

Epithelioid Hemangioendothelioma of Liver; Report of a Case and Review of Literature

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Abstract

Case Report

Hepatic epithelioid hemangioendothelioma (HEH) is an extremely rare tumor of vascular origin. The disease can manifest itself in several symptoms and can be diagnosed by ultrasound, CT scan and MRI. Hepatic capsular retractions due to extensive fibrosis can be identified by CT and MRI. There is no consensus on the treatment of hepatic epithelioid hemangioendothelioma, which ranges from liver resection, hepatectomy, liver transplantation. This case report presents a 45-year-old woman who presented with hepatic epithelioid hemangioendothelioma diagnosed after the onset of jaundice.

Keywords: Hepatic epithelioid hemangioendothelioma; Hepatectomy; Liver tumors.

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INTRODUCTION

Epithelioid hemangioendothelioma (HEH) is an angiocentric vascular neoplasm of soft tissue, characterized by proliferation of endothelial cells with an epithelioid morphology. It can involve lung and mediastinum, thyroid, peritoneum, lymph nodes, bone, palate and liver. We report a case of a patient who presents with an epithelioid hemangioendothelioma discovered by a cholestatic icterus [1].

CASE REPORT

We report the case of a 45-year-old woman with no specific medical history. She presented with right hypochondrial pain dating back to 3 months, along with progressive cholestatic jaundice. On interrogation, a weight loss of 5 Kg was found.

The biological assessment showed a serum bilirubin level of 28 mg/L, hepatic cytolysis (transaminases levels at 3 times normal), and prothrombin level of 92 %. Tumour markers were not elevated.

A CT scan showed a normal sized liver, with hypodense nodular formations weakly enhanced after contrast. These lesions are confluent and located in the left liver as well as certain segments of the right liver; and suggest at first, the aspect of hepatic metastases, but no primary tumour was found. (Fig: 1 and 2).



Fig-1: Abdominal CT scan showing confluent hypodense nodular formations of the right and left liver

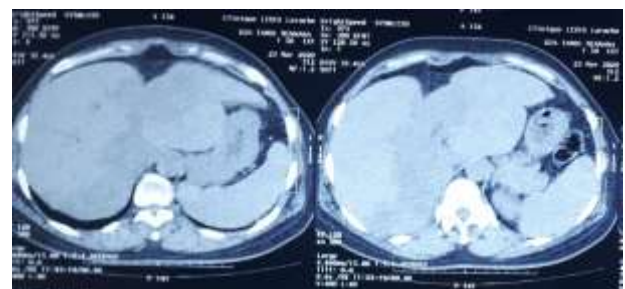


Fig-2: Abdominal CT scan showing the tumor with capsular retraction

An ultrasound-guided biopsy was performed; the histological and immunohistochemical profile was in favour of an epithelioid hemangioendothelioma, with a positive endothelial marker (anti ERG antibody) and a negative epithelial marker (anti CK7 antibody).

DISCUSSION

Epithelioid hemangioendothelioma is a rare primary liver tumor of mesenchymal origin. It usually affects middle-aged subjects (average age: 45) with a slight predominance of women (sex ratio: 3/2), with a reported incidence of one per million population [2].

There are no clear risk factors for the development of HEH.

Presentation of the disease is essentially nonspecific and the patients can have weight loss and right upper quadrant pain. In rare cases, jaundice and hepatic dysfunction may be noted due to replacement of liver parenchyma by the tumor. Tumor markers (N-fetoprotein, carcinoembryonic antigen, CA 19-9) remain normal in cases of HEH [3].

From a macroscopic point of view, the lesions have a multifocal character affecting the two hepatic lobes. They are in the form of nodules of variable size (0.5 to 12 cm) tending to converge. The topography of the lesions is rather peripheral with contiguous capsular extension. The capsular retraction is secondary to the significant fibrosis of the periphery of the nodular lesions. The spared and healthy segments show a compensatory hypertrophy [4].

Microscopically, epithelioid hemangioendothelioma is characterized by a large myxoid fibrous stroma. The key point in pathology diagnosis is based on immunohistochemical labeling: the epithelioid hemangioendothelioma appears positive for at least one endothelial marker and negative for epithelial markers [5].

CT scan or MRI allow finding some radiological signs suggestive of the pathological diagnosis such as capsular retraction, peripheral involvement, the possible presence of calcifications within the lesions and the alternation of atrophy of the tumor liver and hypertrophy of the non-tumor liver. After injection, the hypodense lesions can take on a target appearance with moderate peripheral contrast enhancement. On late acquisitions, the nodules become isodense [6].

There is no agreed treatment strategy for HEH owing to its rarity, heterogeneous status, and variable clinical outcome. The management modalities for patients with liver malignancies include surgical

resections, liver transplantation, tumor embolization, chemotherapy and radiotherapy, thermoablation, percutaneous ethanol injection, and even follow-up without any therapy. In a study of 286 HEH patients, 71 were not given any treatment and the reported mortality was more than 50%. Hence, a “no treatment” or “wait and see” strategy is not feasible. Liver transplantation is an accepted treatment option for HEH and is associated with excellent outcomes. [7-9].

CONFLICT OF INTEREST

The authors have no conflict of interest to declare.

REFERENCES

1. Daniela Redaelli, MBBS A* and Shaista Salman Guraya, PhD ba University of Milan Bicocca, Department of Radiology, Niguarda Hospital, Milan, Italy b College of Medicine, Taibah University, Almadinah Almunawwarah, KSA Received 28 January 2014; revised 30 May 2014; accepted 31 May 2014; Available online 3 September 2014
2. Hertl M, Cosimi AB. Liver transplantation for malignancy. *Oncologist*. 2005; 10(4): 269e281.
3. Ishak KG, Sesterhenn IA, Goodman MZD, Rabin L, Stromeyer FW. Epithelioid hemangioendothelioma of the liver:
4. Miller WJ, Dodd GD 3rd, Federle MP, Baron RL. Epithelioid hemangioendothelioma of the liver: imaging findings with pathologic correlation. *AJR Am J Roentgenol*. 1992; 159:53-7.
5. Makhlof HR, Ishak KG, Goodman ZD. Epithelioid hemangioendothelioma of the liver: a clinicopathologic study of 137 cases. *Cancer*. 1999; 85:562-82.
6. Alomari AI. The lollipop sign: a new cross-sectional sign of hepatic epithelioid hemangioendothelioma. *Eur J Radiol*. 2006; 59:460-4.
7. Kayler LK, Merion RM, Arenas JD, Magee JC, Campbell DA, Rudich SM, et al. Epithelioid hemangioendothelioma of the liver disseminated to the peritoneum treated with liver transplantation and interferon alpha-2B. *Transplantation* 2002; 74(1): 128e130.
8. Nagase M, Ryu M, Kinoshita T, Konishi M, Inoue K, Tiku V, Nawano S. Epithelioid hemangioendothelioma of the liver. *Journal of Hepato-Biliary-Pancreatic Surgery*. 2000 Aug;7(4):443-7.
9. Mehrabi A, Kashfi A, Schemmer P, Sauer P, Encke J, Fonouni H, Friess H, Weitz J, Schmidt J, Büchler MW, Kraus TW. Surgical treatment of primary hepatic epithelioid hemangioendothelioma. *Transplantation*. 2005 Sep 27;80(1S):S109-12.