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CASE SERIES

# 2 Cases of Large Cystic-Solid Perivascular Epithelioid Cell Tumor: Case Report and Literature Review

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**Abstract:** Perivascular epithelioid cell tumors (PEComas) are rare mesenchymal tumors that tend to occur in multiple organs and the biological behavior of it between benign and malignant. They are even less common in the liver and there is still no effective treatment method established. In particular, how to deal with it when the imaging manifestations of the lesion are predominantly giant cystic are a question that we need to investigate. We present PEComas in two female patients with large cystic-solid lesions on ultrasound with clinical signs of nonspecific pain and a large mass in the right upper quadrant. Both patients underwent contrast-enhanced ultrasound (CEUS) and interventional ultrasound to assist in diagnosis and treatment. Depending on the patient's condition, we take a different approach to treatment. To our knowledge, such reports have never been documented before.

**Keywords:** perivascular epithelioid cell tumors, two-dimensional ultrasound, contrast-enhanced ultrasound

## Introduction

Perivascular epithelioid cell tumors (PEComas) are a rare type of mesenchymal tumor that can occur in various abdominal organs, though they are rarely primary in the liver.[1,2] PEComas include angiomyolipoma (AML), lymphangioleiomyomatosis (LAM), clear cell sugar tumor of the lung (CCST), and other very rare tumors in different organs.[3] The diversity of imaging manifestations makes clinical diagnosis challenging, and preoperative imaging along with pathological evaluation of frozen sections are also prone to misdiagnosis.[4] The Folpe criteria, which classify PEComa as benign, malignant, or of uncertain malignant potential, have been used in previous studies and may also be applied to liver tumors.[5] In recent years, reports of such tumors have increased. Studies suggest that imaging features such as well-defined boundaries, peripheral hyperechogenicity, lateral shadowing, large vessels around the lesion, and a "fast-in and slow-out" enhancement pattern may indicate tumor expansion and growth, which can aid in the diagnosis of PEComa.[6] The features of two-dimensional ultrasound (2D-US) and contrast-enhanced ultrasound (CEUS) may provide valuable references for clinical diagnosis, though almost all reported cases involve solid PEComas.[7–9] However, there are few reports on the diagnosis, treatment and management of giant cystic PEComa. Cytotoxic chemotherapy and radiotherapy have been reported to show limited efficacy in treating PEComa.[10] We present 2 cases of large cystic solid masses in the liver, pathologically confirmed as PEComas, with distinctive clinical symptoms and imaging findings. Both cases were diagnosed and treated with the assistance of CEUS and interventional ultrasound. Prior to CEUS examination and interventional puncture, informed consent was obtained from both patients after a thorough explanation of the associated risks and potential complications. The protocol and publication of this case report was approved by the Ethics Committee of Lanzhou University Second

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Hospital. The study was conducted in accordance with the World Medical Association Declaration of Helsinki, and written informed consent was obtained from both patients for the publication of their case details.

## Case Report

The first case was a 32-year-old previously healthy female who was referred to a gastroenterologist due to nonspecific pain and a large mass in the right upper quadrant. The patient had no relevant family history, no history of infection, and no significant weight loss. Laboratory tests revealed normal routine hepatic and renal function, primary liver cancer-associated tumor marker were normal range. 2D-US showed increased liver volume with abnormal intrahepatic echogenicity, revealing a well-defined complex echogenic lesion of approximately 20 cm in size containing a large area in the liver (Figure 1A). Color Doppler flow imaging (CDFI) demonstrated that blood flow signals could be seen around the lesion (Figure 1B). No pathologic lymph nodes were identified during the examination. CEUS was subsequently performed to further characterize the liver lesion. A 2.4 mL dose of the ultrasound contrast agent SonVue (Bracco, Italy) was administered intravenously for the examination. The solid component of the lesion demonstrated heterogeneous hyperenhancement during the arterial phase (20–35 s after injection) (Figure 1C) and remained isoenhancing compared to the surrounding liver parenchyma in the portal (35–120 s) and late phase (2–5 min after injection) (Figure 1D). The enhancement pattern of the solid area is characterized by a benign-like appearance with high enhancement in the arterial phase and persistent enhancement in the portal and delayed phase. Based on the tumor marker results, hepatocellular carcinoma (HCC) or intrahepatic cholangiocarcinoma (ICC) was excluded by CEUS, but no definite diagnosis could be established. Considering the absence of cirrhosis and fever in the patient, an initial diagnosis of focal nodular hyperplasia (FNH) of the liver with cystic changes or hepatic echioncoccosis (HE) was made according to EFSUMB Guidelines for Contrast Enhanced Ultrasound of the Liver.[11] The differential diagnosis at that time included tumors of mesenchymal origin and liver metastasis from a gastrointestinal stromal tumor. A Computed tomography (CT) scan revealed a large, oval, well-circumscribed heterogeneous mass in the liver, the density of the marginal area was higher than that of the center within the lesion (Figure 1E). In the arterial phase of contrast-enhanced CT (CECT), the scan showed marked heterogeneous enhancement at the lesion margins and the presence of malformed blood vessels around the lesion (Figure 1F). The enhancement decreased but still remained higher in density compared to the surrounding liver parenchyma during the portal venous and delayed phases. This enhancement pattern was diagnosed by the radiologist as FNH of the liver with cystic changes.

The other patient was a 64-year-old female who presented with abdominal distension for more than 20 days. 2D-US revealed a 24 cm solid-cystic lesion in the liver, in which the echo transmission of cystic component was poorer than that in the first patient (Figure 2A). Color flow signals were observed around the lesion (Figure 2B). Multiple enlarged lymph nodes were identified in the hilar region of the liver. The CEUS (Figure 2C and D) and CECT findings were similar to those of the first patient (Figure 2E and F). Both $CA_{199}$ and $CA_{125}$ were elevated in these patients.

After liver hydatid disease was ruled out by serological examination, and considering that the lesions were too large and prone to rupture, catheter drainage of cyst fluid was performed in both female patients (Figures 1G and 2G). A total of 3500 mL of brown fluid was drained from the younger patient over one week. The other patient drained a total of 2800 mL dark red fluid, which was suggestive of recent internal hemorrhage. Subsequently, pathological tissue specimens were obtained via percutaneous liver puncture with ultrasound-guided. On histopathologic examination, the tumor was mainly composed of epithelioid cells arranged in a trabecular growth pattern (Figures 1H and 2H). Immunohistochemistry (IHC) staining demonstrated that the tumor cells were diffusely positive for HMB45, Melan A, CD68 and CD34 and were negative for desmin. The histological features and the results of IHC were consistent with a diagnosis of hepatic PEComa.

The younger woman underwent surgical removal of the liver mass after her liver function was appropriately supported according to the specific condition. The older woman's family members, considering the high surgical risk and the non-radical nature of the treatment, opted for conservative management. Consequently, transcatheter arterial chemoembolization (TACE) was performed, followed by conservative treatment with chemotherapy due to the large tumor size. To date, follow-up has shown that the younger patient remains free of tumor recurrence 63 months after surgery, while the older patient passed away 6 months after TACE treatment.

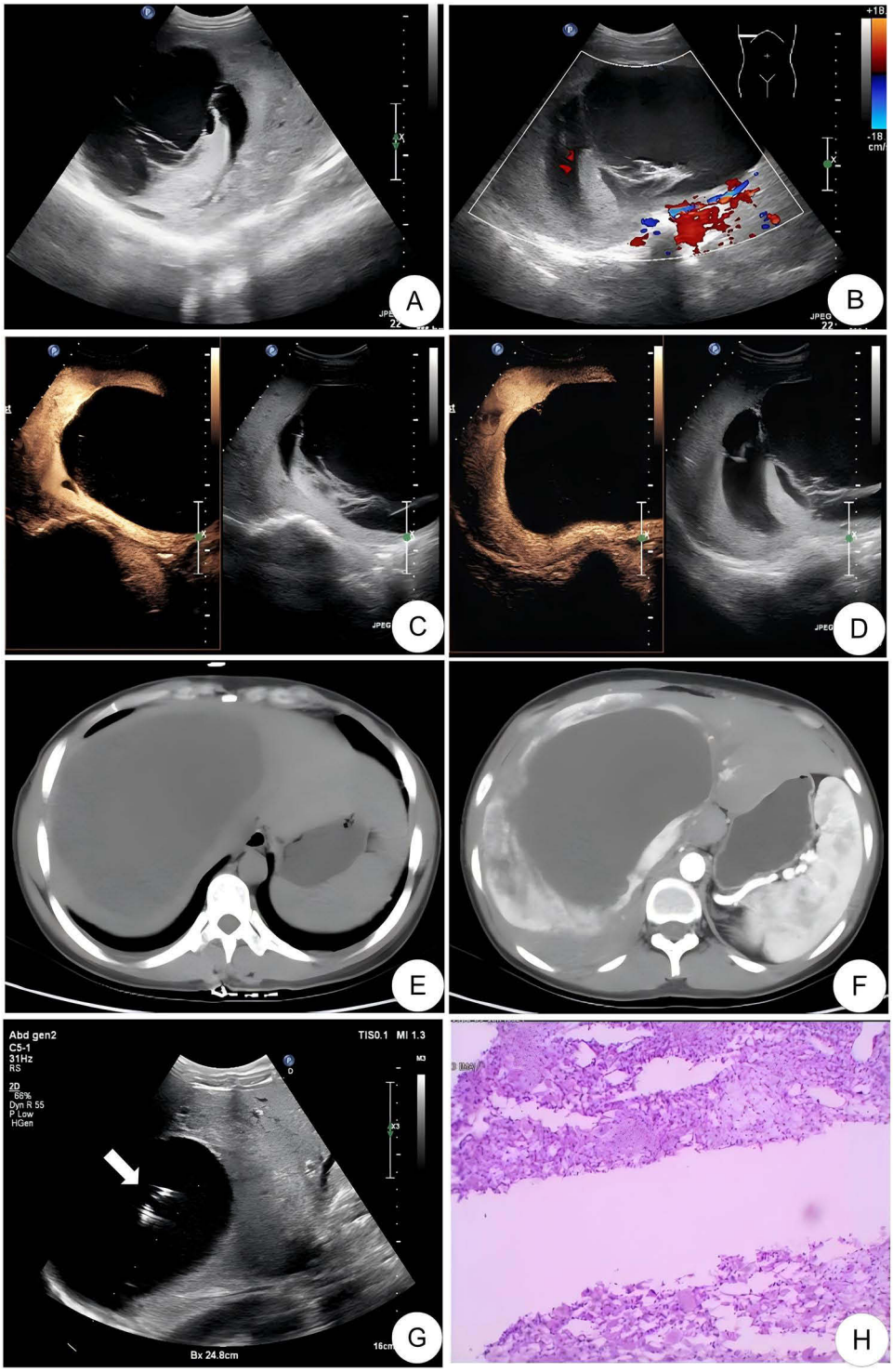


**Figure 1** A 32-year-old woman, 2D-US shows increased liver volume and abnormal intrahepatic echogenicity, as a well-defined complex echogenic lesion of approximately 20 cm with a large cystic area was found in the liver (**A**) with a peripheral short rod-like blood flow signal (**B**). The lesions showed heterogeneous hyperechogenic enhancement in the arterial phase (**C**), and stayed hyper-iso enhancement in comparison to surrounding liver parenchyma in portal venous and late phase (**D**). CT showed a large oval well-circumscribed heterogeneous hypo-density lesion with marginal hyperdensity in liver (**E**). The marginal of lesions demonstrated significantly heterogeneous enhanced and small vascular pattern surrounding the lesion was obviously visible in the arterial dominant phase of CECT (**F**). The drainage tube (white arrow) was inserted into the cystic regions in the lesion (**G**). Histopathology was obtained by percutaneous liver biopsy. Hematoxylin and eosin, magnification 100× (**H**).


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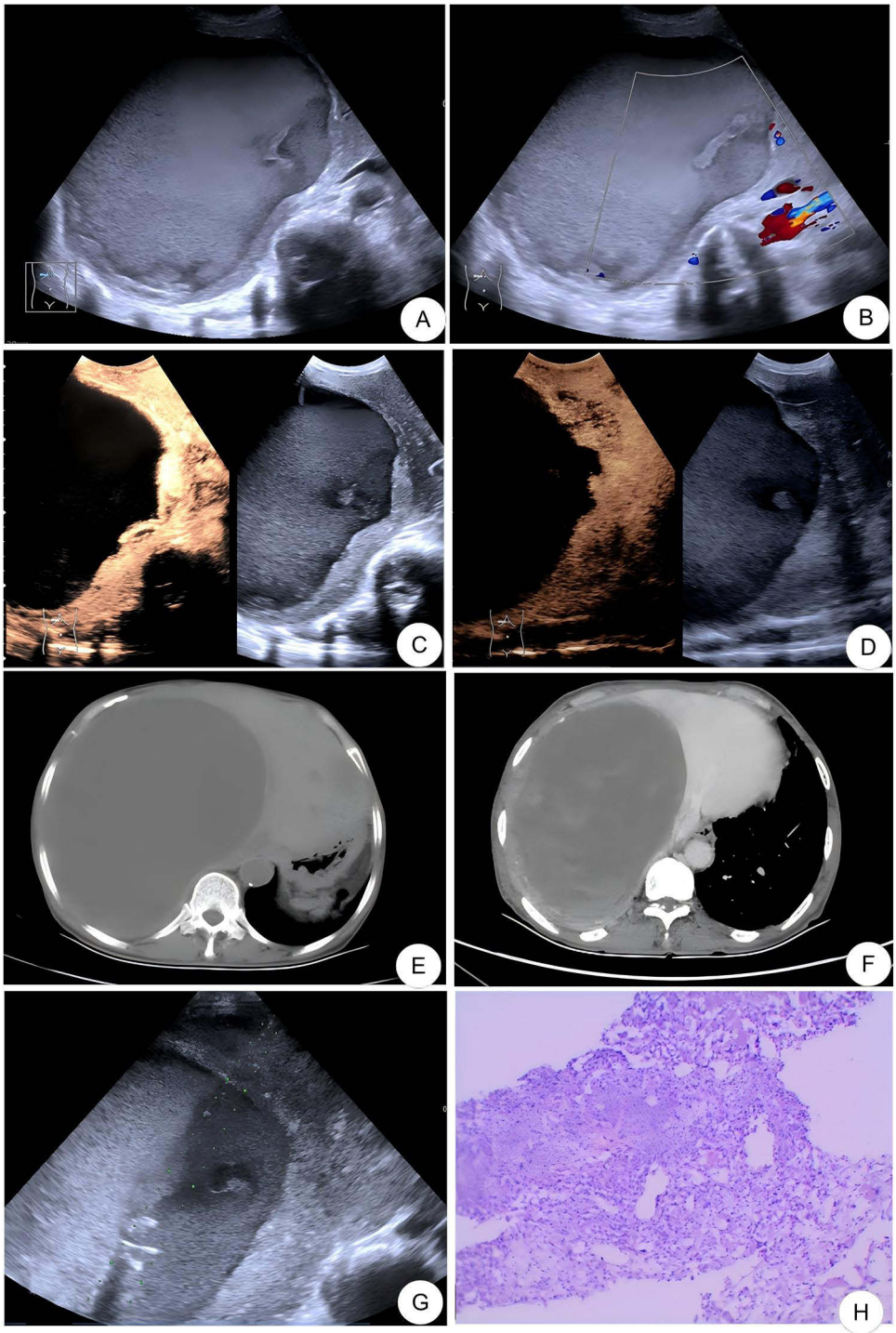


**Figure 2** A 64-year-old woman, 2D-US showed a well-circumscribed 24 cm complex-echo lesion in the right liver containing large cystic areas which presented poor sound transmission (**A**), with spot blood flow signals in the marginal of lesions (**B**). The pattern of CEUS in this patient was similar with the former, heterogeneous hyperechogenic enhancement of solid region in the arterial phase (**C**), and stayed hypo-enhancement in delay phase (**D**). CT scan showed a circular low-density areain the right lobe of the liver (**E**). CECT showed that the lesion margin was significantly enhanced in the arterial stage, and small blood vessel shadows could be seen around the lesion (**F**). The drainage tube (white arrow) was inserted into the cystic regions in the lesion (**G**). Histopathology was obtained by percutaneous liver biopsy. Hematoxylin and eosin, magnification 100× (**H**).

## Discussion

PEComa is a rare mesenchymal tumor characterized by histologically and immunohistochemically distinctive PECs.[12] The concept of PECs, describing an "unusual atypical cell type" with dual melanocytic and myoid differentiation and typically showing a perivascular distribution, was first proposed by Bonetti et al in 1992.[13] In 2013, the World Health Organization (WHO) defined PEComas as mesenchymal tumors composed of distinctive cells that show a focal association with blood vessel walls and typically express both melanocytic and smooth-muscle markers.[14,15] The term "PEComa" unifies a spectrum of rare, histologically diverse tumor types,[16] which can occur in the gastrointestinal tract, gallbladder, lung, bone, and almost any pelvic organ.[17–22] Primary hepatic PEComas appear to be rare, only a few cases of hepatic PEComa have been described worldwide.[23,24] And there was no mention in the literature of ultrasound presenting as a large cystic solid echogenicity. In our study, PEComas were found in the livers of two women, which was consistent with the literature reports that the tumor occurred markedly more frequent in females,[25,26] it illustrates hormones may play an important role in the pathophysiology. Previous studies reported that PEComas were commonly manifested abdominal pain, nausea, indigestion, and loss of appetite, but most patients were asymptomatic or have nonspecific gastrointestinal symptoms.[14] And in these cases, two patients were assumed to have unspecific pain and a large mass in the right upper quadrant, and no history of hepatitis or cirrhosis. Routine laboratory tests were unremarkable in the younger patient, while the other patient showed elevated $CA_{199}$ and $CA_{125}$, suggesting a higher likelihood of malignancy.

The lesions showed as well-defined round mass that may exhibit varying echogenicity on 2D-US and demonstrate a hypervascular pattern on CDFI. In our cases, 2D-US was performed a well-defined approximately 20 cm complex-echoic lesion in the liver containing large cystic areas. The enhanced features of CEUS also suggested that this mass was not a common hepatic tumor, it was easily misdiagnosed, similar to cases previously described in the literature.[27]

In summary, the diagnosis of PEComa is made based on histological examination of the tumor and subsequent additional immunohistochemical examination.[3] We presented two women of primary hepatic PEComa whose imaging findings showed large lesions with cystic areas, one of them presented with swollen lymph nodes, while the other did not, which is consistent with literature reports that some PEComas can exhibit malignant biological behavior.[28] It has also been reported in the literature that a tumor size ≥5 cm indicates a high risk of malignancy.[10] Therefore, younger patients are more likely to have benign or potentially malignant PEComa, whereas older patients tend to be diagnosed with malignant PEComa. Accordingly, although 2D-US cannot definitively determine the pathological type of all tumor, it can be used as a preliminary screening method to indicate space-occupying lesions. Moreover, with the application of CEUS, the diagnostic value of most tumors has been clinically established, which can provide clinicians with important information. Ultrasound-guided puncture biopsy can accurately, safely and effectively obtain a pathological diagnosis. For tumors containing extensive cystic fluid, ultrasound-guided catheter drainage provides useful preparation for subsequent clinical treatment.

## Data Sharing Statement

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

## Ethics Approval and Consent to Participate

This research was approved by the Institutional Review Board at Lanzhou University Second Hospital.

## Patient Consent for Publication

The patient provided written informed consent for the publication of any associated data.

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## Author Contributions

All authors made a significant contribution to the work reported, whether that is in the conception, study design, execution, acquisition of data, analysis and interpretation, or in all these areas; took part in drafting, revising or critically reviewing the article; gave final approval of the version to be published; have agreed on the journal to which the article has been submitted; and agree to be accountable for all aspects of the work.

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## Disclosure

The authors report no conflicts of interest in this work. No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

## References

1. Jia J, Luo J, Pan C-G, et al. Single-center experience in the diagnosis and treatment of hepatic perivascular epithelioid cell neoplasm. *J Clin Transl Hepatol*. 2022;10(1):72–79. doi:10.14218/JCTH.2020.00170
2. Perán Fernández C, de Paco Navaro Á, Castañer Ramón-Llin J, Bertelli Puche J, Sánchez Espinosa A. Perivascular epithelioid cell tumor (PEComa) of the liver. An extremely rare diagnosis. *Rev Esp Enferm Dig*. 2023;115(6):348–349. doi:10.17235/reed.2023.9558/2023
3. Folpe AL, Mentzel T, Lehr H-A, Fisher C, Balzer BL, Weiss SW. Perivascular epithelioid cell neoplasms of soft tissue and gynecologic origin: a clinicopathologic study of 26 cases and review of the literature. *Am J Surg Pathol*. 2005;29(12):1558–1575. doi:10.1097/01.pas.0000173232.22117.37
4. Kou Y-Q, Yang Y-P, Ye W-X, Yuan W-N, Du -S-S, Nie B. Perivascular epithelioid cell tumors of the liver misdiagnosed as hepatocellular carcinoma: three case reports. *World J Clin Cases*. 2023;11(2):426–433. doi:10.12998/wjcc.v11.i2.426
5. Amante MF. Hepatic perivascular epithelioid cell tumors: benign, malignant, and uncertain malignant potential. *World J Gastroenterol*. 2024;30(18):2374–2378. doi:10.3748/wjg.v30.i18.2374
6. Gao X, Tang H, Wang J, et al. Specific imaging features indicate the clinical features of patients with hepatic perivascular epithelioid cell tumor by comparative analysis of CT and ultrasound imaging. *Front Oncol*. 2022;12:908189. doi:10.3389/fonc.2022.908189
7. Xie S, Lu J, Dong J, Shen Z. A case report of a perivascular epithelioid cell tumor in the liver. *Asian J Surg*. 2023;46(11):5092–5093. doi:10.1016/j.asjsur.2023.06.094
8. Estébanez Ferrero B, Cabañó-Muñoz D, Velasco-Albendea FJ, Lorenzo-Liñán MÁ, Reina-Duarte Á. Epithelioid angiomyolipoma of the liver: an incidental diagnosis. *Rev Esp Enferm Dig*. 2022;114(11):684–685. doi:10.17235/reed.2022.8895/2022
9. Zhuang Y, Zeng Y, Ying L, Song C. Epithelioid angiomyolipoma of the liver: a case report. *Asian J Surg*. 2023;46(3):1376–1377. doi:10.1016/j.asjsur.2022.08.124
10. Bleeker JS, Quevedo JF, Folpe AL. "Malignant" perivascular epithelioid cell neoplasm: risk stratification and treatment strategies. *Sarcoma*. 2012;2012:541626. doi:10.1155/2012/541626
11. Dietrich CF, Nolsøe CP, Barr RG, et al. Guidelines and good clinical practice recommendations for Contrast Enhanced Ultrasound (CEUS) in the liver - update 2020 - WFUMB in Cooperation with EFSUMB, AFSUMB, AIUM, and FLAUS. *Ultraschall Med*. 2020;41(5):562–585. doi:10.1055/a-1177-0530
12. Nie P, Wu J, Wang H, et al. Primary hepatic perivascular epithelioid cell tumors: imaging findings with histopathological correlation. *Cancer Imaging*. 2019;19(1):32. doi:10.1186/s40644-019-0212-x
13. Tirumani SH, Shinagare AB, Hargreaves J, et al. Imaging features of primary and metastatic malignant perivascular epithelioid cell tumors. *AJR Am J Roentgenol*. 2014;202(2):252–258. doi:10.2214/AJR.13.10909
14. Son H-J, Kang DW, Kim JH, Han HY, Lee MK. Hepatic perivascular epithelioid cell tumor (PEComa): a case report with a review of literatures. *Clin Mol Hepatol*. 2017;23(1):80–86. doi:10.3350/cmh.2016.0034
15. Rosenberg AE. WHO classification of soft tissue and bone, fourth edition: summary and commentary. *Curr Opin Oncol*. 2013;25(5):571–573. doi:10.1097/01.cco.0000432522.16734.2d
16. Folpe AL, Kwiatkowski DJ. Perivascular epithelioid cell neoplasms: pathology and pathogenesis. *Hum Pathol*. 2010;41(1):1–15. doi:10.1016/j.humpath.2009.05.011
17. Kirste S, Kayser G, Zipfel A, Grosu A-L, Brunner T. Unresectable hepatic PEComa: a rare malignancy treated with stereotactic body radiation therapy (SBRT) followed by complete resection. *Radiat Oncol*. 2018;13(1):28. doi:10.1186/s13014-018-0974-5
18. Lu B, Wang C, Zhang J, et al. Perivascular epithelioid cell tumor of gastrointestinal tract: case report and review of the literature. *Medicine*. 2015;94(3):e393. doi:10.1097/MD.0000000000000393
19. Im S, Yoo C, Jung J-H, Choi HJ, Yoo J, Kang CS. Primary perivascular epithelioid cell tumor in the rectum: a case report and review of the literature. *Pathol Res Pract*. 2013;209(4):244–248. doi:10.1016/j.prp.2013.01.001
20. Zhao J, Teng H, Zhao R, et al. Malignant perivascular epithelioid cell tumor of the lung synchronous with a primary adenocarcinoma: one case report and review of the literature. *BMC Cancer*. 2019;19(1):235. doi:10.1186/s12885-019-5383-0
21. Ramezanpour S, Horvai AE, Zimel M, Bucknor M, Link TM. Fibroma-like perivascular epithelioid cell tumor: a rare case in a long bone. *Skeletal Radiol*. 2021;50(4):821–825. doi:10.1007/s00256-020-03610-w

22. Mor E, Visenzi C, Marasciulo F, Longo FR, Quaglia F. Ultrasound appearance of perivascular epithelioid cell tumor (PEComa). *Ultrasound Obstet Gynecol*. 2021;57(5):844–845. doi:10.1002/uog.22057
23. Tan Y, Zhang H, Wang X-C. Clear cell myomelanocytic tumor of the falciform ligament/ligamentum teres. *Indian J Pathol Microbiol*. 2014;57 (3):453–455. doi:10.4103/0377-4929.138766
24. Zhao LJ, Yang YJ, Wu H, Huang SM, Liu K. Perivascular epithelioid cell tumor of the liver: a case report and literature review. *Eur Rev Med Pharmacol Sci*. 2013;17(12):1665–1668.
25. Tan Y, Zhang H, Xiao EH. Perivascular epithelioid cell tumour: dynamic CT, MRI and clinicopathological characteristics--analysis of 32 cases and review of the literature. *Clin Radiol*. 2013;68(6):555–561. doi:10.1016/j.crad.2012.10.021
26. Ameurtesse H, Chbani L, Bennani A, et al. Primary perivascular epithelioid cell tumor of the liver: new case report and literature review. *Diagn Pathol*. 2014;9:149. doi:10.1186/1746-1596-9-149
27. Della Vigna P, Preda L, Monfardini L, Gorone MSP, Maffini FA, Bellomi M. Growing perivascular epithelioid cell tumor of the liver studied with contrast-enhanced ultrasonography and magnetic resonance imaging. *J Ultrasound Med*. 2008;27(12):1781–1785. doi:10.7863/jum.2008.27.12.1781
28. Bourgmayer A, Nannini S, Bonjean P, Kurtz J-E, Malouf GG, Gantzer J. Natural history and treatment strategies of advanced PEComas: a systematic review. *Cancers*. 2021;13(20):5227. doi:10.3390/cancers13205227