Hepatic Epithelioid Hemangioendothelioma – a Rare Mimicker of Hepatic Metastasis

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Introduction

Hepatic epithelioid hemangioendothelioma (EHE) is a malignant tumor of vascular origin with an estimated incidence of one in a million. The tumor derives its name from the characteristic composition of dendritic and endothelial cells with epithelioid morphology. Mostly asymptomatic, EHE is usually an incidental radiographic diagnosis that can mimic other liver tumors such as metastasis, hepatocellular carcinoma (HCC), angiosarcoma, or cholangiocarcinoma. We present a case of incidental diagnosis of hepatic EHE masquerading as multifocal liver metastasis.

Case Presentation

An 80-year-old man with coronary artery disease presented with 5 days of diarrhea and black stools. He was taking bismuth subsalicylate in an attempt to ease diarrhea. Labs showed mild anemia with hemoglobin (Hb) of 13.1 g/dL with normal platelets and INR. A CT abdomen with contrast showed mild hepatomegaly and multiple space-occupying lesions in the liver concerning for metastatic cancer. The patient thereby underwent an EGD and colonoscopy which was negative for a primary GI malignancy. Ultrasound-guided liver biopsy was then pursued. Immunohistochemical stains performed on the core biopsy showed that the cells were positive for CD31, CD34 and ERG and were negative for cytokeratin-20, CDX-2, TTF-1, arginase, and glypican-3. Focal staining was noted with cytokeratin AE1/AE3 and cytokeratin-7. Special stain for mucicarmine was negative and CAMTA was positive. These findings were consistent with diagnosis of hepatic EHE. His diarrhea resolved and Hb remained stable. He was discharged with outpatient oncology follow-up.

Associated Pathology and Imaging

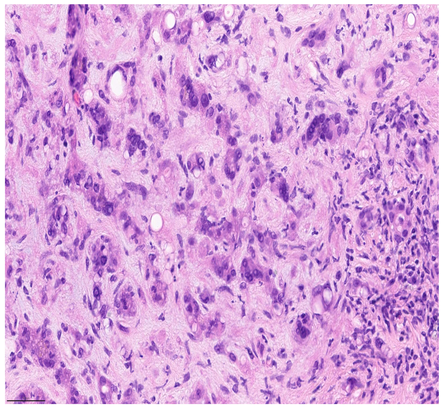

Image 1: Hematoxylin and Eosin staining of liver biopsy in Hepatic EHE

Of note, CAMTA staining was additionally completed on this patient's liver biopsy, but images were unable to be obtained as they were performed at an outside facility.

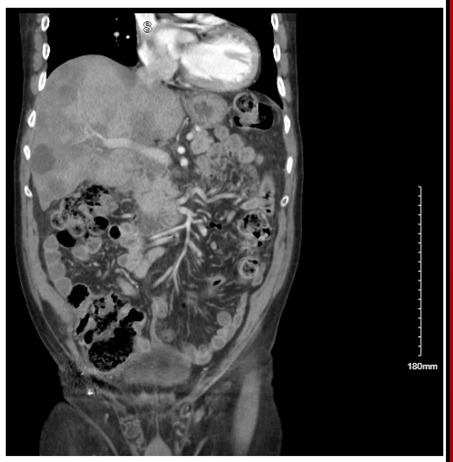

Image 2: Initial CT of the abdomen and pelvis with contrast performed on admission that demonstrates multiple space-occupying lesions in the liver and mild hepatomegaly

Treatment Options

- Treatment options are typically chosen based on the extent of the disease.
- EHE confined within the liver has been treated with liver resection, liver transplantation, radiofrequency ablation, or simply a wait and watch method.
- Patients with extrahepatic involvement have been treated with systemic therapy including a variety of cytotoxic chemotherapy, immune therapy, or targeted therapy.

Discussion

- Hepatic EHE is typically diagnosed incidentally.
- However, the course of EHE may range from indolent to aggressive disease with distant metastases.
- Suspicion for EHE may arise when a patient is discovered to have multiple liver lesions with no identifiable primary cancer.
- Liver biopsy is required for diagnosis as EHE has unique histologic, immunohistochemical, and molecular characteristics.
- EHE usually stains positive for vascular markers, such as factor VIII-related antigen, CD31, and CD34, and negative for cytokeratins, as seen in this case.
- The most common identifiable genetic marker associated with EHE is the translocation t(1;3)(p36.3;q25), resulting in the CAMTA1- WWTR1 fusion product.
- As EHE remains exceptionally rare, prompt diagnosis should be followed by a multidisciplinary discussion for individualized care.

References

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