Hepatic Angiomyolipoma Mimicking Hepatocellular Carcinoma

Alexander Y.F. Chung,¹ Siok-Bian Ng,² Choon-Hua Thng,³ Pierce K.H. Chow and London P.J. Ooi,¹ ¹Department of General Surgery,² Department of Pathology, Singapore General Hospital, and ³Department of Diagnostic Imaging, National Cancer Centre, Singapore.

Angiomyolipoma (AML) is a rare benign lipomatous tumour of the liver. It is typically echogenic on ultrasound, hypodense on computed tomography and hyperintense on magnetic resonance imaging. Its varied imaging appearance is due to the different proportion of the three cell types which make up the tumour. This is a case report of a hepatic AML mimicking hepatocellular carcinoma (HCC) with fatty change in a hepatitis B carrier. Diagnostic difficulty and implications on subsequent management are discussed in the context of an endemic region for HCC. (Asian J Surg 2002;25(3):251–4)

INTRODUCTION

Angiomyolipoma (AML) typically occurs in the kidney and rarely in the liver. Ishak reported the first hepatic AML in 1976 ¹ and, since then, there have been about 100 cases reported in the literature.²,³ With improvements in imaging, its diagnosis can lead to a conservative approach to such lesions, as in the kidney. In a region endemic for hepatocellular carcinoma (HCC), the diagnosis of AML can be difficult. We report a case of hepatic AML and its diagnostic implications.

CASE REPORT

A 67-year-old Chinese gentleman, a hepatitis B carrier with alcoholic liver disease, had a right-sided liver lesion on ultrasound (US) screening. Clinically, he had hepatomegaly with the liver 3 cm below the right costal margin. The α-fetoprotein level was not raised (4.2 μg/L) and liver function tests were also normal.

On US of the hepatobiliary system, there was a large, 5.0 x 5.5 cm, round echogenic mass in the right lobe of the liver (Figure 1). The gallbladder contained gallstones and there were no dilated intrahepatic or extrahepatic ducts.

Dynamic contrast-enhanced computed tomography (CT) of the liver was then performed, showing a 5-cm mass in segment VI of the right lobe, which demonstrated heterogenous enhancement in the arterial phase. There were irregular central areas of fatty attenuation. The lesion was less dense in the portal venous phase and, in the delayed phase, most of the contrast had faded. There was enhancement of the pseudocapsule and internal septae in

Figure 1. Ultrasonography revealed a well-defined echogenic mass in the right lobe of the liver.
the portovenous phase (Figure 2). Radiologically, the lesion was consistent with that of HCC in a hepatitis B carrier. The kidneys were normal.

The patient underwent cholecystectomy and right hemihepatectomy with clear surgical margins. Macroscopically, there was a well-circumscribed, solid, lobulated, tan-white tumour with scattered yellowish areas measuring 5 x 5 x 6 cm (Figure 3). The liver was cirrhotic. Microscopically, the tumour was composed of sheets of polygonal and spindle-shaped cells with clear to eosinophilic granular cytoplasm and nuclei ranging from small and regular to hyperchromatic, multinucleated and bizarre (Figure 4). Scattered lobules of mature adipocytes, capillaries and blood vessels with hyalinized to muscularized walls were also present. The tumour cells displayed focal positivity for HMB-45 (Figure 5), smooth muscle actin and desmin.

The patient recovered uneventfully and was discharged on the sixth postoperative day.

**DISCUSSION**

AML is a rare benign lipomatous tumour in the liver. Unlike AML in the kidney, only 5.8% to 10% of hepatic AML are associated with tuberous sclerosis, though in the setting of the latter, the hepatic lesions may represent...
AML and are multicentric. The tumour is composed of three tissue components: blood vessels, smooth muscle and fat. Extramedullary haemopoiesis is also a distinctive feature.

The unifying factor that aided the histological diagnosis of AML was the introduction of HMB-45, a human monoclonal antibody to melanocytes. In the past, most of these tumours may have been misdiagnosed according to the variants of each cell line. No liver tissue components or primary liver tumours except AML myoid cells are reactive to HMB-45. The presence of cells expressing muscle antigens and HMB-45, as well as fat and blood vessels within the tumour clearly establishes the diagnosis of AML.

The appearance of hepatic AML on imaging varies widely because the relative proportion of vessels, muscle and fat varies widely from tumour to tumour. Definitive preoperative diagnosis is possible and made easier by the use of US, CT and magnetic resonance imaging (MRI). The tumour is usually hyperechoic on US, hypodense (less than –20 Housefield units) on CT and hypervascular on angiography. The fatty component that is crucial for diagnosis is detected as high signal intensity on MRI.

The fatty content of AML varies from less than 10% to more than 90% of the tumour volume. Hence, it can mimic any other tumour with fatty components. A diagnostic dilemma arises in tumours with much lower fatty content. The common differential diagnoses for such lesions that are hypervascular in an endemic region for hepatitis B are haemangiomatous and HCC. Similarly, malignancies with fatty components, e.g., HCC with fatty change, are also difficult to differentiate from AML, as exemplified by this case report. In large HCC with fatty change, the fatty component is localized and is highly echogenic on US, and as a low-density area on CT. On the contrary, small HCC (less than 3.5 cm in diameter) has diffuse fatty change that is harder to differentiate from benign lipomatous tumours.

MRI, with its multiplanar visualization and fat suppression techniques, allows better identification of fat within the tumour. Dynamic CT and MRI have been suggested as reliable methods for differentiating AML from HCC and cavernous haemangioma. From a time density/intensity study curve, AML shows a marked early and prolonged enhancement. This is attributed to the proliferation of blood vessels within the fatty component. The timing of peak enhancement of AML is intermediate between HCC, which has a very early peak, and haemangioma. The enhancement, which is similar to haemangioma, is sustained and prolonged, in contrast to that of HCC. The pattern of enhancement on dynamic visualization also helps to differentiate these lesions. Haemangiomas characteristically demonstrate nodular enhancement with “forest-fire” filling-in effect from the periphery to the centre, whilst HCCs have pseudocapsular enhancement. A combination of CT and MRI with their multi-technique modalities help make a preoperative diagnosis possible and obviates the need for needle biopsy, which can potentially cause tumour seeding.

Non-surgical treatment of AML has been advocated due to its benign nature. This is especially warranted in patients with poor liver reserves and small lesions in deep-seated regions of the liver. Nonetheless, in an endemic region for hepatitis B, one must be cautious about focal masses in the liver and in advocating a conservative approach to these lesions. Close follow-up is still necessary and recommended after a definitive diagnosis of AML.

**REFERENCES**

11. Yoshida H, Itai Y, Ohtomo K, et al. Small hepatocellular...

