pISSN 2384-1095
eISSN 2384-1109

iMRI 2022;26:60-65 https://doi.org/10.13104/imri.2022.26.1.60

iMRI
Investigative Magnetic Resonance Imaging

Hepatic Angiomyolipoma Presenting as a Hyperintense Lesion During the Hepatobiliary Phase of Gadoxetic Acid Enhanced-MRI: a Case Report

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Case Report

Received: August 26, 2021
Revised: September 14, 2021
Accepted: November 11, 2021

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Gadoxetic acid-enhanced magnetic resonance imaging (MRI) has been widely used to detect and characterize focal hepatic lesions. Because gadoxetic acid is a hepatocyte-specific contrast agent, its patterns during hepatobiliary phase enhancement provide useful information for differential diagnoses of focal hepatic lesions.
Hepatic angiomyolipoma (AML) is a rare mesenchymal hepatic neoplasm composed of blood vessels, epithelioid cells, and varying amounts of adipose tissue components. Hepatic AMLs usually show marked hypointensity during the hepatobiliary phase of gadoxetic acid-enhanced MRI as hepatic AMLs are devoid of hepatocytes and fibrotic components.
The present study describes a patient with hepatic AML and an atypical imaging feature. This tumor showed hyperintensity during the hepatobiliary phase of gadoxetic acid-enhanced MRI, mimicking hepatocellular tumors such as hepatocellular adenoma. The hepatobiliary hyperintensity of this lesion was likely due to multifocal entrapped hepatocytes resulting from an intrasinusoidal growth pattern of tumor cells and insufficient hepatic parenchymal enhancement during the hepatobiliary phase of gadoxetic acid-enhanced MRI.

Keywords: Hepatic angiomyolipoma; Gadoxetic acid; Hepatobiliary phase; Magnetic resonance imaging

INTRODUCTION

Gadoxetic acid (Primovist or Eovist; Bayer Healthcare, Berlin, Germany) is frequently used as a contrast agent in magnetic resonance imaging (MRI) of the liver as it provides both hemodynamic and cellular information (1, 2). Since gadoxetic acid is taken up by functioning hepatocytes via organic anion-transporting polypeptide 1B3 (OATP1B3), its signal intensity characteristics during the hepatobiliary phase provide useful information for differential diagnoses of hepatic lesions. Typical focal hepatic lesions containing hepatocytes, including focal nodular hyperplasia, hepatocellular adenoma, and hepatocellular carcinoma, can manifest as hyperintense lesions during the hepatobiliary phase of gadoxetic acid-enhanced MRI (2, 3).

Hepatic angiomyolipoma (AML), a rare benign hepatic neoplasm, is a subtype of PEComa, a mesenchymal neoplasm composed of predominantly epithelioid cells showing variable expression of smooth muscle and melanocytic markers (4). Pathologically, hepatic AMLs contain blood vessels, spindle cells, and varying amounts of adipose tissue components. On imaging, AMLs usually appear as arterial hyperenhancing lesions with or without a detectable fat component in non-cirrhotic liver, mimicking more common hepatic lesions such as focal nodular hyperplasia, hepatocellular adenoma, and even hepatocellular carcinoma (5, 6). Signal intensity characteristics during the hepatobiliary phase of gadoxetic acid-enhanced MRI can provide important clues for differential diagnosis of these arterial hyperenhancing lesions. Hyperintensity during the hepatobiliary phase imaging is indicative of hepatocellular tumors. It excludes the possibility of hepatic AML (2, 3, 5, 6). To the best of our knowledge, all hepatic AMLs described to date have shown low signal intensity during the hepatobiliary phase of gadoxetic acid-enhanced MRI (5, 6). This report describes a 40-year-old woman presenting with a hepatic AML showing hyperintensity during the hepatobiliary phase of gadoxetic acid-enhanced MRI.

CASE REPORT

A 40-year-old woman was referred to our hospital for the evaluation of a hepatic mass incidentally detected on computed tomography (CT), which was performed to determine the cause of recently aggravated fatigue. The patient had no history of viral hepatitis, alcohol consumption, or other liver diseases. Her physical examination was unremarkable. Laboratory tests were also normal, including liver function tests, viral markers, and tumor markers such as alpha-fetoprotein (AFP), prothrombin-induced by vitamin K absence or antagonist-II (PIVKA), and carbohydrate antigen 19-9 (CA 19-9). Drug and family histories were also unremarkable.

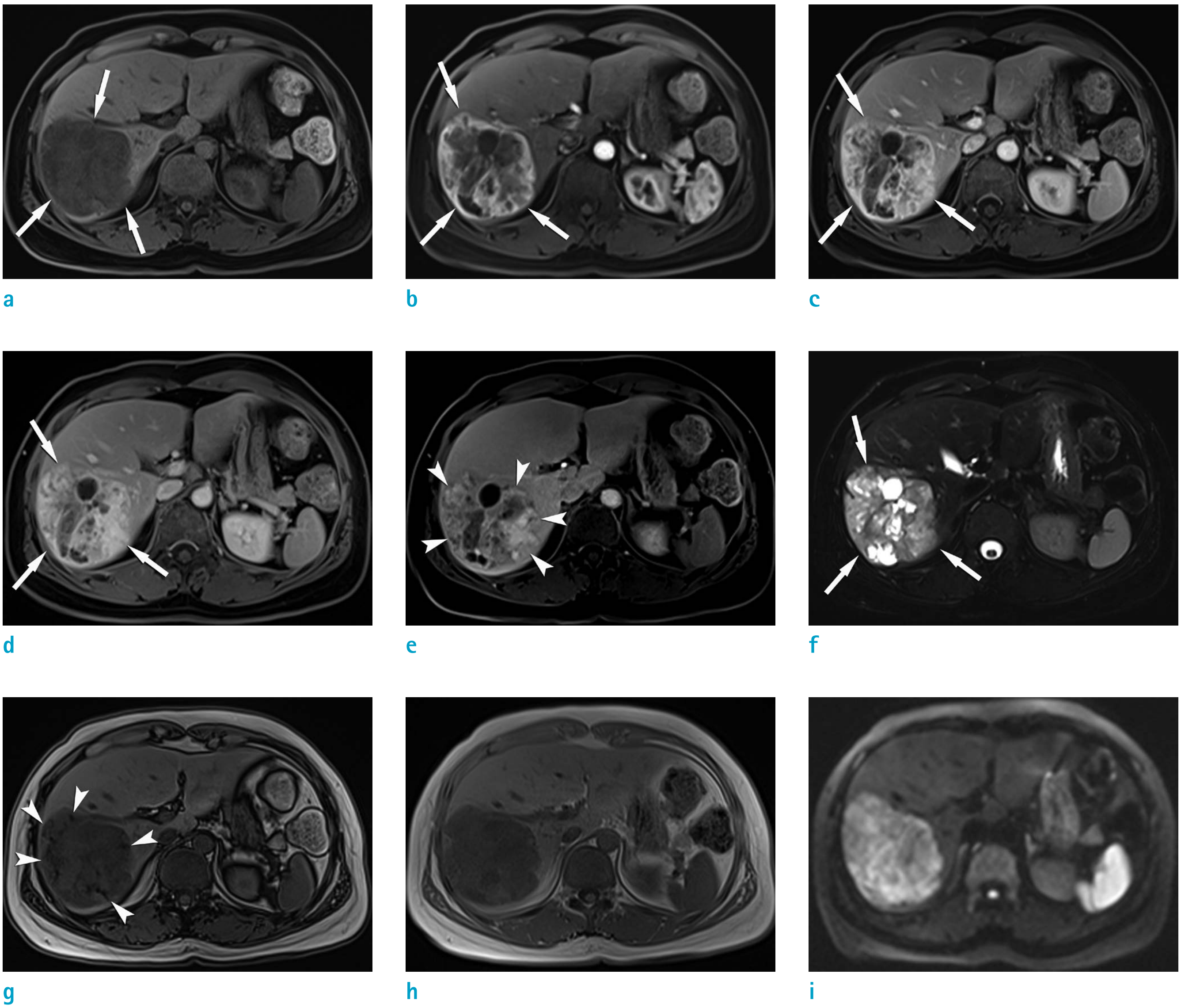
A CT scan showed an 11-cm sized heterogeneously enhancing mass with well-defined borders in the right liver. A subsequent MRI found an arterial phase hyperenhancing mass with a progressive enhancement pattern on portal and transitional phases (Fig. 1a-d). A substantial portion of the enhancing component of the mass showed hyperintensity during the hepatobiliary phase obtained at 20 minutes after contrast injection. This hyperintensity was attributed to the uptake of gadoxetic acid (Fig. 1e). Hepatic parenchymal enhancement during the hepatobiliary phase was insufficient as blood vessels appeared to have the same intensity as the hepatic parenchyma. The mass appeared heterogeneous with some cystic components (Fig. 1f). Small foci of intratumoral fat (Fig. 1g, h) and diffusion restricted areas (Fig. 1i, j) were also observed.

This fat-containing hypervascular mass in a middle-aged woman with a non-cirrhotic liver was originally diagnosed as a hepatocellular adenoma. Although this patient did not have any risk factors for hepatocellular carcinoma, the possibility could not be excluded. Hepatic AML was not included in the differential diagnosis. Because of the size of the lesion and the possibility of malignancy, a right hemihepatectomy was performed without a preoperative biopsy.

Gross examination of the resected specimen showed a well-demarcated, 11-cm sized ovoid mass with extensive hemorrhage. The cut surface of the mass had a soft brownish red color with some multifocal yellowish adipose tissue-like areas and extensive hemorrhage. Microscopically, the mass consisted of characteristic components of hepatic AML, including thick-walled blood vessels, epithelioid cells, and adipocytes without atypical nuclei or mitotic figures (Fig. 2a, b). The surrounding hepatic parenchyma showed mild fatty infiltration. Interestingly, epithelioid tumor cells were identified in sinusoidal spaces with diffusely entrapped hepatocytes, suggestive of an intrasinusoidal growth pattern (Fig. 2b). Immunohistochemical staining for human melanoma black 45 (HMB45) showed that epithelioid tumor cells were diffusely positive (Fig. 2c). Tumor cells also showed patch positivity for smooth muscle actin (SMA), supporting a diagnosis of hepatic AML. The final diagnosis was hepatic AML. Areas of entrapped hepatocytes showed diffusely positive for hepatocyte paraffin 1 (Hep Par 1) (Fig. 2d) and matched with non-HMB45 stained areas on dual staining (HMB 45 and Hep Par 1) (Fig. 2e). The patient's postoperative course was uneventful. She was discharged home at 7 days after the surgery.

DISCUSSION

Signal characteristics of focal hepatic lesions during the hepatobiliary phase of gadoxetic acid-enhanced MRI provide important clues for the identification of lesion components (2, 3). For example, hyperintense lesions are generally considered hepatocellular tumors, although some tumors with abundant fibrotic component have shown mild hyperintensity due to extracellular retention of gadoxetic acid (2). In contrast, hypointense lesions are indicative of non-hepatocellular tumors. Because hepatic AMLs do not contain hepatocytes or dense fibrotic



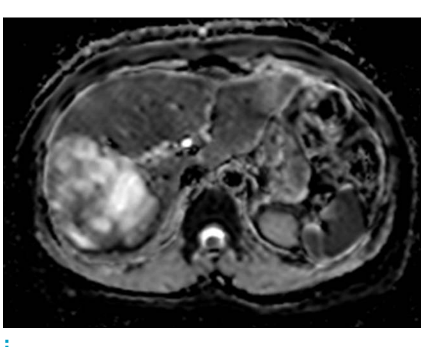


Fig. 1. Gadoxetic acid-enhanced MRI features of the tumor, showing a heterogeneous hypointense mass (arrows) on the noncontrast image (a) in the right lobe of the liver, hyperenhancement on the arterial phase (b) with progressive enhancement patterns on portal phase (c) and transitional phase (d). A substantial portion (arrowheads) of the enhancing component of the mass shows uptake during the hepatobiliary phase (e). On T2WI, the mass (arrows) appears heterogeneous with some cystic components (f). Fat components (arrowheads) showing signal drop on T1-weighted opposed-phase image (g) compared with in-phase image (h) are noted. A part of the lesion shows diffusion restriction on the diffusion-weighted image (b = 800 s/mm^2) (i) and ADC map (j).

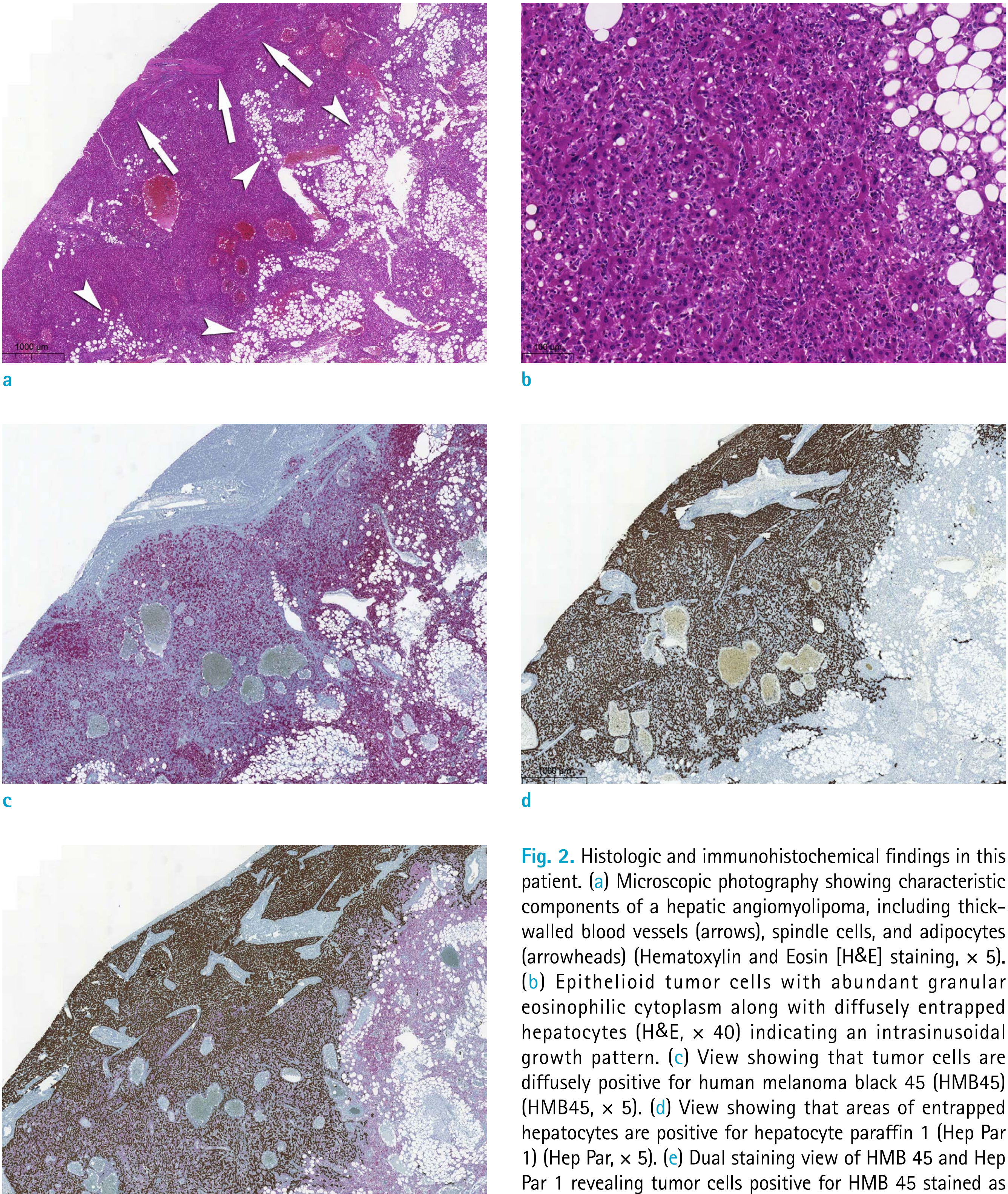


Fig. 2. Histologic and immunohistochemical findings in this patient. (a) Microscopic photography showing characteristic components of a hepatic angiomyolipoma, including thick-walled blood vessels (arrows), spindle cells, and adipocytes (arrowheads) (Hematoxylin and Eosin [H&E] staining, × 5). (b) Epithelioid tumor cells with abundant granular eosinophilic cytoplasm along with diffusely entrapped hepatocytes (H&E, × 40) indicating an intrasinusoidal growth pattern. (c) View showing that tumor cells are diffusely positive for human melanoma black 45 (HMB45) (HMB45, × 5). (d) View showing that areas of entrapped hepatocytes are positive for hepatocyte paraffin 1 (Hep Par 1) (Hep Par, × 5). (e) Dual staining view of HMB 45 and Hep Par 1 revealing tumor cells positive for HMB 45 stained as pale red color are mixed with hepatocytes positive for Hep Par showing brown color (dual staining of HMB 45 and Hep Par 1, × 5).

components, these tumors usually appear hypointense during hepatobiliary phase gadoxetic acid-enhanced MRI. Of 30 hepatic AMLs evaluated to date by gadoxetic acid-enhanced MRI, none has appeared hyperintense during the hepatobiliary phase (5, 6). Contrary to previous belief, the hepatic AML evaluated in this patient presented as a hyperintense mass during the hepatobiliary phase of gadoxetic acid-enhanced MRI. This unique imaging feature can be attributed to an unusual growth pattern, in which tumor cells were present in the hepatic sinusoid. Thus, the tumor contained a substantial number of hepatocytes, which took up gadoxetic acid. This unusual growth pattern, in which tumor cells and non-tumorous hepatocytes are mixed together, has been reported in pathology literature (7, 8). Despite this uncommon growth pattern, the typical pathologic component of hepatic AML with tumor positivity for melanocytic (HMB45) and SMA markers supported the diagnosis of hepatic AML.

In our case, considering imaging features such as arterial phase hyperenhancement and hyperintensity during the hepatobiliary phase, possible differential diagnosis would include hepatocellular adenoma and hepatocellular carcinoma. As the patient did not have known risk factors for hepatocellular carcinoma, hepatocellular carcinoma was not likely. Hepatocellular adenoma was suggested as the primary diagnosis in this case. Differentiation from hepatocellular adenoma did not seem to be possible if only imaging features were considered.

In addition, it should be noted that the enhancement of the non-tumorous hepatic parenchyma during the hepatobiliary phase was insufficient in this patient as hepatic parenchymal enhancement was similar to portal vein enhancement. Although insufficient enhancement of the hepatic parenchyma is usually associated with poor liver function (9), hepatic function was normal in this patient without abnormal pathologic findings in the hepatic parenchyma except for minimal fatty liver. Biliary excretion of gadoxetic acid was normally seen. Mild fatty infiltration in the nontumorous hepatic parenchyma was noted on pathology, which might deteriorate hepatic parenchymal enhancement during the hepatobiliary phase of gadoxetic acid-enhanced MRI (10). However, the degree of fatty liver in our case could not fully explain insufficient enhancement of the liver on gadoxetic acid-enhanced MRI. The hepatobiliary phase hyperintensity of this tumor was likely due to combined effects of insufficient enhancement of non-tumorous hepatic parenchyma and the presence of multifocal entrapped hepatocytes within the tumor. If hepatic parenchymal enhancement was normal, the mass would have looked iso- or hypo-intense during the hepatobiliary phase as the number of hepatocytes within the tumor was smaller than that of normal hepatocytes. When signal characteristics of focal hepatic lesions are considered, we need to determine whether the enhancement of nontumorous hepatic parenchyma is sufficient or not as signal intensity of focal hepatic lesions is a relative feature compared to the adjacent hepatic parenchyma.

In conclusion, this report describes a hepatic AML with an atypical imaging feature. This tumor appeared hyperintense during the hepatobiliary phase on gadoxetic acid-enhanced MRI, mimicking hepatocellular neoplasms. The hyperintensity of this tumor might be due to multifocal entrapped hepatocytes resulting from an intrasinusoidal growth pattern of tumor cells with insufficient hepatic parenchymal enhancement.

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