CASE REPORT

Multiple hepatic angiomyolipomas with a solitary omental angiomyolipoma

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Abstract : Angiomyolipoma (AML) is a rare benign tumor that occurs most commonly in the kidney. Lesions in the liver are usually solitary and multiple AMLs of the liver are extremely rare. Furthermore, extra renal or hepatic AML are rarely found. We report an unusual case of a 34-year-old man with a solitary omental AML and multiple hepatic AMLs. At the age of 23, the patient underwent right nephrectomy and enucleation of a left renal tumor because of bilateral AMLs. At the age of 34, more than 6 lesions in the liver and an enlarged solitary omental AML were discovered. The omental tumor, 50×40mm, 49g, was extirpated ; it was well-defined and encapsulated a soft elastic mass. Histologically it was an epithelioid AML and positive for the melanogenesis-related marker HMB - 45, the same as the earlier right renal tumor. We describe the first case of a solitary omental AML, which had metastasized, and with more than 6 hepatic AMLs. J. Med. Invest. 52 : 218-222, August, 2005

Keywords **:** *angiomyolipoma, liver, multiple, omental tumor, metastasis*

INTRODUCTION

Angiomyolipoma (AML) is a benign tumor occurring most commonly in the kidney and consists of a variable admixture of proliferating blood vessels, adipose tissue, and smooth muscle. The association of these tumors with tuberous sclerosis is well established and bilateral or multiple renal AMLs are often associated with tuberous sclerosis (40%)(1). Multiple liver AMLs have been described in less than ten cases in the literature(2). We earlier reported the first case of omental AML, and mainly described the imprint cytology of the epithelioid AML(3). We now describe a 34-year-old man with an enlarged solitary omental AML and with more than 6 hepatic AMLs.

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CASE REPORT

A34-year-old asymptomatic man was admitted to our hospital due to the enlargement of a tumor in the visceral cavity. At the age 23, the patient underwent right nephrectomy and enucleation of a left renal tumor because of a bilateral renal tumor. Pathological study revealed that the right renal tumor was epithelioid AML because of atypical polygonal cells and the existence of mitotic cells, while the left renal tumor was a conventional AML. At age 29 the patient was found to have multiple hepatic tumors up to 4 cm in diameter. Fine-needle biopsy of the liver tumors (existent in Couinaud Segment VII) revealed conventional AMLs. Because angiography showed the tumors were hypervascular, transcatheter arterial embolization by SMANCS® (Yamanouchi, Tokyo, Japan) was performed. At age 32, abdominal computed tomography revealed a tumor, 2.5cm in diameter, located in the greater omentum beside the antrum of the stomach, and that the liver

tumors that had scarcely changed in size and number. At age 34, 20 months after the last consultation, Magnetic Resonance Imaging (MRI) revealed that the tumor in the visceral cavity had enlarged to 5 cm in diameter; that the liver tumors detected at that time had scarcely changed but three new lesions were detected. The patient had evidence of tuberous sclerosis, but there was no history of epilepsy.

Physical examination on admission revealed evidence of adenoma sebaceum around the nose. An elastic mass, 5cm in diameter, was palpable in the right hypochondrium. The patient had no viral and immunoserological hepatitis markers nor any history of alcohol consumption. Serum squamous cell carcinoma related antigen (SCC) and cancer antigen 72-4(CA72-4) were slightly elevated. Laboratory findings on admission are summarized in Table 1. MRI revealed that the omental tumor was 5cm in diameter with T1-weighted low-intensity and T2-weighted high-intensity with foci of higher intensity lesions in the mass (Fig. 1A , B). Meanwhile, the newly found tumor in Segment VIII was a T1-weighted low-intensity and T2-weighted high-intensity lesion in the mass, and the tumor present in Segment

Table1. Laboratory Findings on Admission

Red blood cell (450-550 $\times 10^4/\mu l$)	413 $\times 10^4$
Hemoglobin (14.0-17.0 g/dl)	12.7
Platelet (15.0-35.0 $\times 10^4/\mu l$)	25.4 $\times 10^4$
Aspartate aminotransferase (10-35 IU/L)	18.00
Alanine aminotransferase (5-40 IU/L)	23.00
Total billirubin (0.1-1.0 mg/dl)	0.70
Total protein (6.5-8.2 g/dl)	6.70
Albumin (3.7-5.1 g/dl)	4.20
Blood urea nitrogen (8-20 mg/dl)	18.00
Creatinine (0.7-1.3 mg/dl)	1.36
Creatinine clearance (70-130 ml/min)	77.00
Sodium (135-146 mEq/L)	142.00
Potassium (3.5-4.8 mEq/L)	4.10
Chloride (98-108 mEq/L)	103.00
ICG retention in 15 min (<10%)	5.90
CEA (<5.0 ng/ml)	1.90
α-fetoprotein (<20 ng/L)	5.00
PIVKA-2 (<20 ng/L)	20.00
SCC (<1.5 ng/ml)	1.59
CA72-4 (<4.0 U/ml)	6.00

ICG, indocyanin green.
CEA, carcinogenic embryonic antigen.
SCC, squamous cell carcinoma related antigen.
PIVKA-2, proteins induced by vitamin K absence or antagonist-II.
CA72-4, cancer antigen 72-4.
Parentheses indicate the normal range.

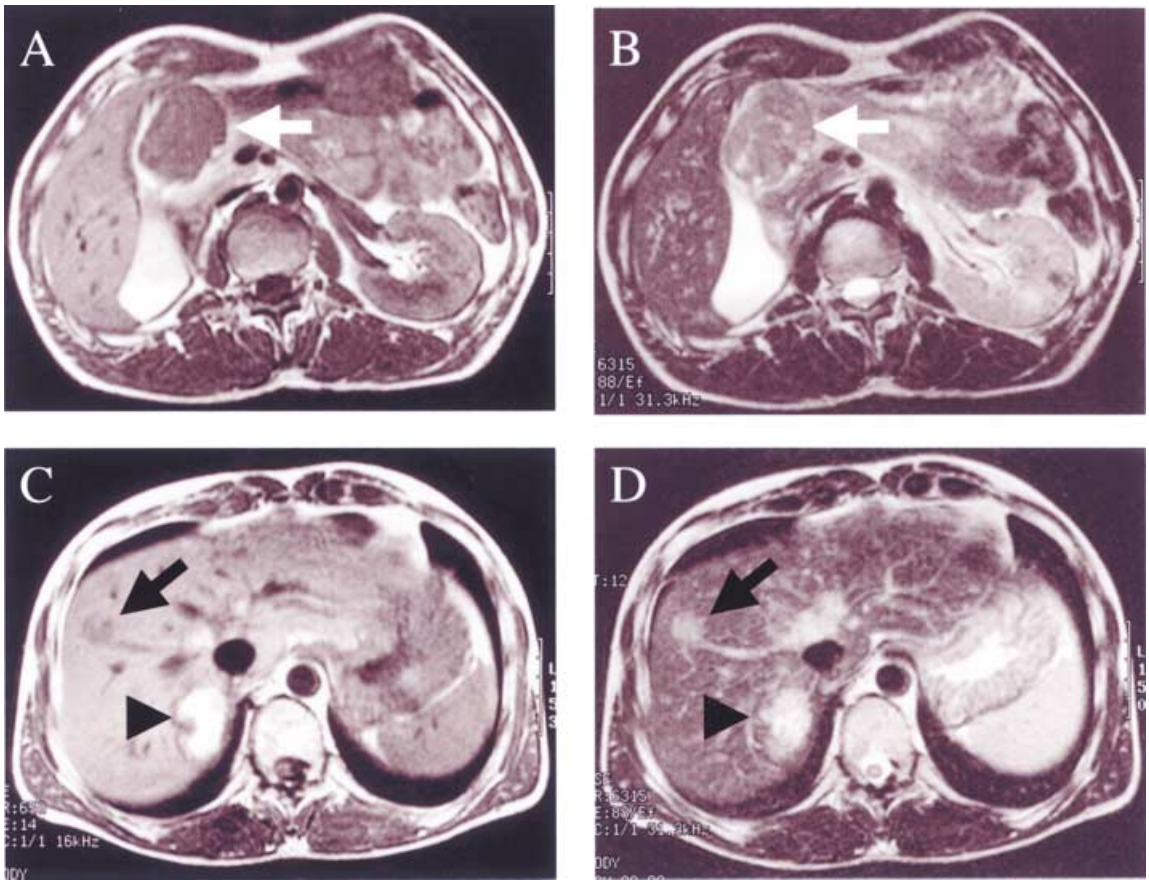

Fig.1 A-D. Magnetic Resonance Imaging (MRI) : The omental tumor (white arrow) is 5 cm in diameter, T 1-weighted low-(A) and T2-weighted high-(B) intensity. C T1-weighted MRI of the liver demonstrates a well-defined and round tumor with a low signal intensity in Segment VIII (black arrow) and high signal intensity in Segment VII (black arrow head). D T2-weighted MRI of the same tumor demonstrates moderately high signal intensity.

VII appeared to be a high intensity mass both T1-and T2-weighted (Fig.1C,D). Angiography showed that the omental tumor was hypervascular, and fed by the gastroduodenal and the branch of supra mesenteric arteries (Fig. 2A,B).

The patient underwent an operation because of the enlargement of the omental tumor and a suspicion of malignancy. There was no peritoneal dissemination and ascites. The tumor was well-defined elastic soft and located in the greater omentum. Intraoperative ultrasonography showed a multiple hyperechoic hepatic mass. The newly appeared tumors in the liver were a hypoechoic mass with a hyperechoic area, which was different from the other mass existent in the liver (Fig. 3A). However, the omental tumor was a hypoechoic mass with a hyperechoic area (Fig. 3B), the same as the newly found tumors in the liver. Needle biopsy was performed on some of the tumors in the liver.

The 50×40 mm omental tumor, weighed 49g, was well-defined and encapsulated a soft elastic mass (Fig. 4A). The cut surface of the tumor revealed 1-mm thick capsules, grayish, focal yellow, and red-brown in color indicating hemorrhage and necrosis (Fig. 4B).

The omental tumor was an epithelioid AML, and positive for HMB-45 antibody (melanocytic cell-specific monoclonal antibody) (Fig. 5) without lymph node involvement. The new lesion in the liver was also an epithelioid AML, and histologically the same as the right renal tumor extirpated before. The other tumors which used to be in the liver were conventional AML.

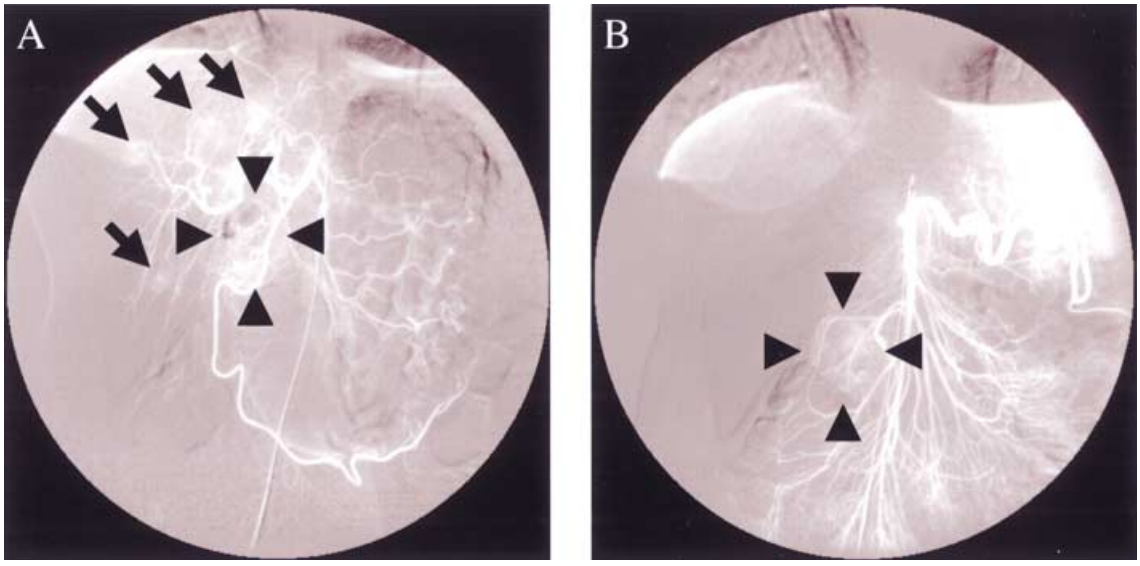

Fig. 2. A, B. Angiography : A Selective celiac arteriography reveals multiple hypervascular tumors with diffuse and homogenous staining in the liver (black arrow), and that the gastroepiploic artery feeds the omental tumor (black arrow head). B Selective supra mesenteric arteriography reveals feeding artery of the omental tumor is also supplied from the branch of supra mesenteric artery.

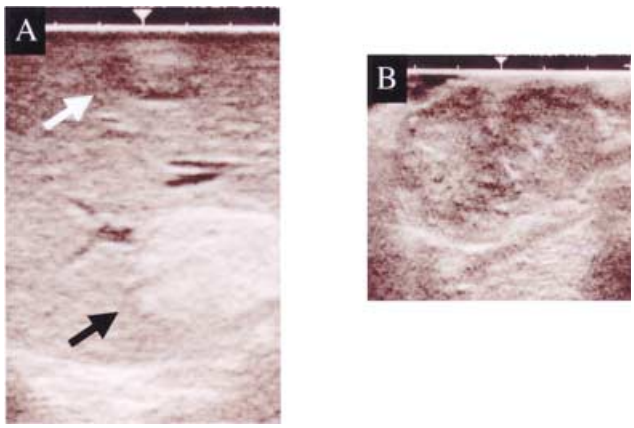

Fig. 3A, B. Ultrasonography : A A newly found tumor in the liver (Segment VIII) (white arrow) is a hypoechoic mass with a hyperechoic area, and different from the other mass existent in the liver (black arrow). B The omental tumor liver is a hypoechoic mass with a hyperechoic area.

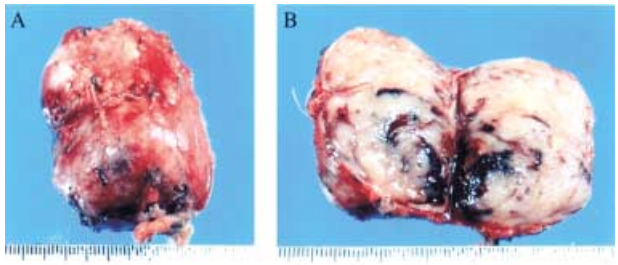

Fig. 4A, B. Resected specimen: A The omental tumor. B The cut surface of the omental tumor.

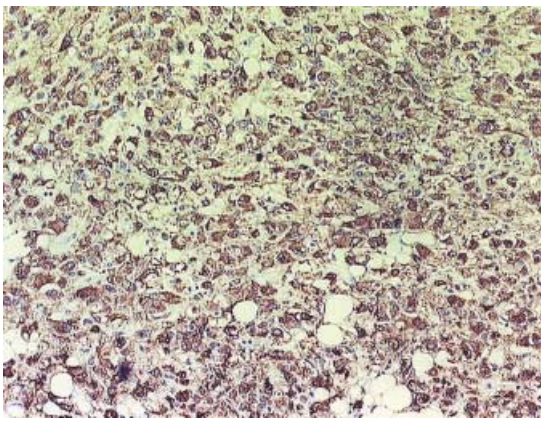

Fig. 5. Histological findings : Some omental tumor cells are positive for HMB-45(melanocytic cell-specific monoclonal antibody) (original magnification ×200).

The patient remains alive 4 years after the last operation, but details are not known because there has been no recent consultation.

DISCUSSION

By immunohistochemical findings an AML is characteristically positive for the melanogenesis-related marker HMB-45(4), it is generally a benign tumor, and even the presence of multiple renal AMLs or localization in regional lymph nodes should be considered as multifocal lesions rather than metastasis (5). Hepatic AMLs share a similar gene expression profile and may differentiate toward activated stellate cells of the liver (6). This might be show hepatic AML is multicentric disease. Hepatic lesions are usually solitary and multiple AMLs are extremely rare. Synchronous in kidney and liver, and based on clinical behavior, it is a multicentric benign disease and there is no evidence of malignancy in the majority of cases (7). Nevertheless, cases of AML including hepatic AML, causing death due to recurrence or metastasis have been described (8-12). The tumor that had newly developed in the liver during the clinical course was also histologically diagnosed as an epithelioid AML. The renal tumor might have metachronously metastasized to the liver. Regardless of multifocal or metastatic, we should keep in mind that some of AML have high malignant potential. The patient underwent removal of the omental tumor 4 years ago ; however, since the patient has not visited our hospital since that surgery, details of his clinical condition are unknown. However, a telephone interview showed that this patient was living well without hospital visits. Based on this clinical behavior, the AMLs here might be benign but have had metastasis. We thus describe the first case of a solitary omental AML, which had metastasized 9 years after radical nephrectomy, and with more than 6 hepatic AMLs. There are no reports of SCC and CA 72-4 being measured as tumor markers of AML. Since these levels were abnormally high in this case, they might be useful as tumor markers. We continue to be interested in the future clinical course of this patient, and expect other reports of omental AMLs to be published.

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