

# Hepatic Epithelioid Hemangioendothelioma – A Findings in Computed Tomography and Ultrasound

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## ABSTRACT

Hepatic Epithelioid Hemangioendothelioma (HEHE) is a very rare vascular tumor of epithelioid origin. This tumor is composed of epithelioid cells and dendritic. HEHE tumor growth leads to hepatic failure and later on death but this tumor is less aggressive as compared to the other tumors of the liver. We present a case on which a 32 years old female patient admitted with the complaints of left side abdominal pain with dyspepsia over 5-6 days. She was then referred to the medical Radio-diagnosis Department for Ultrasonography (USG) and Contrast-enhanced computed tomography (CECT) scan. The diagnosis of USG revealed that complex heterogeneous lesion with internal cystic component was seen in the left lobe of the liver and heterogeneously enhancing lesion with the non-enhancing necrotic area, non-enhancing cystic lesion and minimally enhancing lesion was seen in contrast-enhanced computed tomography (CECT). The clinical diagnosis was confirmed as HEHE and histopathologically correlation was done to confirm the diagnosis.

**Keywords:** Hepatic Epithelioid Hemangioendothelioma (HEHE), Ultrasonography, Computed Tomography.

## INTRODUCTION

Hepatic Epithelioid Hemangioendothelioma (HEHE) is one of the very uncommon malignant tumors of vascular origin with an epithelioid morphology which is composed of both dendritic and endothelial cells.<sup>[1]</sup> Dail and Liebow in 1975, first describe the epithelial lesion of the lung which they named an intravascular bronchioloalveolar tumor. After the details view of the electron microscope and immunohistochemical staining reveals that this is vascular endothelium origin. In 1982, Weiss and Enzinger termed as epithelioid hemangioendothelioma at Armed forces Institute of pathology.<sup>[2-4]</sup> Most of the studies reported that the frequency of occurring this disease is more among women compared to men.<sup>[5,6]</sup> The HEHE disease involved several symptoms which include weight loss, upper right quadrant pain, portal hypertension, few may be present with hepatic failure, and few are asymptomatic.<sup>[7]</sup> The tumor can be multiple appearances involved in both lobes of the liver.<sup>[2]</sup>

## CASE REPORT

A 32 years female patient reported to the General

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Medicine department with complaints of left side abdominal pain with dyspepsia over 5-6 days. She has no history of fever, vomiting, diarrhoea, constipation, urinary complaints, and also no menstrual irregularities. The patient is having a past history of hypothyroidism. Soon after the admission, all the tests were performed i.e. complete blood count (CBC), Urine analysis, Serum creatinine, CECT abdomen and Ultrasonography. The details of the report of CBC and other tests are disclosed as a table no 1.

CBC:

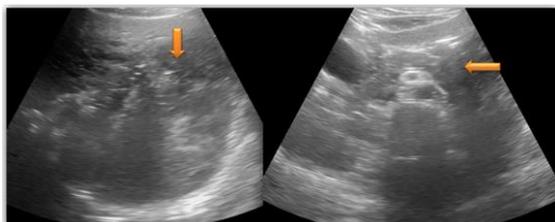
**Table 1: Initial Laboratory findings after a patient admitted into the hospital**

Investigations	Result	Normal Value
Serum Creatinine	0.72 mg/dl	0.8-1.2 mg/dl
Thyroid stimulating hormone (TSH)	42.66 uIU/ml	0.34-5.6 uIU/ml
Haemoglobin	8.84 gm/dl	12-16 gm/dl
Red blood corpuscles(R.B.C)	5.03mill./c.mm	3.8-5.2 mill./c.mm
White blood cells(W.B.C)	4710 cells/c.mm	4000-10000 cells/c.mm
Platelets	2.28 Lakh/c.mm	1.5-4.5 Lakh/c.mm
W.B.C count –		
Differential	77%	40-70 %
Polymorphs	11%	20-40%
Lymphocytes	04%	1-6%
Eosinophil's	08%	2-10%
Monocytes	00%	0-1%
Basophils		
Serum Sodium	134 mEq/L	133-146 mEq/L
Potassium	4.7 mEq/L	3.5-5.4 mEq/L
Chloride	98 mEq/L	97-107 mEq/L

Her vital signs were normal and Complete blood count revealed that the patient is having moderate microcytic hypochromic anemia. Urine Analysis was within normal limits.

#### **Ultrasonography (USG):**

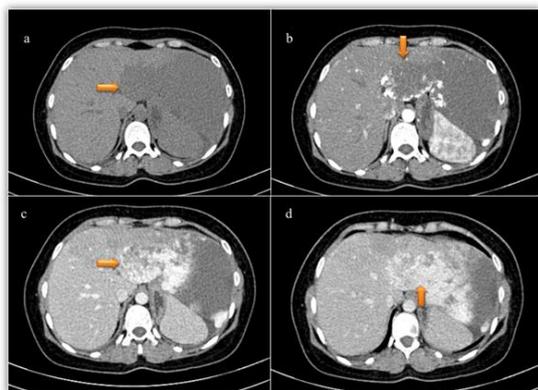
The patient underwent abdominal ultrasonography which demonstrated a large complex heterogeneous lesion of size 11.5 x 13.2 cm in perisplenic – left suprarenal region and also complex lesion with the internal cystic component in the left lobe of the liver [Figure 1].



**Figure 1:** Above ultrasonographic images shows heterogeneous hypoechoic lesion in perisplenic-left suprarenal region.

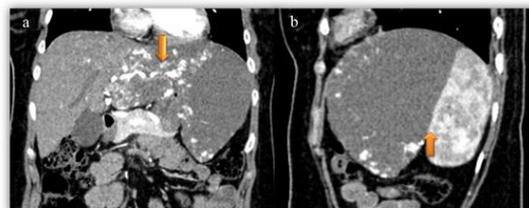
#### **Computed Tomography (CT):**

After ultrasound findings, she received a pre and post-contrast computed tomography scan of the whole abdomen with ultrafast 32 slices computed tomography scanner with 5 mm slice thickness and later on retrospective post-processing of multi planner imaging was done to get coronal and sagittal images. CECT abdomen revealed that approximately 10.8 cm (Anterior-posterior) x 13.6 cm (Transverse) x 12.3 cm (supero-inferior) sized fairly demarcated heterogeneously enhancing lesion with large ill-defined internal non enhancing necrotic areas arising from the left lobe of liver involving segment II and III of the liver. The lesion is reaching up to left lateral anterior abdominal muscles [Figure 2,3].



**Figure 2:** Above Axial sections of upper abdomen shows (a) Non-contrast images - large fairly demarcated lesion which on arterial (b) and venous (c) scan shows – internal ill-defined non enhancing necrotic areas within. On delayed post-contrast images (d) no complete post-contrast opacification of the lesion is appreciated.

Slight centripetal enhancement from the arterial to portal phases on contrast-enhanced CT imaging. Nodular enhancement along with the central vessels. Mass effect is visualized in the form of infero-medial displacement of spleen noted with passive compression of the cardia and body of stomach which resulting in extrinsic luminal narrowing of the stomach.



**Figure 3:** Above coronal (a) and sagittal (b) images shows the extent of the lesion. The lesion is abutting the left lateral anterior abdominal muscles with a mass effect in the form of infero-medial displacement of the spleen.



**Figure 4:** Portal venous phase shows a minimally enhanced lesion in the right lobes of the liver

On the other hand, approximately 2.2 x 2.3 cm sized hypodense, minimally enhancing lesion with the peripheral nodular central non-enhancing pattern of enhancement is visualized in the right lobe of the liver involving segments V and VI of the liver (Fig - 4) and Non enhancing cystic lesion of approximately 0.6 x 0.6 cm sized is seen in the right lobe of the liver (segment V).

The above imaging findings were consistent with the radiological features of hepatic epithelioid hemangi endothelioma.

Histopathological evaluation revealed dendritic and epithelioid cells with intracytoplasmic lumina of the neoplastic cells – confirming the diagnosis.

## **DISCUSSION**

HEHE is a rare malignant tumor with vascular origin arising from endothelial cells. Treska V et al, reported a study based on HEHE in which he reported that the incident rate of this disease is 1 per 1 million. The age range of 30 to 40 years or above is the most common age where this disease frequently affects patients.<sup>[1]</sup> This rare vascular

disease occurs more often in women than men and also present in other tissues like bone, peritoneum, and lung.<sup>[13]</sup> Considering the growth patterns of HEHE it could be classified into three types: nodular, coalescent, and mixed type.<sup>[14]</sup> The differential diagnosis of HEHE includes angiosarcoma, hemangioma, tumors with ‘signet rings’ and ‘lollipop sign’ and other neoplasms.<sup>[8,14]</sup> Mehrabi A et al, review a study on HEHE in which he reported that the most common symptoms of HEHE include hepatomegaly, right upper quadrant pain, weightlessness, weakness, jaundice, fever and fatigability and most of the patients were asymptomatic.<sup>[8,9,11]</sup> Another same type of study reviewed by Afrit M et al, also reported that the patient was suffering for abdominal distension for 2 months with mild dyspnea.<sup>[12]</sup> In our study, the patient was admitted with left side abdominal pain with dyspepsia for 5-6 days. Lyburn ID et al, performed a study based on Hepatic Epithelioid Hemangioendothelioma: MR imaging, Sonographic, and CT. Appearances on which they reported that on sonography most frequently lesion are hypoechoic but mass may be hyperechoic and isoechoic and internal structure of the nodule may be complex and heterogeneous. In CT they disclosed most of the lesion is peripheral, reaching out to the capsular edge. Hepatic parenchymal calcification might be seen. Tumor inclusion maybe with broad confluent masses and barely any detectable indications of the portal or hepatic veins and some tumor nodules show marginal enhancement during the arterial phase. After the contrast-enhanced scan, the tumor nodule may become isodense to the parenchyma of the liver.<sup>[15]</sup> In our study Sonography reveals that the patient is having large complex heterogeneous lesion involving perisplenic – left suprarenal region with the cystic component in the inner side of the left lobe of the liver. On the other hand, Contrast-enhanced Computed Tomography (CECT) shows demarcated heterogeneously enhancing lesion with internal necrosis, slight centripetal enhancement, and nodular enhancement within the central vessels in the left lobe of the liver. Minimally enhancing hypodense lesion with peripheral nodular central non-enhancing pattern and the cystic lesion is noted in the right lobe of the liver.

## CONCLUSION

HEHE is one of the rarest tumors which is very difficult to diagnose and its prognosis is often uncertain. The Ultrasound and CT diagnosis may permit the identification of this tumor at an early stage. In Ultrasonography the lesion appears hypoechoic and in CT the lesion appears hypodense.<sup>[15]</sup> HEHE is one of the less aggressive tumors compared to other tumors in the liver. The accurate diagnosis of HEHE is made by histopathologists by using a minimally invasive

biopsy sample but the diagnosis of HEHE can also be done by the help of imaging technology with good accuracy which is a completely non-invasive procedure.

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