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Original Article

Single-center Experience in the Diagnosis and Treatment of Hepatic Perivascular Epithelioid Cell Neoplasm

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Abstract

Background and Aims: Perivascular epithelioid cell neoplasms (PEComas) are a rare type of mesenchymal neoplasm and their preoperative diagnosis is challenging. In this study, we summarized the experience from a single medical center to study the examinations, clinical presentations, and pathological and histological characteristics of PEComas in the liver in order to optimize overall understanding of the diagnosis and treatment of these neoplasms. ***Methods:*** We conducted a retrospective analysis to investigate the clinical and pathological characteristics as well as imaging presentations of 75 patients diagnosed with hepatic PEComa in The First Affiliated Hospital of Zhejiang University between April 2010 and April 2020. ***Results:*** Among the 75 patients, 52 were women, and the median age was 48 years. Most patients had no specific symptoms, and two were admitted to the hospital for a second time owing to relapse. All patients underwent surgical resection. Histologically, 38 patients had classical angiomyolipoma (AML) and 37 had epithelioid AML. The PEComas were accompanied by positive immunohistochemical expression of HMB45, Melan-A, and smooth muscle actin. Follow-up data were obtained from 47 of the total 75 patients, through October 2020. Two patients had metastasis after surgery. ***Conclusions:*** AML is the most common type of hepatic PEComa. There are no specific symptoms of hepatic PEComa, and serological examinations and imaging modalities for accurate preoperative diagnosis are lacking. Epithelioid AML should be considered a tumor of uncertain malignant potential; however, the prognosis of PEComa after resection is promising.

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Introduction

Perivascular epithelioid cell neoplasms (PEComas) are a rare group of related mesenchymal tumors comprising histologically and immunohistochemically distinct perivascular cells, first proposed by Bonetti *et al*.[1] in 1992. The World Health Organization (WHO) Classification of Tumors formally established them as a new category of tumors in 2002,[2] with the members of this group including angiomyolipoma (AML), lymphangioleiomyomatosis (LAM), pulmonary clear cell "sugar" tumors, and PEComa-NOS. In 2004,[3] the WHO subclassified AML into classical AML (CAML) and epithelioid AML (EAML). EAML is categorized as having malignant potential. In 2013,[4] the WHO defined mesenchymal tumors with perivascular epithelioid cell differentiation as PEComas. These tumors show a focal association with blood vessel walls and commonly express melanocytic and smooth muscle markers. Necrosis or hemorrhage might be observed in these tumors, particularly in large tumors.

PEComas can involve several anatomic sites but the kidney is the most common. Cases that arise from the liver are extremely rare. The imaging presentations of hepatic PEComa are diverse and often lead to misdiagnosis as hepatocellular carcinoma (HCC), hepatic hemangioma, gastrointestinal stromal tumor, undifferentiated liver sarcoma, or other tumors.[5] The gold standard for identification using diagnostic imaging studies is lacking. Here, we present cases of hepatic PEComa to summarize our experience in the diagnosis and treatment of hepatic PEComa.

Keywords: Liver; Perivascular epithelioid cell neoplasms; Diagnosis; Treatment.

Abbreviations: AFP, alpha-fetoprotein; AML, angiomyolipoma; CAML, classical AML; CEA, carcinoembryonic antigen; CK, pancytokeratin; CT, computed tomography; DWI, diffusion-weighted imaging; EAML, epithelioid AML; FNAB, fine-needle aspiration; HBV, hepatitis B virus; HCC, hepatocellular carcinoma; HCV, hepatitis C virus; LAM, lymphangioleiomyomatosis; MRI, magnetic resonance imaging; PEComas, perivascular epithelioid cell neoplasms; SMA, smooth muscle actin; TACE, transcatheter arterial chemoembolization; T1WI, T1-weighted imaging; T2WI, T2-weighted imaging; US, ultrasonography; WHO, World Health Organization.

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Methods

Collection of cases

A total of 75 cases treated between April 2010 and April 2020 were identified in the pathology archive system of the Department of Pathology at The First Affiliated Hospital of Zhejiang University. Hepatectomy had been performed in all patients. Written informed consent was obtained from the patients before the publication of this report to allow for description of their protected health information, including accompanying images.

Clinical data

Clinical data were retrospectively retrieved from the patients' records, including age, sex, location, tumor size, clinical presentation, routine blood tests, liver function tests, hepatitis B virus surface antigen, hepatitis C virus (HCV) antigen, serum tumor markers [such as alpha-fetoprotein (AFP), cancer antigen (CA) 19–9, CA125, and carcinoembryonic antigen (CEA)], imaging presentation, and preoperative diagnosis. Follow-up data were obtained from clinical records or telephonically through October 2020.

Immunohistochemistry

Immunohistochemical staining for HMB45, Melan-A, S-100, smooth muscle actin (SMA), AFP, Hep, CD34, Ki67, CD163, CD68, CD117, TFE3, P53, Syn, CgA, and pancytokeratin (CK) was performed using the Envision method#. All immunohistochemical staining were performed using the BOND-III automated immunostainer (Leica Biosystems, Buffalo Grove, IL, USA).

Hepatic imaging

Patients were subjected to evaluation using imaging modalities. A total of 56 patients underwent computed tomography (CT), 47 underwent magnetic resonance imaging (MRI), and 67 underwent ultrasonography (US). We recorded the following characteristics: lesion morphology; blood vessel invasion; bile duct invasion; and lymphadenectasis.

Statistical analysis

Statistical analysis was performed using SPSS software version 19.0 (IBM Corp., Armonk, NY, USA). Variables were compared using analysis of variance and expressed as the mean ± standard deviation or median. All statistical p values were two-sided, with p values of <0.05 were considered to indicate statistical significance.

Results

Patients and clinical presentation

A total of 75 patients with hepatic PEComa, 52 women (69.3%) and 23 men (30.7%) were included; their median age was 48 years (range: 24–64). Fifty-five patients (73.3%) were asymptomatic, and lesions were found incidentally during regular physical examinations. Overall, 15 patients presented with abdominal discomfort (20%) and 5 patients presented with fatigue (6.7%). Levels of liver function and tumor markers, such as AFP, CEA, CA19-9, and CA125, were normal in the patients. None of the patients had tuberous sclerosis complex or HCV infection; seven patients (9.3%) had a history of hepatitis B virus (HBV) infection, with four having a history of HBV for >10 years and one having a history of hepatitis B that progressed to cirrhosis over a >20-year period. The tumors were located in the right liver lobe in 35 patients (46.7%), the left lobe in 34 (45.3%), both right and left lobes in 3 (4%), and the caudate lobe in 3 patients (4%). With respect to tumor size, the largest lesion measured 14 × 12 × 10 cm; four patients (5.3%) had a lesion diameter of >10 cm and seventeen patients (22.7%) had a lesion size between 5 cm and 9 cm. Twenty-three patients (30.7%) had lesion sizes between 3 cm and 5 cm and thirty-one patients (41.3%) had lesion sizes of <3 cm. Overall, 69 patients (92%) presented with a solitary, well-circumscribed heterogeneous mass and 6 patients (8%) presented with multiple masses. One male patient exhibited recurrence in the right liver and abdominal implant metastasis 3 years after surgery for left renal PEComa; one female patient showed recurrence in the left liver after right hepatic PEComa resection performed 10 years ago, and two patients received transcatheter arterial chemoembolization (TACE) in their caudate lobe before surgery (Table 1).

Imaging presentation

Patients underwent imaging examination. All images demonstrated that the lesions were round or oval with clear borders. There was no blood vessel or bile duct invasion, except in one patient who had extensive abdominal metastasis and a lesion in the liver with no clear boundary. Most patients who underwent US had a hyperechoic or hypoechoic mass.

On CT examination, low density signal was usually observed in the scanning period, and the lesions were unevenly enhanced in the arterial phase and were still enhanced in the portal and delay phases. Additionally, the lesions often contained fat.

On MRI examination, most patients exhibited a slightly hypointense or isointense signal on T1-weighted imaging (T1WI), slight hyperintense signal on T2-weighted imaging (T2WI), and unevenly hyperintense signal on diffusion-weighted imaging (DWI). The enhanced images showed significant and homogeneous enhancement during the arterial phase, which indicated a relatively hypervascular, heterogeneous composition. This enhancement gradually weakened during the portal venous and equilibrium phases, and insufficient enhancement was observed in the late parenchymal phase.

Taken together, the diagnostic accuracy was 30.5%, 31.9%, and 10.4% within the groups of patients who underwent CT, MRI, and US examinations, respectively. According to the consistency of the combined diagnosis, the diagnostic accuracy with two imaging methods was 7.4%, 18.2%, and 31% within the US + CT, US + MRI, and CT + MRI groups, respectively. The combined diagnostic accuracy of these three imaging methods was 7.4%. The imaging characteristics were inaccurate for diagnosing these PEComas (Table 2).

Pathological findings

Thirty-eight patients had CAML and 37 patients had EAML; the tumors were circumscribed, either unencapsulated or encapsulated. The cut surface was soft, with color ranging from yellow to dark red, and areas of necrosis were present in four patients. Routine histopathological examination with hematoxylin and eosin staining was performed. Immunohistochemically, the positive expression rate of HMB45 was

Table 1. Demographic characteristics of patients with hepatic PEComa

	Cases (*n*=75)	Female (*n*=52)	Male (*n*=23)	*P*-value
Age (Mean ± SD)	48.61±9.70	46.77±9.25	52.78±9.60	0.01
Symptom, *n* (%)				
No symptom	55 (73.3)	39 (75.0)	16 (69.6)	0.01
fatigue	5 (6.7)	4 (7.7)	1 (4.3)	0.18
Abdominal discomfort	15 (20)	9 (17.3)	6 (26.1)	0.44
Location of tumors, *n* (%)				
Right lobe	35 (46.7)	30 (57.7)	9 (39.1)	0.01
Left lobe	34 (45.3)	20 (38.5)	11 (47.8)	0.11
caudate lobe	3 (4)	0	3 (100)	1
Both side	3 (4)	3 (5.8)	0	1
Lesion number, *n* (%)				
solitary	69 (92)	47 (90.4)	22 (95.7)	0.01
multiple	6 (8)	5 (9.6)	1 (4.3)	0.10
Tumor size, *n* (%)				
≥10cm	4 (5.3)	2 (3.8)	2 (8.7)	1
9–5 cm	17 (22.7)	12 (23.1)	5 (21.7)	0.09
3–5 cm	23 (30.7)	16 (30.8)	7 (30.4)	0.06
<3 cm	31 (41.3)	20 (38.5)	11 (47.8)	0.11
Blood test				
WBC (Mean ± SD)	5.49±1.71	5.47±1.86	5.53±1.36	0.91
N% (Mean ± SD)	60.57±9.62	59.90±10.57	62.09±7.08	0.39
TB (Mean ± SD)	12.54±7.50	11.99±7.40	13.75±7.76	0.38
DB (Mean ± SD)	4.34±2.62	4.16±2.63	4.74±2.59	0.40
IB, median	7.10 (5.0,10.0)	7.00 (4.90,9.78)	8 (5,12.15)	0.27
Ca199, median	7.3 (4.2,13.90)	9.7 (6.2,15.20)	4.75 (2.08,11.95)	0.01
Ca125, median	8.8 (5.6,15.70)	9.35 (6.28,16.50)	7.85 (4.85,12.80)	0.29
CEA, median	1.6 (1.1,2.2)	1.5 (1,2.2)	1.75 (1.40,2.38)	0.12
HBsAg, *n* (%)				
positive	7 (9.3)	3 (5.7)	4 (17.4)	0.71
negative	68 (90.7)	49 (94.2)	19 (82.6)	0.01

CEA, carcinoembryonic antigen; HBsAg; hepatitis B surface antigen; PEComas, perivascular epithelioid cell neoplasms; WBC; white blood cell.

98.4%, that of SMA was 93.2%, and that of Melan-A was 98.2%. For S100, the expression rate was 14.3% in general, and in hepatocytes was 4.3% (Fig. 1). The tumor cells were negative for AFP, CD117, CK, and TFE3 (Table 3).

Microscopically, CAML comprised a mixture of thick-walled blood vessels, different spindle cells, and mature adipose tissue. EAML comprised epithelioid cells with pale, clear, eosinophilic, or foamy cytoplasm. Mitotic figures were relatively rare. The tumor cells were different in size and irregularly arranged. The cytoplasm was rich with red staining, the nucleus was large and oddly shaped, and the nucleolus was prominent (Fig. 2).

Treatment and follow-up

In our study, most patients underwent partial hepatectomy, and the follow-up study was completed for 47 patients. The follow-up continued until the end of the study. During the follow-up period, one patient whose liver tumor size was 12 × 11 × 8 cm with necrosis had a mediastinal metastasis 5 years after surgery; another patient whose liver tumor size was 1.7 × 1.5 cm had a lung metastasis 4 years after surgery, and two patients had EAML.

Discussion

PEComas are mesenchymal tumors with perivascular epithelioid cell differentiation comprising distinct cells that show a focal association with blood vessels and walls; they usually express melanocytic and smooth muscle markers.[4] The "PEComas group" comprises AML, CCST, LAM, CCMMT, and other NOS. AML is the most common pathological type;

Table 2. Diagnostic accuracy of imaging modalities in patients with hepatic PEComa

Imaging methods	Cases (*n*)	Simultaneous diagnostic accuracy (*n*/%)
US	67	7 (10.4)
CT	56	17 (30.5)
MRI	47	15 (31.9)
Combined two imaging examinations		
US+CT	54	4 (7.4)
US+MRI	44	8 (18.2)
CT+MRI	29	9 (31)
Combined three imaging examinations		
US+CT+MRI	27	2 (7.4)

CT, computed tomography; MRI, magnetic resonance imaging; PEComas, perivascular epithelioid cell neoplasms; US, ultrasonography.

it includes CAML and EAML, and other pathological types are relatively rare.[2] PEComas are predominantly diagnosed in the uterus or kidney and rarely occur in the liver.[5] Hepatic PEComa usually presents a solitary, well-circumscribed heterogeneous mass in patients with no potential liver diseases. A higher frequency of multiple masses can occasionally accompany tuberous sclerosis;[6] however, we did not find tuberous sclerosis in our patients, and most patients (92%) presented with solitary lesions. Four patients with EAML and three with CAML had HBV infection; one patient with cirrhosis was diagnosed with HCC and received TACE treatment before hepatic surgery. Hepatic PEComa is mostly asymptomatic, and serological test results are normal. Owing to the highly variable histological composition of hepatic PEComa, these tumors often do not have typical imaging characteristics; thus, their diagnosis is difficult.

Clinically, the differential diagnosis of a hepatic lesion before surgery primarily depends on imaging examinations, and how to make a correct preoperative diagnosis with imaging examinations is worth investigating. Another study has described the morphological imaging characteristics of hepatic PEComa on CT, MRI, or US.[5] It has been suggested that dynamic imaging is the most reliable method for differentiating AML from HCC[7,8] because AML can be easily distinguished from hepatocytes with adipose tissue. Previous reports have suggested that contrast-enhanced CT or MRI is a useful diagnostic modality for hypervascularity and arteriovenous characteristics in PEComas.[9–11] Researchers have now defined PEComas as neoplasms without adipocytes, thereby distinguishing them from classical AML.[12] The di-

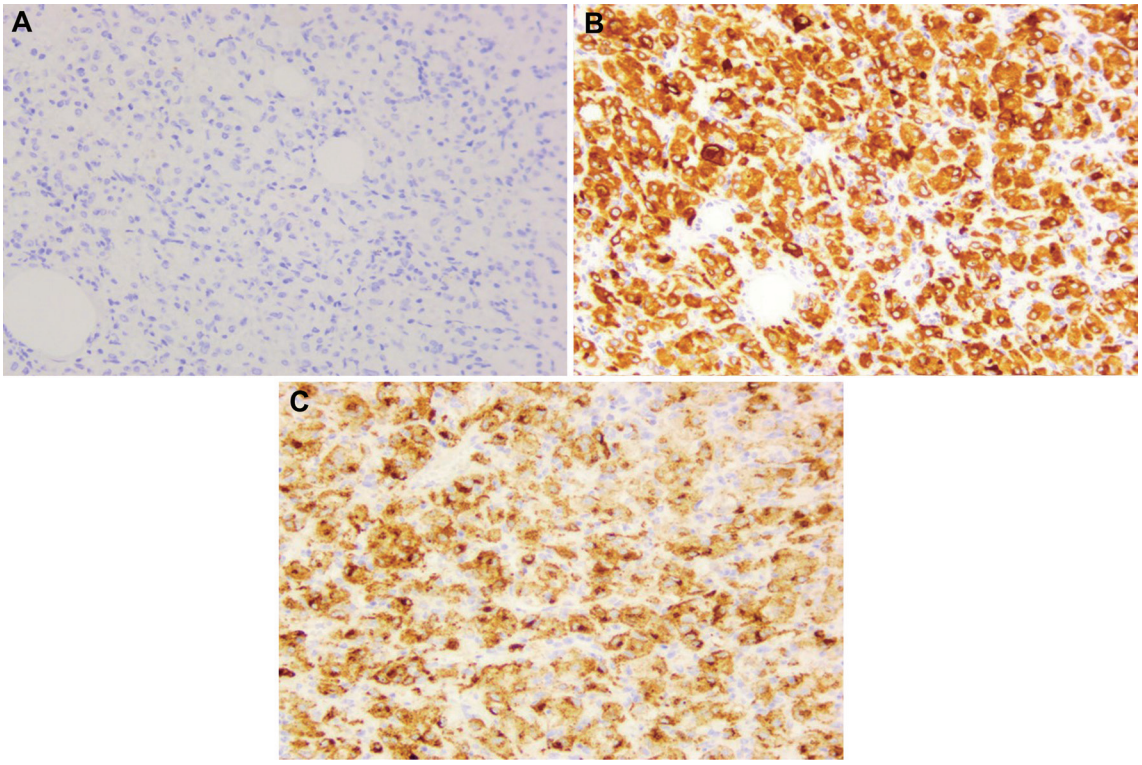

Fig. 1. Histological appearance of PEComa. (A) immunohistochemical analysis showed negative expression of hepatocytes and (B) strong and diffuse expression of Melan-A (C) and HMB45 in most patients (original magnification, ×400). PEComas, perivascular epithelioid cell neoplasms.

Table 3. Immunohistochemistry

	Cases		Male (*n*=23)		Female (*n*=52)	
	Count (*n*/%)	Positive (*n*/%)	Count	Positive (*n*/%)	Count	Positive (*n*/%)
AFP	24/33.8	0	8	0	16	0
Hepatocyte	49/69	2 (4.3)	15	2 (13.3)	34	0
Ki 67	29/40.9	27 (93.1)	7	6 (85.7)	22	21 (95.5)
CD34	28/39.4	20 (71.4)	6	2 (33.5)	22	16 (72.7)
CD163	1/1.4	1 (100)	1	1 (100)	0	0
CD68	2/2.8	1 (50)	2	1 (50)	0	0
CD117	8/11.3	0	1	0	7	0
Syn	1/1.4	1 (100)	1	1 (100)	0	0
CGA	1/1.4	1 (100)	1	1 (100)	0	0
CK	38/53.5	0	9	0	29	0
HMB45	61/85.9	60 (98.4)	18	18 (100)	43	42 (2.3)
Melan A	56/78.9	55 (98.2)	16	15 (93.8)	40	40 (100)
SMA	44/62	41 (93.2)	11	9 (81.8)	33	32 (97.)
S-100	28/39.4	4 (14.3)	6	6 (100)	22	4 (18.2)
TFE3	3/4.2	0	1	0	2	0
P53	4/5.6	2 (50)	0	0	4	2 (50)

AFP, alpha-fetoprotein; CK, pancytokeratin; SMA, smooth muscle actin.

agnostic accuracy of liver PEComa using imaging modalities is low and that of CT combined with MRI is only 20%.[13,14] In our study, the diagnostic accuracy of CT combined with MRI was 31%. Contrast-enhanced CT and MRI of PEComa showed that the lesions were significantly and heterogeneously enhanced in the arterial phase, continually enhanced in the portal venous phase, and slightly hypodense in the delayed phase, which exhibits a broad spectrum of imaging findings.

Hepatic PEComa can be diverse based on echogenicity of blood flow in or around lesions. For cases with fat, particularly in lesions with fat comprising >50% of the tumor, AML can be easily diagnosed. Diagnosis of tumors with no or little fat is challenging.[15] Most tumors show low signal intensity on T1WI and high signal intensity on fat-suppressed T2WI and DWI images on MRI. Unfortunately, these findings are not specific in several tumors.[16] Previous reports have suggested hypervascularity and arteriovenous connections as the features of PEComa. The current study demonstrated the utility of MRI in the early diagnosis of liver PEComa. Combining our cases, we created a form to summarize the characteristics of different hepatocyte tumors to distinguish PEComas (Table 4).

PEComas are positive for melanocytic and muscle markers, including HMB45, Melan-A, and SMA,[4] but negative for other markers, including AFP, CD117, CK, and TFE3. Detection of TFE3 protein expression is recommended for PEComas because TFE3-positive PEComas are associated with

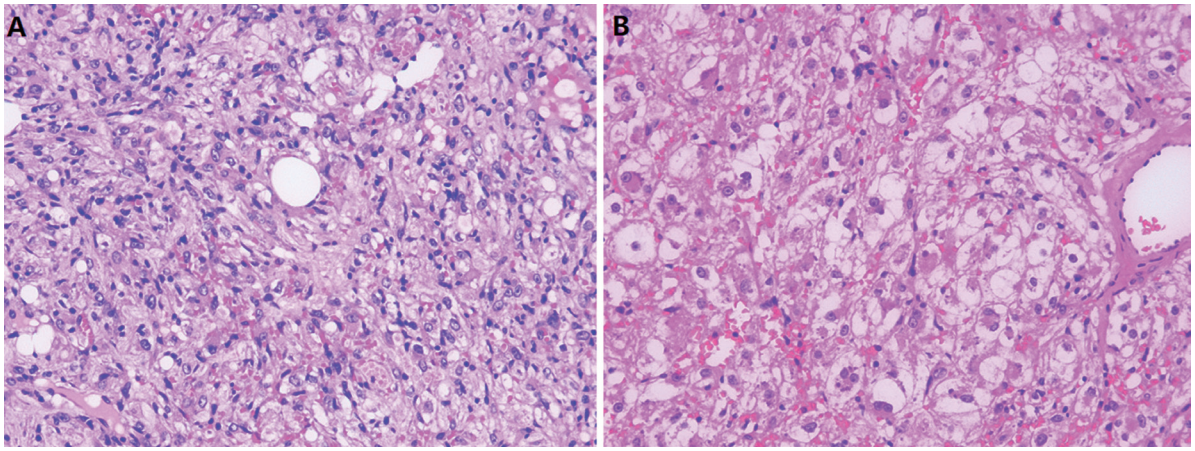

Fig. 2. Hematoxylin-eosin staining of PEComa. (A) CAML comprised a mixture of thick-walled blood vessels, spindle cells, and mature adipose tissue (original magnification, ×200). (B) EAML comprised light eosinophilic cells with clear eosinophilic, transparent, or granular cytoplasm. The cytoplasm was rich with red staining, the nucleus was large and oddly shaped, and the nucleolus was prominent (original magnification, ×200). CAML, classical AML; EAML, epithelioid AML; PEComas, perivascular epithelioid cell neoplasms.

Table 4. Imaging characteristics of different tumors

	Pathological features	Enhancement reason	Blood supply	Plain MRI	Arterial phase	Portal phase	Venous phase	Enhanced form
AML	Central stellate scar, no liver lobules, bile duct fibrosis	Composed of blood vessels and fibrous tissue	Small arteries	T2W1 is hyperintense or slightly hyperintense; T1W1 is isointense or slightly hypointense	Heterogeneity strengthening	Fades away, strengthened area continues	Isointense or hypointense	Fast in and slow out
Hemangioma	Blood-filled vascular cavity and fibrous separation	Blood pool filling	Hepatic artery	T2W1 is hyperintense; T1W1 is slightly hypointense	Peripheral nodular enhancement	Extending toward the center	Filling reinforcement	Fast in and slow out
FNH	Abnormal arrangement of normal stem cells, central stellate scar, tubule structure	Scars are composed of blood vessels and bile ducts	Small arteries; blood is eccentric	T2W1 is isointense or slightly hyperintense; T1W1 is isointense or slightly hypointense (center high)	Clearly uniform strengthening, low center	Quick removal, low center	Isointense, strengthened at the center	Fast in and out, center delay strengthened
Hepatocellular adenoma	Hepatocytes are hypertrophied and lipid-bearing, bile ducts are absent and can be malignant, and 1/3 have pseudocapsule	Absence of portal vein and bile duct, easy bleeding	Subcapsular arteriole blood flow is concentric	Easy to bleed, signal diversification, envelope is hypointense	Significantly strengthened	Isointense or hypointense	Isointense or hypointense	Fast in and out
Lamellar fibrocarcinoma	There is a central scar and fibrous separation, which can be accompanied by calcification and necrosis	Contains many fiber separations	Both arteries and portal veins exist	T2W1 is hyperintense; T1W1 is slightly hypointense. (center low)	Early periphery strengthening	Further strengthening	Low periphery, strengthened center	Fast in and slow out
Metastasis carcinoma	Multiple blood spread	Tumor angiogenesis	Small arteries	T2W1 is hyperintense; T1W1 is slightly hypointense	Early edge enhancement	Quick removal	Isointense or hypointense	Fast in and out

AML, angiomyolipoma; MRI, magnetic resonance imaging; T1WI, T1-weighted imaging; T2WI, T2-weighted imaging.

poor prognosis.[17,18] In our study, TFE3 protein was detected in only three patients and these patients had a good prognosis.

Classical AMLs are universally regarded as slow-growing benign tumors. However, it is becoming increasingly clear that some PEComas should be considered tumors of uncertain malignant potential, such as EAML. Malignant PEComas often show the following characteristics: epithelioid cells ≥70%; tumor size of >5 cm; invasion of blood vessels; ≥2 mitoses per 10 high-powered fields; pathological mitosis and necrosis; and tumor aggressiveness.[19]

Among our patients with EAML, three patients had necrosis, two patients were admitted to the hospital for the second time owing to a relapse, one had undergone surgery for left renal PEComa 3 years earlier, and one had undergone right hepatic PEComa resection 10 years earlier.

Most authors agree that the criteria for malignancy and the biological behavior of PEComas have been insufficiently established. Indeed, PEComas feature several biological behaviors, including benign as well as malignant behavior and uncertain malignant potential. Folpe *et al*.[20] performed statistical analysis and found that the behavior of PEComas in the gynecologic tract and soft tissue correlated with a tumor size of >5 cm, infiltration, high nuclear grade and cellularity, mitosis in >1/50 high-power fields, necrosis, and vascular invasion. PEComas showing two or more worrisome histological features should be classified as aggressive. In our cases, most of the patients (41.3%) had tumor sizes of <3 cm. We should take into account that tumors of >10 cm with necrosis tend to become malignant.

Because this disease is rare, it is difficult to perform a clinical, therapeutic trial, and therefore the treatment for hepatic PEComa remains controversial. The variable proportions of the different components of hepatic PEComa make diagnosis difficult before surgery, and the majority of patients undergo surgical resection. Furthermore, postoperative complications or recurrence have rarely been reported.[21] In our study, all patients underwent surgical resection, including laparotomy and laparoscopic partial hepatectomy. Two patients had received TACE before the surgery. Two patients relapsed after surgery, including one male patient whose liver tumor size was 12 × 11 × 8 cm with necrosis, who had mediastinal metastasis 5 years after surgery and pathological necrosis, and one other male patient, whose tumor presented as a solitary mass with a size of 1.7 × 1.5 cm and who had lung metastasis 4 years after the surgery. Pathological examination showed that the tumor had vascular invasion. Both patients received chemotherapy in other hospitals. Yang *et al*.[14,22] recommended the performance of fine-needle aspiration (FNAB) combined with HMB45 and Melan-A staining in all asymptomatic patients with lesions of <5 cm and without serological abnormalities, when hepatic PEComa is suspected. If FNAB is performed and the pathology indicates a benign pattern, then observation and imaging follow-up are recommended.[22] For patients with unresectable malignant PEComas, O'Malley *et al*.[23] recommended that applying targeted therapy, such as mTORC1 inhibitors, may have a specific effect.

Most of these patients were found incidentally during a regular physical examination. In our study, some patients presented with abdominal discomfort, and four had a history of a liver mass for >2 years. They were diagnosed with AML and then followed by observation. We found that the tumors had gradually increased to >5 cm at 2-year follow-up and that the patients visited the hospital upon experiencing abdominal discomfort. The patients had a good prognosis after surgery. This indicates that patients with PEComas of <5 cm can be administered conservative treatment. However, we found one patient with a solitary mass of <5 cm who had a lung metastasis 4 years after the surgery. Pathological examination showed that one worrisome histological feature was the invasion of blood vessels. Further research must allow accurate prediction of the biobehavior of this lesion and establish criteria for discrimination between malignant and benign tumors. In our study, most patients who underwent surgical resection had a good outcome; therefore, surgical resection may be the best choice for hepatic PEComa therapy.

Our study also had some limitations. The efficacy and prognosis were not compared with different treatments, such as conservative therapy, TACE, or surgery. Although we sorted different liver tumor characteristics according to MRI, owing to PEComa heterogeneity, the application of the characteristics described in Table 4 may have some limitations. When imaging diagnosis is challenging, FNAB should be used to make an auxiliary diagnosis.

In conclusion, AML is the most common type of PEComa in the hepatic tissue. There are no specific symptoms of hepatic PEComa, and serological examinations and imaging modalities for accurate preoperative diagnosis are lacking. EAML should be considered a tumor of uncertain malignant potential. However, the prognosis of PEComa after resection is promising.

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Conflict of interest

The authors have no conflict of interests related to this publication.

Author contributions

Study concept and design (SZ, JY), acquisition of data (JJ, JL, CGP), analysis and interpretation of data (GG, MF, BZ, LL), drafting of the manuscript (JJ, JL), critical revision of the manuscript for important intellectual content (SZ, JY), and administrative, technical, or material support, study supervision (SZ, JY).

Data sharing statement

No additional data are available.

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