Fat-deficient hepatic angiomyolipoma: A radiological challenge

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Case report

A 25-years-old woman presents with progressive onset of a right-upper quadrant and epigastric abdominal pain associated with a weight loss.

At physical examination, a large abdominal mass is palpable in the epigastric region. Neither jaundice nor temperature is noticed. Standard biological tests are normal. Hepatitis B antigen and anti-hepatitis C antibody are negative and the serum level of α-fetoprotein is within the normal range.

Computed Tomography (CT) scan of the abdomen (Fig. 1) displays a voluminous (20 cm), heterogeneous but well-delimited mass arising from the right hepatic lobe. The lesion contains numerous ectasic vessels and nodular areas of arterial enhancement associated with delayed venous washout are depicted. There is no sign of macroscopic vascular involvement.

Magnetic Resonance Imaging of the liver (Fig. 2) does not display any fat component within the mass; therefore, the spectrum of hepatic fat-containing lesions is not considered in this case.

Given these features, the diagnosis of fibrolamellar carcinoma is considered. Primary histological findings on fine needle core biopsies are suggestive of hepatocellular carcinoma (HCC), with potential areas of fibrolamellar organization. Urgent liver transplantation is decided, based on the surgical challenge for a complete resection and the absence of vascular involvement or extra-hepatic spread.

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2211-5684/$ — see front matter © 2013 Éditions françaises de radiologie. Published by Elsevier Masson SAS. All rights reserved.
http://dx.doi.org/10.1016/j.diii.2013.03.018
Figure 1. Axial iodine-enhanced CT images of the liver at arterial phase (a) and portal venous phase (b) display a large hepatic mass with heterogeneous enhancement. Nodular areas of arterial enhancement and delayed wash-out are clearly demonstrated.

Figure 2. Axial in-phase (a) and out-of-phase (b) T1-weighted MRI images demonstrate the absence of fat component within the mass.

Macroscopic findings (Fig. 3) display a friable, well-delimited, partially encapsulated tumor. Its cut surface is reddish. Neither macroscopic nor microscopic fat are displayed. Microscopic findings and immunohistochemical analysis using anti-HMB45 antibodies (Figs. 4 and 5) definitively establish the diagnosis of a benign, fat-deficient epithelioid angiomyolipoma.

Discussion

Angiomyolipoma is a benign mesenchymal tumor composed of smooth muscle cells, thick-walled vessels and mature adipose tissue in various amounts that mostly affects the kidneys, especially in adults with tuberous sclerosis.

The hepatic location is rare, with about 200 cases reported in the English literature in 2003 [1]. Depending on the dominant cell type, hepatic angiomyolipoma (HAML) can be subcategorized into epithelioid, spindle and intermediate forms [2]. Very rare cases of malignant transformation have been described.

It has been established that fat-deficient HALM, as in our case, are difficult to differentiate from HCC in non-cirrhotic liver, as most tumors present as a well defined, hypervascular enhancing mass on arterial phase followed by washout...
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Fat-deficient of present because hepatic could pattern, and because of the absence of macroscopic fat. One of the major imaging finding for differentiating both entities could be the tumor capsule, which is absent in HAML, and present in 60–82% of HCCs [3].

Even if the diagnosis may be established by fine needle biopsy, a diagnostic dilemma can be posed to pathologists because of the overlapping histological features with other hepatic neoplasms, especially HCC [4], as illustrated in our case. Immunochemistry using HMB-45 is therefore a fundamental tool in establishing the final diagnosis [5].

Surgical management may be considered in symptomatic patients [6], in cases when malignancy can not be ruled out or in cases of rapidly-growing tumor [1]. Nevertheless, liver transplantation is not the modality of choice and could have been avoided in our patient if adequate immunohistochemical study had been performed.

Conclusion

HAML should be considered in the presence of a fat-containing lesion, but the diagnosis may be all the more difficult to evoke that the lesion lacks the fatty component, as illustrated here.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References