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HEPAR LOBATUM CARCINOMATOSUM

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Key-word: Lung neoplasms, secondary

Background: A 59-year-old woman was admitted for rapid health degradation, with abdominal pain, icterus and ascites. She was known for a two years long-standing breast cancer aggravated progressively by multi-organ metastases. The patient was treated by hormonal therapy (Tamoxifen followed by Exemestane) and chemotherapy (5-Fluorouracil – Epirubucin – Cyclophosphamide then Cisplatine – 5-Fluorouracil).

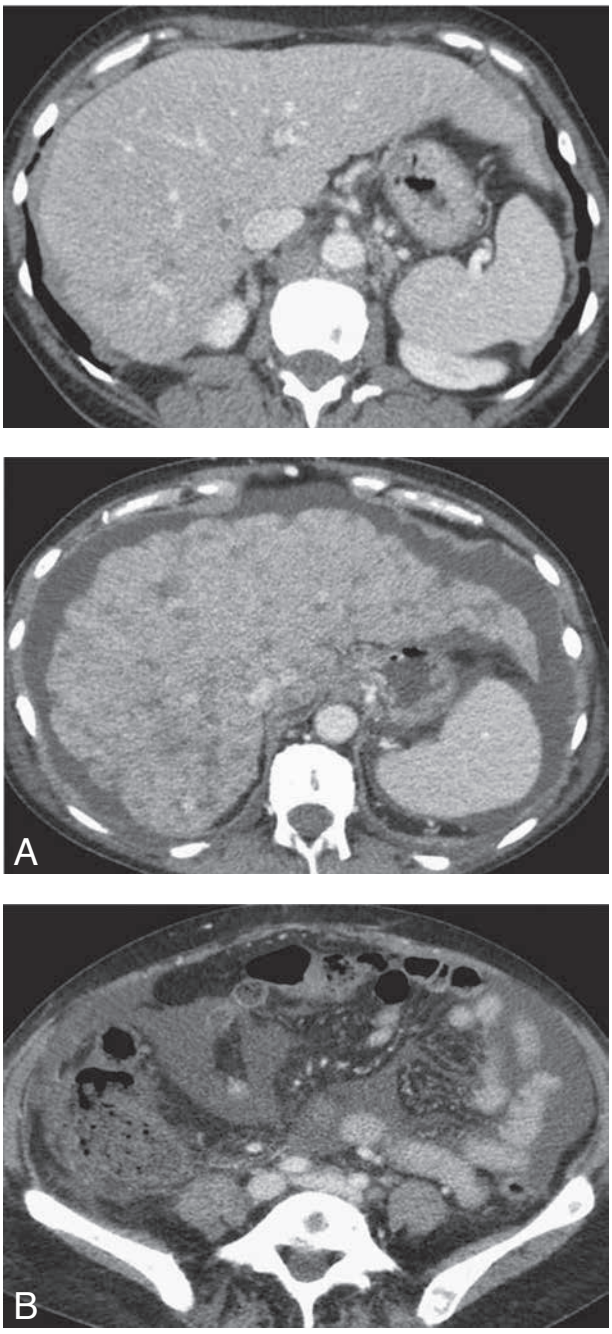

Fig. 1 / 2A / 2B

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Work-up

Axial contrast-enhanced CT scan, section at the level of the liver (January 2011) (Fig. 1), showed a normal sized and smooth liver. Small metastases are already observed in the right hepatic lobe. No ascites or portal hypertension is present.

Axial contrast-enhanced CT scan of upper abdomen (August 2011) (Fig. 2) shows on section at the level of the liver (A) a highly dysmorphic and heterogeneous liver with capsular retractions, lobulated pattern and multiples hypodense lesions, complicated by ascites. On section at mid-abdominal level (B), features of portal hypertension are demonstrated with ascites, enlarged mesenteric veins and congested wall of the right colon. Notice also the presence of multiple bone metastases.

Radiological diagnosis

Based on the radiological findings and the clinical history, the diagnosis of *hepar lobatum carcinomatosum* was made.

Discussion

Hepar lobatum carcinomatosum (HLC) is an acquired non-cirrhotic liver dysmorphy associated with liver metastases of carcinoma, most often breast carcinoma.

Hepar lobatum, characterized by a coarsely nodular lobulated liver parenchyma by deeply grooved scars, was first described in association with tertiary syphilis. Since 1924, this entity was occasionally reported in association with carcinomatous liver metastases and thus renamed *hepar lobatum carcinomatosum*.

At pathology, diffuse metastatic involvement of the liver is observed in HLC, characterized by the presence of fibrous septa originating from the organ surface and running deep into the parenchyma. These septa contain scattered metastatic cells. HLC can be observed independently of neoplasic progression or apparent regression, and independently of chemotherapy of the metastatic carcinoma.

On imaging, HLC must be suggested in case of rapid (months) dysmorphic transformation of a previously smooth and normal sized liver into a liver with capsular retractions and lobulated pattern, in the absence of etiologic factor for cirrhosis (hepatitis B or C, alcohol or drug consumption, hemochromatosis,...).

Metastatic liver lesions can be observed or not. MRI demonstrates scarring and progressive fibrosis of the liver, demonstrated by late parenchymal enhancement after IV Gadolinium injection.

The oncologic history is usually already known, but HLC can occasionally be the initial manifestation of an occult metastatic carcinoma.

The physiopathology of HLC remains controversial. First authors hypothesized a desmoplastic stromal response in reaction of metastatic cells invasion. Later, it has been proposed that HCL could be a certain form of healing and scarring after tumor regression, since several cases have been reported following chemotherapy. More recently, it has been suggested that vascular injuries could play a major role in HCL development: firstly, direct diffuse invasion and obstruction of intra-hepatic portal and/or hepatic venous branches has been reported in most of cases. This peripheral vascular invasion can be responsible for a congestive effect on liver, leading to its gradual shrinkage with typical venocentric distribution of fibrosis and development of portal hypertension. Secondly, chemotherapy could be an additional factor due to toxic vascular lesions.

In summary, HLC could result from the combination between an important stromal reaction to carcinomatous cells associated to intrahepatic vascular tumoral obstruction, potentially aggravated by chemotherapy vascular toxicity.

HLC has a poor prognosis, even in case of apparent tumoral regression after chemotherapy.

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