Perivascular epithelioid cell tumors (PEComas) belong to a family of mesenchymal neoplasms that include angiomylipoma, clear-cell “sugar” tumors of the lung and other organs, lymphangiomatosis, and a rare group of morphologically and immunophenotypically similar lesions that may affect soft tissues, viscera, and bones (1-3). Perivascular epithelioid cells were named by Bonetti et al. (4) to describe the epithelioid cells found in these neoplasms. PEComas, which are immunoreactive with melanocytic markers, have been reported to occur at almost all body sites, and have a predilection for the liver with diphilic cytoplasm, and perivascular distribution. The uterus is the most prevalent reported site of involvement. Primary hepatic PEComas are extremely rare, and only 12 cases have been reported before (5-15), most being monotypic or epithelioid angiomylipomas. Most cases of PEComa are benign, but two cases of metastatic PEComa have been reported (6). Histological features of the liver have been misdiagnosed by imaging as focal nodular hyperplasia, adenoma, or hepatocellular carcinoma. No angiographic findings of PEComas have been previously reported.

We report a case of PEComa of the liver diagnosed by ‘early washout’ of the lesions on computed tomography (CT) and angiographic scans.

Case report

A 51-year-old woman with a history of ovarian cancer received an oophorectomy 34 years earlier and wedge resection for a gastric tumor (intramuscular gastrointestinal stromal tumor [GIST]) with submucosal leiomyoma) 3 years earlier. The patient had no history of tuberous sclerosis. She had poor appetite, with body weight loss of 6 kg in 1 month. Abdominal ultrasound showed a mixed-echoic mass in the left lobe of the liver. CT scan of the abdomen showed a 9-cm well-defined heterogeneously enhancing mass lesion in segments 2 and 3 of the liver. The mass was enhanced in the hepatic arterial-phase scan with early washout in the portal venous-phase in the lateral segment of the liver. Angiography revealed a hypervascular tumor. US-guided rapid biopsy of the mass was performed. On histological examination, central necrosis was found in the part of the capsule (Fig. 1). The tumor displaced the left gastric artery and compressed the stomach. Hepatocellular carcinoma (HCC) was diagnosed and treated with surgery. At surgery, a well-encapsulated elastic-to-firm tumor measuring 9.0 × 8.8 × 5.8 cm was found in the left hepatic lobe, and hepatic S2 and S3 bisegmentectomy was performed. On histological examination, central necrosis was found in the part of the capsule showing low density on the CT scan. The parenchymal tumor resembled a PEComa, and histological examination of tumor sections revealed numerous tortuous vessels with radiating sheets, nests, and fascicles of epithelioid and spindle cells displaying clear to granular eosinophilic cytoplasm, round to oval nuclei, prominent nucleoli, and 1-2 mitotic figures per 10 high-power fields. Focal multinucleated giant cells and cytologic atypia were observed (Fig. 3). The tumor cells were diffusely positive for HMB-45, vimentin, and smooth muscle actin; focally positive for S-100 protein, and negative for cytokeratin, desmin, CD31, CD117, and CD34, immunohistochemically confirming the diagnosis of hepatic PEComa (4). The patient had no tumor recurrence at her 9-month follow-up visit.

Discussion

The clinical presentation of hepatic PEComa has no specificity. The symptoms and signs are similar to those of other tumors arising from the liver. Generally, indigestion, loss of appetite, nausea, body weight loss, and intermittent colic pain can occur, and on physical examination, tenderness to palpation and liver enlargement may be observed. These tumors have occurred in female patients aged from 13 to 70 years (median age: 55) (5-15). Of the reported 12 cases (5-15), 6 cases occurred in the left lobe (7-9, 11, 14, 15), 4 in the right lobe (5, 6, 10, 12), 1 in the caudate lobe (7), and 1 in the ligamentum teres (13). Tumor sizes ranged between 1.5 and 20 cm. Four patients had normal levels of serum AFP and were negative for hepatitis B surface antigen and anti-hepatitis C virus antigen (6, 10, 12, 15); reports of the other 8 cases did not mention this. Two patients had a history of melanoma (13, 14), and 1 had a history of GIST (8). Four had no history of tuberous sclerosis.

Our patient had a history of GIST in the stomach 3 years prior to diagnosis of PEComa in the liver. Carloss et al. (8) reported a case of PEComa coexisting with GIST. Loss of heterozygosity (LOH) in chromosome 16p (harboring the TSC2 locus) has been detected in some PEComas, but it has not been investigated in GISTs (1, 3). Based on this finding, we conclude that the coexistence of these two lesions in our case is a coincidance.

PEComas are characterized by their perivascular location, often with a radial arrangement of cells around the vascular lumen. Typically, the cells in PEComas are mainly epithelioid in the immediate perivas-
The PEComas in these 12 cases had no fat component, hemorrhage, cystic component, or central scarring on CT or MRI. Necrosis was present in large but not small PEComas. In 4 cases, the diagnosis of PEComa was biopsy proven, and complications were not mentioned. The resections of these 12 tumors ranged from tumorectomy to hemihepatectomy (5-15). In most cases, PEComas are benign, and patients have good postoperative prognosis and no recurrence after several months of follow-up. However, in 2 of the 12 cases, the PEComas metastasized (5, 10). Thus, PEComa is potentially malignant, and surgery is required when biopsy-proven PEComa is diagnosed.

Folpe and colleagues (3) suggested diagnostic criteria for malignant PEComa, including a size greater than 8.0 cm, mitotic count of more than 1 per 50 high-power fields, and necrosis; a division into benign, uncertain malignant potential, and malignant categories has been based on the presence of none, 1, 2, or 3 of these criteria, respectively. Infiltrative growth or edges, marked hypercellularity, and marked nuclear pleomorphism/ataxia may be secondary features suggesting aggressive behavior or malignancy.

According to the American Association for the Study of Liver Diseases, HCC can be diagnosed by imaging studies and AFP elevation without confirmation by biopsy. Therefore, most centers rely on imaging studies (CT, MRI, and angiography) instead of biopsy for the diagnosis of HCC, but nonhistologic (imaging) criteria are not standardized. Consequently, PEComa may be misdiagnosed as HCC and mistakenly treated with transcatheter arterial chemoembolization.

In summary, if a hepatic tumor is found in a female patient who is a non-HBV and non-HCV carrier with normal AFP level and no alcoholic liver cirrhosis, tissue biopsy should be done before treatment so that PEComa in the liver is not misdiagnosed as HCC and then treated with transcatheter arterial chemoembolization.

References