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Review Article

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THE HEPATIC LYMPHANGIOMA: ANOTHER BENIGN LIVER TUMOR TO KNOW

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ABSTRACT

Hepatic lymphangiomas are a category of benign tumors secondary to a malformation of the hepatic lymphatic system and usually coexist with other visceral lymphangiomas. They are extremely rare in adults and uncommon in children. The clinical presentation is very polymorphic, most often of accidental discovery. The differential diagnosis is essentially made with other liver tumors, especially malignant ones. The prognosis of hepatic lymphangioma is good and patients do not need additional treatment after surgical resection. We present through this work the clinical case of a 48-year-old female patient with no notable pathological ATCD who consulted for atypical hepatic pain. The radiological examination revealed a malignant hepatic mass with negative tumor markers. A biopsy was performed which confirmed the diagnosis of hepatic lymphangioma.

KEYWORDS: Lymphangioma; liver tumor.

1. INTRODUCTION

Hepatic lymphangiomas are rare benign neoplasms characterized by congenital cystic dilatation of lymphatic vessels in the liver parenchyma, and usually occur as part of systemic lymphangiomatosis.[1] Most lymphangiomas are located in the head, neck, and axillae, where loose connective tissue allows easy expansion of lymphatic channels. Intra-abdominal cases account for less than 5% of all lymphangiomas.[2-4]

A solitary hepatic lymphangioma in an adult is extremely rare and after the first description by Ziegler in 1892, only a few cases have been documented.[5-6]

They have been classified into three types: simple lymphangioma, cavernous lymphoma and cystic lymphoma. The fluid component of a lymphangioma can be serous or chylous, depending on the location. In the presence of hemorrhage or infection; it may become bloody or purulent.[7-8]

Most hepatic lymphangiomas are detected during routine physical examinations. Occasionally, abdominal pain is caused by compression. An abdominal MRI or CT scan may be used to help differentiate it from similar lesions.

Lymphangiomas are benign tumors but always carry a risk of malignant transformation. There is also a risk of rupture or compression due to continued enlargement. Surgical resection of lymphangiomas is the standard treatment. For huge or multiple angiomas, liver transplantation should be considered.[8-9]

2. Medical Observation

The patient was 48 years old and had no notable pathological history. She came to us for the management of a heaviness pain in the right hypochondrium dating back 2 years before her consultation, without any particular irradiation, progressively worsening without any calming factors, evolving in a context of conservation of the general condition. The general examination revealed a hemodynamically stable patient; apyretic; anicteric; WHO 0; BMI 24.

The abdominal examination showed tenderness in the right hypochondrium without hepatosplenomegaly or palpable mass. A complete biological workup was requested showing a hemoglobin at 13.7g/dl; white blood cells at 8500/mm3; platelets at 252,000/mm3. The PT was 77% with a correct liver workup: GOT at 30 ui/L; GPT at 15 ui/L; BT at 6 mg/L; BC at 5mg/L; GGT22ui/l PAL 69ui/l and an Albumin at 46 g/L.

We completed with tumor markers that came back correct: AFP is 1.7 ng/mL; CA19-9 is 3 U/mL; and CEA is 1.68 ng/mL.

An abdominal CT scan was done "figure 1" showing a normal-sized liver, with a poorly limited, heterogeneous tissue infiltrate on the left liver, which was globally hypodense at the different times, enhanced in the center at portal time, retracting the capsule and deforming the hepatic contours measuring 82x60mm in diameter, which could be related to a fibro-lamellar HCC, an intra-hepatic cholangiocarcinoma, or any other origin that should be considered, in particular an atypical hemangioma.

The case was discussed at the digestive PCR and the decision was to perform a liver biopsy.

The anatomopathological study "figure 3" concluded to a vascular tumor proliferation made of capillaries of variable size; the latter are lined with elongated endothelial cells with hyperchromatic nuclei; the vascular structures are separated from fibrous tissue. An immunohistochemical study was carried out: the anti CD 31 and CD 34 antibodies came back negative on the vascular structures with a Ki67 estimated at 2% and thus the diagnosis retained is that of hepatic lymphangioma given the negativity of the vascular markers.

The decision of the staff was a therapeutic abstention with radiological surveillance by a hepatic MRI annually.

The initial abdominal MRI "figure 2" showed a normal-sized liver, with regular contours, located on the left liver, with a lobulated and irregular tissue mass, described as T1 hyposignal, T2 hypersignal, heterogeneous, with diffusion on hypersignal, with lumpy enhancement, homogenizing at late times, delimiting areas of fluid, measuring 9.5x7.2x8 cm, its ADC was measured at 1.8

The annual abdominal MRI check-up showed a stability of the previous imaging. Patient currently stable; has occasional right hypochondrium pain pain with on-demand symptomatic treatment; follow-up MRI scheduled soon.

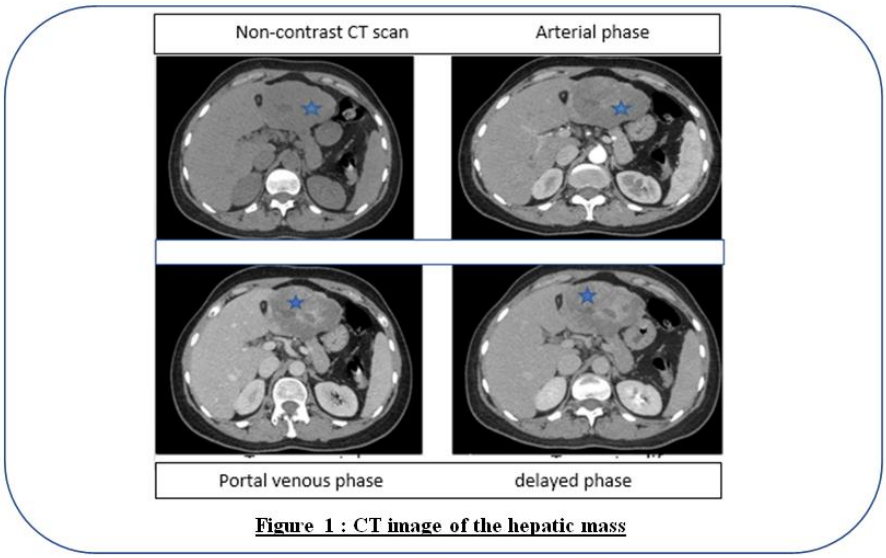

Figure 1 : CT image of the hepatic mass

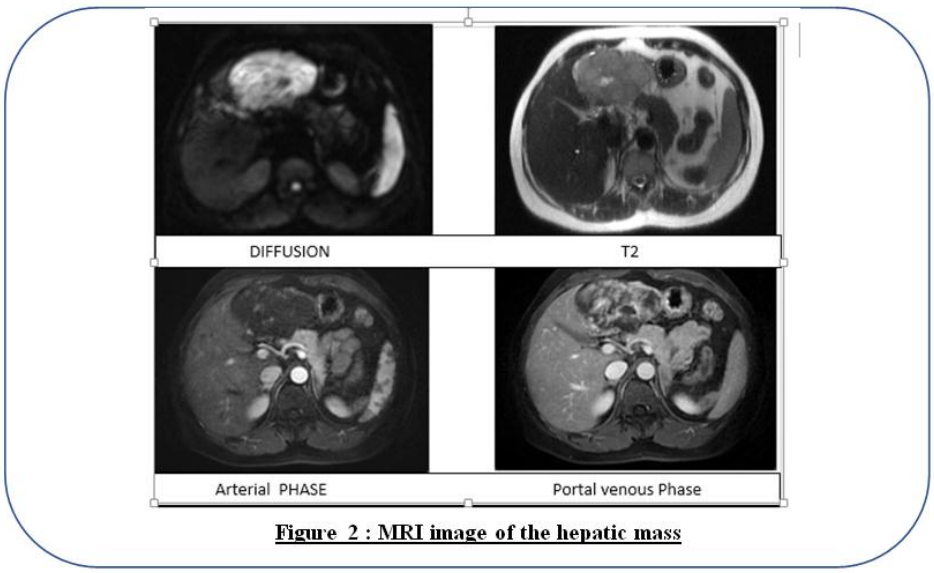

Figure 2 : MRI image of the hepatic mass

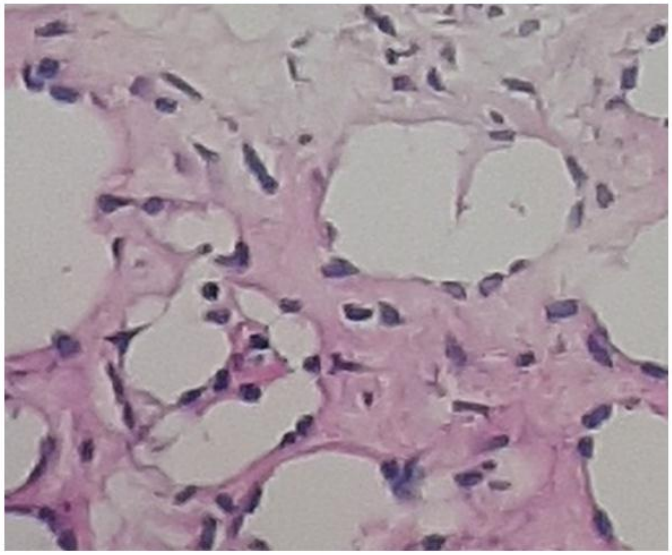

Figure 3 : Microscopic aspect during liver biopsy

3. DISCUSSION

Hepatic lymphangioma is rarely seen in routine practice, so no consensus is currently available. It could be misdiagnosed as malignant liver tumors or hepatic cysts, hence the interest of pathological study.[10]

The exact pathogenesis of lymphangioma is unknown, although congenital abnormality of lymphatic tissue development with dilatation of abnormal ducts and lymphatic obstruction is considered the main cause. Other pathogeneses discussed include trauma, inflammation and fibrotic processes, as well as disorders of vascular endothelial permeability.[12-13]

Most lymphangiomas are present at birth and may become symptomatic before the second year of life.[11] The average age of discovery is 30 years with a slightly higher incidence in women.[1-10-14]

The diagnosis in our patient was made at the age of 48 years.

The clinical presentation is nonspecific, and most cases are presented as abdominal pain or discomfort caused by the mass effect on adjacent organs or the mass on the affected side. Some cases present with no symptoms and are detected by routine medical examination.[15-16]

The ultrasonographic aspects of hepatic lymphangioma are varied, ranging from an anechoic, unicystic, or multiloculated mass containing cystic and solid components.[10]

The CT scan objectifies a lesion of low density overall and especially for the unilocular cystic mass and which does not enhance on arterial time. For the mixed-component multilocular mass, enhancement is observed for the septum at arterial time but not for the cystic parts. Based on these characteristics, the lesion can be successfully identified before surgery.[5-16]

Magnetic resonance imaging (MRI) of hepatic lymphangioma is not the first-line examination; when requested it usually shows a heterogeneous multi-lobulated mass with low signal intensity on T1-weighted images and high signal intensity on T2-weighted images.[10-18]

The initial diagnosis should be differentiated from simple cyst, cystadenoma, cystadenocarcinoma, hydatid cyst, sclerosing hemangioma, or a hepatocellular carcinoma.[10]

Although CT or MRI provide obvious features, it is still difficult to make the diagnosis of hepatic lymphangioma; therefore, pathological examinations have been suggested as the only safe way to identify the nature of the liver mass.[19]

Histologically, lymphangiomas are classified into three subtypes according to the congenital dilatation of the lymphatic ducts: capillary (super-microcystic), cavernous (microcystic) or cystic (macrocystic).[20-21]

Histologic features of cystic lymphangiomas include the following: flat endothelial lining of the cyst rather than cuboidal epithelium; lymphoid aggregates in the cyst wall; and smooth muscle in the cyst wall. In addition to these histologic features, the endothelial lining cells generally react positively with CD31 or factor VIII-related antigen but negatively with cytokeratin on immunohistochemical staining. Unlike hepatic lymphangiomas, biliary cystadenomas typically present as a cyst with a single wall of cuboidal to columnar epithelium supported by thick fibro-conjunctive tissue

with papillary folds in the cyst. In addition, biliary cystadenocarcinoma is identified by the presence of cytologically malignant proliferating epithelium.[20]

The treatment for hepatic lymphangioma is complete resection. Partial resection with cleaning of cystic fluid and needle aspiration certainly leads to recurrence.

The efficacy of sclerosing agent injection, which is effective for a simple liver cyst, is unknown.[22-23-24]

The use of immunosuppressive agents to treat liver lymphangioma or lymphangiomatosis has not been approved.[25]
Patients with unresectable giant liver lymphangiomas or with severe liver function impairment may be treated with orthotopic liver transplantation,[20-23-26]

Although the prognosis of solitary hepatic lymphangioma is favorable after surgical resection, it is recommended to continue follow-up even after transplantation given the risk of post-transplant recurrence.[23-25]

Conflicts of Interest
No conflict of interest for authors.

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