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

Quiste ciliado hepático, diagnóstico diferencial de lesiones hepáticas del segmento IV

Ciliated hepatic foregut cysts (CHFC) are an uncommon pathology. While their histopathologic characteristics are pathognomonic, these lesions are clinically and radiologically indistinguishable from hepatic cystic neoplasms. Their potential for malignization requires surgical resection, so CHFC should always be included in the differential diagnosis of hepatic lesions.1-4

We report the case of a patient with colorectal cancer (CRC) and a single synchronous hepatic lesion that was suspicious for metastasis, treated by simultaneous laparoscopic resection of the sigma and liver. The hepatectomy specimen was compatible with CHFC.

The patient is an 82-year-old male with no prior medical history of interest and a recently diagnosed adenocarcinoma of the sigma. Thoracoabdominal extension computed tomography (CT) scan showed evidence of a simple cyst in liver segment VIII and an enhanced solid lesion measuring 16 mm in segment IV that was suggestive of metastasis (Fig. 1).

In this context of a patient with CRC and resectable solitary synchronous hepatic metastasis, a multidisciplinary committee decided to conduct a combined laparoscopic resection of the sigmoid and hepatic lesions.

After having completed the laparoscopic sigmoidectomy without incident, laparoscopic hepatic ultrasound was done. We identified the simple cyst in segment VIII as well as a 15 mm lesion in segment IV that was hypoechoic and cystic, whose content presented different echogenicity than the simple cyst. No other lesions were identified.

Ultrasound-guided laparoscopic partial hepatectomy was completed without incident. When the surgical specimen was opened, we observed a cystic lesion with a gray mucoid content and wide resection margins. The patient was discharged on the seventh day post-op.

The pathology study described a cystic structure covered in pseudostratified epithelium of a respiratory type, with cilia and isolated goblet cells. In the periphery of the nodule, fibrosis was observed with a proliferation of reactive pattern duct elements, all of which were compatible with CHFC (Fig. 2).

CHFC are rare lesions, with no more than 100 published cases, although their incidence is growing, probably due to the improved quality of diagnostic techniques.5

In most cases, CHFC present as solitary subcapsular lesions that are small in size (mean diameter 4 cm) and located in the center of the liver, mainly in segment IV (although other sites have been reported in the literature6-7). This location could be explained by the fact that, during early embryonic development, bronchial remains from the proximal intestine could become trapped in the liver (derived from the caudal intestine).1

CHFC usually present in middle-aged patients. They are generally asymptomatic or associated with some abdominal discomfort, probably due to their location, which causes distension in Glisson’s capsule.1 In adults, large-sized lesions can cause symptoms of compression and portal hypertension.7

The definitive radiologic diagnosis is not simple due to the variability in the appearance of CHFC (which seems to be attributed to the variable content of the cyst), so combined imaging studies are recommended in order to increase their diagnostic precision.5

On ultrasound, even though they may present a solid appearance, CHFC are usually seen as unilocular, hypoechoic cystic lesions.2 On baseline CT, they are hypodense lesions that do not uptake contrast, and in up to one-third of cases they can present as solid-looking lesions,3 as occurred in our patient. On magnetic resonance imaging (MRI), they are hyperintense in T2, while in T1 they present varying densities.

Cytology studies of samples obtained by fine-needle aspiration (FNA) can be useful to confirm the diagnosis with a positive predictive value of 76%.5 The histology of CHFC is pathognomonic, as its wall is comprised of 4 perfectly defined layers: (1) ciliated pseudostratified columnar epithelium, (2) connective tissue, (3) smooth muscle, and (4) fibrous tissue.1,5

Fig. 1 – Computed tomography with contrast, showing a simple cyst in segment VIII and a “solid” lesion in segment IV corresponding with CHFC (white arrow).

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From an immunohistochemical standpoint, these lesions express general and specific markers of foregut structures (cytokeratin 7 or 19), while the most specific markers of the caudal intestine are generally negative. Most cases express thyroid transcription factor 1 (TTF-1).⁸ CHFC are usually benign, although malignization has been described in 3%–5% of cases in the form of squamous cell carcinoma, mainly in larger lesions, with poor survival results.⁴ Therefore, the treatment of choice of these lesions is surgery.⁵ As other authors have demonstrated⁹ and as occurred in our case, the laparoscopic approach is a feasible option and recommendable due to the small size of these lesions as well as their central hepatic and subcapsular location. We should emphasize that, although the location and size of the lesion detected in our patient was characteristic of CHFC, the finding of a solid hepatic lesion in the context of colorectal oncologic disease required a differential diagnosis with metastasis since 15%–25% of patients with CRC present synchronous metastases.¹⁰ Complementary diagnostic tests (MRI, PET or FNA) could have fine-tuned the diagnosis, although the surgical indication for resection would have prevailed in any event.

Conflict of Interests

The authors have no conflict of interests to declare.

REFERENCES

Enteral Nutrition in Crohn’s Disease With a High Output Enteroatmospheric Fistula

Nutrición enteral en enfermedad de Crohn con fístula enteroatmosférica de alto débito

Enteroatmospheric fistulae (EAF) are a subgroup of enterocutaneous fistulae (ECF) in patients with laparostomy or open abdomen. They are characterized by being superficial, with a high volume of discharge and surrounded by viscera or granulation tissue. These factors can lead to a situation of metabolic and water-electrolyte imbalance, sepsis and severe malnutrition.

The conservative approach in functional short bowel syndrome associated with a complete jejunal fistula, using the reimplantation of the proximal fistula discharge through the distal jejunostomy, provides good results. In the physiological approach that we propose, we combine the reimplantation of the discharge with artificial enteral nutrition, which achieves improved nutritional state, a reduction in comorbidities and greater recovery of the intestinal mucosa, thereby facilitating the surgical closure of the EAF.

We present the case of a 19-year-old male with penetrating/stenosing Crohn’s disease (CD), diagnosed 11 years earlier, with perianal involvement and ileocecal disease, which had had a terminal colostomy due to severe stenosis of the sigma 3 years earlier. He had presented with pancolitis and steroid-dependent extensive ileitis, refractory to various lines of biological treatment and currently treated with ustekinumab, with favorable clinical, radiological and endoscopic response, but stenotic changes persisted at the ileal level. The patient was hospitalized due to abdominal pain secondary to ileal stenosis and reactivation of his CD. During hospitalization, he presented symptoms of bowel obstruction that required emergency surgery, involving ileocecal resection after meticulous adhesiolysis. On the 10th day post-op, the patient was re-operated due to bowel leak, observing a catastrophic abdomen with involvement of multiple loops, so we opted for damage-control surgery and laparostomy assisted with negative pressure therapy for later surgical revision or a “second look”.

In the postoperative period, a complete fistula persistence was observed with high discharge with the afferent and efferent loops in the proximal jejunum, which resulted in a situation of functional short bowel syndrome. Initially, parenteral nutritional support was initiated, but worsened hepatic function was observed, with cytolysis of multifactorial etiology (fasting, infection, total PN, medication).

Given the situation of important malnutrition (41 kg, BMI: 15.74 kg/m²) and comorbidity associated with the use of parenteral nutrition, we considered optimizing the enteral nutritional support by means of a physiological model.


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