



Very Rare Liver Tumor: PEComa Case Report with and a Review of Literature

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Abstract

Perivascular epithelioid cell tumors (PEComas) are very rare mesenchymal tumors. In addition to being rare in general, they are even rarer in the liver. There are various subtypes and there is only one case of liver PEComa reported in the clear cell type in the literature to date. We present the second clear cell type liver PEComa in this case by reviewing the literature data. The hypodense, smooth-edged, ovoid lesion was detected on computed tomography (CT) for performing abdominal pain in a 41-year-old female patient. Magnetic resonance imaging (MRI) was then performed for lesion characterization. The lesion was hypointense on T1-weighted imaging (T1WI), hyperintense on T2-weighted imaging (T2WI). In dynamic phases, it showed marked enhancement on the arterial phase and capsular enhancement with central washout on the portal and late venous phases. The posterior branch of the right portal vein extended into the mass. The lesion was excised and the pathological result was epithelioid clear cell subtype of PEComa. Although the imaging findings are generally nonspecific and certain diagnosis is made histopathologically, radiologists should consider PEComa in the differential diagnosis in the presence of intensely enhanced lesion on the right lobe in female patients. Also, the “large vessel sign” may help in the diagnosis.

Keywords

- ▶ perivascular epithelioid cell tumors liver
- ▶ computer tomography
- ▶ magnetic resonance imaging

Introduction

Perivascular epithelioid cell tumors (PEComas) are very rare mesenchymal tumors. PEComas are composed of distinctive cells that show association with blood vessel walls and usually express melanocytic and smooth-muscle markers.¹ The PEComa tumor group includes tumors such as angiomyolipoma (AML), clear cell “sugar” tumor, lymphangiomyomatosis (LAM), and clear cell myomelanocytic tumor (CCMMT). They are far more common among women.²

PEComas show a wide anatomical distribution, but most arise from the kidney, retroperitoneum, uterus, and pancreas

in the abdomen.^{1,3} Liver PEComas are extremely rare. They are usually discovered incidentally and benign in character.⁴

In this study, we aimed to present an incidentally detected liver PEComa and its common radiological features by reviewing the literature data.

Case Report

A 41-year-old female patient had an incidental lesion in the liver on contrast-enhanced abdominal computed tomography (CT) performed for abdominal pain. The hypodense lesion was 35 × 30 mm in size, smooth-edged, and ovoid

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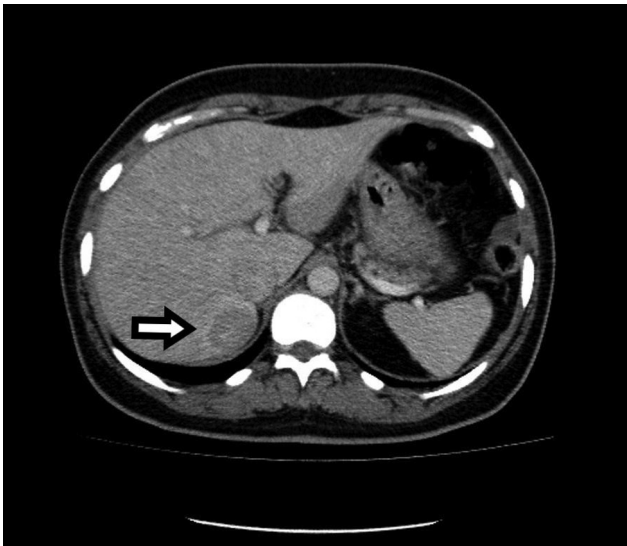


Fig. 1 Contrast-enhanced computed tomography (CT) image showing an ovoid, smoothly margined, hypodense mass on the right lobe of the liver. Arrow is showing the lesion.

(► **Fig. 1**). Dynamic contrast-enhanced magnetic resonance imaging (MRI) was then performed for lesion characterization. The lesion was hypointense on T1-weighted images (T1WI) and hyperintense on T2-weighted images (T2WI), and there was no signal loss in fat-suppressed sequences (► **Fig. 2**) but there was restricted diffusion (► **Fig. 3**). In dynamic phases, the lesion showed marked enhancement in the arterial phase. Capsular enhancement with central washout was detected in the portal vein and late venous phases. In the hepatobiliary phase examination, the lesion was found to be hypointense (► **Fig. 4**). In the liver parenchyma, there was no finding in favor of chronic liver parenchymal disease and it was remarkable that the posterior branch of the right portal vein extended into the mass (► **Fig. 5**). The patient's hematological and biochemical parameters and tumor markers were within normal limits. With the preliminary diagnosis of hypervascular liver mass, the lesion was excised. Pathologically, HMB45, actin, vimentin, and melanin were positive. Findings were consistent with PEComa and epithelioid clear cell tumor subtype. So far, the patient has been followed for 66 months with no local recurrence or distant metastasis.

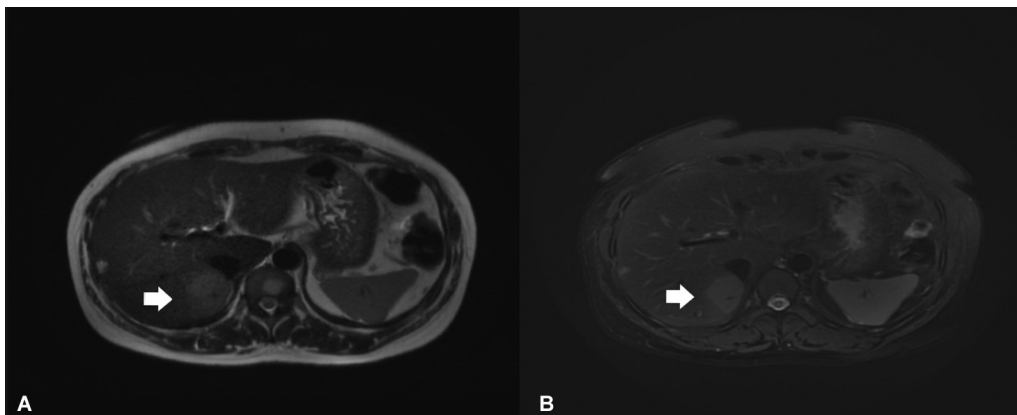


Fig. 2 (A) Axial T2-weighted images showing the high-signal mass lesion in the right of liver. (B) Axial fat-suppressed T2-weighted images not showing the suppression of the signal. Arrows are showing the lesion.

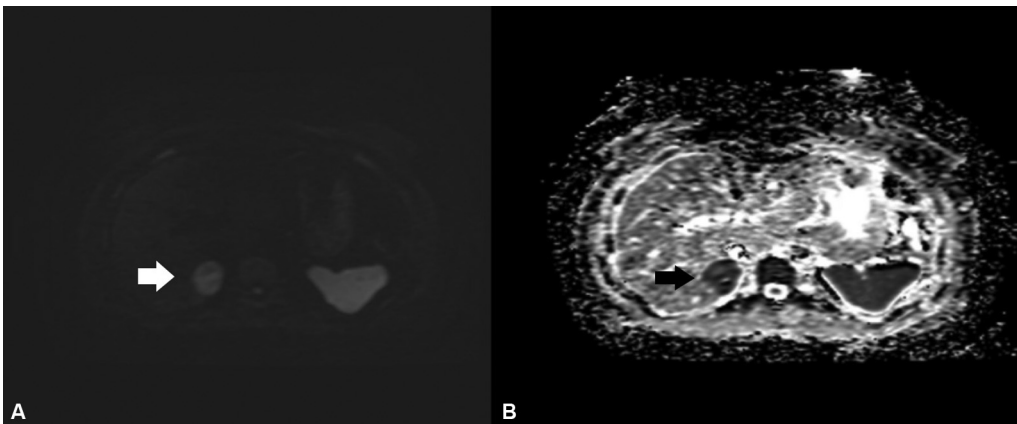


Fig. 3 Diffusion-weighted imaging (DWI) magnetic resonance imaging (MRI). (A) DWI and (B) apparent diffusion coefficient (ADC) map: a significant restricted diffusivity of the liver mass (white arrow in A and black arrow in B) is detected. Arrows are showing the lesion.

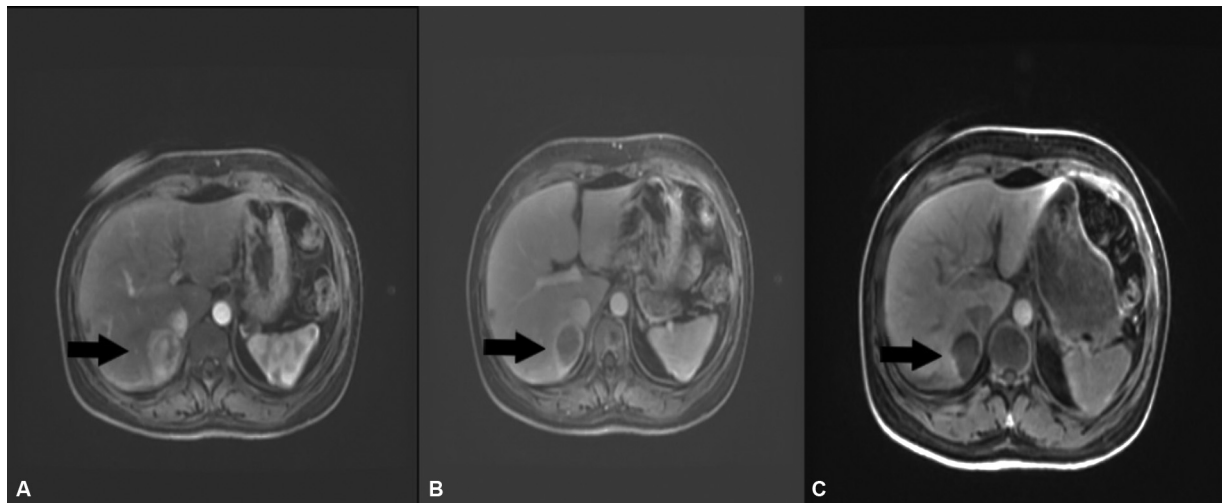


Fig. 4 Perivascular epithelioid cell tumor (PEComa) in segment VII demonstrating (A) bright enhancement at the arterial phase, (B) capsular enhancement at the portal phase, and (C) no enhancement at the hepatobiliary phase. Arrows are showing the lesion.

Table 1 Review of the liver PEComa cases reported in the literature

Study	Age (y)	Gender	Hepatic lobe	Size (cm)	Symptom/finding	Histologic	Malign/benign
Fang et al ²²	56	F	Left	5.1	Abdominal pain	PEComa	Benign
Fang et al ²²	63	F	Left	None	Abdominal pain	PEComa	Benign
Svajdler et al ²³	55	F	Left	3.5	None	PEComa	Benign
Larbcharoensub et al ²⁴	31	F	Right	1.8	Abdominal pain	CCMMT	Benign
Zimmermann et al ¹⁷	53	M	Right	8	Abdominal pain	PEComa	Benign
Paiva et al ²⁵	51	F	Left	0.8	Abdominal pain, weight loss	PEComa	Benign
Strzelczyk et al ⁶	50	F	Right	17	Abdominal pain	Clear cell sugar PEComa	Benign
Priola et al ²⁶	36	F	Left	11	Abdominal pain	PEComa	Benign
Jafari et al ²⁷	53	F	Left	7.5	Hemorrhage	PEComa	Malign
Cheung et al ²⁰	53	F	Right	10	Abdominal pain	PEComa	Benign
Zhao et al ¹²	50	M	Right	10	Abdominal pain	PEComa	Benign
Yu and Tang ²⁸	41	F	Right	2.2	Abdominal pain + fever	PEComa	Benign
Patra et al ²⁹	50	F	Right	24	Abdominal pain	Pigmented PEComa	Benign
Shen et al ⁹	55	M	Right	1.6	Incidental	PEComa	Benign
Khaja et al ³⁰	51	M	Right	None	Incidental	PEComa	Benign
Liu et al ¹³	25	F	Right	1.8	Incidental	PEComa	Benign
Ameurtesse et al ¹⁰	63	F	Left	8	Abdominal pain	PEComa	Benign
Khan et al ¹⁵	61	M	Right	7	Incidental	PEComa	Benign
Abhirup et al ¹¹	72	F	Both	10	Abdominal pain	PEComa	Malign
Arribas Jurado et al ³¹	45	F	Right	4	Abdominal pain	PEComa	Benign
Schaeffer and Poulin ⁴	49	F	Left	2.9	Abdominal pain	PEComa	Benign
Son et al ¹	56	F	Right	4.5	Incidental	PEComa	Benign
Kirnap et al ³²	22	F	Right	16	Incidental	PEComa	Benign
Voulgari et al ³³	47	F	Both	7	Abdominal pain	PEComa	Benign
Matrood et al ³⁴	51	F	Right	2	Incidental	PEComa	Benign
Kou et al ¹⁸	37	M	Right	2.5	Abdominal pain	PEComa	Malign
Kou et al ¹⁸	70	F	Left	5	Incidental	PEComa	Benign
Kou et al ¹⁸	30	F	Right	5	Abdominal pain	PEComa	Benign
Harwal et al ¹⁹	27	F	Right	10	Abdominal pain	PEComa	Benign
Perán Fernández et al ³⁵	74	F	Right	3.5	Incidental	PEComa	Benign

Abbreviations: CCMMT, clear cell myxoid melanocytic tumor; PEComa, perivascular epithelioid cell tumor.

Discussion

PEComa can originate in almost any part of the body. Liver PEComa was classified by the World Health Organization (WHO) in 2002 as a mesenchymal tumor of the liver.⁵ AML is the most common PEComa tumor in the liver and kidney. In our case, PEComa was a subtype of clear cell tumor, which is very rare in the liver PEComa tumor family. Clear cell PEComa is more common in middle-aged women and in the lung.⁶ Extrapulmonary clear cell PEComas have been described in various organs such as the uterus and pancreas.^{7,8} We found only one case of clear cell type PEComa in liver parenchyma in the literature.⁶ In this case, the tumor was observed in the right lobe and in a middle-aged female patient similar to our case.

Up to now 30 liver non-AML PEComa cases have been reported in the literature (►Table 1). The mean age was calculated as 49.2 (range: 32–75) years. Six of the patients were males, and 24 were females. The average tumor size was about 5.9 cm (range: 0.8–17). It can be stated that liver PEComas are seen in a wide range of sizes. Nine of 30 masses in the literature were incidentally detected. The mass was detected in the right lobe in 19 cases, in the left lobe in 9 cases, and in both lobes in 2 cases. Histopathologically, only two cases were reported as malignant. According to these findings, PEComas are generally benign. They are detected as incidental. Clinical symptoms are nonspecific findings such as abdominal pain, swelling, and nausea. A mass was detected incidentally in our case. As in our case, PEComas occur more commonly in middle-aged female patients and in the right lobe.

PEComas have nonspecific CT and ultrasound findings. In MRI, they show high signal intensity on T2WI and low signal intensity on T1WI.^{1,9–13} Heterogeneous enhancement in the arterial phase was noted in the literature.^{9,14–17} Heterogeneous contrast enhancement may vary depending on the amount of fat and vascular and muscle cells it contains. The presence of washout in the portal and venous phases raises suspicion for hepatocellular carcinoma (HCC).^{4,8,17,18} It can also restrict diffusion like HCC.^{1,10} Hypervascular liver tumors such as focal nodular hyperplasia, hemangioma, and adenoma are included in the differential diagnosis. As the name implies, PEComas have abundant vascularity in or around the tumors. Therefore, they may show intense contrast enhancement and close vessel relationship, especially in contrast phases.^{19,20} In the study of Gao et al, “large blood vessel” was observed in approximately 80% of liver PEComas.²¹ In our case, the mass was observed to be associated with the right portal vein branch (►Fig. 5). Vascular feeding of PEComas may be helpful in the differentiation of HCC. In HCC, it may be associated with arteries and in PEComas with veins.¹

The definite diagnosis of PEComa is made histopathologically. Its histopathological features are diagnostic, and as its name signifies, it includes a group of cells with positive perivascularly located myocyte and melanocytic markers.

The appropriate treatment option is surgery. They usually have a benign course and rarely relapse-metastasize.

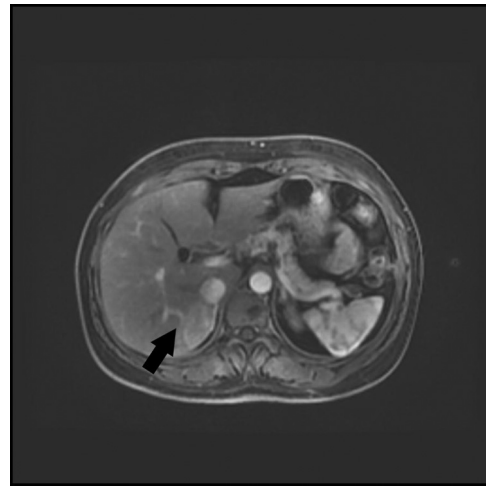


Fig. 5 A portal venous phase magnetic resonance (MR) scan demonstrating a vessel with direct contact to the right main portal vein (PV) and mass. Arrow is showing the vessel.

Conclusion

The incidence of PEComas is increasing due to the increase in imaging. Although the imaging findings are generally non-specific, radiologists should consider PEComa in the differential diagnosis in the presence of intensely enhanced lesion in the right lobe in a female patient. Also, the “large vessel sign” may help in the diagnosis.

Author Contributions

C.Y. was responsible for the conceptualization, data curation, investigation, and writing the original draft. E.G. was responsible for formal analysis, investigation, review, supervision, and editing.

Consent for Publication

Informed consent was obtained from the patient prior to the study.

Funding

None.

Conflict of Interest

None declared.

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