Primary perivascular epithelioid cell tumor (PEComa) of the liver is a very rare tumor that originates from mesenchyma. Gastrointestinal tract with perivascular distribution is the most common anatomic sites of these tumors. Only few cases of hepatic PEComa have been described so far. Malignant PEComas exhibit aggressive behavior with poor prognosis, making early diagnosis crucial. Hereby, we report a 79-year-old female with unusually located mass in the liver. A partial curative hepatectomy has been done, and PEComa was diagnosed histopathologically. No evidence of recurrence was observed during the 6-month follow-up.

**Keywords:** Computed tomography, liver, magnetic resonance imaging, perivascular epithelioid cell tumor

**Case Report**

A 79-year-old female who was previously healthy admitted to our hospital with a 1 year history of constant dull right upper quadrant pain and tenderness. All laboratory tests including tumor markers were in normal limits. There was not any story of hepatitis or chronic liver disease. Abdominal ultrasound revealed a hypoechoic solid lesion that located in the right and caudate lobe of liver with irregular border. MDCT examination reveals a liver mass with 52 mm × 43 mm diameters located in segment VIII and caudate lobe of the liver [Figure 1]. The lesion showed heterogeneous

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enhancement in arterial phase with hypointense central area also. The mass become hypodense to isodense to adjacent parenchyma on delayed venous phase. The region of nonenhancing hypointense area was likely to represent central necrosis, which was frequently seen in malign PEComas. The patient underwent MRI, after first assessment with MDCT for better evaluation of vascular invasion. In MRI, the tumor had intermediate isointense signal on T1-weighted images and heterogeneously hyperintense on T2-weighted images relative to the liver parenchyma [Figure 2]. There was not seen any fat component in the lesion. On gadolinium (Gd)-enhanced images, the mass showed significant and heterogeneous enhancement. Nonenhancing hypointense area in tumor center representing necrosis. Inferior vena cava (IVC) invasion with tumor thrombus into the lumen is also demonstrated, especially coronal Gd-enhanced images [Figure 3]. There was also marked diffusion restriction of lesion on diffusion-weighted images as totally hypointense signal in apparent diffusion coefficient maps [Figure 4].

Calcification and/or hemorrhage were not detected in the lesion. Metastasis in other systems was also not found. The patient consented to tumor biopsy after radiologic assessment. The patient underwent biopsy of the liver mass, and pathology revealed a hepatic perivascular epithelioid cell tumor or “PEComa.” Histologic findings were a mixture of large epithelioid cells and clear-to-eosinophilic granular cytoplasm [Figure 5]. There was severe nuclear atypia with large nucleoli and increased mitotic activity detected microscopically. However, immunochemistry study indicated that tumor cells were positive for human melanoma black (HMB)-45 marker. Following the surgery, the patient has not shown any evidence of recurrence for 6 months.

**DISCUSSION**

PEComa was first proposed in 1992 by Bonetti et al. and also classified by the World Health Organization in 2002 as a mesenchymal tumor.[4] However, perivascular epithelioid cells were first defined by Apitz in 1944. PEComas are tumors that consist of epithelioid cells with histologically and immunohistochemically specific markers for melanocytes and smooth muscles.[5] This tumor had extensive family with various cytologic subtypes that include classic and epithelioid angiomyolipoma, clear cell tumor of the lung, lymphangioleiomyomatosis, clear cell tumors of the retroperitoneum, rectum, heart, pancreas, and uterus, and also myomelanocytic clear cell tumor of the ligamentum teres in liver.[6]

PEComas are predominantly seen in women in the first three decades (3–29 years) of life. These tumors are mostly asymptomatic in early stages. However, it may cause vaguely pain once they grow to moderately large size. Liver PEComas have a tendency to locate at falciform ligament/ligamentum teres. Folpe et al. reported seven cases with liver PEComas located within or abutting near the falciform ligament or ligamentum teres.[7] PEComas may be located in another place of liver rarely as seen in this case report. Besides, in this case, the mass was located at the medial aspect of the liver with the invasion of IVC. In literature, it was not concluded any IVC invasion of PEComa until today.
Hepatic PEComas usually exhibit distinct margins. In general, PEComas have been tend to exhibit benign behavior in the majority of the cases. However, several malignant PEComas have been reported.[3,5,6] It was concluded that tumor was >5 cm, and concomitant necrosis was criteria for malignancy.[8] In another study, Tan and Xiao detected malign PEComas range accepted from 2.5 to 8.5 cm with a mean of 4 cm.[9]

Ultrasound, computed tomography (CT) and MRI are not sufficiently sensitive to enable exact diagnosis of PEComa because of its nonspecific radiologic characteristics. Furthermore, distinct criteria between benign and malign types of PEComas have not been defined in most of the studies until today.[10,11]

CT and MRI studies should be made with contrast enhanced for exact evaluation of liver PEComas. Typically, lesion reveals heterogeneous enhancement on arterial and early portal phase of contrast-enhanced CT and MRI.[6] The imaging characteristics of hepatic PEComa may commonly mimic malignant liver tumors such as hepatocellular carcinoma or metastases. Hypervascularity and also arteriovenous connections could be responsible for misdiagnosis in some cases. On the other hand, chronic liver disease is cooccurrence with liver PEComas in the elderly. If the liver parenchyma is normal appearance, hepatocellular carcinoma should not be considered, firstly. Literature reviews suggest that hepatic PEComas were detected in livers without a background of cirrhosis or hepatitis, also.[9] Nonenhancement areas in the mass with low attenuation generally show internal necrosis, which is frequently due to the rapid growth of PEComas.

Hepatic PEComas are mostly isointense to mildly hypointense to muscle on T1-weighted images on MRI. If the presence hemorrhagic components into the mass, it could be seen as hyperintense on fat-suppressed T1-weighted images. Heterogeneous hyperintense signal is characteristic sign on T2-weighted images for PEComas, especially for malign ones. Besides, some regions in the lesion with T2-shortening brightness could be seen due to internal protein component or hemorrhage.[10,11] Tumor margins with well demarcated are better seen on T2 images. Contrast-enhanced sequences always should be added to MR studies in PEComas.

Enhancement characteristics of PEComas are very variable; a recent study concluded that PEComas tend to show intense enhancement.[11] However, PEComas may show persistent enhancement in late phases in some cases, which gives rise to mistake as benign masses such as focal nodular hyperplasia and hemangioma.[6,9]

Exact diagnostic type of PEComa depends on the pathology and also immunohistochemistry. PEComa cells show similarity to epithelioid cells with dilated vascular channels and contain eosinophilic cytoplasm. Nevertheless, all of the PEComas nearly are diagnosed with positive melanocytic markers and smooth muscle markers in studies.[4,12] In this case report, the tumor was positive for HMB-45 marker also.

Today, surgery with adequate margins remains the gold standard for treatment of PEComas, especially in malignant ones.

CONCLUSION

PEComa is detected increasingly in liver. Dynamic and multiplanar imaging should be done with MDCT and MRI at invasive liver lesions. If an invasive lesion
was found in liver without a background of hepatitis or cirrhosis, liver PEComa should be considered in differential diagnosis. The exact diagnosis of PEComa depends on the pathologic observations basically. Surgery appears to be necessary for a total cure. These tumors show benign behavior mostly, and the prognosis seems good after surgery.

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Conflicts of interest
There are no conflicts of interest.

References
1. Martignoni G, Pea M, Reghellin D, Zamboni G, Bonetti F. PEComas: The past, the present and the future. Virchows Arch 2008;452:119-32.
2. 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;13:5537-9.
3. Lao IW, Yu L, Wang J. Malignant perivascular epithelioid cell tumor (PEComa) of the femur: A case report and literature review. Diagn Pathol 2015;10:54.
4. Liu D, Shi D, Xu Y, Cao L. Management of perivascular epithelioid cell tumor of the liver: A case report and review of the literature. Oncol Lett 2014;7:148-152.
5. Abhirup B, Kaushal K, Sanket M, Ganesh N. Malignant hepatic perivascular epithelioid cell tumor (PEComa) - Case report and a brief review. J Egypt Natl Canc Inst 2015;27:239-42.
6. Phillips CH, Keraliya AR, Shinagare AB, Ramaiya NH, Tirumani SH. Update on the imaging of malignant perivascular epithelioid cell tumors (PEComas). Abdom Radiol (NY) 2016;41:368-76.
7. Folpe AL, Goodman ZD, Ishak KG, Paulino AF, Taboada EM, Meehan SA, et al. Clear cell myomelanocytic tumor of the falciform ligament/ligamentum teres: A novel member of the perivascular epithelioid clear cell family of tumors with a predilection for children and young adults. Am J Surg Pathol 2000;24:1239-46.
8. Bleeker JS, Quevedo JF, Folpe AL. “Malignant” perivascular epithelioid cell neoplasm: Risk stratification and treatment strategies. Sarcoma 2012;2012:541626-38.
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;37:781-7.
10. Khaja F, Carilli A, Baidas S, Sriharan A, Norford S. PEComa: A perivascular epithelioid cell tumor in the liver-A case report and review of the literature. Case Rep Med 2013;2013:904126.
11. Tirumani SH, Shinagare AB, Hargreaves J, Jagannathan JP, Hornick JL, Wagner AJ, et al. Imaging features of primary and metastatic malignant perivascular epithelioid cell tumors. AJR Am J Roentgenol 2014;202:252-8.
12. Zimmermann A, von der Brelie C, Berger B, Kappeler A, Candinas D. Primary perivascular epithelioid cell tumor of the liver not related to hepatic ligaments: Hepatic PEComa as an emerging entity. Histol Histopathol 2008;23:1185-93.