Perivascular Epithelioid Cell Tumor (PEComa) of the Liver Diagnosed by Contrast-enhanced Ultrasonography

Reiko Akitake, Hiroyuki Kimura, Satoru Sekoguchi, Hideki Nakamura, Hiroshi Seno, Tsutomu Chiba and Sotaro Fujimoto

Abstract

Perivascular epithelioid cell (PEC) is a unique cell which expresses both myogenic and melanocytic markers, and forms PEComa. A 36-year-old woman presented with a 35 mm-diameter liver tumor. MRI showed poor fat component in the tumor. Contrast-enhanced ultrasonography using the newly developed enhancing reagent, Sonazoid, clearly demonstrated early-phase enhancement of the tumor and rapid drainage of the reagent to veins, suggesting a PEComa. Lateral segmentectomy of the liver was performed. Histologically, epithelioid tumor cells around the vessels were immunostained with both HMB-45 and α-smooth muscle actin, confirming the diagnosis of PEComa. No recurrence has been found for 18 months following the operation.

Key words: PEC tumor, epithelioid angiomyolipoma, HMB45, α-smooth muscle actin, Sonazoid

(Inter Med 48: 2083-2086, 2009)  
(DOI: 10.2169/internalmedicine.48.2133)

Introduction

Perivascular epithelioid cell tumor (PEComa) is a new classification category established in the World Health Organization Classification of Tumors in 2002 (1). Perivascular epithelioid cell (PEC) is a hybrid tumor cell immunoreactive with both melanocytic (HMB-45 and/or Melan-A) and smooth muscle (α-smooth muscle actin and/or desmin) markers, the normal counterpart has not been identified (2). Here we present a case of PEComa in which contrast-enhanced ultrasonography using the newly developed enhancing reagent, Sonazoid, revealed hypervascularity and an arteriovenous shunt in the tumor and was useful for the preoperative diagnosis.

Case Report

A 36-year-old woman was admitted to Kyoto First Red Cross Hospital in March of 2007 for further examination of the liver tumor which was detected by ultrasonography in the annual health check. She had been asymptomatic and no tumor was observed in the annual health check in 2005.

Physical examination showed no abnormalities. Hematological and chemical studies, including tumor markers such as α-fetoprotein and carcinoembryonic antigen, gave normal results. Hepatitis virus markers, such as hepatitis B surface antibody, hepatitis B surface antigen and hepatitis C antibody, were all negative. Conventional ultrasonography revealed well-demarcated isoechoic tumor with a diameter of 35 mm in the segment 2 of the liver (Fig. 1a, left). Power Doppler ultrasonography showed that the marginal low-echoic lesion of the tumor was composed by displaced vessels around the tumor (Fig. 1a, right). Contrast-enhanced ultrasonography using a newly-developed enhancing reagent, Sonazoid, showed that the tumor was enhanced in early arterial phase and the reagent rapidly flowed into drainage veins in no more than one second after the infusion, which indicated hypervascularity and arteriovenous shunt in the tumor (Fig. 1b, c). In magnetic resonance imaging (MRI) and computed tomography (CT), our patient could not undergo contrast-enhanced studies because of the presence of bronchial asthma. In non-enhanced MRI, the tumor showed low intensity on T1-weighted image and high, but not extremely high, intensity on T2-weighted image (Fig. 2a-e). There was no significant difference in the intensity of out-of-phase and...
Figure 1. (a) left; Ultrasonography showed a well-demarcated isoechoic tumor with a diameter of 35 mm in segment 2 of the liver. right; Power Doppler ultrasonography revealed that displaced vessels encircled the tumor. (b, c) Sonazoid contrast-enhanced ultrasonography showed early influx of arterial blood into the tumor (b; arrow) and rapid drainage of the reagent (c; arrow) suggesting existence of arteriovenous shunt. Note good enhancement of the tumor (b, c; arrowheads) and intratumor vessels (c; arrowheads) indicating hypervascularity of the tumor.

Researchers at the University of Verona first suggested the term perivascular epithelioid cell (PEC) in 1996 (3), and the World Health Organization defined PEComa as ‘a mesenchymal tumor composed of histologically and immunohistochemically distinctive perivascular epithelioid cells’ in 2002 (1). ‘PEComa family’ is now composed of epithelioid angiomyolipoma, clear cell ‘sugar’ tumor, lymphangioleiomyomatosis and clear cell myomelanocytic tumor of the fallopian ligament/ligamentum teres (1, 2). However, there still remain confusion and controversy on the use of terms ‘PEComa’, epithelioid angiomyolipoma and ‘classic’ angiomylipoma (2). These three tumors are found ubiquitously in various organs, and a considerable number of cases have been reported in the liver (2, 4-7). Among them, to our knowledge, only seven cases of the liver have been reported using the term ‘PEComa’ (4-7). Researchers at the University of Verona define PEComa (or epithelioid angiomyolipoma) as a neoplasm without adipocytes, and discriminate them from classic angiomyolipoma (2). According to their definition, we used the term PEComa in this case report.

Here, we presented a liver PEComa case in which dynamic study of MRI and CT could not be performed because of the presence of bronchial asthma and contrast-enhanced ultrasonography was alternatively useful for preoperative diagnosis. Diagnostic imaging of PEComa shows a wide variety of patterns (4, 6, 7). Previous reports have suggested hypervascularity and arteriovenous in contrast-enhanced CT as a feature of PEComa (4, 6, 7). In the present case, Sonazoid contrast-enhanced ultrasonography was useful to show early influx into the tumor and rapid drain-
Figure 2. (a, b) T1-weighted MRI showed a hypointense tumor. There was no significant intensity difference in out-of-phase (a) and in-phase (b). (c) T2-weighted MRI showed a hyperintense tumor. (d) Fat-suppressive MRI did not show suppression of hyperintensity of the tumor, indicating poor fat component. (e) SPIO-MRI demonstrated low uptake of SPIO into the tumor. (f) Pre-contrast CT demonstrated a low-density tumor.

Figure 3. (a) At surgery, the tumor was an elastic hard mass projecting from the lower surface of the liver. (b) Gross appearance of the tumor was a smooth, light brown mass containing partial hemorrhage.

age to veins of arterial blood. In addition, power Doppler ultrasonography, which revealed displaced vessels around the tumor, was useful to rule out typical hepatocellular carcinoma with fibrotic capsule. Taken together with MRI find-
Figure 4. (a) Hematoxylin and Eosin staining showed large polygonal cells with enlarged nuclei and abundant cytoplasm in the tumor. ×400 (b, c) Immunostaining showed strong and diffuse expression of HMB-45 only in the tumor cells (asterisk in b, ×100: c, ×400) (d) Immunohistochemistry showed expression of α-smooth muscle actin in some of the tumor cells (×400).

ings which showed poor fat component in the tumor, we first suspected PEComa.
To diagnose PEComa definitely, histological findings are essential (1, 2). PEComa cells are characterized by their perivascular location, often with radial arrangement around the vascular lumen. Typically, tumor cells are epithelioid and spindle-shaped resembling smooth muscle cells, and tend to have abundant clear to eosinophilic pale granular cytoplasm. The most important histological finding is positive immunostaining with both melanocytic (HMB-45 and/or Melan-A) and smooth muscle (actin and/or desmin) markers. In the present case, those histological findings, especially positive staining with both HMB-45 and smooth muscle actin, were critical for the final diagnosis.

PEComa is considered as a benign tumor, but there have been several reports that showed distant metastasis even after surgical resection of the original tumors (1, 2, 5). Also in the present case, a close and long-term follow-up may be necessary due to its rapid growing history.

References