Annals of Clinical Case Studies

ISSN: 2688-1241

Case Report

Contrast-Enhanced Ultrasonography Imaging Features of Perivascular Epithelioid Cell Tumor (PEcoma)

Terracciano Fulvia[1], Marra Antonella[1], Covelli Claudia[2] and Perri Francesco[1]*

[1]Department of Gastroenterology and Endoscopy, IRCCS, Fondazione Casa Sollievo della Sofferenza, Italy

[2]Pathology Unit, IRCCS Fondazione Casa Sollievo Della Sofferenza, Italy

Abstract

We report a case of primary hepatic PEComa presenting as a single mass in a 48-year-old woman.

Keywords: Liver mass; Transabdominal ultrasound; Biopsy; Histology

Introduction

Perivascular epithelioid cell tumors, or PEComas, are rare mesenchymal neoplasms composed of distinctive epithelioid cells, which are immunoreactive for both smooth muscle and melanocytic markers [1]. These tumors can arise anywhere in the body, but develop predominantly in the uterus, skin, upper aero-digestive tract, urinary bladder and intestinal tract. In the liver, primary PEComas are particularly rare. Here, we report a case of primary hepatic PEComa presenting as a single mass in a 48-year-old woman.

Case Presentation

A 48-year-old woman was admitted to our Institution for an asymptomatic liver mass that was incidentally detected on ultrasonography performed for follow-up.

The patient was affected by systemic lupus erythematosus, diagnosed almost 30 years ago, in treatment with corticosteroids and immunosuppressive drugs, and had a completely excised malignant melanoma of the left thigh 6 years ago, with a Breslow's depth of 2 mm and a negative sentinel lymph node. In addition, the patient underwent a segmental liver resection for a solid mass in 2014, which was diagnosed as angiomyolipoma. The physical examination was negative, and laboratory investigations including liver function tests, hepatitis panel and tumor markers were normal.

Abdominal ultrasonography revealed a homogeneously hypoechoic mass in the segment 2 of the liver, measuring 28 mm × 25 mm, with a thin hyperechoic halo (Figure 1), and a hyperechogenic area of focal steatosis anterior to the portal bifurcation in segment 4. Power Doppler ultrasonography showed that the marginal low-echoic lesion was composed of displaced vessels around the tumor. The right hepatic lobe showed the presence of multiple hypoechoic lesions of 8 mm-15 mm in diameter, with no evidence of hyperechoic halo. No dilated peripheral bile ducts or blood infiltration were observed.

Contrast-Enhanced Ultrasonography (CEUS) using SonoVue contrast agent, showed that the lesion in the segment 2 was homogeneously IPeR enhanced in early arterial phase with isoehancement in portal and venous phases (Figure 2).

On contrast-enhanced Computed Tomography (CT) scan, the lesions showed a strong heterogeneous enhancement in the arterial phase with a slight decrease during the portal phase, and a weak washout pattern in the venous phase. Neither lymphadenopathy nor portal vein involvement was present.

On MRI, the mass at segment 2 was hypointense on T1-weighted images, while the other lesions were isointense. A strong enhancement during the arterial phase without wash-out in the venous phase was observed in all lesions. Only the lesion in the left lobe was isointense with hyperintense halo in the delayed phase, while the others remained isointense.

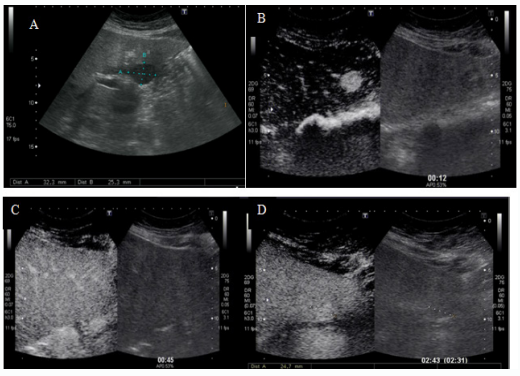

Figure 1: (a). Ultrasonography showed a well-demarcated isoechoic tumor with a diameter of 30 mm in segment 2 of the liver. (b). Sonovue contrast-enhanced showed early homogeneously in arterial phase (C). Isoehancement in portal (d). Venous phases.

Citation: Fulvia T, Antonella M, Claudia C, Francesco P. Contrast-Enhanced Ultrasonography Imaging Features of Perivascular Epithelioid Cell Tumor (PEcoma). Ann Clin Case Stud. 2023; 5(2): 1073.

Publisher Name: Medtext Publications LLC

Manuscript compiled: Feb 08th, 2023

***Corresponding author:** Terracciano Fulvia, Division of Gastroenterology and Endoscopy, IRCCS, Fondazione Casa Sollievo Della Sofferenza, Viale Cappuccini 1, 71024 San Giovanni Rotondo, Italy, Tel: +39-3393185405; E-mail: terracciano74@hotmail.com

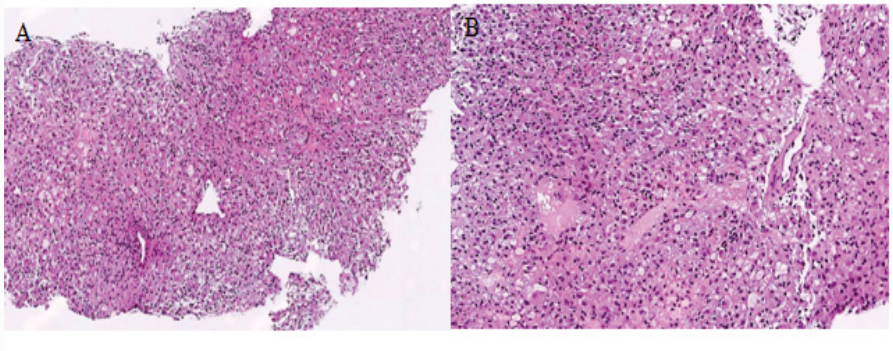

Figure 2: (a). The lesion consists of nests of epithelioid cells with abundant granular eosinophilic, sometimes clear cytoplasm. The nests are surrounded by thin-walled vessels (Hematoxylin and Eosin, 7X). (b). Higher magnification shows round nuclei and inconspicuous nucleoli in the neoplastic cells. No pleomorphism, mitotic figures or necrosis are seen (Hematoxylin and Eosin, 18X).

Due to the previous diagnosis of malignancy, the discordance between CT and MRI and the different contrast characteristics, we performed an ultrasound-guided fine needle biopsy of the mass at segment 2.

Microscopically, the biopsy consisted of a fragment of unremarkable liver tissue and fragments of a neoplasm composed of epithelioid cells with abundant eosinophilic and granular cytoplasm, arranged in a nested pattern. No atypia, mitoses or necrosis were observed. There was no evidence of adipose tissue or thick-walled blood vessels. On immunohistochemistry, the neoplastic cells were positive for HMB45, melan A, α-Smooth Muscle Actin (SMA); negative for S100, SOX10, TFE3, and pan-cytokeratin (Figure 3). On the basis of these findings, a diagnosis of PEComa was rendered.

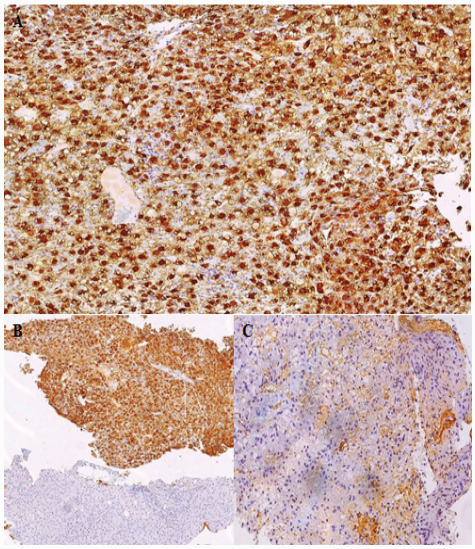

Figure 3: On immunohistochemistry, the neoplastic cells characteristically express the melanocytic markers HMB45 (a). 11X, and Melan-A (b). 6X, as well as the alpha-Smooth Muscle Actin (a-SMA) marker (c). 28X. in Figure 3B, a fragment of background liver parenchyma is also present.

Discussion

The term PEComa was first introduced by Zamboni et al. [2] to describe a group of neoplasms composed of histologically and immunohistochemically distinctive perivascular epithelioid cells. The PEComa family includes angiomyolipoma (AML), Clear Cell 'Sugar' Tumor of the lung (CCST), Lymphangioleiomyomatosis (LAM), clear cell myomelanocytic tumor of the falciform ligament/ligamentum teres (CCMMT) and unusual clear cell tumors of the pancreas, rectum, abdominal serosa, uterus, vulva, thigh, and heart. Angiomyolipomas (AMLs) are a subtype of PEComa arising almost exclusively in the liver, kidney and pancreas, and are characterized by an admixture of epithelioid cells, adipocytes and thick-walled blood vessels, in variable proportions.

Primary hepatic PEComas are very rare. Most are sporadic and solitary lesions, with only 5%-10% arising in patients with tuberous sclerosis, more frequently in a multifocal fashion [3].

Clinical manifestations of hepatic PEComas usually are minimal. Gastrointestinal symptoms, such as abdominal pain, abdominal discomfort, nausea or vomiting may be present, especially when the tumor grows in size and causes localized pressure effect [4].

Diagnosis with imaging techniques only can be challenging because of the different distribution of adipose tissue, irregular vessels and smooth muscle cells, and therefore histological sampling is needed for a definite diagnosis.

On ultrasound examination, different patterns of echogenicity have been described in the Literature: hyperechoic [5-7], isoechoic [8], hypoechoic [9], and even with mixed echogenicity [10]. In one report, the lesion appeared hypoechoic with a thin, slightly hyperechoic halo [4], similarly to our case. With regard to the contrast-enhanced ultrasound, two studies described the contrasting behavior of hepatic PEComas. In 2008 Della Vigna et al. [5] after injecting a sulfur hexafluoride microbubble as a contrast medium (SonVue) described the tumor as hyphenated in early arterial phase, isoechoic in the portal phase, and hyperechogenicity in the equilibrium phase [5]. In the study of Akitake, the lesion was studied with a newly developed contrast-enhancing reagent, Sonazoid [8], and it showed early influx into the tumor and rapid drainage of arterial blood to veins suggesting the existence of arteriovenous shunt. In these two studies, both reagents highlighted the hyperenhancement in the arterial phase and the hypoenhancement in the portal and delayed phases of the tumor. These characteristics were the same in our patients as well.

From a radiological point of view, PEComas of the liver show a variety of patterns. Almost all lesions reported in the Literature show hypervascularity and arteriovenous connections in contrast-enhanced CT scan [2], and appeared hypointense on T1-weighted images, had low-signal on T2-weighted images, and had no fat signal on magnetic resonance imaging MRI [5,7-9,11]. After injecting

gadolinium, the lesions usually present a strong enhancement in the arterial phase, with slight attenuation during the portal phase and remained hypointense in the late parenchymal phase. In most of the cases, PEComas follow a benign course; however, criteria for identifying potential malignant behavior remain unclear.

In a study of PEComas of soft tissue and gynecologic origin, Folpe et al. [12] identified the following as high-risk features: a diameter greater than 5 cm; an infiltrative growth pattern; high nuclear grade and cellularity; a mitotic rate >1/50 per High Power Field; necrosis or vascular invasion. According to the Authors, the presence of two or more high risk features would define a PEComa as malignant [12]. Similarly, in the gastrointestinal tract, marked nuclear atypia, diffuse pleomorphism, and mitotic activity were described as the strongest predictors of malignant behavior [13]. In our case, the lesion was 2 cm in diameter and had no mitosis, necrosis or atypia.

Nevertheless, specific radiological or histopathological features for risk stratification of primary hepatic PEComa have not yet been defined.

Therefore, because of limited data on tumor behavior, complete excision and follow-up are indicated in all cases.

References

1. WHO Classification of Tumours Editorial Board. Soft tissue and bone tumours. Lyon, France: International Agency for Research on Cancer. 5th ed. 2020;3.
2. Zamboni G, Pea M, Martignoni G, Zancanaro C, Faccioli G, Gilioli E, et al. Clear cell "sugar" tumor of the pancreas. A novel member of the family of lesions characterized by the presence of perivascular epithelioid cells. Am J Surg Pathol. 1996;20(6):722-30.
3. WHO Classification of Tumours Editorial Board. Digestive system tumours. Lyon, France: International Agency for Research on Cancer. 5thed. 2019;1.
4. Tang D, Wang J, Tian Y, Li Q, Yan H, Wang B, et al. Hepatic perivascularepithelioid cell tumor: case report and brief literature review. Medicine (Baltimore). 2016;95(51):e5572.
5. Vigna DP, Preda L, Monfardini L, Gorone MSP, Maffini FA, Bellomi M. Growing perivascularepithelioid cell tumor of the liver studied with contrast enhancedultrasonography and magnetic resonance imaging. J Ultrasound Med. 2008;27(12):1781-5.
6. Yamasaki S, Tanaka S, Fujii H, Matsumoto T, Okuda C, Watanabe G, et al. Monotypic epithelioid angiomyolipoma of the liver. Histopathology. 2000;36(5):451-6.
7. Xie L, Jessurun J, Manivel JC, Pambuccian S. Hepatic epithelioid angiomyolipoma with trabecular growth pattern: a mimic of hepatocellular carcinoma on fine needle aspiration cytology. Diagno Cytopathol. 2012;40(7):639-50.
8. Akitake R, Kimura H, Sekoguchi S, Nakamura H, Seno H, Chiba T, et al. Perivascular epithelioidcell tumor (PEComa) of the liver diagnosed by contrast-enhanced ultrasonography. Intern Med. 2009;48(24):2083-6.
9. Tan Y, Xiao EH. Hepatic perivascular epithelioid cell tumor (PEComa): dynamic CT, MRI, ultrasonography, and pathologic features--analysis of 7 cases and review of the literature. Abdom Imaging. 2012;37(5):781-7.
10. Strzelczyk JM, Durczynski A, Szymanski D, Jablkowski M, Dworniak D, Sporny S. Primary perivascular epithelioid cell tumor (PEComa) of the liver: report of a case. Surg Today. 2009;39(10):916-92.
11. Fang SH, Zhou LN, Jin M, Hu JB. Perivascular epithelioid cell tumor of the liver: a report of two cases and review of the literature. World J Gastroenterol. 2007;13(41):5537-9.
12. Folpe AL, Mentzel T, Lehr HA, 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):1558-75.
13. Doyle LA, Hornick JL, Fletcher CDM. PEComa of the gastrointestinal tract: clinicopathologic study of 35 cases with evaluation of prognostic parameters. Am J Surg Pathol. 2013;37(12):1769-82.