Brief Report

Hepatic perivascular epithelioid cell tumor in three patients

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ABSTRACT: Perivascular epithelioid cell tumor (PEComa) is a rare, soft tissue tumor that can occur in various locations. The present report included three patients (one male and two females; age range, 25-51 years) with hepatic PEComas. The collected data included the clinical manifestations, diagnosis, management, treatment, and prognosis. Since it is difficult to diagnose hepatic PEComas by imaging, the patients were diagnosed by tumor tissue examination such as immunohistochemistry, which was positive for HMB-45, Melan-A, and SMA on all slides. The tumor was composed of diverse tissues including smooth muscle, adipose tissue, and thick-walled blood vessels. During the follow-up period, one of the tumors was malignant (double-positive for CD34 and Ki-67) and recurred 3 months after surgery. In addition, malignant hepatic PEComas were reviewed in the literature, indicating that the majority of hepatic PEComas are benign, but few hepatic PEComas exhibit malignant behaviors in older female patients (>50 years of age) with abdominal discomfort and pain, larger tumor size (>10 cm), or positive staining for CD34 and Ki-67. In conclusion, there is no effective method to diagnose PEComas. Currently, the diagnosis of PEComas depends on immunohistochemical staining. Tumor resection and close follow-up are the principal methods for the management of PEComas.

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KEY WORDS: perivascular epithelioid cell tumor, hepatic; diagnosis; hepatectomy; recurrence

Introduction

Perivascular epithelioid cell tumor (PEComa) is classified as a mesenchymal tumor that is composed of histologically and immunohistochemically distinctive perivascular epitheliod cells according to the World Health Organization (WHO).[1] PEComas are found in a variety of locations, including the uterus, vulva, rectum, heart, breast, urinary bladder, abdominal wall, pancreas, retroperitoneum, and liver[2, 3] and most commonly, in the kidneys. Hepatic PEComa is rare[4, 5] and therefore, our knowledge about it is limited. Indeed, the biological features, diagnosis, and treatment of primary hepatic PEComas have not been elucidated. In the present study, we described the clinicopathologic features, diagnosis, treatment, and follow-up of three pathologically-proven patients with primary hepatic PEComas treated in our center. Furthermore, we reviewed and compared the characteristics of malignant hepatic PEComas with those reported in the literature.

Methods

The three patients with PEComa were pathologically confirmed at the First Affiliated Hospital of Nanjing Medical University between January 2010 and December 2014. Demographic data, clinical history, and clinical presentation, including gender, age, family history, symptoms, hepatic function, tumor markers, ultrasonography and computed tomography (CT) findings were collected. All of the patients underwent hepatectomy. Tumor specimens were taken and fixed with 10% neutral formaldehyde, then embedded in paraffin. The specimens were

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sectioned at 4 μm thicknesses and stained with hematoxylin and eosin (HE). SMA, HMB-45, Melan-A, S-100, E-cad, Ki-67, Desmin, Hep-1, CK8/18, CD31, CD34, CK7, and AFP in the specimens were detected by immunohistochemistry (IHC). The specimens were analyzed independently by one pathologist. The histopathologic diagnosis was made according to the WHO classification of tumors of the liver and intrahepatic bile ducts.[6] All of the patients were followed up after hepatectomy independently by one researcher. In addition, the patients with malignant hepatic PEComas reported elsewhere were searched and reviewed from the PubMed database.

Results

Clinical history and presentation

The demographic data of the three patients with PEComa are shown in Table 1. Hepatic PEComas are rare, and usually occur in patients with tuberous sclerosis[5] or Li-Fraumeni syndrome.[7] But both diseases did not appear in our patients. The specimens were obtained from one male and two female patients, who were at age of 25-51 years. They were not symptomatic and received no preoperative treatment. Liver function tests and tumor markers were normal in all patients. One patient was hepatitis B virus (HBV)-positive. All patients had a single tumor, which was resected subsequently. According to the WHO classification of tumors of the liver and intrahepatic bile ducts, the most important diagnostic criterion was the presence of HMB-45 and Melan-A positive myoid cells.

Imaging features

All patients underwent ultrasonography (Fig. 1A) and CT examinations (Fig. 1B, C, and D). Sonographically, the tumors appeared as homogeneous or heterogeneous hyperechoic masses. CT scan showed well-demarcated masses with low-density heterogeneous areas in all

Table 1. Demographics and clinical characteristics

Variables	Patient 1	Patient 2	Patient 3
Gender	Female	Female	Male
Age (yr)	51	30	25
Family history	No	No	No
Symptom	No	No	No
Hepatitis	Positive (HBV)	Negative	Negative
Liver function	Normal	Normal	Normal
Tumor marker			
AFP (ng/mL)	1.3	4.0	2.3
CA19-9 (U/mL)	6.7	14.5	9.7
CEA (ng/mL)	2.4	0.8	1.8
Tumor			
Number	1	1	1
Size (cm)	8.0	2.5	8.0
Location	VI	VIII	VI
Boundary	Still clear	Clear	Clear
PD	FNH	HCC	HCH
Treatment	Hepatectomy	Hepatectomy	Hepatectomy
Follow-up (mon)	6	8	36

HBV: hepatitis B virus; PD: preoperative diagnosis; FNH: focal nodular hyperplasia; HCC: hepatocellular carcinoma; HCH: hepatic cavernous hemangioma.

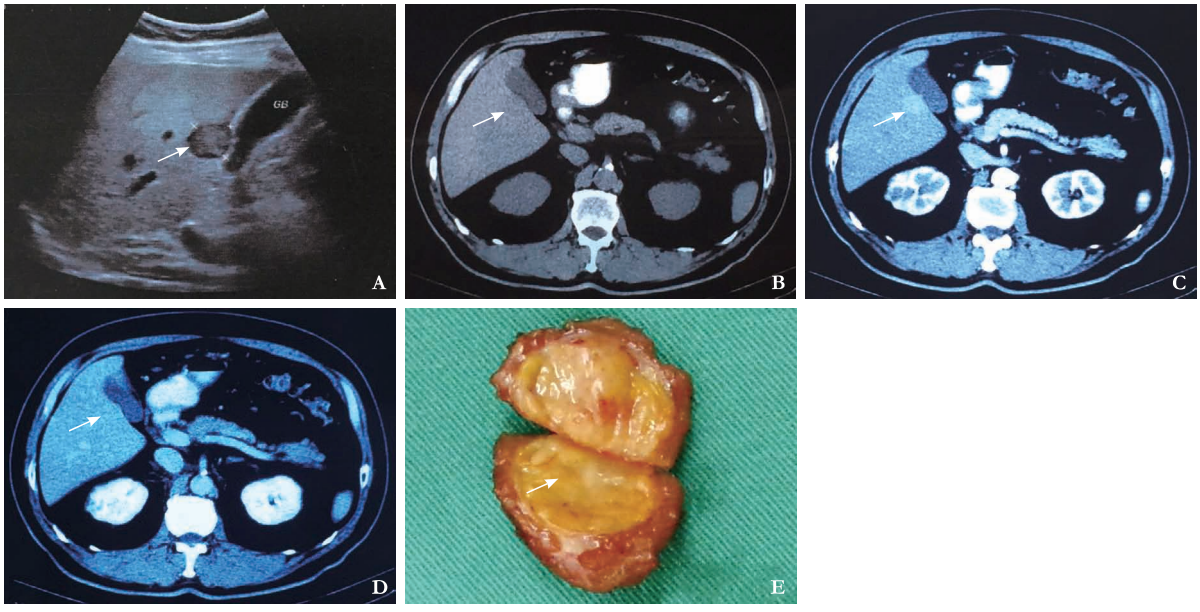

Fig. 1. Images and specimens of hepatic PEComas. **A**: ultrasonography of PEComa (arrow); **B** (plain CT scan) (arrow), **C** (arterial phase) (arrow), and **D** (venous phase): CT scan of hepatic PEComa (arrow); **E**: Operative specimens of hepatic PEComas (arrow).