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CASE REPORT

Hepatic angiomyolipoma with early drainage veins into the hepatic and portal vein

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Abstract
Hepatic angiomyolipoma (AML) is a rare stromal tumor composed of variable admixtures of thick-walled vessels, smooth muscles and adipose tissue. One of the specific radiological findings of hepatic AML is an early drainage vein noted via enhanced computed tomography (CT). We report a case of hepatic AML showing early drainage veins into both the hepatic and portal vein. The case involved a 46-year-old woman who was referred to our hospital because of a giant hepatic tumor. CT revealed well-enhanced 14 cm and 1 cm tumors in the left and right lobes, respectively. Magnetic resonance imaging demonstrated the existence of adipose tissues in the larger tumor. Hepatic arteriography revealed early drainage veins draining into both the hepatic and portal vein. Based on a diagnosis of hepatic AML, left hepatectomy and partial hepatectomy were performed. Pathology revealed both tumors as hepatic AML based on human melanoma black-45 immuno-positivity. Hepatic AML with early drainage veins into both the hepatic and portal vein is rare. The dilated and retrogressive vein drains the abundant arterial blood flow of the tumor. The finding of early drainage veins into not only the hepatic vein but also the portal vein should be helpful for diagnosing hepatic AMLs.

Keywords Hepatic angiomyolipoma · Early drainage vein · Hepatic vein · Portal vein

Introduction

Angiomyolipoma (AML) is a rare stromal tumor composed of variable admixtures of thick-walled blood vessels, smooth muscles and adipose tissue [1]. AML commonly arises in the kidney, while the liver is a less common site of origin [2]. Because of the advance in imaging modalities, hepatic AML has become well recognized in recent years. One of the characteristic findings of hepatic AML is an early drainage vein into the hepatic vein on contrast-enhanced computed tomography (CT) and magnetic resonance imaging (MRI) [3–5]. Hepatic AML rarely shows early drainage veins into the portal vein [4, 6]. To our knowledge, there is no report of hepatic AMLs with early drainage veins into both the hepatic and portal vein. We, herein, present a case of hepatic AML showing early drainage veins into both the hepatic and portal vein.

Case report

A 46-year-old woman was referred to our hospital with a suspicion of giant hepatic tumor based on a finding of upper stomach deformation via barium contrast gastrography. On physical examinations and symptoms, there was no typical symptom related to tuberous sclerosis complex. Contrast-enhanced CT showed two well-enhanced tumors of 14 cm and 1 cm in the left and right hepatic lobe, respectively. An early drainage vein into the hepatic vein was detected around the tumor in the left hepatic lobe. Further, a dilated and tortuous vessel was found inside the left hepatic lobe tumor (Fig. 1).

Angiography of the left hepatic artery revealed dilated and tortuous vessels inside the tumor in the early phase.

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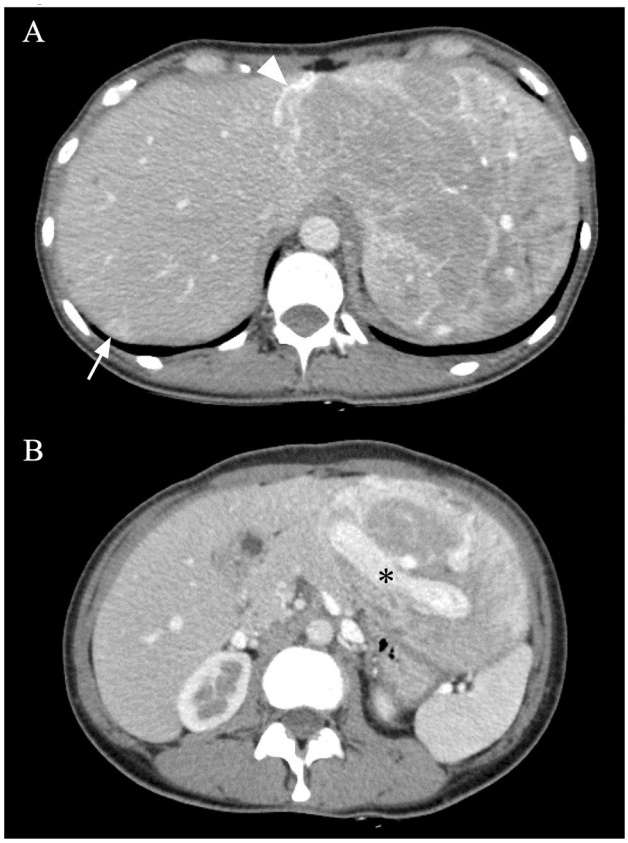

Fig. 1 Contrast-enhanced computed tomography (CT) findings. **a** Abdominal CT showed a large hepatic tumor in the left lobe and another small hepatic tumor in the right lobe (arrow). An early drainage vein into the hepatic vein from the tumor may be seen (arrowhead). **b** A dilated and tortuous vessel (asterisk) can be seen inside the tumor in the left hepatic lobe

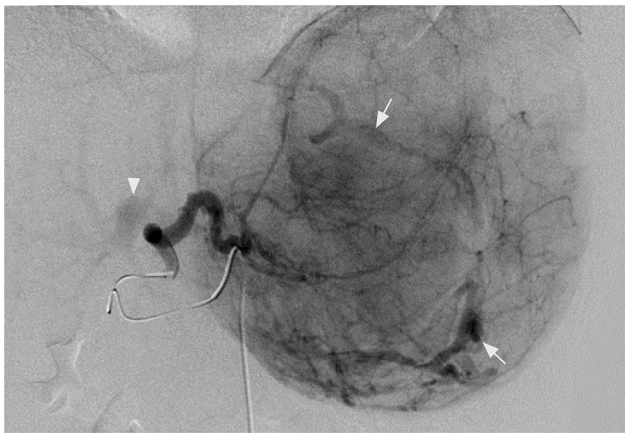

Fig. 2 Findings of left hepatic arteriography. In the early phase, dilated and tortuous vessels (arrows) were noted inside the tumor. The portal umbilical portion (arrowhead) may also be seen

The portal umbilical portion was noted in the early phase, suggesting that the above-mentioned dilated vessel was connected to the portal vein (Fig. 2). In the delayed phase of CT during proper hepatic angiography, the left lobe tumor was heterogeneously enhanced. Further, dilated and tortuous vessels as well as the portal umbilical portion were noted (Fig. 3).

MRI revealed a hypointense tumor in the left lobe containing high- and low-intensity areas in T1-weighted imaging in-phase and out-phase, respectively (Fig. 4), suggesting that the tumor had an adipose tissue component.

Positron emission tomography imaging using an ^{18}F-fluorodeoxyglucose (FDG) tracer revealed that both hepatic tumors exhibited a low FDG uptake.

Blood examination showed slight anemia (red blood cell 3.92×10^4/μL, hemoglobin 9.8 g/dL, hematocrit 32.5%). Tests for all evaluated markers of hepatic viruses were negative, and tumor markers such as carcinoembryonic antigen, cancer antigen 19-9, alpha-fetoprotein, and protein induced by vitamin K absence-2, were within normal ranges.

Based on a diagnosis of multiple hepatic AML, left hepatectomy and partial hepatectomy of the right hepatic lobe were performed. The operation time was 268 min and blood loss was 420 g. The patient was discharged on the 16th day postoperatively without any complication.

Gross examination of the resected specimen revealed a soft, well-circumscribed large tumor, measuring $15 \times 14 \times 8$ cm, bulging outward from the liver left lobe. The cut surface of the tumor consisted mainly of whitish nodules with a band-shaped blackish area containing a dilated and tortuous blood vessel with thrombus (Fig. 5a). Microscopic examination of the tumor revealed a predominant dense proliferation of epithelial-like to spindle-shaped cells with clear to eosinophilic cytoplasm (Fig. 5b). Swollen and eosinophilic large myoid cells were also observed on some occasion (Fig. 5c). In blackish area, many cystic spaces filled with blood, i.e., the "peliosis hepatis" -like structures were conspicuous, and foci of extramedullary hematopoiesis were often seen in this area (Fig. 5D). Thick wall blood vessel and adipocytes were mixed in some places (Fig. 5E). Immunohistochemical examination revealed that tumor cells were positive for human melanoma black-45 antigen (HMB-45) (Fig. 5F), melan-A/MART1 and alpha smooth muscle actin (Fig. 5g), while negative for Hep Par-1. The mitotic figures were very rare [approximately 1–2/50 high power fields (HPFs)]. There was no venous and serosal invasion of tumor cells. The pathological findings of AML from right lobe partially resected were the same to those of main AML from left lobe. Based on the above findings, these tumors were diagnosed as multiple hepatic AMLs.

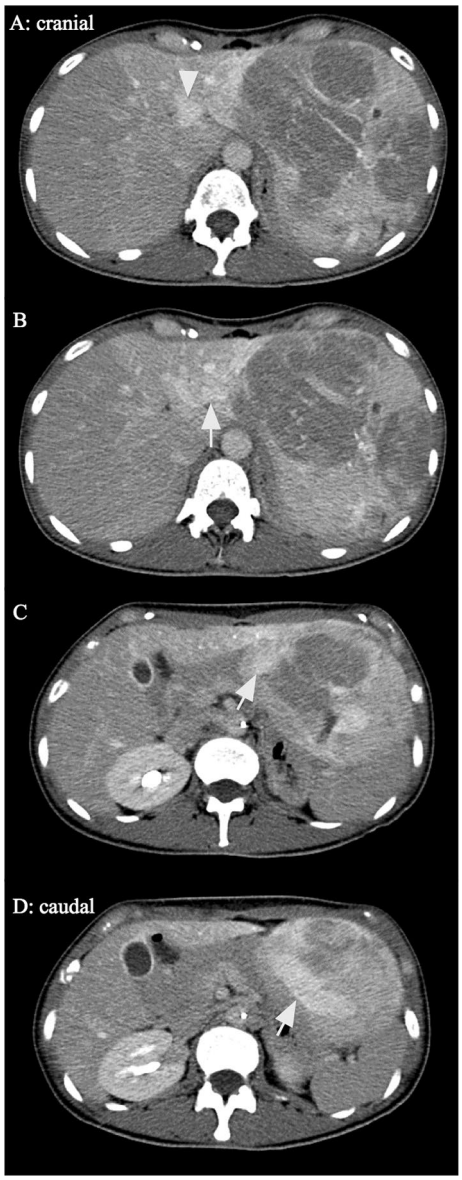

Fig. 3 Computed tomography (CT) findings during the proper hepatic arteriography. The tumor was heterogeneously enhanced. Dilated tortuous vessels (arrows), as well as the portal umbilical portion (arrowhead) are shown. **a–d** CT sections cranial–caudal sequence

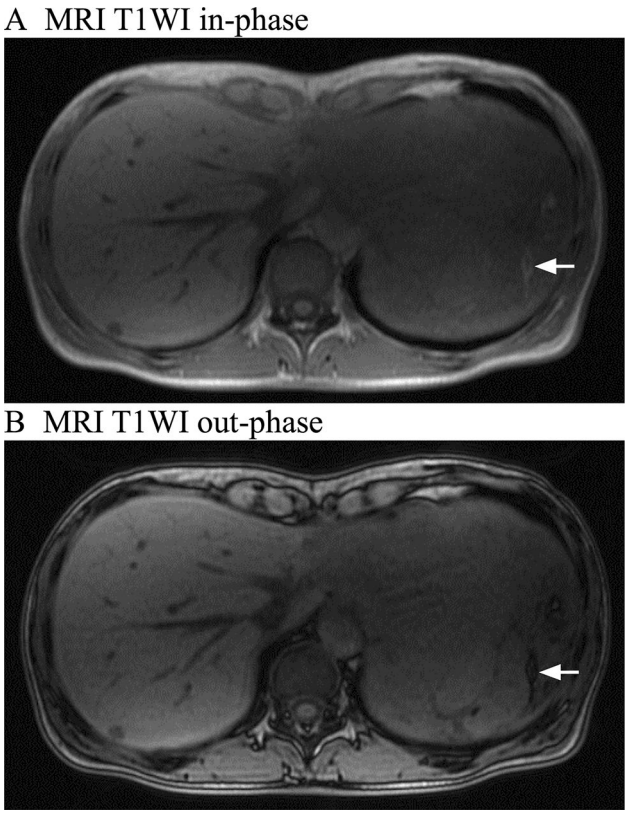

Fig. 4 Findings of abdominal magnetic resonance imaging. **a** The tumor in the left hepatic lobe is hypointense and contains sporadic high-intensity areas, noted via T1-weighted in-phase imaging (arrow). **b** The above-mentioned high-intensity areas became hypointense in the T1-weighted out-phase image (arrow)

Discussion

AML is rare stromal tumor composed of variable admixtures of thick-walled blood vessels, smooth muscles, and adipose tissues [1]. AML commonly arises in the kidney, while the liver is a less common site of origin. Hepatic AML was firstly reported by Ishak [2], and hepatic AML has been identified often in recent years because of advances in imaging modalities. Although AML was previously regarded as a variant of hamartoma, it has recently been considered a tumor of perivascular epithelioid cells (PEComa), exhibiting dual myomatous and lipomatous differentiation and melanogenesis [7, 8]. Immunostaining of HMB-45, which is a monoclonal antibody against melanocyte, is a useful method for the pathology-based diagnosis of hepatic AML, since hepatic tumors, except AML and the metastasis of malignant melanoma, do not show positivity for HMB-45 [9, 10].

Typical hepatic AMLs have several characteristic findings on radiological examinations [11–13]. These findings include (1) early staining in the arterial phase and wash-out of enhanced medium in the equivalent phase on enhanced radiological examinations and (2) a component of adipose tissue in the tumor on CT and MRI. Radiological diagnosis of hepatic AML is occasionally difficult since the proportion of adipose tissue in the tumor greatly varies, ranging from 5 to 90% [1, 14]. In particular, differentiating AML from hepatocellular carcinoma (HCC) with a fatty component is challenging. However, early drainage vein around the tumor is very helpful for diagnosing hepatic AML [3–5], since peritumoral early drainage vein and tortuous tumoral vessels on MRI are rarely seen in HCCs [15]. Such peritumoral vessels are supposed to work as drainage routes for the abundant tumor arterial

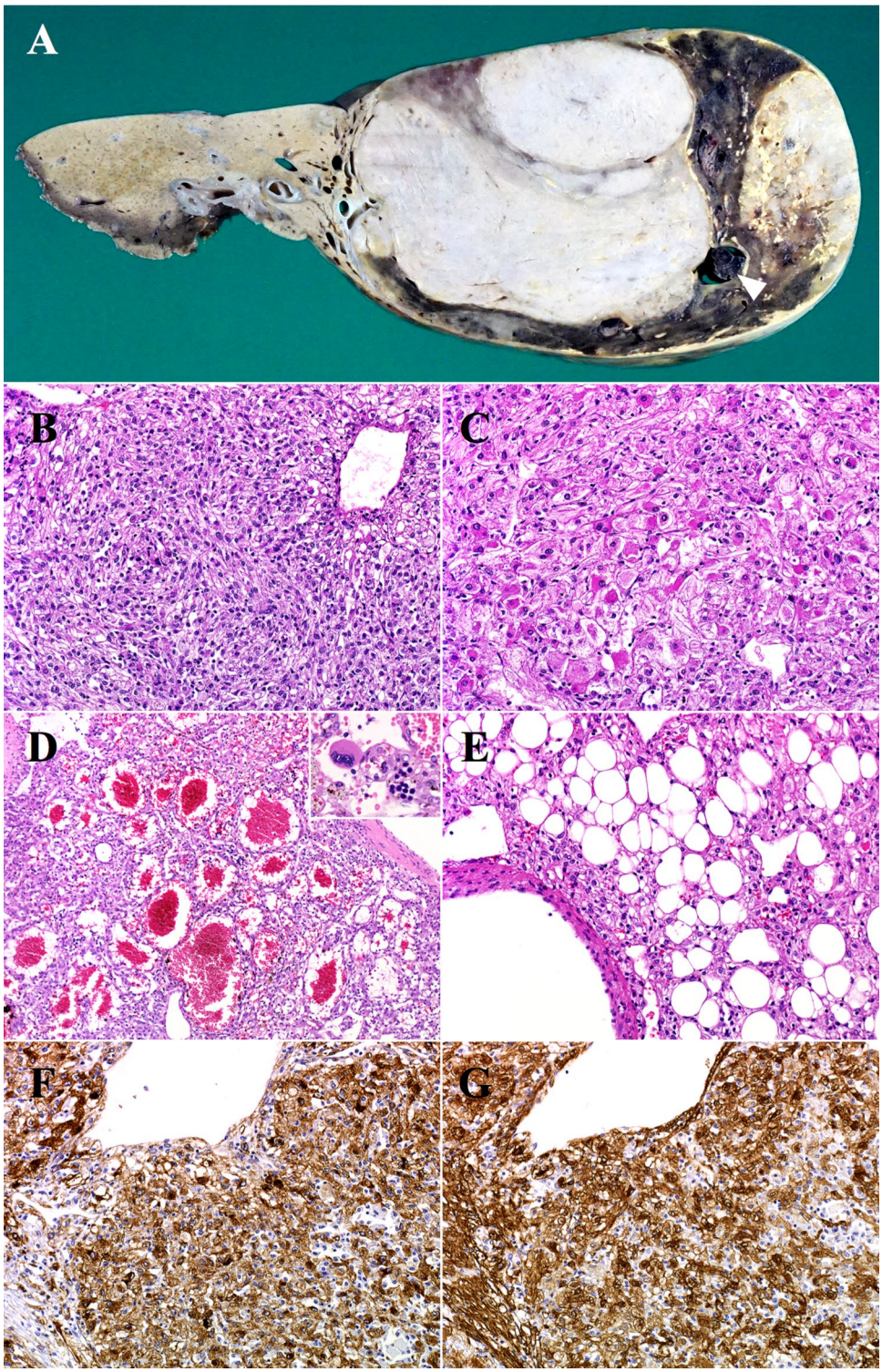

Fig. 5 Macroscopic and histological findings of the resected specimen. **a** Grossly, a well-circumscribed large tumor, bulging outward from the liver, consists mainly of whitish nodules with blackish area in between and at periphery. The arrowhead indicates dilates dilated vessel with thrombus. **b** Microscopically, the tumor consists mainly of epithelial-like to spindle-shaped cells with a clear to eosinophilic cytoplasm. **c** Swollen and eosinophilic large myoid cells are occasionally also observed. **d** The "peliosis hepatis"-like structure is conspicuous in blackish area, and extramedullary hematopoiesis is often seen in this area (inset). **e** Thick wall blood vessels and adipocytes are mixed in some places. **f, g**: Tumor cells are immunohistochemically positive for human melanoma black-45 antigen (HMB-45) and alpha-smooth muscle actin (SMA)

flow [16]. Among 36 AMLs in the literature, including the present case, most of the early drainage veins flowed into the hepatic vein, and drainage into the portal vein was rare [4, 6] (Table 1). Only the present case showed drainage routes into both the hepatic and portal vein. Possible reasons for the drainage routes into both veins are follows: (1) since AML in the left hepatic lobe was very large and intratumoral arterial flow was abundant, the drainage into the hepatic vein was insufficient and (2) an intratumoral arterio-portal communication (shunt) had been formed during the development of AML. The blood flow from AML in left lobe would trend toward umbilical portion and

Table 1 Reported cases with early drainage veins in hepatic angiomyolipoma

References	Number of cases	Detection modality	Drainage into
Murakami et al. [17]	1	DSA	HV
Yoshimura et al. [14]	2	DSA	HV
Zheng and Kudo [3]	2	DSA, CED, US	HV
Jeon et al. [4]	5	MDCT	HV
Iwao et al. [16]	4	DSA	HV
Wang et al. [15]	16	MRI	HV
Yoshioka et al. [5]	1	DSA, CT	HV
Jeon et al. [4]	3	MDCT	PV
Saito et al. [6]	1	MDCT, MRI	PV
Present case	1	MDCT, DSA	HV and PV

DSA digital subtraction angiography, *CED* contrast-enhanced Doppler, *US* ultrasonography, *MDCT* multidetector computed tomography, *MRI* magnetic resonance imaging, *CT* computed tomography, *HV* hepatic vein, *PV* portal vein

right portal vein because the revascularization of round ligament of liver was not detected in CT and angiography.

Although most hepatic AMLs are benign, some have malignant potential [18–23]. In pathological and immunohistological examination, the mitotic activity exceeding 1/50 high power field and the deficiency of the c-kit expression are related to the malignant potential of AML [18, 20, 21, 24, 25]. However, such pathological findings are unpredictable under radiologic modalities. Folpe et al. reported that recurrence or metastasis of PEComa (another name for AML) of soft tissue or in tissue of gynecologic origin is strongly associated with a tumor size exceeding 8 cm [26]. Taking these into consideration, we decided to resect the tumor in the present case. Although Ki-67 labeling index was 1–2%, the expression of c-kit was low and the tumor in the present case could have had malignant potential. Thus, periodic postoperative check-up will be necessary in this case.

In conclusion, we reported a rare case of hepatic AML with early drainage routes into both the hepatic and portal vein. The dilated and retrogressive vein perfused into the portal vein as well as into the hepatic vein to drain the abundant arterial blood flow of the tumor. The finding of early drainage veins into the portal vein should be helpful for diagnosing hepatic AML with a narrow hepatic vein.

Compliance with ethical standards

Conflict of interest Ryota Kiuchi, Takanori Sakaguchi, Ryo Kitajima, Satoru Furuhashi, Makoto Takeda, Takanori Hiraide, Yoshifumi Morita, Takasuke Ushio, Rei Ishikawa, Satoshi Baba and Hiroya Takeuchi declare that we they have no conflict of interest.

Human/animal rights All procedures followed have been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments.

Informed consent Informed consent was obtained from all patients for being included in the study.

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