Diagnosis and treatment of hepatic angiomyolipoma in 26 cases

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Abstract

AIM: To summarize the experience of the diagnosis and treatment of hepatic angiomyolipoma (HAML).

METHODS: The clinical, imaging and pathological features, and treatment strategies of 26 patients with HAML treated at the authors’ institute between October 1998 and January 2003 were retrospectively analyzed. All the patients received liver resection and were followed up till the study. Immunohistochemical assays were performed with a panel of antibodies.

RESULTS: There was an obvious female predominance (21:5), and most of the patients (18/26) had no symptoms. Heterogeneous high echo was found in ultrasonography and punctiform or filiform vascular distribution pattern was found in color Doppler-sonography in most of the lesions (21/26). All of the 5 lesions further enhanced with Levovist showed early and prolonged enhancement. At contrast-enhanced spiral CT, the soft-tissue components of 24 lesions were markedly enhanced in the arterial phase and 18 lesions remained enhanced in the portal venous phase. MRI was performed in 9 patients, and showed hypointensity on T1-weighted images and heterogeneous hyperintensity on T2-weighted images. Histopathologically, all lesions were composed of adipose tissues, smooth muscle and blood vessels with different proportions. Most lesions showed positive immunohistochemical staining for HMB45 (26/26), A103 (24/26) and SMA (24/26). All of the 26 patients showed a benign course with no sign of recurrence.

CONCLUSION: Preoperative radiological diagnosis of HAML is possible. The demonstration of intratumoral fat and central vessels is helpful in the diagnosis. HMB45, A103 and SMA are promising markers for pathologic diagnosis of HAML, and surgical resection is effective for the treatment of HAML.

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INTRODUCTION

Hepatic angiomyolipoma (HAML) is a rare benign mesenchymal neoplasm of the liver. Since its first description by Ishak in 1976[1], not more than 200 cases have been reported in the English literatures[2,3]. However, with recent progress in imaging diagnostic techniques, the reported cases of HAML are increasing in number, and the significance of accurate diagnosis is becoming more important clinically. The purpose of this study was to investigate the clinical, imaging and pathological features of HAML and to summarize our experience in the diagnosis and treatment of this disease.

MATERIALS AND METHODS

Patients and clinical data
Twenty-six patients with HAML were surgically treated in Liver Cancer Institute of Fudan University from October 1998 to January 2003. There was a marked female predominance (21/26) and the mean age was 44.3 with a range of 31 to 64 years. Most of the patients (18/26) had no symptoms and were detected incidentally by medical check-up. Seven of 26 patients had symptoms caused by tumor oppression and one patient had slight fever as a chief complaint. The average tumor size at detection was 6.1 cm ranging from 1.5 to 15 cm. None of them was found complicated with a diagnosis of tuberous sclerosis and renal AML. Concomitant hepatic hemangioma was found in one patient. None of them had the history of hepatitis virus infection. Serum alpha-fetoprotein (AFP) levels were all within normal limits.

Imaging examinations
All of the patients underwent ultrasonography, color Doppler-sonography and computer tomography (CT) examinations. Nine patients also received magnetic resonance imaging (MRI) examination.

Treatment and follow-up
Limited partial liver resections were performed in 19 patients, left lateral lobectomy in 4, left hemihepatectomy in 2, and right hemihepatectomy in 1. All the patients have been followed up till the study.

Pathological and immunochemical assays
Routine histopathological examination with hematoxylin and eosin staining was performed. Immunohistochemical studies were performed by the EnVisionTM method using a panel of antibodies (HMB45, A103, smooth muscle actin, S100, Vimentin and CK8) in all of the tumor tissues.

RESULTS

Imaging features
Most lesions (21/26) showed heterogeneous high echo in ultrasonography, and punctiform or filiform vascular distribution pattern in color Doppler-sonography. Five lesions were further enhanced with Levovist, and all of them were found to have early and prolonged enhancement.

In contrast-enhanced spiral CT examination, the soft-tissue components of 24 lesions were markedly enhanced in the arterial phase, and 18 lesions remained enhanced in the portal venous phase. MRI was performed in 9 patients, hypointensity or hyperintensity was found on T1-weighted images and heterogeneous hyperintensity on T2-weighted images.
Histopathological and immunochemical characteristics

The tumors were well circumscribed but no obvious capsule could be found. The non-tumorous liver parenchyma was normal, and no cirrhosis was found. All tumors were composed of adipose tissues, smooth muscle and blood vessels in different proportions. In immunohistochemical studies, most tumors were positive for HMB45, SMA, A103, and Vimentin. However, for CK8, negative results were observed.

**Tissue components in the tumor**

The tissue components in the tumor were highly variable from case to case, and even between different lesions. So, although diagnosis may be suggested, histological confirmation remains mandatory.

**Diagnosis**

Angiomyolipoma, which occurs relatively frequent in kidney, is a rare benign mesenchymal neoplasm of the liver. The tumor size of HAML at the first diagnosis is variable, ranging from 0.1 cm to ≥36 cm. Clinically, most of the patients have no symptoms and are detected incidentally by medical check-ups. Patients with large tumors usually have some symptoms caused by tumor compression. The diagnosis of HAML depends on imaging examination.

According to our experience, typical performance of HAML is a smoothly contoured heterogeneous high echo lesion, with a well-defined border separating it from adjacent normal hepatic tissues by ultrasonography and punctiform or filiform vascular distribution pattern by color Doppler-sonography. In further enhanced imaging with Levovist, the tumor showed early and prolonged enhancement. The lesions appeared as hypodense, and adipose dense could be found in pre-contrast CT scans. In the arterial phase, the soft-tissue components of the lesions were markedly enhanced and central vessels could be found. In the portal venous phase, the lesions remained in hypodense, and adipose dense could be found in pre-contrast CT scans. In the arterial phase, the soft-tissue components of the lesions were markedly enhanced and central vessels could be found. In the portal venous phase, the lesions remained in hypodense, and adipose dense could be found in pre-contrast CT scans. 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