MRI diagnosis of multifocal epithelioid hemangioendothelioma of the liver
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Case Type: Clinical Cases
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Patient: 37 years, female

Clinical History:
37-year-old woman with right upper quadrant abdominal pain, low grade fever and weight loss.

Imaging Findings:
37-year-old woman presented with a four-month history of right upper quadrant abdominal pain low grade fever and 5 kg weight loss. The physical examination revealed painless hepatomegaly. Laboratory values including aspartate aminotransferase, alanine aminotransferase, bilirubin and alpha-fetoprotein (AFP) levels were all within the normal ranges except for increased alkaline phosphates 620U/l, gamma-glutamyl-transpeptidase 300U/L and mild hypoalbuminaemia 3g/dl. She underwent abdominal CT outside which revealed conglomerate mass lesions suspicious for metastasis in the liver. Due to elevated choledastic liver enzymes and for further characterization of the lesions, MRI of the abdomen was ordered. Multiple planar MRI of the abdomen was performed on 1.5-T unit (GE Medical Systems, Milwaukee, Wis) using T1-weighted, T2-weighted and dynamic gadolinium-enhanced technique. The patient was subjected to dynamic Ultrasound US guided percutaneous liver biopsy

Discussion:
Epitheloid hemangioendothelioma EHE of the liver is a very rare clinical entity first described by Weiss and Enzinger in 1982. The tumor is often an incidental finding. It usually affects adult women (61%) and presents as multiple hepatic nodules. Its etiology is unknown. However association with oral contraceptives and a possible hormonal influence on the development of EHE have been suggested, as has exposure to vinyl chloride and a possible etiology from major trauma to the liver. These tumors demonstrate progressive sclerosis, hyalinization, and calcification in up to 50% of patients. With progression of the disease, the hepatic EHE nodules often coalesce, as they grow, usually in the periphery of the liver resulting in diffuse disease. Cases of EHE have been misdiagnosed as cholangiocarcinoma, fibrolamellar hepatocellular carcinoma, sarcoma and metastatic carcinoma. Radiological findings, especially MRI help in reaching the diagnosis of EHE. It readily depicts the internal architecture of EHE better than does CT. MR findings of coalescent peripheral hepatic masses with capsular retraction are highly suggestive of hepatic epithelioid hemangioendothelioma. The capsular retraction is mostly due to tumor fibrous reaction which indents the liver capsule. Normal tumor markers, capsular retraction of the liver adjacent to hepatic tumor is highly suggestive of epithelioid hemangioendothelioma, as other peripheral malignant tumors shows protrusion of the adjacent liver margin along with abnormal markers, biliary obstruction causes atrophy may may mimic retraction but biliary dilatation is more suggestive, cirrhosis with fibrosis may mimic but its morphological characteristic is explainable. Two histological types—nodular and diffuse have been described. Histologically, EHE
may mimic other vascular tumors and metastatic carcinomas. Demonstration of the vascular or endothelial origin of
the tumor is critical for diagnosis and requires immunostaining for endothelial markers, including factor VIII–related
antigen, CD31, and CD34. Although tumor growth may be progressive, lead to hepatic failure, extra hepatic
metastasis and death, the prognosis is considered more favorable than that of other hepatic malignancies. Thus
keeping an eye of suspicion and familiarity with the radiological features and histological characteristics of this
tumor, we could definitely facilitate the accurate diagnosis and thereby avoid unnecessary interventions.

**Differential Diagnosis List:** Multifocal Epithelioid Hemangioendothelioma of the liver

**Final Diagnosis:** Multifocal Epithelioid Hemangioendothelioma of the liver

**References:**


Figure 1

Description: Histopathology shows cords of spindle to epithelioid cells in a myxoid stroma and prominent vacuoles, some of them containing red blood cells. Hematoxylin and eosin stain; original magnification, 50×. Origin:
**Description:** Transverse in-phase spoiled gradient echo image with TR/TE = 120/4.2 shows the presence of multiple round lesions of moderately low signal intensity in the lateral and medial segment of the liver and one subcapsular lesion within the right hepatic lobe peripherally within the anterior segment abutting the liver capsule. **Origin:**
Description: Transverse post-gadolinium T1-weighted (120/2.3, 90° flip angle) during (c) equilibrium phase shows slightly more enhancement and peripheral hyperintensity compared with the (a) arterial and (b) portal venous phases. Origin:
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**Figure 4**

**a**

Description: MRI reveals capsular retraction of the liver capsule anterior to the nodules (arrows).

Origin:

**b**

Description: MRI reveals capsular retraction of the liver capsule anterior to the nodules (arrows).

Origin: