The Korean Journal of Internal Medicine
Vol. 10, No. 1, January, 1995

A Case of Huge Solitary Angiomyolipoma of the Liver

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A 32-year-old male patient, diagnosed as a hepatic solitary angiomyolipoma post-operatively, is reported. The tumor was well definedly inhomogenous fat density mass on ultrasonography, computerzied tomography(CT) and magnetic resonance imaging(MRI). The lesion was hypervascular on arteriography. The diagnosis was confirmed by an extended right lobectomy and histological examination.

Key Words : *Angiomyolipoma, Liver*

INTRODUCTION

Angiomyolipoma of the liver is an extremely rare benign tumor. This tumor is not uncommon in the kidney, and many cases of renal angiomyolipoma are associated with tuberous sclerosis. But most hepatic angiomyolipoma was not associated with tuberous sclerosis and only 33 cases had been reported in the English literature before 1993[1-20]. Several authors have recently suggested that diagnosis may be established with relative certainty by recognition of the characteristic features of this tumor using imaging modalities such as ultrasonography, computed tomography, magnetic resonance imaging and angiography[3,6,13]. But because this tumor is generally regarded as a mesenchymal hamartoma consisting of blood vessels, smooth muscle and fatty tissue, various features were shown according to the imaging method employed. So its definite diagnosis requires excision and histologic confirmation. This paper shows a case of huge solitary 25 cm sized angiomyolipoma of the liver diagnosed post-operatively. which express smooth muscle actin as well as melanoma markers such as HMB-45 and S-100

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CASE REPORT

A 32-year-old male presented with a 3 month history of right upper quadrant abdominal discomfort and indigestion These symptoms had occurred especially after exercise such as a foot -ball game. physical examination revealed enlarged liver to 10cm below the right costal margin. There was no stigmata or family history of tuberous sclerosis The liver function tests revealed an elevated alkaline phosphatase of 277 IU/L(normal <237) and gamma glutamyl transpeptidase of 80 IU/L(normal <50). Serum aminotransferase activities were within normal limits. Serum HBsAg, HBsAb and anti-HCV were negative. Serum alpha fetoprotein was also within normal limit. Ultrasonography demonstrated a huge hyperechoic round mass with a well defined smooth contour and several small hypoechoic foci in it(Fig. 1).

There was also a posterior echo attenuation by the mass. CT showed an enhanced round mass with multiple irregular fat dense areas and sharply defined margin(Fig. 2). Hepatic MRI revealed inhomogenous well-capsulated solid mass occupying the entire right lobe. On T_1 weighted image, the mass showed hyper-and isointensity lesion compared to normal liver, and bright signal intensity was shown on T_2 weighted image. On dynamic study, the mass was demonstrated to be inhomogenous and showed prolonged enhancement, which pattern seemed to be atypical

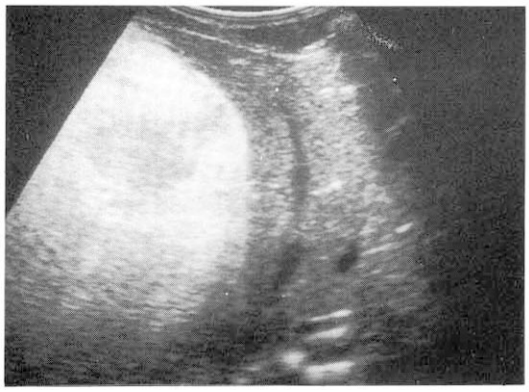

Fig. 1. US: A huge hyperechoic round mass with well-defined swooth contour and several small hypoechoic foci in it.

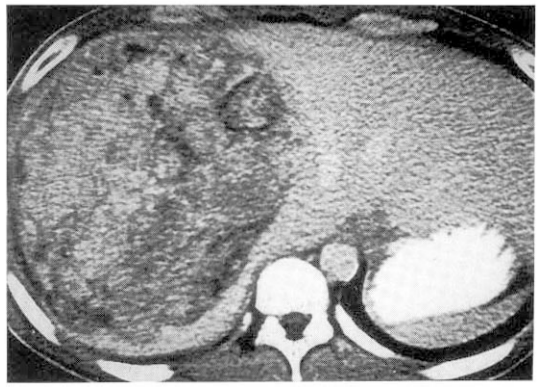

Fig. 2. CT: An enhanced round mass with multiple irregular fat dense areas and sharply defined margin.

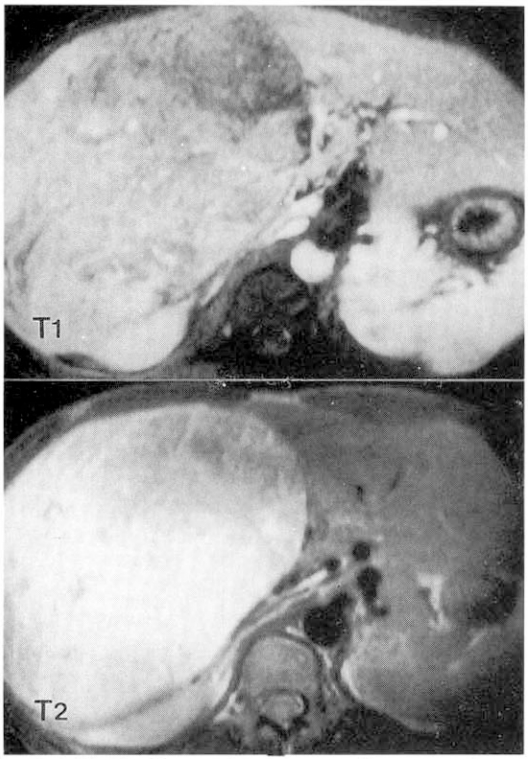

Fig. 3. MRI: On T_1 weighted image, the mass shows hyper-and isodensity lesion compared to normal liver, but on T_2 weighted image it shows bright signal intensity.

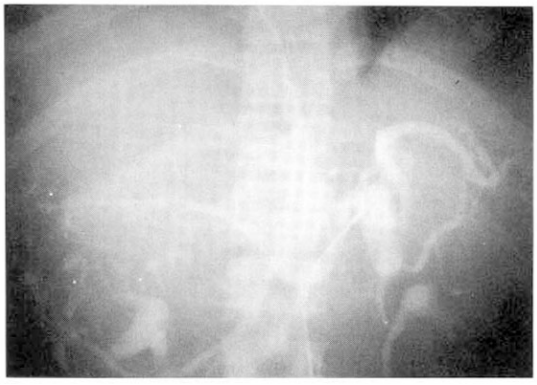

Fig. 4. Angio graphy: celiac angiography demonstrated a large hyper-vascular tumor and dilated hepatic artery and vascular pooling in the lower lateral portion.

for hemangioma(Fig. 3). Celiac angiography demonstrated a large hypervascular tumor without feeding vessels or nodular staining. Displacement and stretching of the intragepatic portal veins were demonstrated on the post-arterial portogram(Fig. 4).

With the diagnosis of atypical hemangioma of the liver, an exploratory laparotomy was per formed and extended right lobectomy was carried out. The resected specimen was measured 29.0×22.0×6.5cm and weighed 2,500g(Fig. 5). The cut surface of the specimen showed 25.0×20.0cm soft, dark reddish and partly yellowish tumor completely surrounded by capsule(Fig. 6). Histologically, the tumor composed of adipose tissue intermingled with areas of epithelioid smooth muscle cells. Multiple thick walled vessels, many of which are abnormal arteries, as well as capillaries and veins, are noted throughout. There are also scattered foci of extramedullary hematopoiesis(Fig. 7). On immunohistochemistry, smooth muscle actin as well as melanoma markers such as HMB-45 and S-100 were

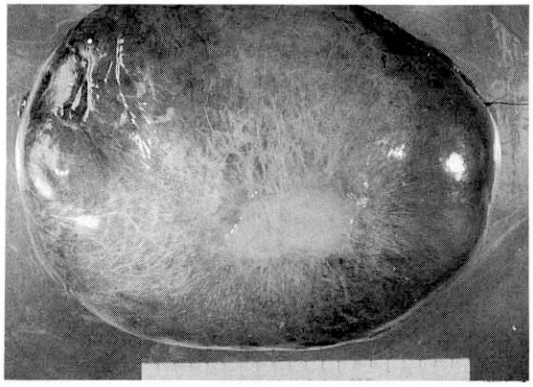

Fig. 5. The resected specimen was measured 29.0×22.0×6.5cm and weighed 2,500g.

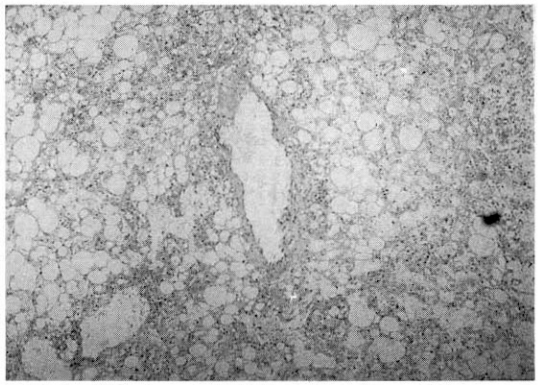

Fig. 7. A. The tumor is mainly composed of fatty tissue and blood vessels (H & E, ×100)

Fig. 6. The cut surface of the specimen revealed 25.0×20.0 cm soft, dark reddish and partly yellowish tumor completely surrounded by capsule.

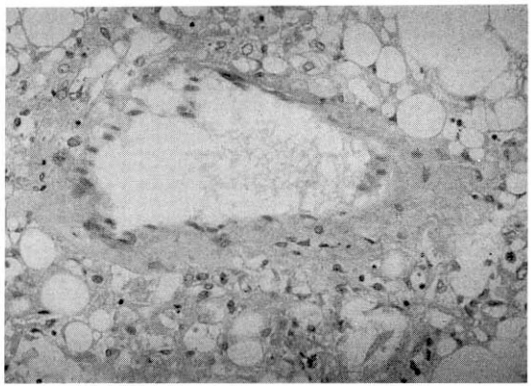

Fig. 7. B. Mulfiple thick walled vessels, many epitheloid smooth muscle cells and scattered foci of extra medullary hematopoiesis are seen (H & E, ×400).

expressed.

DISCUSSION

With the exception of hemangiomas, benign angiomyolipoma of the liver is an extremely rare tumor, especially when compared with that of the kidney. To our knowledge, only 33 cases have been reported in English literature[1-20].

In most of these cases, diagnosis was made post-operatively, except for a few cases, since a pre-operative diagnosis, especially differentiation from malignant tumor, was difficult[1-15]. Since this lesion is benign in nature, it would be preferrable to differentiate it from a malignant vascular tumor unless surgery is inevitable. Its diagnosis requires evaluation by means of imaging modalities and histologic confirmation. But, by reviewing literature, it was very difficult to diagnose hepatic angiomyolipoma by imaging methods only, even though the radiological characteristics of this tumor are well known. This is because of various problems associated with the diagnostic imaging of hepatic angiomyolipoma. With ultrasonography, hepatic angiomyolipoma can not be differentiated from hemangioma, focal nodular hyperplasia, focal fatty changes, some hepatocellular carcinomas containing fatty changes or other hyperechoic masses[24, 25]. Although the CT attenuation value is thought to be diagnostically useful[3, 7], the relative proportions of the various tissue components vary in this tumor, it is well known that the fat content of hepatic angiomyolipoma varies considerably, ranging from less than 10% to more than 50%[4]. Therefore, the CT attenuation value also varies, and it may be difficult to detect the three components-blood vessels, smoth muscle and fatty

tissue by CT scan. The MRI appearance of hepatic angiomyolipoma is a hyperintense mass on T_1-weighted spin-echo pulse sequence. Hyperintense imaging of the lesion on both T_1 and T_2 weighted sequences in this case suggested that they contained a fatty component[9,13,19]. But its finding is not sufficient to make a confirmative diagnosis. Angiography, as well as CT arteriography and MRI, is also useful in the recognition of the vascular componet within the nodule, and in differentiating it from cavernous hemangioma, but is still non-specific.

Particularly, it is very difficult to diagnose focal lesion using imaging modalities when it is small. On the other hand, ultrasonographically guided fine needle biopsy(FNAB) is generally considered to be an important procedure for rapid diagnosis of angiomyolipoma[20-23]. The distinctive FNAB findings can make a definite diagnosis without laparotomy for tissue diagnosis. Histologically the tumor has three or four componets, namely blood vessels, smooth muscle, fat and hematopoietic tissue. However, these elements are variable in proportion and distribution. In the largest series, described by Goodman and Ishak[4], the smooth muscle component is the most prominent, consisting of both spindle and epithelioid cells. The epithelioid cells were polygonal or rounded and were found singly, in clusters or in sheets. Immunohistologically, these cells expressed HMB-45 in the central condensed cytoplasm and actin in a perimenbranous fashion[26-29]. The adipose tissue also reactive for S-100 protein as in this case presented[12]. The size of hepatic angiomyolipoma in the literature was variable from 1 cm to 18cm in diameter. This case may be the single largest one ever reported in the literature. Although all the resected hepatic angiomyolipoma have been cured and there is no evidence of malignant potential, the lesion requires surgical excision if symptoms referrable to the tumor are present and there is a risk of spontaneous rupture due to its locatjon on the surface of the liver.

ACKNOWLEDGEMENT

The authors thank Dr. K. G. Ishak and his collegues in the Department of Pathology, AFIP, Washington, D. C., for their pathological confirmation and special stains.

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