Hepatic Angiomyolipoma: a case report

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ABSTRACT

We experienced a rare case of hepatic angiomyolipoma (AML). A 66-year old Japanese female presented inhomogeneous echogenic lesion in the lateral segment of the liver on ultrasonography. Contrast computed tomography (CT) revealed early arterial enhancement within the lesion that stayed hyperdense in the equilibrium phase. Magnetic resonance imaging (MRI) demonstrated a non-homogeneous and partially high intensity mass on both T₁- and T₂-weighted images. Selective hepatic digital subtraction angiography (DSA) showed the lesion to be inhomogeneously hypervascular, supplied via branches of the left hepatic artery. The patient underwent elective left hemihepatectomy. Microscopic findings demonstrated that the tumor was composed of fat cells, blood vessels, and smooth muscle cells. Most of the spindle cells were immunoreactive to homatropine methylbromide 45 (HMB-45), alpha-smooth muscle actin and Melan-A/MART-1. Morphological pattern and immunophenotype were consistent with hepatic angiomyolipoma.

Key words: Angiomyolipoma, Liver

Hepatic angiomyolipoma (AML) is a quite rare mesenchymal tumor. It is a usually benign lesion that consists of three components in variable proportions: smooth muscle, fat and blood vessels. We present a benign hepatic AML, which was found during a regular health checkup.

CASE REPORT

A 66-year old Japanese female presented an inhomogeneous echogenic lesion in the lateral segment of the liver on ultrasonography performed for a regular health checkup. She had no abdominal complaint. She had undergone lumpectomy for a benign phyllodes tumor one year before. Pertinent laboratory data were all within normal limits and hepatitis B and C viral infection tests were negative. Biochemistry showed that tumor markers were within normal ranges, with an alfa-fetprotein level of 5.0 ng/ml (normal range < 20.0 ng/ml), PIVKA-2 level of 16 mAU/ml (normal range < 30.0 mAU/ml) and carcinoembryonic antigen level of 3.3 ng/ml (normal range < 4.0 ng/ml). There was no evidence of tuberous sclerosis.

Contrast computed tomography (CT) revealed an 80 × 60 mm inhomogeneous lesion, containing an area with CT number under -20 Hounsfield Units (HU) in the lateral segment of the liver. There was early arterial enhancement within the lesion that stayed hyperdense in the equilibrium phase. There was no portal vein or hepatic vein invasion (Fig. 1).

Magnetic resonance imaging (MRI) demonstrated a non-homogeneous and partially high intensity mass on both T₁- and T₂-weighted images. The fat suppression image confirmed the fatty nature of the mass. Selective hepatic digital subtraction angiography (DSA) showed the lesion to be inhomogeneously hypervascular, supplied via branches of the left hepatic artery. The radiologic findings were not typical of hepatocellular carcinoma and hepatic angiomyolipoma was suspected as most likely. The patient underwent elective left hemihepatectomy. Gross examination of the resected specimen revealed an 80-mm yellow tumor, which was smooth externally, globular, and...
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Fig. 1. Contrast computed tomography (CT) demonstrated early arterial enhancement within the lesion (A) that stayed hyperdense in the equilibrium phase (B).

Fig. 2. Most of the spindle cells were immunoreactive to homatropine methylbromide 45 (A), and alpha-smooth muscle actin (B).

well circumscribed. Microscopic findings demonstrated that the tumor was composed of fat cells, blood vessels, and smooth muscle cells. Most of the spindle cells were immunoreactive to homatropine methylbromide 45 (HMB-45), alpha-smooth muscle actin and Melan-A/MART-1 (Fig. 2). Morphological pattern and immunophenotype were consistent with hepatic angiomyolipoma. The patient recovered uneventfully and was discharged two weeks after the operation. She has subsequently been observed as an outpatient for thirty six months without any symptoms.

DISCUSSION

Angiomyolipoma (AML) is a mixed mesenchymal tumor, which usually occurs in the kidney but rarely in the liver. Ishak described the first hepatic AML in 1976. Hepatic AML usually occur in non-cirrhotic livers, and present as a solitary hepatic tumor. Multiple tumors are rare and only four cases of multiple hepatic angiomyolipomas have ever been described. It is reported that 40 to 50% of renal AML is associated with tuberous sclerosis; however, in hepatic AML, tuberous sclerosis is an associated finding in 6%. About two-thirds of patients are reported to have symptoms, with abdominal pain as the most common symptom with an incidence of 60%. Preoperative diagnosis relies on imaging study including CT scan, magnetic resonance imaging. The majority of hepatic AML show a density component of less than -20 HU on CT, indicating the presence of adipose tissue. It is suggested that the amount of fat tissue within a hepatic AML can range from 5 to 90%. Variable proportions of fatty tissue in the tumors may result in varied appearance on CT scan.

Dynamic study has been suggested as a reliable method for differentiating AML from fat containing hepatocellular carcinoma. It is suggested that early and prolonged enhancement of the lesion with its characteristic time-density curve is significant for hepatic AML.

In angiographic examination, it is reported that the tumor was hypervascular in 90% of cases. Although angiography is not a specific diagnostic modality for differentiation, it is useful to detect the vascular supply to the lesion and thereby facilitate surgical resection.
Histologic features of hepatic AMLs are vascular hyperplasia and smooth muscle and fat cells. It is thought to be a member of the PEComa family, a group of tumors derived from the perivascular epithelioid cells (PEC). These cells are characterized by the coexpression of muscle and melanogenois markers and, as such, stain immunohistochemically with alpha smooth muscle actin and HMB-45. The case we describe displays these typical immunohistochemical phenotypes. It was suggested that occasional follow up would be sufficient, when the diagnosis of hepatic AML has been established. However, instance of malignant transformation of hepatic AML has been reported recently. It was a tumor with vascular invasion and a fatal outcome. The authors pointed out the possibility that malignant AML has been misdiagnosed as a hepatocellular carcinoma in the past and highlighted the importance of immunohistochemistry and electron microscopy in diagnosing this type of tumor. Complete surgical excision is thought to be curative. Long term follow-up of our patient would be needed to monitor for any recurrence lesion.

(Received January 27, 2010)
(Accepted March 4, 2010)

REFERENCES