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Case Reports

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Hepatic Epithelioid Hemangioendothelioma: A Rare Malignant Lesion

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Background

- First described in 1982 Epithelioid Hemangioendothelioma is a rare, low to intermediate grade malignant lesion of vascular origin
- This tumor usually occurs in the soft tissues of the extremities and visceral organs, commonly the liver
- Hepatic Epithelioid Hemangioendotheliomas (HEHE) have an incidence of less than 0.1 cases per 100,000
- HEHE usually affects females in a 3:2 predominance
- Clinical manifestations are nonspecific and many patients remain asymptomatic
- Tumor marker levels are often normal

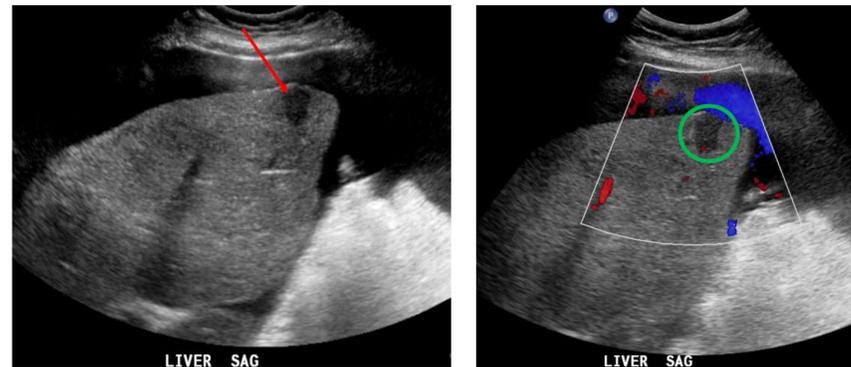
Case Presentation

- 34 y/o male with a past medical history of deep venous thrombosis who presented with increased shortness of breath and weight gain. Patient also noted increased abdominal girth.
- Symptoms had been worsening over a period of months
- No significant family history. No history of alcohol use.
- Physical exam showed obesity and ascites

Diagnostic Workup and Clinical Course

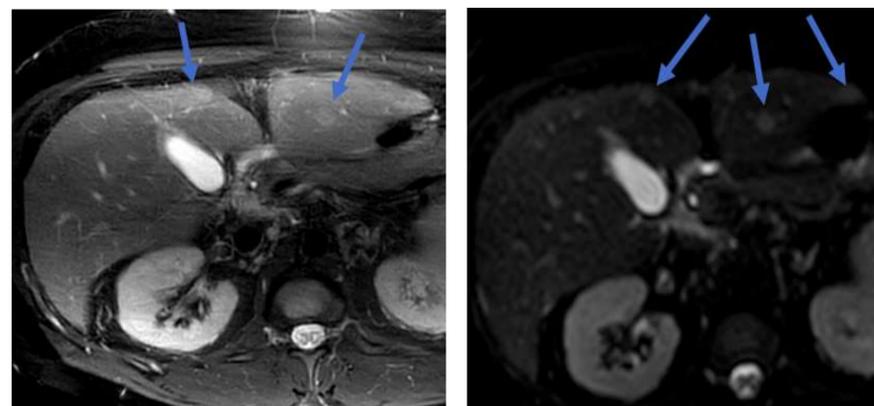
- Laboratory testing:
 - HCV Ab and HBsAg negative
 - ANA, AMA, AMSA negative
 - AFP 1.5 ng/mL
 - Normal liver function tests
 - MELD-Na score of 17
- Ultrasound ordered and showed steatosis and multiple small incompletely characterized liver masses
- Given workup, concern for non-alcoholic steatohepatitis related cirrhosis with possible hepatocellular carcinoma
- Multiphasic CT and liver biopsy negative for malignancy but showed evidence of venous outflow obstruction
- Angiogram demonstrated severe stenosis of the inferior vena cava at the right atrial junction which was treated with angioplasty, resulting in temporary relief in symptoms
- Patient returned with increasing ascites
- Repeat MRI showed multiple ring enhancing masses thought to be metastases or abscesses
- Biopsy showed vascular neoplasm consistent with epithelioid hemangioendothelioma
- Patient was then referred to the hepatobiliary transplant service for transplant consideration

Ultrasound

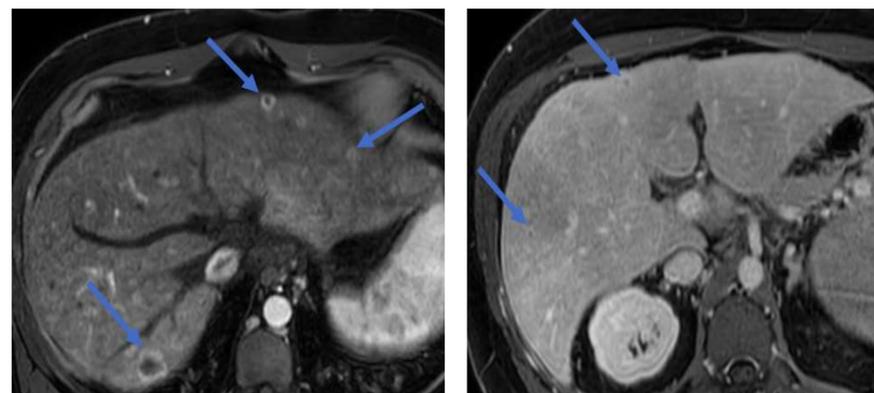


Sagittal images of the left hepatic lobe show a peripheral hypoechoic lesion measuring 2cm with no internal flow on color doppler

MRI



Axial T2-weighted fat saturation image (left) shows multiple hyperintense lesions. Axial Diffusion weighted image (right) at the same level shows these lesions restrict diffusion.



T2 weighted post contrast axial slice of the upper abdomen (left) shows these lesions demonstrate ring like peripheral enhancement. Five minute delayed post contrast axial image (right) shows these lesions demonstrate central filling.

Imaging Characteristics

- The imaging manifestations of HEHE are variable and depend on the stage of disease at presentation
- If found early, HEHE may present as a solitary lesion, however most patients present with advanced disease and will have multiple peripheral, coalescent lesions with capsular retraction
- On unenhanced CT, HEHE lesions are typically hypoattenuating without calcifications. Enhanced CT may show the characteristic “lollipop sign” caused by hepatic or portal vein branches which terminate within the edges of lesions.
- On MRI, HEHE lesions are typically hypointense on T1-weighted images and have heterogeneously increased signal intensity on T2
- The most common pattern of enhancement is a peripheral halo with an occasional thin peripheral hypointense rim
- Ferumoxide-enhanced T2 images can be used to distinguish tumor margins.
- Many reported cases of biopsy proven HEHE do not present with many of these characteristic imaging findings

Discussion

- Given its variable appearance, 60-80% of these lesions are initially misdiagnosed
- Underlying comorbidities such as cirrhosis, found in only 1% of HEHE cases, can also confound diagnosis
- Differentiating HEHE from its differential diagnoses such as hepatic metastatic carcinoma, cholangiocarcinoma, and other liver vascular tumors like hepatic angiosarcoma or cavernous hemangioma requires histopathological staining
- No standard of treatment for HEHE exists given its rarity
- As patients often present with multifocal disease, orthotopic liver transplantation is often performed
- Adjuvant chemotherapy and radiation are also a part of the treatment regimen
- The overall 5 year survival rate for patients treated with liver transplant is 83%.

References

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