



Hepar Lobatum Carcinomatosum Associated With Metastatic Breast Cancer: An Unusual Cause of Liver Dysmorphism

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Abstract

Hepar Lobatum Carcinomatosum (HLC) is an acquired hepatic distortion which generally refers to liver abnormalities in tertiary syphilis. It is an exceptional liver dysmorphism associated with liver metastases of carcinoma, most often breast carcinoma. It is a non-cirrhotic hepatic disease. We report the case of a 58-year-old woman with HLC resulting from metastatic mammary carcinoma in the liver. We analyse through this observation the clinical, radiological and histological characteristics of this entity.

Keywords

Hepar lobatum; Breast; Cancer; Metastases

Introduction

Hepar Lobatum Carcinomatosum (HLC) is an unusual clinical finding. It is an acquired liver dysmorphism with lobulated contour and linear depressions mostly known as the end-stage of tertiary syphilis [1]. HLC have been described in association with liver metastases of carcinoma in many cases [2]. That is resulting from invasive metastatic breast cancer in most cases [3].

Case Presentation

A 58-year-old woman was followed-up for carcinoma of the breast, which was staged T4bN1M1 with multiple liver metastases. Breast Biopsy led to a diagnosis of an invasive ductal carcinoma grade III, Her2 positive, hormone-receptor-negative with a ki-67 index of 18%. She was initially treated by chemotherapy (5-Fluorouracil – Epirubicin-Cyclophosphamide). After 5 courses, the patient complained of hepatalgia and major asthenia. The physical examination showed hepatomegaly and a right upper quadrant abdominal pain. Last blood tests revealed thrombocytopenia and liver tests showed increased serum levels of aspartate and alanine aminotransferases which were respectively four and three times the upper normal limit. Low prothrombin ratio (58%) and increased glutamyl-transpeptidase which was fourteen times the upper normal limit were observed. The level of CA 15-3 was 584.6 UI/ml. There was no clinical or serological evidence for hepatitis B or C virus infection. Moreover, there was no history of alcohol or drug consumption. CT scan revealed a heterogeneous and dysmorphic liver with capsular retractions, bosselated contour and a major atrophy of the right liver lobe. Hepatic nodules were observed in the left lobe. Imaging revealed a tumor regression of metastases of the left liver lobe (Figure 1). Magnetic resonance imaging supported the diagnosis of Hepar Lobatum Carcinomatosum. A CT-guided liver biopsy was performed. Histological examination of the liver biopsy showed a proliferation with trabecular and glandular architecture invading the liver sinusoids and obstructing the biliary tract. No fibrosis was observed. There were no granulomas, giant cells or features indicating coexisting chronic hepatitis. The patient is still on second line chemotherapy by Capecitabine. A baseline CT scan was performed before starting Capecitabine showing dysmorphic liver with hypertrophy of segments VIII, VII, VI and IV with an extensive fibrosis supporting the diagnosis of Hepar Lobatum Carcinomatosum (Figure 2).

Discussion

In this observation, we made the diagnosis of hepar lobatum by the association of hepatic dysmorphism, the evocative clinical context (liver metastases of breast cancer) and

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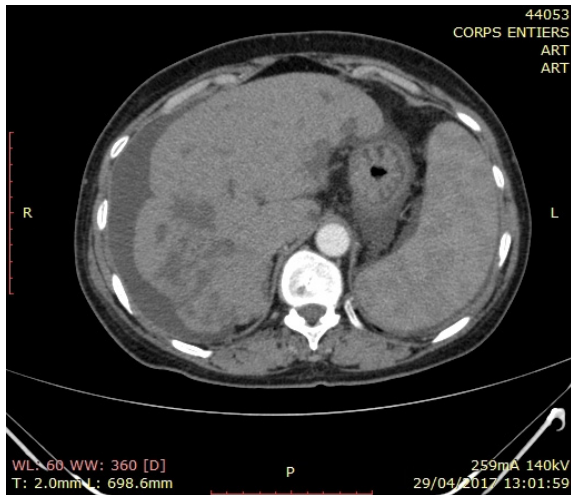


Figure 1: CT scan of the liver showing a heterogeneous and dysmorphic liver with bosselated contour and a major atrophy of the right liver lobe

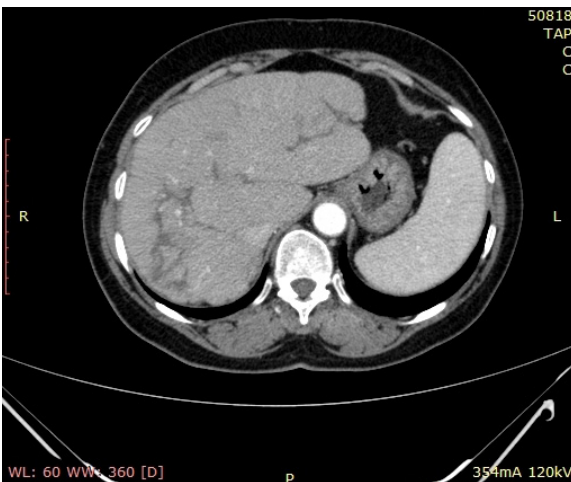


Figure 2: CT scan showing dysmorphic liver with hypertrophy of segments VIII, VII and IV

the physical examination (hepatomegaly and a right upper quadrant abdominal pain). The absence of risk factors for chronic liver disease (no alcohol consumption, no serological evidence for hepatitis B or C virus) dismisses the diagnosis of liver cirrhosis. This hypothesis was confirmed by the sequential data of the computed tomography and MRI that reported dysmorphic liver with capsular retractions and the histological examination. Hepar Lobatum (HL) is a non-cirrhotic hepatic dysmorphism. It is mostly known as the end-stage of tertiary syphilis [1]. Since 1924; HL was reported in association with liver metastases of carcinoma [2]. In most cases, that is resulting from invasive metastatic mammary carcinoma [3]. Liver metastases from the stomach, rectum has also been associated to that pathology [4-5].

The clinical manifestations of liver disease observed in patients who developed HLC have included jaundice, hepatomegaly, ascites, and esophageal varices [6]. The HL can be revealed by ruptured esophageal varices [7].

A variety of chemotherapy agents have been used in patients reported to have developed HLC including alkylating agents (Cyclophosphamide), cytotoxic antibiotics (Doxorubicin, Mitomycin, Mitoxanthrone), antimetabolites (Methotrexate, 5-fluorouracil), and plant alkaloids (Vinblastine, Paclitaxel, Docetaxel) [7]. In our

case, liver dysmorphism was detected in CT scan after 5 courses of Cyclophosphamide/Epirubicin/5-Fluorouracil (FEC regimen).

Liver imaging made the diagnosis in most cases showing liver with capsular retractions and lobulated pattern. A dysmorphic transformation of a previously smooth and normal sized liver is usually observed in the absence of etiologic factors of cirrhosis. Signs of portal hypertension are frequently observed in HLC. MRI may also be useful to support this diagnosis, showing large fibrous bands [8].

Histopathological examination usually shows fibrous septa containing scattered tumor cells running deep in the parenchyma [9]. Intrahepatic vessels, mostly sinusoidal and portal are invaded by metastatic cells causing an occlusion. Heterogeneous liver and atrophy of the invaded segments are a result of the vascular injury. The loss of sinusoidal wall integrity can be associated with peri sinusoidal fibrosis [4,8].

HLC has a poor prognosis. The expected survival is usually less than 1 year [9].

Conclusion

Hepar lobatum carcinomatosum resulting from metastatic carcinoma is a very rare phenomenon. This particular presentation could result from the combination between vascular injury caused by metastatic cells and chemotherapy agents. HLC has a poor prognosis.

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