Hepatic angiomyolipoma: A radiological dilemma

Abstract

We present a case of giant hepatic angiomyolipoma (HAML) in a 54-year-old female. Clinical manifestation of this entity is vague and imaging plays a major role in diagnosis. The presence of fat and efferent draining hepatic vein with early arterial enhancement persisting into the portal venous phase favors diagnosis of HAML. However, radiological findings can be confusing as in this case, and final diagnosis is made by histopathology and immunohistochemistry with positivity for human melanoma black-45.

Key words: Early efferent draining vein, Fat poor angiomyolipoma, Hepatic angiomyolipoma, Hepatic phleboliths, Human melanoma black-45.

INTRODUCTION

Angiomyolipoma (AML) is an uncommon benign hamartomatous hepatic mass lesion, containing blood vessel (angioid), smooth muscle (myoid), and mature fat (lipoid). Three hundred cases have been reported in literature till now. Liver is the second most common site of presentation after kidney. They have varied imaging findings and are differential for fat-containing lesions of the liver. They are positive for human melanoma black-45 (HMB-45) but negative for hepatocyte paraffin-1 (Hepar-1). We present a case of atypical giant AML which mimicked a hemangioma with the presence of an early efferent draining vein.

CASE REPORT

A 54-year-old female patient was referred to our institute with a history of pain abdomen on and off and weakness for 1 month. The patient underwent ultrasound and contrast-enhanced computed tomography (CECT) elsewhere with provisional diagnosis of hepatocellular carcinoma. On examination, the patient had mild epigastric tenderness and no hepatomegaly. There were no other positive examination findings. Routine blood investigations were unremarkable. Hepatitis B surface antigen and hepatitis C antibody were negative; alpha-fetoprotein level was normal.

On ultrasonography (USG), the patient had a well-defined hypoechoic lesion which was homogenous with multiple punctate foci of calcification and mild vascularity. The patient was subjected to CECT examination of the abdomen which showed a well-circumscribed lesion involving segment IV-A, II, and VIII of the liver abutting the segments IV-B and III measuring 10.3 cm × 7.5 cm × 5.7 cm. Few discrete foci of calcification were noted within the lesions [Figure 1]. The lesion derived arterial supply from the left gastric artery and partly from the common hepatic artery. The lesion showed moderate enhancement on arterial phase [Figure 2].

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The radiological features of HAML may vary according to its histological components. AML can be histologically classified on the basis of fat content into mixed, lipomatous (≥70% fat), myomatous (≤10% fat), and angiomatous types.\(^4\)

The typical findings in imaging of HAML are as follows:

On USG, tumor is either usually echogenic or heterogeneous due to lipomatous and myomatous tissue or less commonly hypoechoic due to angiomatous tissue. Hyperechoic lesion mimics hemangioma.\(^5\)

On CT, homogenous or heterogeneous lesion with low attenuation value (<-20HU) significantly enhances in the arterial phase and prolongs enhancement in the portal phase. The presence of an early efferent draining vein which drains into the hepatic vein eventually is likely to be associated with hemangiomyolipoma and was present in our case.\(^6\)

On magnetic resonance imaging (MRI), lesion is hyperintense on the T2W image and hyper- or hypo-intense on the T1W image depending on the fat component. In- and out-phase imagings are useful in detecting fat. The early enhancement in the arterial phase followed by the prolonged enhancement in the portal phase is observed on contrast-enhanced dynamic MRI.\(^7\)

Nevertheless, HAML is sometimes difficult to diagnose based on imaging studies unlike renal AML; 50% of HAMLs lack considerable fat content.\(^4\)

The diagnosis of HAML should be confirmed with FNAC or biopsy. Definite pathologic diagnosis of this tumor is usually made by identification of the three different components of smooth muscle cells, adipose tissue, and blood vessels.

HMB-45 positive staining of myoid cells is characteristic.\(^8\)

Differential diagnosis includes hemangioma, hepatocellular carcinoma, lipoma, and focal steatosis.

The effective therapy of HAML is surgical resection.\(^9\) Several authors advocated a conservative approach in the treatment of HAML. Ding et al. proposed the indication for resection as follows:
Kapali, et al.: Hepatic angiomyolipoma: A radiological dilemma

a. All symptomatic patients should receive surgical resection
b. Tumors >6 cm in size
c. Tumors show extrahepatic growth and risk of rupture
d. Tumors show a tendency to grow
e. Findings of diagnostic imaging and/or biopsy cannot make a definitive diagnosis.

Complications of AML include post op recurrence, spontaneous rupture, malignant transformation, disseminated intravascular coagulation, and Budd-Chiari syndrome.

CONCLUSION

Hepatic angiomyolipoma is an uncommon hepatic lesion. Presence of fat is an important finding to diagnose HAML. However, 50% of patients have fat poor HAML, in them the identification of venous drainage of lesion into hepatic veins instead of portal vein is a pointer towards the diagnosis.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES


