoscopy revealed a tumour located at 20 cm from the anal verge, and biopsy confirmed an adenocarcinoma. A thoraco-abdominal CT scan was performed, showing a simple cyst in segment VIII and a single solid non-enhancing hepatic lesion measuring 16mm, affecting segment (seg) IV suggestive of synchronous liver metastasis (Figure 1).

In this setting of colorectal cancer with a unique small synchronous liver metastasis, a combined laparoscopic resection of the sigmoid colon and the hepatic lesion was planned. After the laparoscopic sigmoidectomy was uneventfully completed, an intraoperative laparoscopic hepatic ultrasonography (US) was accomplished. The simple cyst in seg. VIII was identified and in seg. IV, a 15mm-lesion was identified: it showed as hypoechoic and apparently cystic with a filling material, ecographically different to that in the simple cyst. No other nodules were identified and the suspicion was a cystic metastasis of CRC. The hepatic lesion in seg. IV was therefore, removed by an US-guided laparoscopic partial hepatectomy. Once completed, in order to assure surgical margins, the specimen was opened revealing a cystic lesion with grayish mucinous content with wide margins. Postoperative course was uneventful and the patient was discharged in postoperative day 7.

The histopathological analysis of the specimen showed a cystic nodule lined by a pseudostratified ciliated columnar epithelium with isolated goblet cells. Within the cyst wall, isolated muscle fibers were also found. Diagnosis of ciliated cyst was firmly set (Figures 2, 3 & 4).

Figure 1: Enhanced CT scan showing a simple cyst in segment VIII and “solid” lesion in segment IV corresponding to CHFC.

Figure 2: Low power view of the CHFC, well-demarcated from the surrounding liver parenchyma. (Hematoxylin-eosin).
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Discussion

Ciliated hepatic cysts or ciliated hepatic foregut cysts are rare lesions with increasing incidence probably due to better imaging diagnostic techniques [2].

In most of the cases, CHFC present as small solitary lesions (mean diameter 4cm), located mainly in the nearby of segment IV, although other locations have also been reported [3,4]. This location is justified by its hypothesized embryological origin in the foregut as a result of an entrapment, during the early embryonic development, of bronchial sprouts (from the cranial foregut), in the liver (caudal foregut) [5].

They usually present in middle-aged patients, commonly as asymptomatic lesions (except in large lesions with compressive symptoms), or associated to abdominal discomfort, probably due to its subcapsular location which causes distension of the Glisson’s capsule [2]. In infants a biliary communication is not uncommon [6] and in adults, complications have been reported such as portal hypertension or obstructive jaundice [7,8].

Diagnosis can be made radiologically although it may not be certain and, combined imaging techniques are recommended to increase the diagnostic accuracy [2] and thus avoiding misdiagnosis, as happened in our patient.

On US, although it may appear as a solid-like lesion, CHFC typically show as a hypoechoic unilocular cyst [9]. On unenhanced CT-scan, CHFC are hypodense with no enhancement after contrast injection and in one third of the patients (including our case), they may show a solid tumor appearance [10]. On MRI they are typically hyperintense in T2 with variable densities in T1 [2]. FNA (Fine needle aspiration) cytology could be an useful to confirm diagnosis, with a positive predictive value of 76% [2].

Histologically, CHFC are easily diagnosed as the cyst wall is composed of 4 layers consisting of 1) pseudostratified ciliated epithelium, 2) connective tissue, 3) smooth muscle and 4) fibrous tissue [2,4]. These lesions have been shown to express general epithelial markers and also specific keratins of foregut structures (cytokeratin 7 or 19), while those more specific of hindgut are usually negative. Furthermore, most cases express the thyroid transcription factor 1 (TTF-1) [11].

Although CHFC have mainly a benign course, malignant transformation to squamous cell carcinoma can occur, with a reported incidence of 3-5%, and poor survival results [12,13]. Given these data, once diagnosed, CHFC should be surgically excised [2] and laparoscopic approach represents a feasible and advisable option due to its anterior subcapsular location and small size, as demonstrated in our patient and in previous publications [1,14].

It should be noted that although the location and size of the lesion were characteristic of CHFC, in our patient, the finding of a single solid hepatic lesion in the context of a sigmoid neoplasm, turns the differential diagnosis of metastasis in mandatory, since 15-25% of the patients with colorectal cancer present with synchronous liver metastasis [15]. Complementary imaging techniques could probably have helped in our case to avoid this mis-diagnosis, although surgical excision would have been recommended anyway in this case.

References


