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Case Report

Hepatic epithelioid angiomyolipoma: a malignant potential tumor and literature review

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Abstract: Hepatic epithelioid angiomyolipoma (HEAML) is an extremely rare primary hepatic tumor which is easily misdiagnosed as hepatocellular carcinoma or other tumors before surgery. It is a special kind of angiomyolipoma that is rarely reported in the liver. Patients with HEAML may recurrence or metastasis after surgery. The research on prognosis of the disease is poor due to its malignant potential. The epidemiology, clinical diagnosis, etiology and treatment of the disease remain challenging. It's important to improve the recognition of HEAML. Here, we report a woman who had complained of upper bellyache for 3 months and she was confirmed as primary HEAML by histology and immunohistochemistry after a laparoscopic hepatectomy. The patient had survived well for 1 year since surgery and long-term follow-up was continued till now. Previous studies and our experience suggested that surgical excision is the reasonable treatment of HEAML. Lastly, we have a brief discussion about HEAML and review the relevant literature.

Keywords: Epithelioid angiomyolipoma, liver, immunohistochemistry, pathology, laparoscopic hepatectomy

Introduction

Hepatic epithelioid angiomyolipoma (HEAML) is an unusual disease. To our knowledge, there were only a few cases about HEAML and most of them were case reports. The disease has non-specific symptoms and imaging characteristics. Computed tomography (CT) usually shows enlargement of the liver with multiple heterogeneous masses. Immunohistochemical staining reveals that human melanoma black 45 (HMB-45) is positive. The diagnosis of the disease remains challenging. In order to improve the recognition and treatment of HEAML, we described a 37-year-old female patient with HEAML and reviewed the literature.

Case report

A 37-year-old female with a 3-month history of pain in the upper abdominal quadrant was admitted to our hospital for treatment. Discomfort and pain were not related to meals, defecation or change in position, and could be tolerated. Following the occurrence of these symptoms, she had lost 3 kg in weight. On physical examination, it was significant for upper abdominal tenderness. The patient had a radical mastectomy because of the left breast cancer and. There was no significant evidence of tuberous sclerosis complex (TSC) and no family history of liver disease.

Laboratory examinations on admission revealed: blood tests were all within normal ranges; serum tumor markers, α-fetoprotein (AFP) was 15.09 μg/L, and HBsAg was positive. Computed tomography (CT) imaging of the abdomen at our institution revealed a 7.7×7.2 cm nodule in the left hepatic lobe (**Figure 1**). Multidetector spiral CT during arterial (a) and portal phase (b) of hepatic contrast enhancement demonstrated a large heterogeneous mass posteriorly in the left lobe of the liver. It was considered as a kind of primary hepatic carcinoma or breast cancer liver metastases. A laparoscopic hepatectomy was then performed not only for the purpose of curing the disease, but also for histopathologic diagnosis.

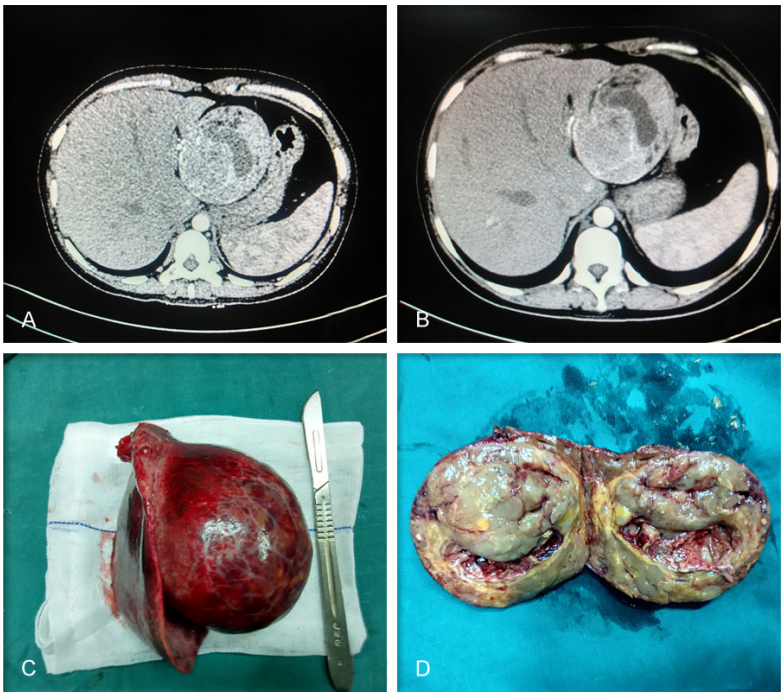

Figure 1. Multidetector spiral CT during arterial (A) and portal phase (B) show a tumor in the left hepatic lobe; (C) The tumor specimen 7.2×8.3 cm; (D) Cross-section of the specimen. CT and specimens images of the case of Hepatic epithelioid angiomyolipoma.

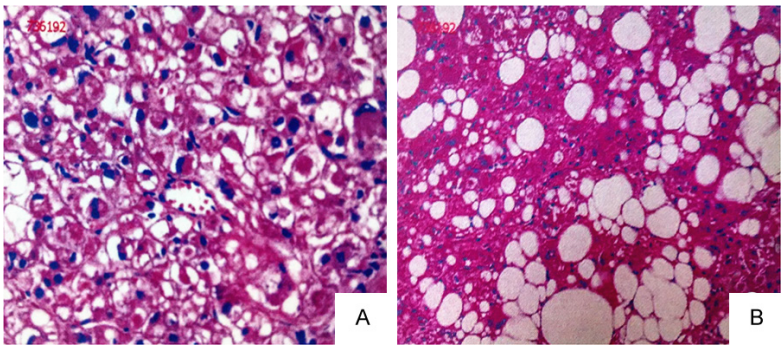

Figure 2. A. The lesion was lined by sheets of epithelioid cells (H and E, ×400); B. Scattered fat cells and a few thick-walled blood vessels (H and E, ×400). Pathological images of the case of Hepatic epithelioid angiomyolipoma.

The tumor was well encapsulated and demarcated with a largest diameter of 8.3 cm (**Figure 1C**). Cross-section of the specimen were variably gray to light-brown and nodular gray (**Figure 1D**). Then, a histopathologic diagnosis proved that it was a primary lesion of the liver. Microscopically, the lesion was lined by sheets of epithelioid cells, which were large with clear cytoplasm and prominent perinuclear eosinophilic condensations (**Figure 2A**). Scat-tered fat cells and a few thick-walled blood vessels were also seen (**Figure 2B**). Immunohistochemical analysis showed that the tumor cells were positive for HMB-45 (**Figure 3A**), SMA (**Figure 3B**), S100 (**Figure 3C**), Vim (**Figure 3D**) and CD68 (**Figure 3E**), but negative for CK (**Figure 3F**), EMA (**Figure 3G**), Des (**Figure 3H**) and Ki-67 (3%+) (**Figure 3I**).

The patient left the hospital after 7 days with no complications, and she was monitored with abdominal rep-eat CT after hospital discharge. She had survived well for 1 year after surgery with no abdominal uncomfortable and the follow-up still continued.

Discussion

Hepatic epithelioid angiomyolipoma (HEAML) was first described by Yamasaki in 2000 [1], it's generally thought to be one member of the PEComa family (a perivascular epithelioid cell tumor family originates from AML and some other tumors) [2]. The pathogenesis of the disease is still unclear. To our conclusion on the relevant papers, more women suffered from HEAML than men, at a ratio of 5:1. The disease is easily misdiagnosed with hepatocellular carcinoma (HCC) or other liver tumors [3], because it has non-specific symptoms such as abdominal pain or distension, discomfort and vomiting. The majority of the cases were discovered incidentally upon physical examination, but our patient had an upper bellyache possibly caused by tumor's compression. The patient may suffer from bellyache and the tumor would have a greater malignant potential when the tumor was large, but it is easily ignored when the tumor is small. Some papers described that 6%-10% of HEAML patients had associated tuberous sclerosis complex (TSC) [4], but the patient in this study was not related to TSC but with a 10-year his-

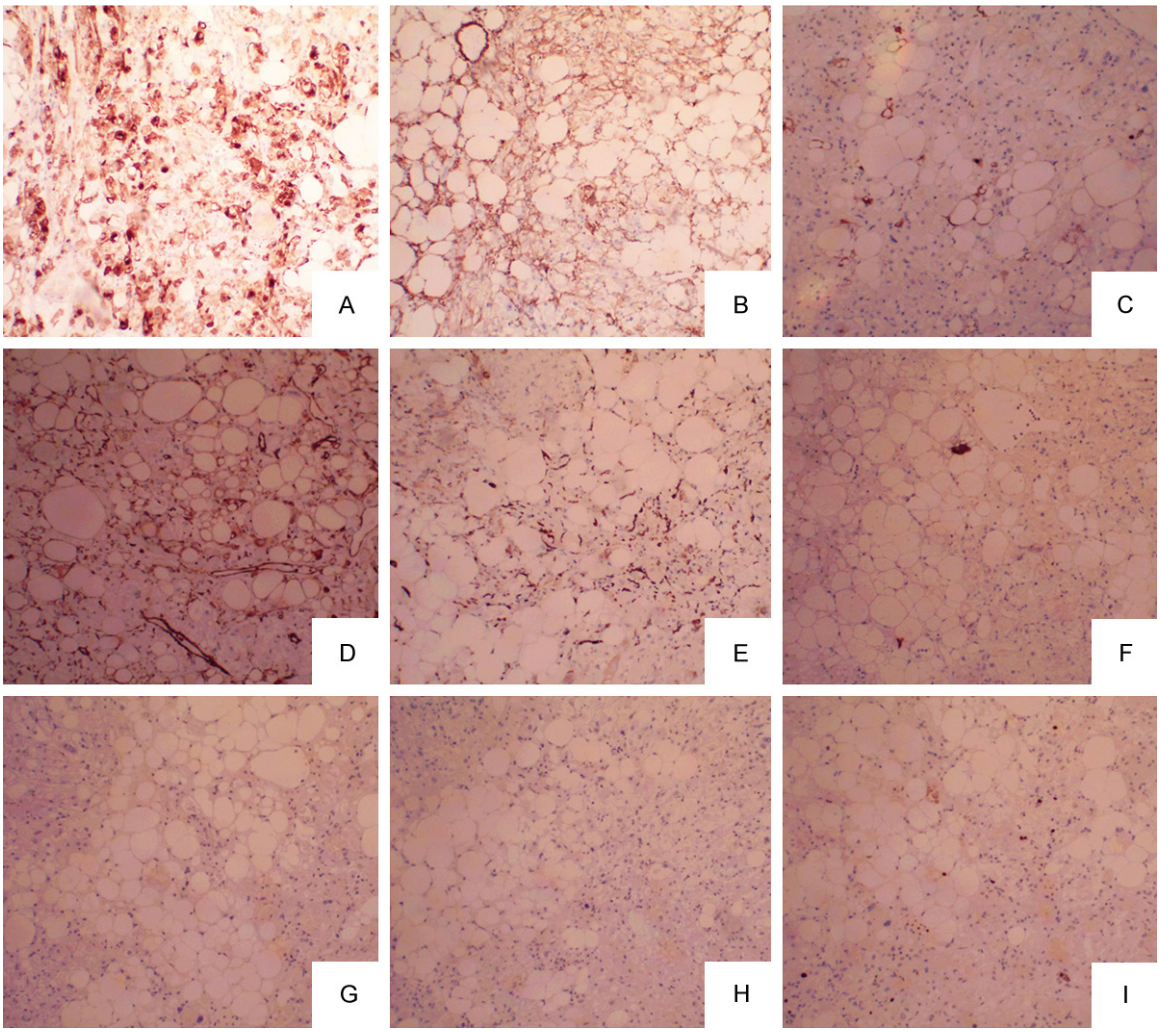

Figure 3. IHC staining images of the tumor, positive for HMB-45 (A), SMA (B), S100 (C) Vim (D) and CD68 (E); negative for CK (F), EMA (G), Des (H) and Ki-67 (I) (×400). IHC staining images of the case of Hepatic epithelioid angiomyolipoma.

tory of HBsAg positive. Thus it's worthwhile to research the link between hepatitis virus and HEAML.

Based on previous reports, laboratory blood tests usually showed within normal ranges, and serum AFP rose a little in our report. HEAML always presents with less typical imaging manifestations because of the less of fat cells, but more smooth muscle and vascular, and it's easily misdiagnosed with HCC. It is reported that most cases of HEAML were obviously enhanced in the early arterial phase in dynamic enhanced Computed tomography [5], but showed low density in the portal venous phase and delayed phase in magnetic resonance imaging [6]. Our search conformed the imaging features. But it is difficult to discriminate between HEAML and HCC. Alatassi [7] reported a confirmed case by fine-needle aspiration biopsy, but it is a serious problem that hemorrhage and peritoneum metastasis caused by the method. It is difficult to diagnose before surgery. Our patient was misdiagnosed as HCC or breast cancer liver metastases before surgery. Lastly she was diagnosed with primary HEAML by histopathologic and immunohistochemical after a laparoscopic hepatectomy. Pathology is thought as the only definite diagnostic criteria. HEAML is usually a solitary but multifocal tumor, and the presence of epithelioid cells is important for the diagnosis of HEAML. It could be defined as an EAML if epithelioid cell components are higher than 10% while others believed this

standard should be as high as 50%, or even more than 90% [8]. It's reported that HEAML cell is positive for cell markers including HMB-45 and SMA, but negative for EMA and CK, the expression of Ki-67 could indicate its malignancy by immunohistochemical staining. This is regarded as the most important criterion for the diagnosis of HEAML. And HEAML should be diagnosed by pathology after surgery.

HEAML is usually thought as a benign tumor for its biological behavior in several series of case reports, but HEAML has malignant potential and may metastasize to other organs [9], and we have no evidence to determine whether the same prognostic parameters of renal EAML are applicable to HEAML. But Liu thought it is a malignant potential tumor by analyzing the clinicopathological data and imaging results of HEAML cases ever reported [10]. Surgery is the mainly effective method to cure HEAML, and a laparoscopic hepatectomy is the first choice for its advantage to patients. There is little literature about chemotherapy and radiation therapy on HEAML. Considering the potential risk of malignant changes of HEAML, a long-term follow-up should be performed. In our report, the patient had survived well for 1 year after surgery with no recurrence and metastasis.

Summarize

This case report together with the previous literature review provide an alert for clinicians about primary hepatic epithelioid angiomyolipoma. Early diagnosis is important for it because of its malignant potential, the right treatment should be acted, and laparoscopic hepatectomy is a good choice for the patients.

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Disclosure of conflict of interest

None.

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