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Ectopic hepatocellular carcinoma arising from the peritoneum

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El paciente ha dado su consentimiento informado para la publicación de este caso.

ABSTRACT
Ectopic hepatocellular carcinoma is a rare entity. Most cases are asymptomatic and are occasionally found during autopsy or laparoscopy. They may sometimes cause relevant clinical problems such as abdominal pain or intra-abdominal bleeding. In this clinical case report, we review the literature in order to decipher the case of a 68-year-old female with an ectopic hepatocellular carcinoma that arose from the peritoneum. The patient was diagnosed after being studied due to a mild asymptomatic hypertransaminasemia.

Key words: Hepatocellular carcinoma. Ectopic liver. Alpha-fetoprotein. Epiplon/Omentum.

INTRODUCTION
Ectopic liver (EL) is a rare entity defined as the presence of normal liver parenchyma separated from the liver, due to aberrant migration of the hepatic tissue during embryological development. The incidence of ectopic liver has been reported to be between 0.24% and 0.47% (2-4,7). It may occur at different anatomical locations. The most common are the hepatic ligaments or the gallbladder, which is the most frequent location. However, it may also be located in areas further from the liver such as the pancreas, adrenal glands, peritoneum, retroperitoneum, testicles or thorax (1,5,7). The presentation is usually clinically silent, but it may occasionally compress the adjacent organs and cause intra-abdominal bleeding or hepatocarcinogenesis (1). An ectopic liver can result in the development of benign lesions. However, more frequently these lesions are malignant, including hepatocellular carcinoma (HCC).

**CASE REPORT**

We describe the case of a 68-year-old female with no relevant past medical history, except for dyslipidemia. A mild elevation in aminotransferases (AST 58 UI/l, ALT 74 UI/l) was identified in a routine laboratory test and therefore, an abdominal ultrasound was performed (Fig. 1) that identified a solid and rather heterogeneous lesion of 6 x 5 cm in the right hemi-abdomen. Abdominal computed tomography (CT) scan showed (Fig. 2) the presence of an 8 x 6 x 4 cm solid and heterogeneous vascularized tumor with a polylobulated aspect that was located on the right hemi-abdominal mesenteric fat, which was suspicious of a mesenchymal/stromal origin. There were no radiological signs of cirrhosis or portal hypertension, the liver was a normal size and contour, and the spleen was also normal in size. The patient was asymptomatic, without abdominal pain, weight loss or any other digestive symptoms (ECOG 0). The tumor markers in serum (CEA, CA 125 and CA 19.9) were within the normal range. A diagnostic laparoscopy showed heteroplasia of the juxtacolic greater omentum next to the transverse colon, with three satellite lesions. One of these lesions was located between the II liver segment, lower gastric curvature and the left diaphragmatic pillar, whereas the two others were located at the jejunal and ileal level. Thus, a right colectomy was performed with a double resection in wedge of the gut and an excision of the lesion located between the liver, stomach and
The anatomopathological study of the surgical section (Fig. 3) identified moderately differentiated hepatocellular carcinoma on the ectopic liver, specifically in the mass located between the liver segment and the diaphragmatic pillar. Invasion of the venous vessels was also observed, which did not affect the thickness of the intestinal wall. Tissue corresponding to the ectopic liver was not observed in the remaining two lesions. However, vascular invasion was visualized in this tissue and was therefore considered as a metastatic mass from the parent lesion.

Immunohistochemical studies were positive for Heppar-1, Glypycan 3, CK 18, alpha-inhibin and polyclonal CEA. Staining for CK 7, CK 8, CK 20, CD34, c-KIT, vimentin, chromogranin, CDX2, HMB45 and Melan-A was negative. Ki67 was graded at 20%, with a heterogeneous pattern.

Alpha-fetoprotein levels were measured after surgery and were high at 651 ng/ml. HBV, HCV and autoimmune tests were negative. The postoperative course was uneventful and the patient remains asymptomatic, with normal alpha-fetoprotein levels and no radiological signs of recurrence two years after surgery.

DISCUSSION
The ectopic liver has the same risk factors for hepatocarcinogenesis as the liver (1). However, risk factors such as HBV, HCV or liver cirrhosis do not appear to be associated in its pathogenesis. In fact, only two of 21 cases (9.5%) described by Arakawa et al. were infected by the hepatitis viruses (2). It is estimated that only 27-32% of ectopic hepatocellular carcinoma are associated with liver cirrhosis (4), suggesting that ectopic liver is more prone to neoplastic transformation. The reason why ectopic liver is particularly predisposed to carcinogenesis remains elusive (5,6).

However, one theory is that the defective arterial supply and venous and biliary drainage could predispose to neoplastic transformation, prolonging exposure to carcinogenic substances or impairing reparative nuclear mechanisms (1-3). The majority of HCC in ectopic livers are reported in Asian patients, it is less frequent in Caucasians (1) and is mainly located in the gallbladder.
The prognosis of these patients is significantly better and with lower recurrence rates than in patients with ordinary HCC, especially if the tumor is completely resected (4,5). Surgical treatment is the preferred option as it has a high curative rate in the majority of cases (5). With regard to the three cases reported by Leone et al. (1), two were alive without any signs of recurrence four years after surgery.

In conclusion, hepatocellular carcinoma in the ectopic liver should be considered as a possible diagnostic option in patients with an intra-abdominal tumor and high alpha-fetoprotein serum levels, even if there are no signs of chronic liver disease.

REFERENCES
Fig. 1. Abdominal ultrasound examination showing a solid and rather heterogeneous hypoechoic tumor of 6 x 5 cm, located in the right hemi-abdomen.
Fig. 2. A. Abdominal CT scan shows the liver with a normal size and morphology, without signs of liver cirrhosis. B. There was also a solid and heterogeneous vascularized tumor with a polylobulated aspect, located on the right hemi-abdominal mesenteric fat.
Fig. 3. A. Metastasis from a hepatocarcinoma in the meso colon. B. Microscopic image showing ectopic liver tissue, with a large portal space similar to a hepatic hilum surrounded by hepatocellular carcinoma. HE40x.