A misleading hepatic tumour : epithelioid angiomyolipoma

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Abstract

Hepatic angiomyolipoma (HAML) is a rare, benign mesenchymal neoplasm composed of varying amounts of smooth muscle cells, adipose tissue, and vessels. Its morphological diversity often poses diagnostic problems. In this paper, the authors report a peculiar case of epithelioid HAML mimicking histologically hepatocellular carcinoma with focal areas resembling inflammatory pseudotumour. A 57 year-old male patient presented with abdominal pain and discomfort. Non enhanced CT scan demonstrated a heterogeneous hypodense mass located in segment II and IV of the liver. Hepatocellular carcinoma was suspected and the patient underwent left lobectomy. Histologically, the tumour was mainly composed of epithelioid cells arranged in trabeculae and sheets (50% of the tumour surface) admixed with mature fat cells (20%) and thick-walled blood vessels. Lymphocytic aggregates and clusters of foamy histiocytes were focally found in the stroma (30%). Most of the epithelioid tumour cells were immunoreactive to homatropine methylbromide 45 (HMB-45) and smooth muscle actin. Morphological pattern and immunophenotype were consistent with epithelioid HAML. (Acta gastroenterol. belg., **2012**, 75, **443-445**).

Key words : epithelioid angiomyolipoma, liver, HMB-45, immunohistochemistry.

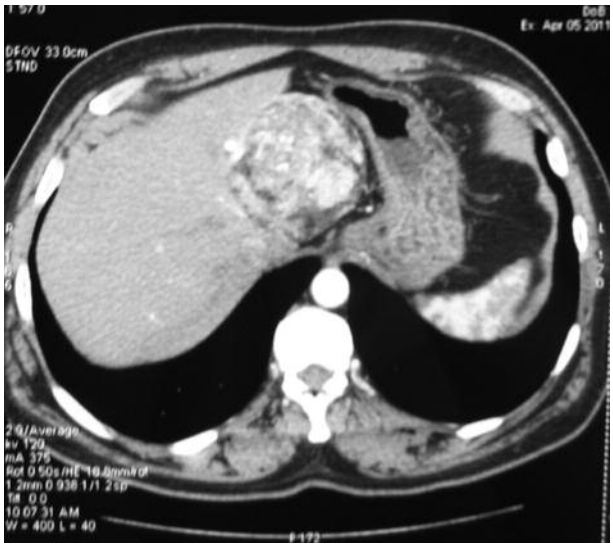

Fig. 1. — Magnetic resonance imaging demonstrating a heterogeneous and partially high intensity mass.

Introduction

Hepatic angiomyolipoma (HAML), a rare benign mesenchymal neoplasm, is a constituent of the group of perivascular epithelioid cell tumours called PEComa (1, 2). The considerable morphological heterogeneity of this tumour often poses diagnostic difficulty. In this paper, the authors report a new and peculiar case of HAML displaying an admixture of inflammatory and trabecular patterns that raised diagnostic confusion with hepatocellular carcinoma and inflammatory pseudotumour.

Clinical history

A 57-year-old previously healthy man with no stigmata of tuberous sclerosis, presented with a two-month history of abdominal pain and discomfort. Physical examination was unremarkable. His liver function tests were normal and serologic assay for viral hepatitis was negative. Abdominal ultrasonography showed a hyperechoic mass in segments II and IV of the liver. CT scan revealed a well delineated heterogeneous and hypodense liver mass in segments II and IV measuring 82 mm across. Magnetic resonance imaging (MRI) demonstrated a non-homogeneous and partially high intensity mass on both T1- and T2-weighted images (Fig. 1). Because of uncertain nature of the lesion and the possibility of malignancy, left lobectomy was performed. Grossly, the hepatic resection specimen contained a well-circumscribed soft and friable tumour measuring 8 × 7.5 × 5 cm in size with variegated appearance on cut section including yellowish fatty areas, haemorrhage and focal necrosis (Fig. 2). Histological examination of the surgically resected specimen showed that the tumour consisted of predominant epithelioid cells arranged in sheets and trabeculae (50% of the tumour surface) (Fig. 3) admixed with mature fat cells (20% of the tumour surface) and few thick-walled blood vessels. Lymphocytic aggregates as well as clusters of foamy histiocytes were focally found in the stroma accounting for approximately 30% of the tumour surface (Figs. 4a and 4b). No foci of extramedullary hematopoiesis were identified. Immunohistochemical studies revealed intense positive staining of epithelioid

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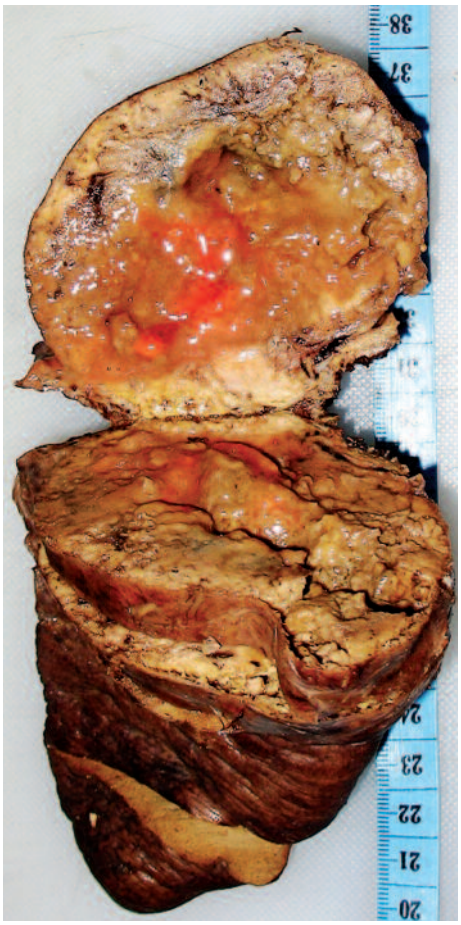

Fig. 2. — Gross findings : circumscribed tumour with variegated appearance on cut section including yellowish fatty areas, haemorrhage and focal necrosis.

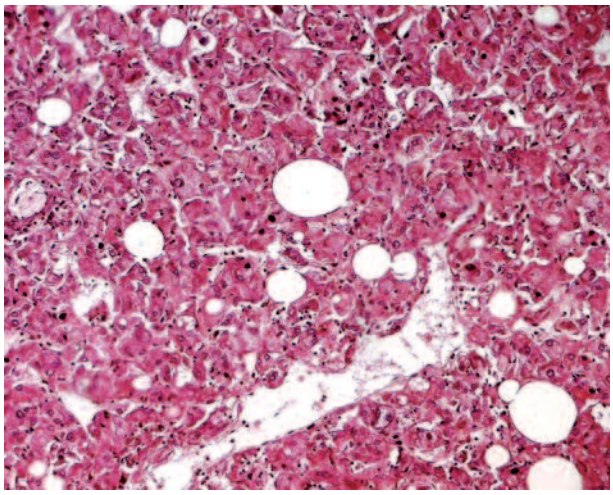

Fig. 3. — Polygonal epithelioid cells are arranged in solid sheets and trabeculae admixed with a few adipocytes. (Hematoxylin & eosin, original magnification ×40).

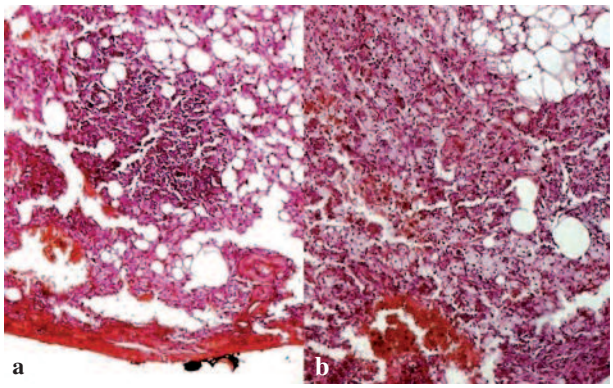

Fig. 4a-b. — Lymphocytic aggregates and clusters of foamy histiocytes were identified in the stroma (Hematoxylin & eosin, original magnification ×25).

tumour cells for HMB-45 and actin (Fig. 5) but they were negative for cytokeratin, desmin and melan A. The final histological diagnosis was epithelioid hepatic angiomyolipoma. Postoperative course was uneventful and the patient is still at follow-up.

Discussion

Since its initial description by *Ishak* in 1976, more than 200 cases of HAML have been reported in the English literature (3,4). Although imaging studies may help in the diagnosis, the definitive diagnostic tool of HAML remains the histological examination of the surgically resected lesion coupled with immunohistochemical stains. However, because of the morphological pleomorphism of HAML, histologic diagnosis of this entity may be challenging in some cases. In fact, misdiagnosis of this tumour has been focused on in various articles (5). Hepatic angiomyolipomas may demonstrate a variety of histologic patterns depending on the variable proportion of adipose tissue, myoid cells and vasculature. The smooth muscle is the only specific diagnostic component, comprising mainly epithelioid cells in sheets and, to a lesser extent, spindle-shaped cells arranged in bundles (6). Tumours composed predominantly of epithelioid cells have been referred to as epithelioid angiomyolipoma as it was the case in our patient. Extramedullary haematopoiesis is a frequent and characteristic feature of hepatic tumours but was not identified in our case. According to the line of differentiation and the predominance of tissue components, the tumours are subcategorized into mixed, lipomatous (≥ 70% fat), myomatous (≤ 10% fat), and angiomatous types. The mixed type is the most common (5,6). The other histologic patterns described in the literature include trabecular, pelioid, and inflammatory pattern. Of these, inflammatory or pelioid pattern usually presents as a focal finding within the tumour but very rarely they may become the predominant pattern creating great diagnostic confusion with other tumours (7). In our case, the tumour showed an admixture of several patterns in variable proportions including the inflammatory (30%), the trabecular (50%) and the mixed patterns (20%). Immunohistochemically,

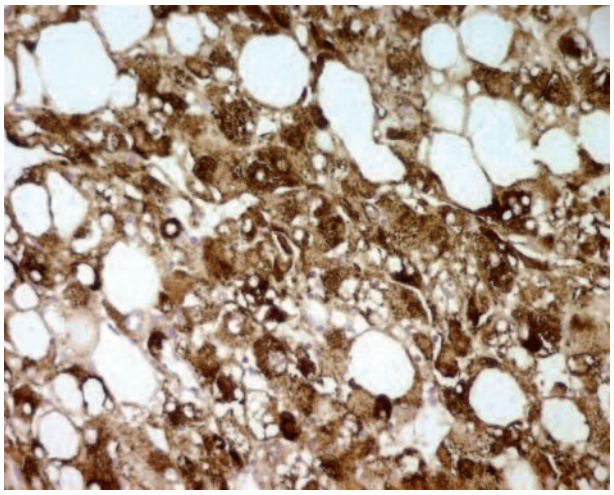

Fig. 5. — Tumour cells demonstrating HMB-45 immunoreactivity (Immunohistochemistry, original magnification ×40).

the defining myoid cells are consistently positive for HMB-45 and other melagonesis markers. S-100 protein, CD 117, actin, desmin and vimentin expression is variable (6). There are many entities in the differential diagnosis of HAML, and the distinction can become problematic as the adipose component decreases. Some of the lesions considered in the differential diagnosis include hepatocellular carcinoma, hepatic adenoma, leiomyoma, hepatoblastoma (especially when hematopoietic elements are present), melanoma, angiosarcoma, and gastrointestinal stromal tumors. Many features of HAML can lead to a misdiagnosis of hepatocellular carcinoma, such as polygonal cells in a trabecular arrangement as it was the case in our patient, nuclear pleomorphism, eosinophilic globules, and tumor necrosis (5,6). However, hepatocellular carcinomas usually express Hep par 1 and do not express smooth muscle or melanoma antigens. Inflammatory HAML may be confused with inflammatory pseudotumour and inflammatory myofibroblastic tumour (7). In our case, the tumour displayed a prominent inflammatory background focally mimicking inflammatory pseudotumour. The treatment of HAML is hepatectomy for large tumours and conservative follow-up for small ones (8). Most HAML behave in a benign fashion although malignant HAML has been reported in the literature (9). Because of the small number of cases, the clinical features of this tumour are not fully known yet. Therefore, as recommended in previous reports, long-term follow up is necessary even after curative surgical treatment (10).

In conclusion, pathologists should be aware of the existence of the several subtypes of angiomyolipomas, especially when these tumours occur in the liver. Reactivity for HMB- 45 is the best available marker for the diagnosis of HAML and can help the pathologist to avoid a regrettable misdiagnosis.

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