

# Perivascular epithelioid cell tumor of the liver: a case report and literature review

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**Abstract.** – Perivascular epithelioid cell tumors (PEComas) are mesenchymal tumors containing variable component of smooth muscles, fat and vessels. They occurred pretty rare in the liver and no characteristic of imaging data have been demonstrated up to now. We herein present a case of a 58-year-old man with a hepatic PEComa which was correct diagnosed through immunohistochemical confirmation with HMB-45. The clinicopathological and imaging features of hepatic PEComa were analyzed retrospectively. This is the first paper which demonstrated the characteristic of imaging data among liver PEComas and other liver lesions. However, the diagnostic criteria of PEComa depends on pathology. Hepatectomy is the effective treatment.

*Key Words:*

Liver neoplasms, Perivascular epithelioid cell, Computed tomography, Imaging data features.

## Introduction

Perivascular epithelioid cell tumors are defined by the WHO as unusual mesenchymal tumors which present histologically and immunohistochemically distinctive perivascular epithelioid cells<sup>1</sup>. From our review of the related literature, PEC-derived tumors have been documented in various anatomical locations, including the uterus, skin<sup>2,3</sup>, liver/falciform ligament, upper aero-digestive<sup>4</sup>, bone<sup>5</sup>, pancreat<sup>6</sup>, colon<sup>7</sup> et al. The uterus is the predominant site. PEComas of the liver are rare and occur frequently in women in their fifth decade<sup>1,8</sup>. However, no characteristic of liver PEComas' imaging data have been demonstrated. Herein, we describe a case of a 58-year-old man with a hepatic PEComa and showed its features of the contrast enhancement computed tomography (CT). This case emphasizes the importance of imaging data for correct pre-operative diagnosis in liver PEComa.

## Case Report

A 58-year-old man complained of abdominal distention for over 10 days. Physical examination showed upper abdominal tenderness without rebound-tenderness. A mass was touched in the right hypochondriac region. The laboratory examinations such as blood, stool and urine routines were normal. Not only liver function parameters but also serum tumor markers including AFP were within normal limits. Both HBsAb and HBcAb were positive.

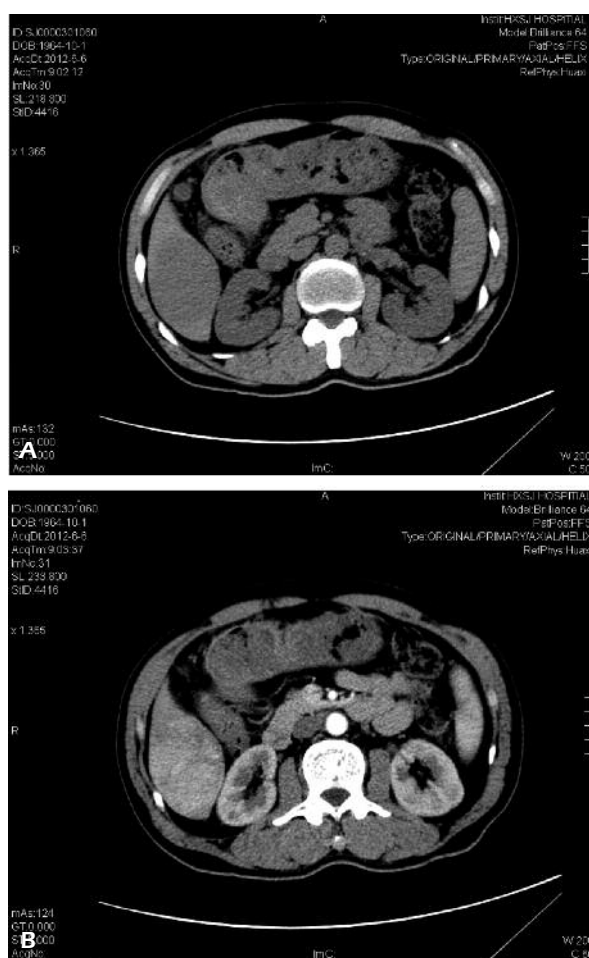
Ultrasonography revealed a well-circumscribed heterogeneous hypoechoic mass in the right hepatic lobe. On pre-contrast Computed tomography (CT), a huge heterogeneous and lower density lesion with a well demarcated margin, sized 7.6 x 5.2 cm, was found in the inferior segment of right lateral lobe of the liver (Figure 1A). The lesion was obvious and heterogeneous enhancement on arterial phase and being slightly hypodense on the portal venous phase (Figure 1B).

The diagnosis was hepatocellular carcinoma (HCC) pre-operative. The patient underwent right IV segmentectomy in our hospital.

Gross examination demonstrated a 6 x 5 x 4 cm, solitary lesion with a well-demarcated margin. The mass was smooth and soft. Cut surface was white-tan to gray-red. The hepatic tissues around the mass were gery yellow with nodular cirrhosis. The pathological diagnosis was established as PEComa of the liver.

On microscopic examination, there were a majority of epithelioid and spindle tumor cells with eosinophilic cytoplasm arranged disorganized. The nuclei often contained discernible nucleoli, and necrosis was inconspicuous. Particularly, many lymphocytes and fibroblasts/myofibroblasts were invaded in the neoplasm (Figure 2A).

Immunohistochemical study showed that the lesions were positive for HMB-45 (Figure 2B), MART-1, while SMA and ALK-1 were focally positive. The neoplastic cells failed to stain with antibodies against S100, CK, EMA, Happer-1 (Figure 2C).



**Figure 1.** **A**, Pre-contrast CT scan revealed a lower density lesion in the right lobe of liver. **B**, Contrast-enhanced CT scan demonstrated the hyperintense mass with a well-demarcated tumor margin.

The correct diagnosis was liver perivascular epithelioid cell tumor. The patient recovered uneventfully and was discharged two weeks after surgery.

## Discussion

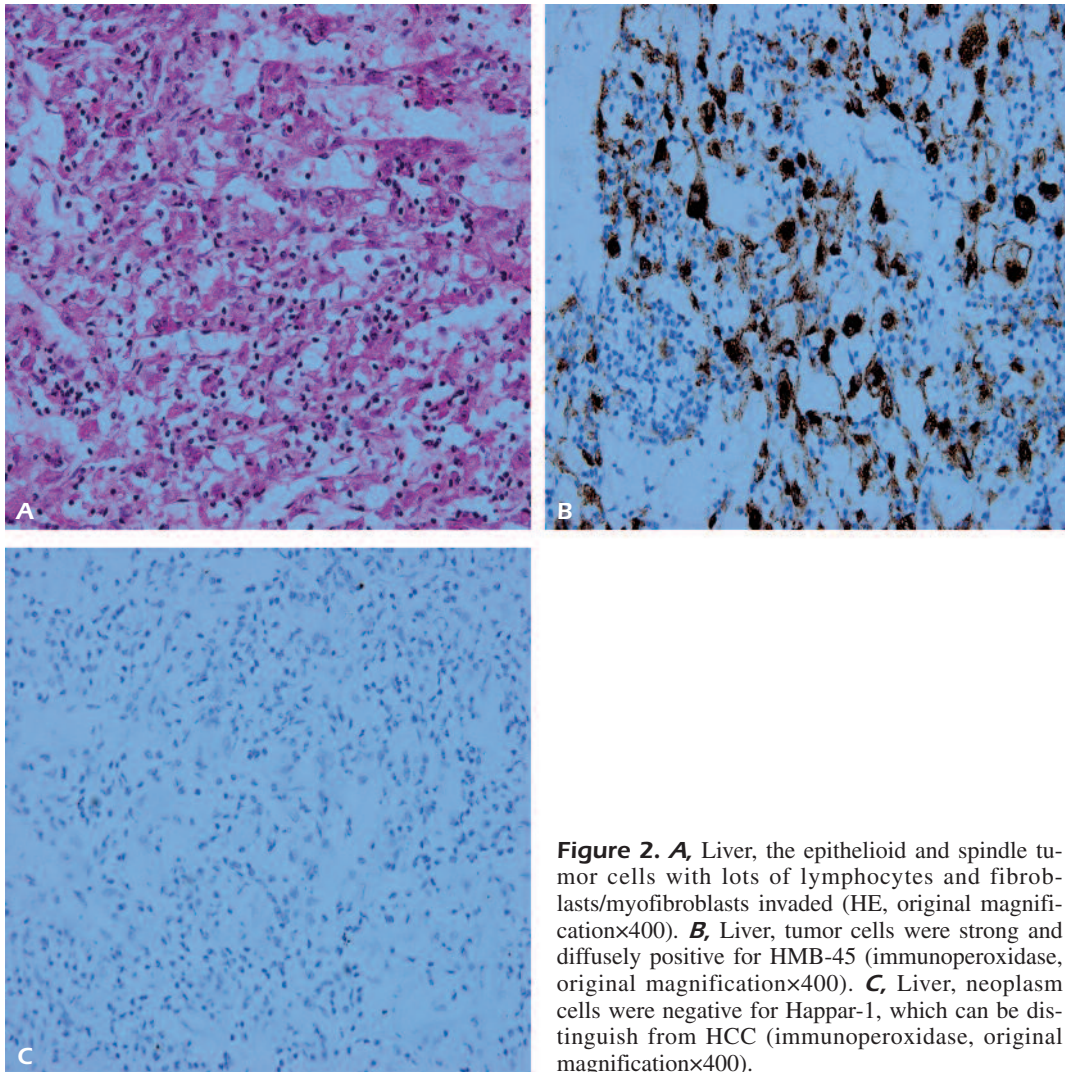
PEComas can be found in various organs and can occur at any of the age, but the median age is 43 years. A strong female predominance (4:1 female-to-male ratio) was documented in the latest review<sup>2</sup>. Hormones may play a role in their pathophysiology.

Hepatic PEComa has no specificity in clinical presentation. The tumor was often found incidentally in physical examinations. As the growing size of the lesion, the disease would be sympto-

matic in some patients. Abdominal pain or distress, followed by malaise and upper abdominal mass or hepatomegaly, were the most common symptoms<sup>9</sup>. Sometimes, a painless mass could be touched in the soft tissue.

Hepatic PEComa is a type tumor which often mimicking other liver lesions such as hepatocellular carcinoma (HCC), hepatic metastasis, focal nodular hyperplasia (FNH) and liver hemangioma. The combination of imaging data of ultrasonography, contrast enhancement CT, magnetic resonance imaging (MRI) is useful for the preoperative diagnosis of hepatic PEComa. On ultrasonography, most space-occupying lesions show heterogenous low echo. The presence of fat components within the tumor were demonstrated as hypodense area. However, variable relative proportions of fatty tissues in the neoplasm, from less than 10% to over 90% of the lesion volume, result in various imaging performances<sup>10</sup>. Contrast enhancement CT has higher accuracy than ultrasonography. According to Yan et al's<sup>11</sup> review, a total of 75% of neoplasms enhanced inhomogeneously on arterial phase scanning with punctual or curved vasculature be seen. HCC usually marked early in the arterial phase and decreased rapidly in the portal venous phase. However, when HCC contains partly fatty metamorphosis, the difference needs to be made via histology. HCC are positive for Haparr-1 and most of the AFP is in a high level, which can be distinguish from PEComa. FNH and HAML have the similar enhancement pattern<sup>11</sup>. During the delayed phase, both of them show a prolonged enhancement. But the central scar of FNH demonstrate no enhancement in the arterial phase. Last but not the least, the blood vessels in HCC or FNH often located in the periphery of the neoplasms. As to liver hemangioma, MRI could be useful because of the characterization imaging, namely classic "light" on the T2-weighted imaging.

Although the diagnostic criteria of PEComa depends on pathology. We hope to establish some diagnostic criteria including the imaging data such as CT in the future. PECs frequently have the coexpression of muscle markers (smooth muscle actin, desmin, muscle myosin, and pan-muscle actin) and melanocytic markers (HMB-45, Melan-A/MART-1, microphthalmia transcription factor). Cytokeratins and S-100 protein are usually negative. In Fole et al<sup>12</sup> review, all the 61 cases of PEComas were HMB-45 positive, while cytokeratin negative.



**Figure 2.** **A**, Liver, the epithelioid and spindle tumor cells with lots of lymphocytes and fibroblasts/myofibroblasts invaded (HE, original magnification×400). **B**, Liver, tumor cells were strong and diffusely positive for HMB-45 (immunoperoxidase, original magnification×400). **C**, Liver, neoplasm cells were negative for Happar-1, which can be distinguish from HCC (immunoperoxidase, original magnification×400).

Tumor resection seems to be the mainly and effective treatment for primary and malignant PEComas. In this case, the neoplasm did not demonstrate infiltrative growth over 9 months, yet a close and long-term clinical follow-up is suggested.

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#### Conflict of Interest

None.

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