

## CASE REPORT

# Hepatic epithelioid angiomyolipoma with arterioportal venous shunting mimicking hepatocellular carcinoma : report of a case

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**Abstract :** A patient with hepatic epithelioid angiomyolipoma (Epi-AML) with arterioportal venous shunting, who was successfully treated by a laparoscopic left lateral sectionectomy, is presented herein. AML is an uncommon benign neoplasm of the liver. Tumors composed predominantly of epithelioid cells have been subcategorized into Epi-AML, and the treatment strategy for Epi-AML is currently undetermined. There are no reports describing Epi-AML with arterioportal venous shunting to date. An arterioportal venous shunting of the liver tumor was suggested to be one of the malignant signs of the liver tumor. It would be important to differentiate Epi-AML with arterioportal venous shunting from hepatocellular carcinoma and hypervascular metastatic tumors. Minimally invasive resection, such as laparoscopic hepatectomy, for patients having Epi-AML with arterioportal venous shunting may be recommended. *J. Med. Invest.* 60 : 262-266, August, 2013

**Keywords :** *hepatic epithelioid angiomyolipoma, arterioportal venous shunting, differential diagnosis from hepatocellular carcinoma, laparoscopic hepatectomy*

## INTRODUCTION

Angiomyolipoma (AML) is a rare tumor in the liver composed of varying mixtures of smooth muscle, fat cells, and abnormal, thick-walled blood vessels. Depending upon the dominant cell type, AML can be subcategorized into epithelioid, spindle, and intermediate forms (1). In 2000, epithelioid AML (Epi-AML) was first described in the liver<sup>2</sup>. Recently, several case reports of hepatic Epi-AML have

been described or reviewed in the literature (2-5). Differential diagnosis of Epi-AML from hepatocellular carcinoma (HCC) and the metastatic sarcomatoid variant of renal cell carcinoma are considered to be difficult.

Liver tumors with arterioportal venous shunting have generally been recognized to be malignant. However, several authors have argued that arterioportal venous shunting is not always a sign of malignancy (6-9).

We herein present a case of hepatic Epi-AML with significant arterioportal venous shunting that could barely be differentiated from HCC and successfully treated with minimally invasive laparoscopic hepatectomy.

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## CASE REPORT

A 46 year-old man without symptoms had a hypoechoic mass in the left lateral section. Under the diagnosis of HCC, based on the findings of an enhanced computed tomography (CT) scan, he was referred to our department for surgical treatment.

Laboratory tests revealed a leukocyte count of  $5800 / \text{mm}^3$ , 16.4 g/dL hemoglobin, a platelet count of  $26.6 \times 10^4 / \text{mm}^3$ , 4.5 g/dL albumin, 0.9 mg/dL total bilirubin, 0.2 mg/dL direct bilirubin, 38 IU/L serum alanine transaminase, and a prothrombin time of 10.4 s. The patient was negative for hepatitis B surface antigen, hepatitis B surface antibody, hepatitis B e antigen, hepatitis B e antibody, and hepatitis B core antibody, hepatitis B virus DNA, hepatitis C antibody and hepatitis C virus RNA. The indocyanine green dye retention rate after 15 minutes was

10.9 % (normal range < 10 %). All serum tumor markers of  $\alpha$ -fetoprotein (AFP), AFP-L3 and des- $\gamma$ -carboxy prothrombin (DCP) were within normal limits.

Dynamic multi-detector computed tomography (MDCT) showed a hypervascular mass, measuring 27 mm in diameter, in the arterial phase (Fig. 1A). In the portal phase, the tumor had almost washed out the contrast agent, but a weak contrast-enhancing effect had been sustained due to arteriportal venous shunting (Fig. 1B), and almost showed isodensity with the liver tissue in the delayed phase (Fig. 1C). T1-weighted magnetic resonance imaging (MRI) revealed low intensity (Fig. 2A) and T2-weighted MRI showed high intensity (Fig. 2B). Gd ethoxybenzyl diethylenetriamine pentaacetic acid (Gd-EOB-DTPA)-enhanced MRI in the hepatocyte phase revealed low-intensity areas in the tumor 20 min after Gd-EOB-DTPA administration (Fig. 2C).

1-A

1-B

1-C

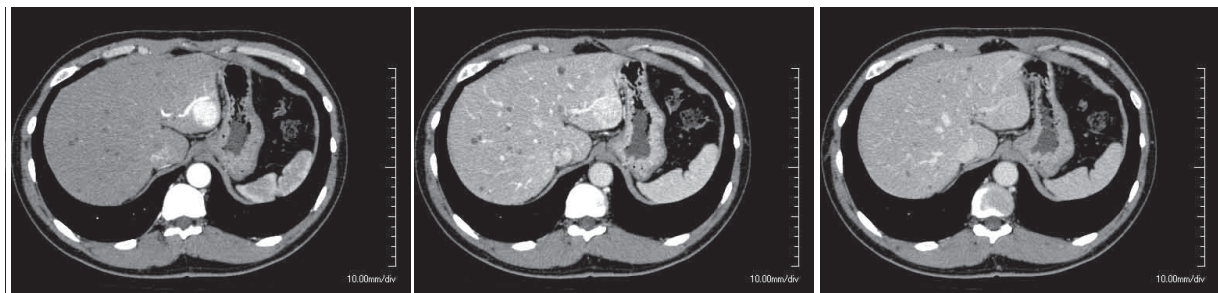


Figure 1 : Dynamic multi-detector computed tomography (MDCT).

1-A : Arterial phase showed a hypervascular mass in segment 2.

1-B : Portal phase showed it had almost washed out the contrast agent but a small contrast-enhancing effect had been sustained due to arteriportal venous shunting.

1-C : Delayed phase almost showed isodensity with the liver tissue.

2-A

2-B

2-C

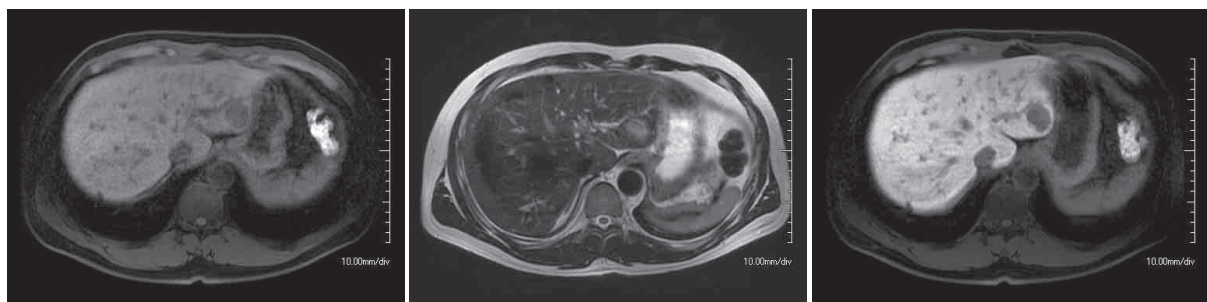


Figure 2 : Magnetic resonance imaging (MRI).

2-A : T1-weighted MRI revealed the tumor was detected with low intensity area.

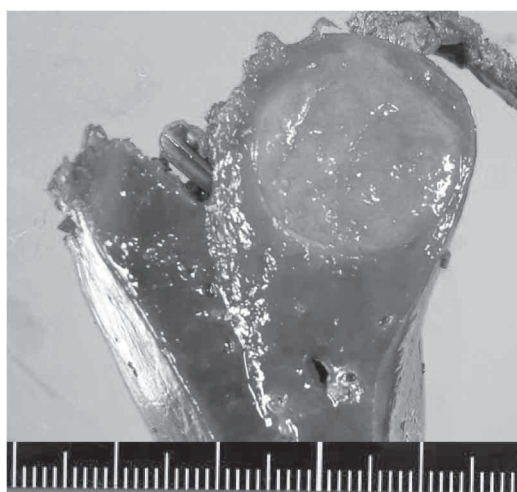
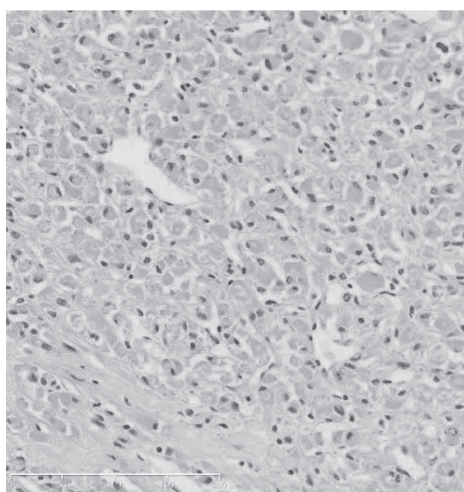
2-B : T2-weighted MRI with high intensity area.

3-C : Gd ethoxybenzyl diethylenetriamine pentaacetic acid (Gd-EOB-DTPA)-enhanced MRI in the hepatocyte phase, 20 min after Gd-EOB-DTPA administration, revealed low-intensity areas in the tumor.

We could not exclude the possibility of HCC with arterioportal venous shunting, and performed laparoscopic left lateral sectionectomy.

The resected specimen appeared to be a tuberous tumor, measuring some  $25 \times 22$  mm in diameter, on the cutting surface, and there were no macroscopic evidence to suggest the presence of arterioportal venous shunting (Fig. 3A). In our histopathological findings, the hematoxylin-eosin (HE) stain revealed

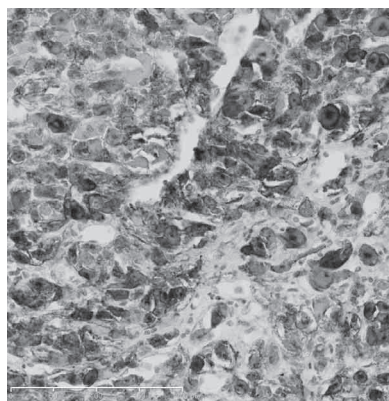
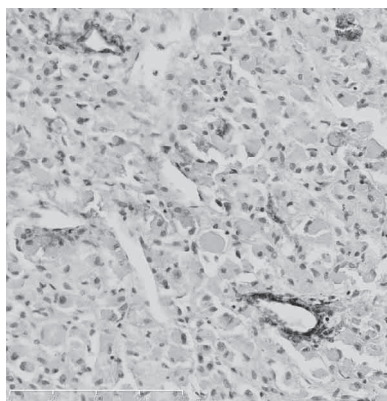
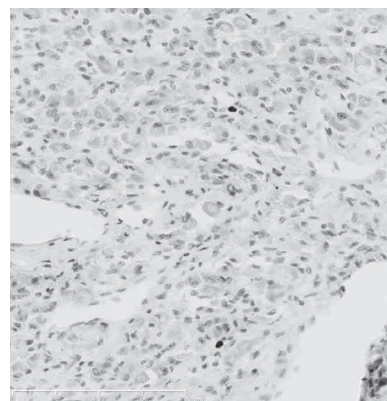
many cells with acidophilic cytoplasm and a lower nuclear-cytoplasmic ratio in the tumor tissues (Fig. 3B). Furthermore, immunohistochemical analysis revealed a high expression of melanoma-associated antigen (HMB45) (Fig. 4A), partial expression of  $\alpha$  smooth muscle actin ( $\alpha$ -SMA) (Fig. 4B) and almost no expression of MIB-1 (Fig. 4C). Following these results, the final pathological diagnosis was determined to be Epi-AML.

**3-A****3-B**

**Figure 3 :** Macroscopic and histopathological findings.

3-A : Resected specimen appeared to be a tuberous tumor, measuring some  $25 \times 22$  mm in diameter, on the cutting surface.

3-B : Hematoxylin-eosin (HE) staining revealed there were many cells with acidophilic cytoplasm and a lower nuclear-cytoplasmic ratio in the tumor area.

**4-A****4-B****4-C**

**Figure 4 :** Immunohistochemical analysis.

4-A : High expression of melanoma-associated antigen (HMB45).

4-B : Partial expression of  $\alpha$  smooth muscle actin ( $\alpha$ -SMA).

4-C : Almost no expression of Mindbomb E3 ubiquitin protein ligase 1 (MIB-1).

## DISCUSSION

AML is a rare tumor most commonly found in the kidneys, followed by the liver. Besides these organs, it has also been identified in the uterus, retroperitoneum, mediastinum, renal capsule, nasopharynx, penis, vagina, colon, skin, parotid gland, lung, and spleen (3, 10). A case of hepatic AML was first reported in 1976 (11). By 1999, 110 cases of hepatic AML had been published in the literature (1), while a rare variant, Epi-AML, was first described in 2000 (2). Considering from previous reports, hyper vascularity with central punctiform or filiform vessels is suggested to be a characteristic radiographic feature for hepatic Epi-AML (12, 13), however, there were no such findings in the present case. Histologically, Epi-AML is reported to be strongly positive for HMB45, which is the most sensitive immunohistochemical marker for AML, and weakly to moderately positive for  $\alpha$ -SMA or desmin (2, 4, 13).

Arterioportal venous shunting is considered to be one of the signs of a malignant liver tumor, and the incidence of arterioportal venous shunting was reported to be 20-30% in HCC (14). We have previously reported that hepatic resection for hemangiomas with arterioportal venous shunting might be recommended due to the high risk of rupture and the possible progression of portal hypertension (15). The route of arterioportal shunting is considered either transsinusoidal or transvasal (7). Transsinusoidal route occurs in cases of cirrhosis or the Budd-Chiari syndrome and results in retrograde hepatofugal flow in portal branches. On the other hand, in transvasal route, portal flow often remains hepatopetal. The transvasal route occurs in cases of HCC, metastases, shock, hepatic arterial obstruction, and many other conditions. To the best of our knowledge, there have been no reports of Epi-AML with arterioportal venous shunting so far and it is difficult to assume the relation between its tumor growth and arterioportal venous shunting.

Typical AMLs are regarded as universally benign tumors and often grow slowly; however, Epi-AMLs are regarded as tumors of uncertain malignant potential. Indeed, rare cases of Epi-AML with tumor recurrence, and vascular invasion have recently been reported (16, 17). Surgical resection should thus be considered for all symptomatic patients. Conservative management with close follow-up is suggested in asymptomatic patients with tumors with a small mass (< 5 cm) that are diagnosed through fine-needle aspiration biopsy (18).

Since hepatic Epi-AML with arterioportal venous shunting is barely differentiated from HCC and potentially malignant, minimally invasive hepatectomy may be recommended.

## CONFLICT OF INTEREST

The author and all co-authors declare no conflict of interest related to this case report.

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