

Malignant hepatic epithelioid hemangioendothelioma: case of a rare liver tumor mimicking metastasis

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A 30-year-old female presented to our hospital with a fever. She was diagnosed with urinary tract infection and bacteremia. After completing intravenous antibiotics, she began having persistent diarrhea. Computerized tomography (CT) of the chest and abdomen was performed to explain the underlying cause of her symptoms.

CT scan of the liver showed two well-defined nodular masses in the peripheral aspect of the segment 2

liver parenchyma, with minimal associated capsular retraction (Fig. 1). Additionally, in the chest, predominantly subcentimeter bilateral pulmonary nodules were noted within the lung parenchyma, which was suspicious for metastases (Fig. 1). Liver function test and tumor markers were all within normal limits. Serological tests presented negative results.

Magnetic resonance imaging (MRI) of the abdomen was performed for further characterization.



Figure 1. On contrast-enhanced computed tomography (CT) scan: **AB.** A slightly enhanced target-like nodular mass measuring 4.0 x 3.5 cm in the peripheral aspect of liver segment 2, with minimal associated capsular retraction; **CD.** Two well-defined slightly enhancing target-like nodular masses measuring 5.0 x 4.0 and 4.0 x 3.5 cm in the peripheral aspect of segment 2 liver parenchyma with minimal associated capsular retraction. On axial images of contrast-enhanced chest CT: **EF.** predominantly subcentimeter bilateral pulmonary nodules (white arrows) within the lung parenchyma

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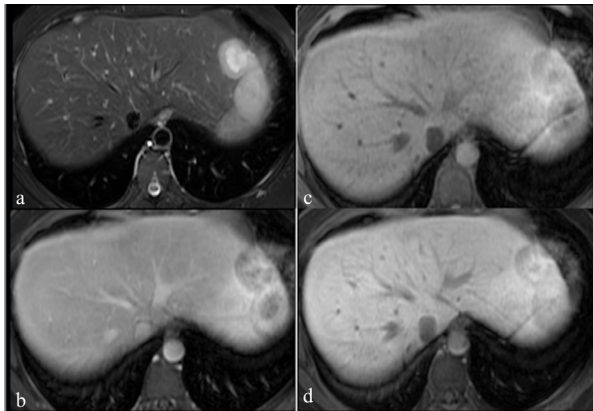


Figure 2. Contrast-enhanced magnetic resonance imaging (MRI) of the abdomen. **A.** T2 heterogeneously hyperintense, well-defined masses in the peripheral aspect of segment 2; **B.–D.** T1 predominantly hypointense well-defined masses in the peripheral aspect of liver segment 2 with target type contrast enhancement and minimal capsular retraction

Lesions appeared as well-defined masses in the peripheral aspect of segment 2 with target type contrast enhancement and minimal capsular retraction (Fig. 2).

CT-guided biopsy of the lesions revealed that the patient’s tumor cells were diffusely positive for CD34, ERG, and CAMTA1, which was suggestive of epithelioid hemangioendothelioma. Additional CT-guided lung biopsy from the left lower lobe nodule demonstrated that cells were positive for ERG and CD31, supporting the diagnosis.

Partial hepatectomy was performed since the condition only involved segment 2 of liver parenchyma. The patient received chemotherapy for the metastatic lung nodules. The patient appeared well at the latest follow-up without signs of recurrence on the remaining liver.

Hepatic epithelioid hemangioendothelioma (HEHE) is a rare tumor with an incidence less than 0.1% per 100,000 [1]. It has a greater female incidence with a peak around patients aged 30–40 years old. Its prognosis has a relatively favorable, but overall variable and unpredictable course.

These tumors might be asymptomatic or symptomatic. Most common signs and symptoms include right upper quadrant pain jaundice, hepatosplenomegaly, and Budd-Chiari syndrome.

Different spectrums of tumor growth may be observed including the more common nodular form where multiple liver nodules coalesce to form large,

confluent masses over time, and very rarely in diffuse or extensive form.

On CT, HEHE lesions are characteristically seen as multiple low attenuated and subcapsular localized masses. Target-like enhancement patterns or peripheral enhancement may be seen [2].

MRI commonly shows T1 hypointense T2 hyperintense lesions with either a peripheral halo or a target-type enhancement on post-contrast sequences [3]. Lesions demonstrate a peripheral halo or a target-type enhancement pattern with a thin peripheral hypointense rim [3].

Differential diagnoses include angiosarcoma, peripheral cholangiocarcinoma, treated malignancy, and focal confluent fibrosis. Because long-term survival is possible, this tumor must be distinguished from other malignant tumors such as adenocarcinoma and angiosarcoma.

The definitive diagnosis is only possible with histopathological examination. At least one endothelial marker (factor VIII-related antigen, CD34 or CD31, or both) is positive in tumors, while epithelial markers (cytokeratin and carcinoembryonic antigen) are negative [3].

Metastasis has been reported in 30% of patients at presentation, most commonly in the lungs [4].

The treatments of choice are radical surgical resection or orthotopic liver transplantation.

Herein, we highlight the importance of considering hepatic epithelioid hemangioendothelioma when multiple liver lesions in a largely peripheral distribution with associated capsular retraction are detected. Differential diagnosis of HEHE from other malignant liver tumors is important due to its relatively good prognosis.

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