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| Title | A case of hepatic angiosarcoma supplied by both hepatic artery and portal vein |
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| Citation | Fukushima Journal of Medical Science. 52(1): 13-19 |
| Issue Date | 2006-06 |
| URL | http://ir.fmu.ac.jp/dspace/handle/123456789/181 |
| Rights | © 2006 The Fukushima Society of Medical Science |
| DOI | 10.5387/fms.52.13 |
| Text Version | publisher |

[Case Report]

A CASE OF HEPATIC ANGIOSARCOMA SUPPLIED BY BOTH
HEPATIC ARTERY AND PORTAL VEIN

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(Received October 5, 2005, accepted December 9, 2005)

Abstract : Liver neoplasms, whether they are benign or malignant, are usually fed by the hepatic artery. We experienced a case of hepatic angiosarcoma supplied by both hepatic artery and portal vein. Since there are currently no specific laboratory tests to diagnose hepatic angiosarcoma, it is important to detect suspect cases from imaging features. This unique hemodynamic property was detected on computed tomography (CT) during hepatic arteriography and CT during hepatic arteriportography. If any imaging examinations indicate the liver tumor to be fed by the portal vein, hepatic angiosarcoma should be suspected.

Key words : Hepatic angiosarcoma, Portal venous supply, CTA, CTAP

INTRODUCTION

Hepatic angiosarcoma is a rare disease that accounts for only 0.013-0.02% of all autopsy cases^{1,2)} and 0.2-1.8% of all primary malignant liver tumors^{1,3)}. Although some radiological imaging features of this disease have been discussed^{4,5)}, the diagnosis is still difficult to make because of its rarity and variety of imaging

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patterns. Sometimes the tumor is first diagnosed as hemangioma, and later found to be an angiosarcoma on the basis of changes in size and appearance of symptoms⁶. Therefore, distinctive imaging features need to be accumulated for correct diagnosis. The imaging features we describe here are considered to reflect the infiltrating pattern of the tumor that, we suggest, causes it to be fed by both hepatic artery and portal vein.

CASE REPORT

A 59-year-old man suffering from heart burn went to a clinic and was examined by abdominal ultrasound (US) and CT, which showed multiple liver tumors. He was referred to our hospital for further examinations. He was not infected with hepatitis B or C virus. He drank alcohol only on occasion. For a few years before being referred, he had had his liver checked by US, because the level of γ -glutamyltransferase was found to be increased, but no abnormal lesions were detected. He had no exposure to the chemicals associated with angiosarcoma, such as thorotrast, arsenic and vinyl chloride⁷⁻⁹. Laboratory data on admission showed throm-

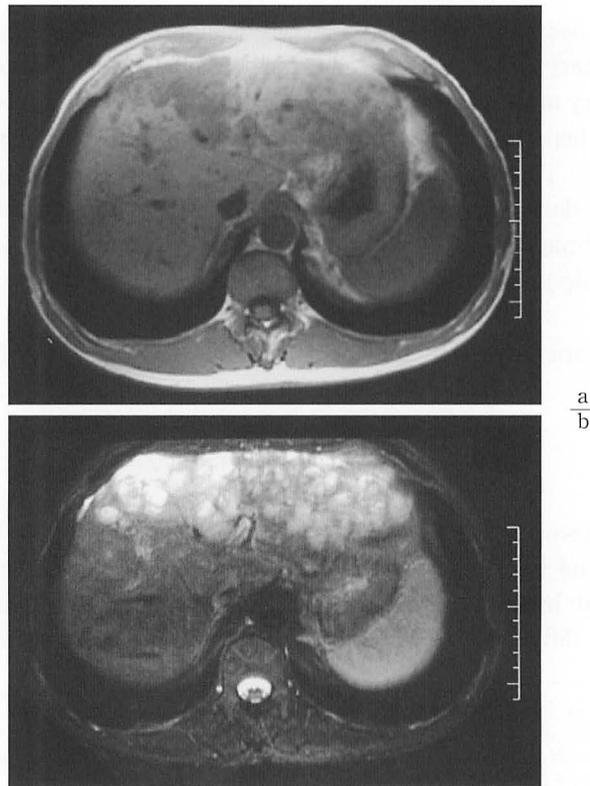


Fig. 1. (a) T1-weighted MRI. Multinodular hypointensity lesions spreads S2, S3, S4.
(b) T2-weighted MRI. Lesions show hyperintensity.

bocytopenia of $114,000/\mu\text{l}$ (normal $125,000\text{--}370,000/\mu\text{l}$), lowest normal prothrombin time (70.7%, normal $>70\%$), slightly increased activated partial thromboplastin time (41.1 s., normal 23–37 s.), decreased hepaplastin (54.2%, normal $>70\%$), and slightly increased fibrinogen/ fibrin degradation products ($11.9\ \mu\text{g}/\text{ml}$, normal $<10\ \mu\text{g}/\text{ml}$). These data suggested the impairment of coagulation system. Aspartate aminotransferase (59 IU/l, normal 9–35 IU/l), alanine aminotransferase (71 IU/l, normal 5–40 IU/l), γ -glutamyltransferase (985 IU/l, normal <50 IU/l), alkaline phosphatase (818 IU/l, normal 85–345 IU/), and total bilirubin (2.0 mg/dl) were all increased. Protein induced by vitamin K absence or antagonist-II (PIVKA-II), α -fetoprotein (AFP), and CA19-9 were all within normal range.

Abdominal US revealed multiple nodular lesions in the liver, most of them in the S2, 3, and 4 segments. They were basically hyperechoic, but some showed mixed echo textures. Doppler US with contrast material, a mixture of galactose and palmitic acid, showed the turbulence of blood flow suggesting that both hepatic artery and portal vein were feeding some lesions.

On contrast-enhanced CT images, the tumor was enhanced in the arterial phase, and the left portal vein was depicted simultaneously, indicating arterioportal shunt-

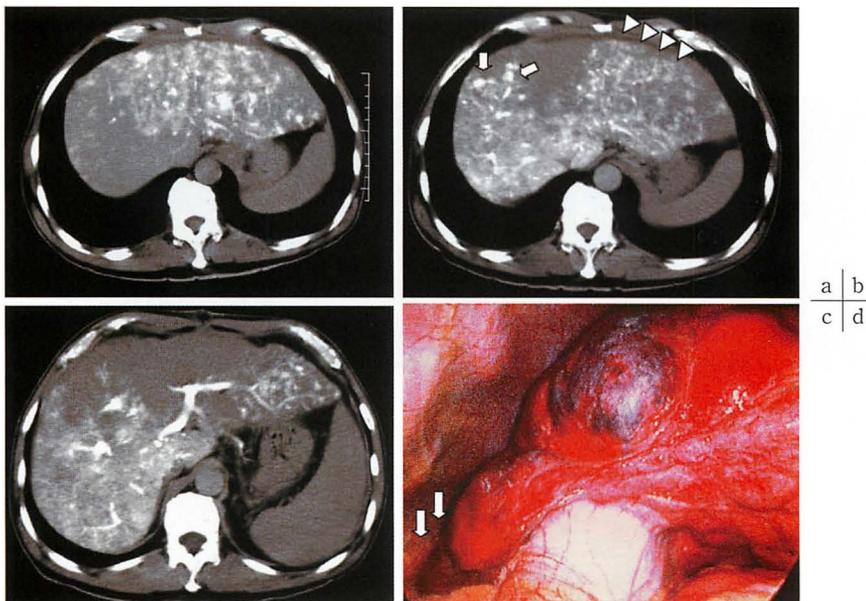


Fig. 2. (a) CTA. Some lesions are depicted as clear nodular lesions, but most of the lesions are enhanced blurrily. (b) CTAP. Lesions in S2 and S3 are enhanced (arrowheads). Two nodular lesions in S4 are also enhanced (arrows), but the large area in S4 shows defect. (c) CTAP. The portal vein penetrates through the tumor. It looks like the vessel is "entrapped" by the tumor. (d) Laparoscopy. A dark-red main tumor surrounded by multiple red nodular lesions is observed in S4. Normal liver structure is seen only as a small part in the lower left (arrows).

ing.

The tumor showed low intensity on T1-weighted magnetic resonance imaging (MRI) images (Fig. 1a), and high intensity on T2-weighted images (Fig. 1b). Superparamagnetic iron oxide (SPIO)-enhanced MRI was also performed, and the tumor showed high intensity on T2-weighted images. From these findings, we suspected the tumor to be HCC.

Angiography, which was performed to understand the hemodynamic properties, showed multiple nodular lesions, a so-called cotton wool-like appearance. CT during hepatic arteriography (CTA) enhanced most of the lesions (Fig. 2a). Some lesions were depicted as clear nodular lesions as seen on MRI images, but most of the lesions were enhanced blurrily. CT during hepatic arteriography (CTAP) also enhanced some lesions, especially lesions in S2 and S3 (Fig. 2b), which was a rare finding because most liver tumors are fed by the hepatic artery. The portal vein was almost completely surrounded by the tumor, but the vein penetrated through the lesions without proximal occlusion. It looked as if the portal vein was “entrapped” by the tumor (Fig. 2c).

Exploratory laparoscopy was performed to observe the liver surface and to gain samples for histological evaluation. The liver surface exhibited irregularities with multiple nodular lesions (Fig. 2d). Two small lesions in S3, about 1.5 cm each in diameter, were excised. Histologically, the portal triad structures and residual

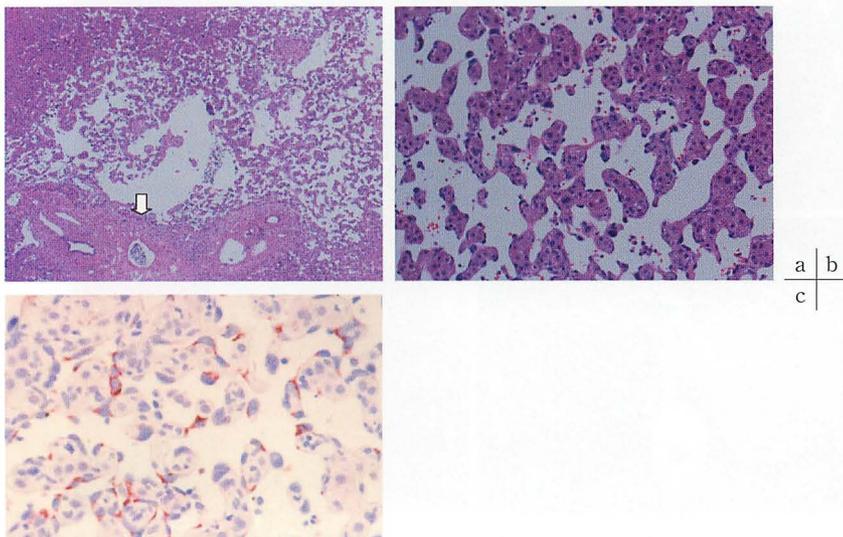


Fig. 3. Pathology of the liver tissue. (a) Tumor cells proliferate without completely replacing the portal spaces. Intact liver structure is seen in the upper left part of the figure. However, tumor cells infiltrate along the sinusoids throughout this area. An AVM-like structure is seen in the bottom (arrow). (HE, $\times 40$) (b) Spindle cells infiltrate along dilated sinusoids. Atrophic hepatic cells are still found. (HE, original magnification, $\times 100$) (c) Spindle cells are positive for factor VIII-related antigen. (Factor VIII-related antigen immunostain, $\times 100$)

atrophic hepatocytes survived the massive invasion of tumor cells without being completely replaced. Intact liver structures were found among dilated sinusoids not only in the peripheral of the tumor but also in the center of it, although the tumor cells infiltrated throughout these structures. In some portal spaces, randomly connected arteries and veins formed the arteriovenous malformation (AVM)-like structure (Fig. 3a). Proliferation of various-sized spindle cells along dilated sinusoids with atrophic liver-cell cords was observed. These cells exhibited marked pleomorphism with elongated and hyperchromatic nuclei (Fig. 3b). Immunohistochemically, the tumor cells were reactive for factor VIII-associated antigen (Fig. 3c). One or two mitoses per high power field (HPF) were found, and the MIB-I index was 15%. Based on these findings, the final diagnosis of high-grade hepatic angiosarcoma was made.

The patient received two times of transcatheter arterial injections with 30 mg of farnorubicin, 20 mg of cisplatin, 6 mg of mitomycin C and 500 mg of 5-fluorouracil. The disease constantly progressed and the patient died of liver failure after 5 months.

DISCUSSION

Kojiro and colleagues reported that angiosarcoma has two basic tumor growth patterns, sinusoidal and solid. There is no significant correlation between the histological and macroscopic features¹⁰. In the present case, histological findings showed proliferation of tumor cells along sinusoids with various degrees of sinusoidal dilatation, the sinusoidal growth pattern. No solid growth pattern was found in the specimen. Portal spaces were left utterly surrounded by tumor cells without being completely destroyed, which suggested that the tumor proliferated rapidly into the parenchyma along the sinusoids and slowly into the portal spaces. The slow invasion into the portal spaces may have spared the portal supply for a relatively long time which, we suggest, caused dual feeding of the tumor. The lesions enhanced on CTAP may be the reflection of these histological features. Most lesions in S4 were fed by the hepatic artery. Presumably, the portal spaces were completely replaced by tumor cells in these lesions. Since no tissue sample was obtained from this part of the liver, it remains no more than a speculation. The entrapped portal vein seen on CTAP is another unique feature supposedly reflecting the growth pattern. Since some case reports also show CT images of the portal vein surrounded or entrapped by the tumor^{11,12}, this feature may be a good clue for suspecting hepatic angiosarcoma.

CT and MR imaging of hepatic angiosarcoma has been studied to determine the features of this tumor. But we could find only one case report of hepatic angiosarcoma (diffuse type) which explained the findings on CTA and CTAP in detail¹³. In this previous report, the tumor was also happened to be fed by both hepatic artery and portal vein. The author concluded that the continuous infiltration of atypical

cells into the dilating sinusoids was probably the reason for sustaining portal venous feeding. Although there are other liver lesions, such as early HCC, adenomatous hyperplasia and regenerative nodule, reported to be enhanced by CTAP¹⁴⁻¹⁶⁾, they are usually accompanied by chronic liver diseases; on the other hand, most hepatic angiosarcoma cases are not. It is also important to differentiate a hepatic angiosarcoma from other angiomatous lesions, such as peliosis hepatis and epithelioid hemangioendothelioma, by CTA and CTAP, but that has not been discussed in the literature.

In addition to the imaging features, thrombocytopenia and slightly increased fibrinogen/ fibrin degradation products were suggestive of a tumor of vascular origin. Unlike disseminated intravascular coagulation (DIC) due to progressive malignant disease, thrombocytopenia in this patient fluctuated from 70,000/ μ l to 150,000/ μ l during the period of hospitalization without DIC therapy. The platelet count was 147,000/ μ l on the day before the patient's death. We suggest it was consumption coagulopathy due to the angiosarcoma.

CONCLUSION

We present a case of hepatic angiosarcoma with a unique hemodynamic detected on CTA and CTAP. These examinations are very useful for investigating hemodynamic properties in detail. If a liver tumor is found to be fed by the portal vein, hepatic angiosarcoma should be suspected.

ACKNOWLEDGMENT

We thank Dr. M. Unagami for his expert advice on pathological evaluation.

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