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Heparlobatum Carcinomatosum is a Rare Entity not to be ignored

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Abstract **Case Report**

Hepar lobatum carcinomatosum corresponds to a major hepatic dysmorphia on non-cirrhotic liver acquired during the evolution of a carcinoma, most often of mammary origin, metastasized to the liver. It is often associated with portal hypertension. It is the consequence of invasion of portal veins by tumor cells and of a desmoplastic reaction in contact with the tumor. This is a complication with a poor prognosis but which seems to be amenable to early detection and appropriate management. We report the case of two patients followed for multi-metastatic invasive breast carcinoma under palliative chemotherapy and hormonal therapy and whose evolution was marked by the radiological characterization of a pseudo cirrhosis aspect during the radiological evaluation of their response to treatment.

Keywords: Hepar lobatum carcinomatosum, liver dysmorphia, CT, portal hypertension.

INTRODUCTION

Heparlobatum carcinomatosum, pseudo cirrhosis, or metastatic cirrhosis designate a rare radiological entity described mainly in the context of metastatic breast cancer to the liver previously treated with chemotherapy and hormonal therapy.

It is characterized by an aspect of acquired hepatic dysmorphia resulting in a liver with irregular contours, lobulated, with a macro nodular aspect with atrophy of certain segments and hypertrophy of other hepatic segments, and this, in a random way with evidence of lesions of metastatic origin which may be missing after macroscopic regression of the lesions or in case of radiologically occult sinusoidal tumor infiltration.

Heparlobatum carcinomatosum despite its designation as pseudo cirrhosis is frequently complicated by portal hypertension and rapidly progressive hepatocellular insufficiency leading to the same unfortunate clinical consequences of cirrhosis with a rapid evolution leading to death in a few months.

The etiopathogenic of this entity is still poorly understood involving a phenomenon of desmoplastic reaction, an intense fibrous reaction in contact with the metastatic lesion under the effect of chemotherapy molecules, but also a direct vascular lesion attack by the formation of tumor thrombi or indirectly under the effect of chemotherapy agents leading to ischemic phenomena with atrophy, compensated by nodular hyperplasia of preserved segments.

This is a complication with a poor prognosis but which seems to be possibly improved by early detection with adequate management.

We report the case of two patients followed for multi-metastatic invasive breast carcinoma under palliative chemotherapy and hormonal therapy and whose evolution was marked by the radiological characterization of a pseudo cirrhosis aspect during the radiological evaluation of their response to treatment.

CASE REPORTS

Case No: 1

This is a 55-year-old female patient, followed for infiltrating ductal carcinoma since March 2020 treated by surgery: left mastectomy and lymph node curage, radiotherapy, and adjuvant chemotherapy, under hormonal therapy, having developed liver and bone metastases in June 2021.

She was put on palliative chemotherapy, and in March 2022 (eight months later) the disease partially regressed with the appearance of a dysmorphic liver, reduced in volume, with irregular and lobulated contours, associated with abundant ascites without splenomegaly, suggesting a heparlobatum

carcinomatosum appearance with ascitic decompensation.

Case No: 2

This is a 34-year-old female patient who has been followed since February 2022 for HER2-positive locally advanced invasive ductal carcinoma with carcinomatous mastitis and multi-metastatic disease of the lung, bone, and liver. The patient was put on palliative chemotherapy and anti-HER 2. The evolution was marked four months later by the appearance of hepatic dysmorphia with a marked lobulated contour in the left liver without any other associated sign showing the beginning of a pseudo-cirrhosis.

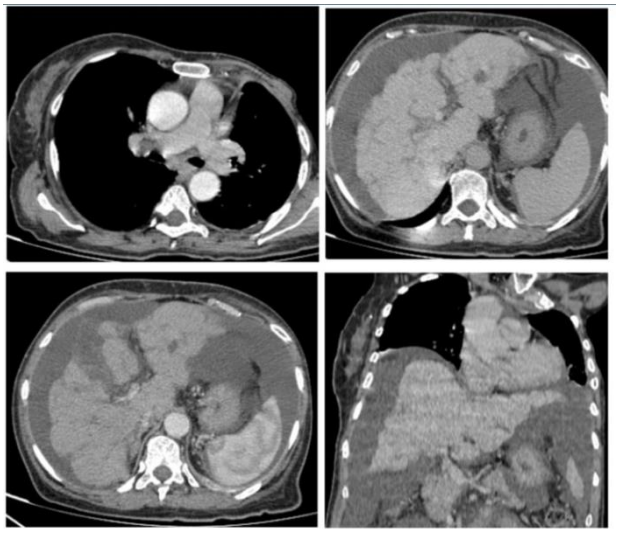

Figure 1: Patient followed for metastatic left breast ductal carcinoma under palliative chemotherapy who developed 8 months after the start of chemotherapy a volume-reduced dysmorphic liver with the lobulated appearance of its contours about a pseudo-cirrhosis.

Note the abundant ascites.

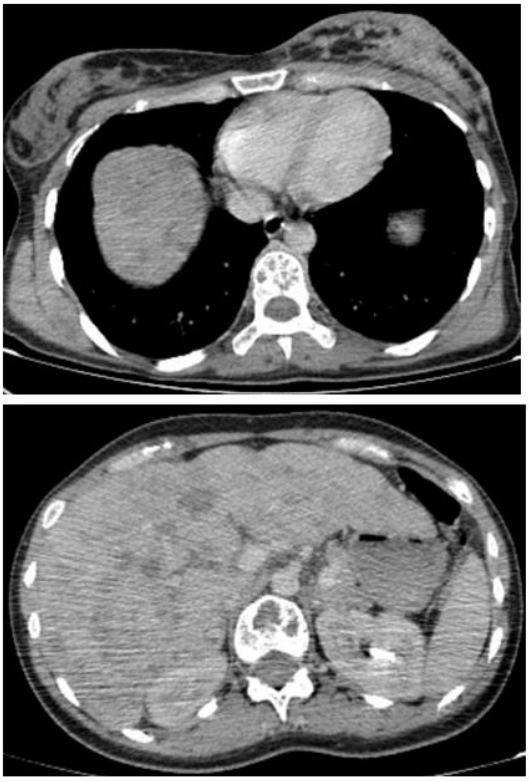

Figure 2: A 34-year-old patient, followed for multi-metastatic ductal infiltrating carcinoma under palliative chemotherapy and anti-HER 2, with the appearance of a hepatic dysmorphia at the control CT scan four months after the beginning of the treatment with a multinodular liver with lobulated, irregular contours, testifying to a pseudo cirrhosis at the beginning.

DISCUSSION

Heparlobatum was first described in the setting of tertiary syphilis as a deformation of the liver whose contours become the irregular, lobulated, seat of crevices and linear depressions, resulting from the healing of syphilitic gums.

This aspect was then described radiologically in the context of metastatic livers, mainly of breast cancer, after systemic treatment by chemotherapy, taking the name of heparlobatum carcinomatosum.

The term pseudo cirrhosis is also used for an imaging appearance reminiscent of a cirrhotic liver in the absence of histopathological criteria for hepatic cirrhosis, but also in a patient with no history of chronic liver disease or risk factors related to it (hepatitis B, hepatitis C, alcoholism, metabolic disease) with a relatively rapid onset of morphological abnormalities not consistent with the evolution of a liver with chronic liver disease towards cirrhosis.

A review of the literature and meta-analysis on the subject, published in 2022, reported that the average time between the diagnosis of liver metastases and the onset of pseudocirrhosis was 8 months [1].

An observational study published in 2007 on the frequency of liver contour abnormalities and signs of portal hypertension in patients followed for liver metastatic breast cancer and receiving chemotherapy reported that 75% of the patients followed developed variable morphological abnormalities such as limited shrinkage, diffuse shrinkage, and diffuse nodularity after 15 months of follow-up [2].

Cases of pseudo cirrhosis have been reported with other cancers, notably pancreatic, digestive, and thyroid, but more sporadically [3-8].

It is also noted that cases of pseudo cirrhosis of the liver revealing an unknown and untreated occult metastatic breast cancer have been reported [9, 10].

The strong association of pseudocirrhosis with systemically treated liver metastatic breast cancer is of unknown cause [11].

The etiopathogenic of this acquired hepatic dysmorphia, pseudo cirrhotic remains poorly understood.

It is thought to be due to a desmoplastic reaction in the liver parenchyma, in contact with liver metastases, secondary to the administration of chemotherapy molecules with the formation of a fibrous scar at the site of the metastatic lesion associated with fibrous septa going from the surface of the organ to the depth, resulting in capsular retraction and irregularity of the contours [12].

Direct vascular lesions due to the formation of tumor thrombi within the portal veins or indirect lesions linked to the effect of chemotherapy molecules have also been evoked, leading to ischemia of the hepatic parenchyma and therefore to atrophy of the segments concerned, against compensatory hypertrophy of other hepatic segments with the formation of hyperplastic nodules of regeneration [12-14].

This suggests the need for diffuse asymmetric hepatic metastatic involvement associated with a slow lesion evolution allowing the compensatory hyperplasia phenomenon.

The portal hypertension syndrome variably associated with hepatic pseudocirrhosis would be related to the compressive effect of the hyperplastic regeneration nodules but also to the tumor infiltration of the hepatic vessels [13, 15].

Clinically, patients may be asymptomatic, develop signs of portal hypertension (ascites, splenomegaly, collateral circulation, esophageal varices) or present a picture of liver failure.

The early diagnosis of this pseudo cirrhosis and the introduction of discontinuation of chemotherapy would allow the regression of liver dysmorphia [4].

This would improve the prognosis of these patients, which remains generally poor with a lower overall survival than in metastatic patients who have not developed pseudo cirrhosis and particularly in patients with pseudo cirrhosis associated with signs of PH such as varicose veins [16].

The regression of liver dysmorphia reported in this case suggests that it was probably secondary to a nodular hyperplasia phenomenon rather than to a fibrous (desmoplastic) reaction to the tumor lesions. The limitation is the lack of histopathological data.

The time interval between the installation of pseudo cirrhosis and death is on average 2 months according to Villani *et al.*, [1].

CONCLUSION

Heparlobatum carcinomatosum is a rare complication, most often reported during metastatic breast cancer to the liver treated with chemotherapy and hormonal therapy. Nevertheless, it remains a formidable complication, significantly impacting the overall survival of these patients. Radiologists' and clinicians' knowledge of this entity would allow its early detection and thus better management.

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