

RADIOLOGIC PATHOLOGIC CORRELATION / *Gastrointestinal imaging*

## Radiologic-pathologic correlation in liver angiomyolipoma in a 68-year-old woman

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### KEYWORDS

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PEComa

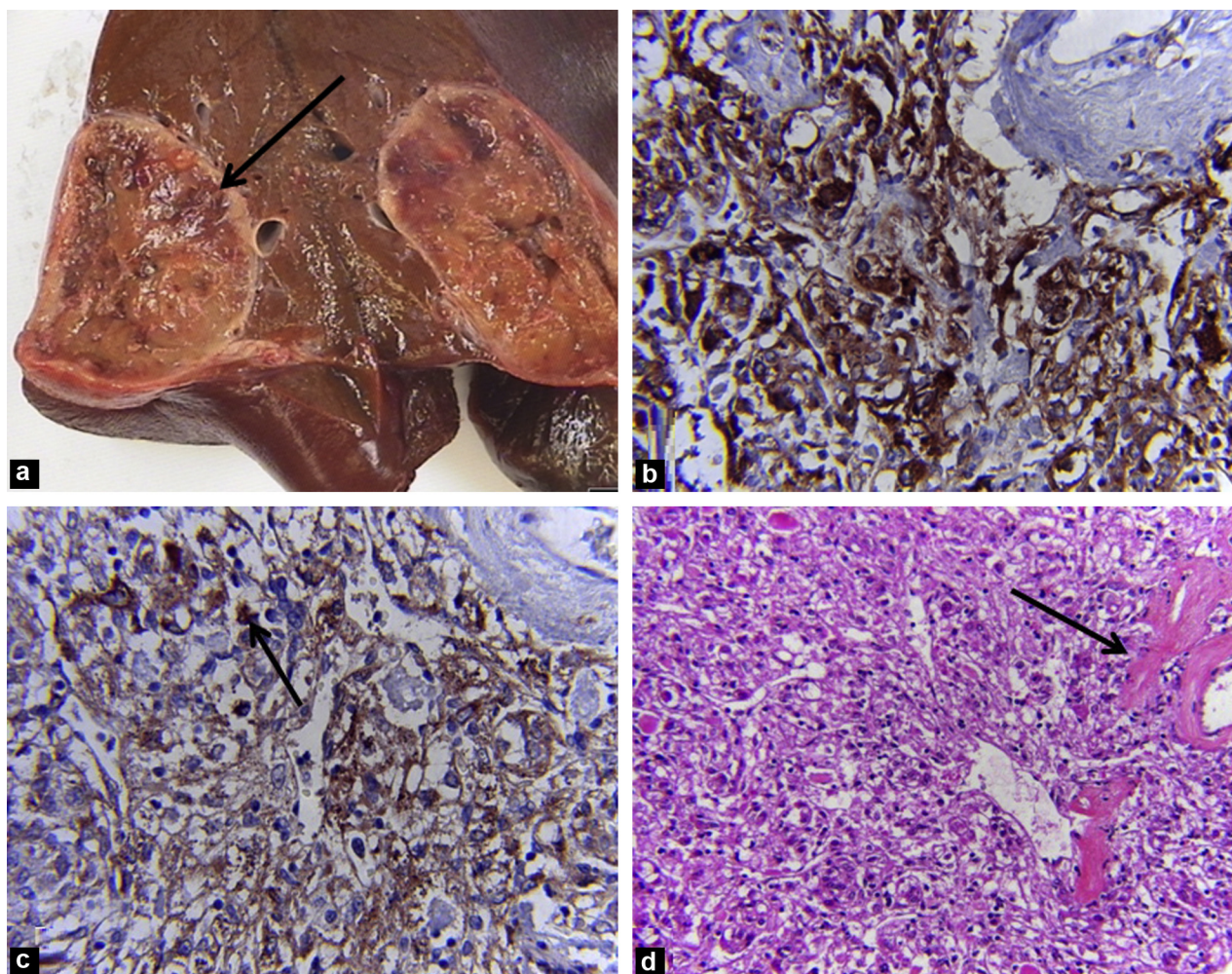
### Case presentation

A 68-year-old woman with no remarkable past history, had a chest, abdominal and pelvic CT to search for a potential cause of rhizomelic pseudopolyarthrititis. A focal liver lesion was found in the subcapsular area of segment V of the liver. The lesion had a largest axial diameter of 48 mm. The lesion was difficult to characterize because of limited acquisition in the portal phase. On MR imaging, the liver lesion had regular contours with increased heterogeneous hyperintense signal on T2-weighted images, reduced T1 signal and intense heterogeneous early gadolinium uptake with no visible washout. The in-phase and out-of-phase sequences did not show fatty component.

The patient was asymptomatic and her liver profile was normal. As a result of the atypical imaging presentation, she had percutaneous liver biopsy from which a histological diagnosis of Edmonson Grade II hepatocellular carcinoma (HCC) was made. The lesion was resected with a bisegmentectomy. Macroscopic examination showed a well delineated, encapsulated, soft homogenous and brownish-colored subcapsular tumor. On microscopic examination tumor proliferation was present with three different components, including epithelioid smooth muscle cells, adipocytes and vascular structures with hyalinized walls. The largest component was made of smooth muscle cells. On immunohistochemistry, the tumor cells intensely and diffusely expressed melanocyte antigens HMB45, Melan-A and, to a lesser extent, smooth muscle antigens and smooth muscle actin antigen. The liver parenchyma distant to the lesion was healthy and the final diagnosis was that of a hepatic angiomyolipoma developing in a healthy liver (Figs. 1–3).

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**Figure 1.** a: macroscopic findings: well delineated, encapsulated (arrow), brownish subcapsular tumor measuring  $5 \times 5 \times 3$  cm; b: microscopy  $\times 40$ , smooth muscle cells labeled in brown with Melan A; c: microscopy  $\times 40$ , cytoplasmic labeling with HMB 45 (arrow); d: microscopy  $\times 20$ : dystrophic vascular walls (arrow).

## Discussion

Many focal hepatic lesions are discovered incidentally in a healthy liver. Although CT and ultrasound cannot provide a definite diagnosis, MR imaging often has a pivotal role prior to any further invasive diagnostic procedure.

Many hypervascular benign and malignant tumors can develop in a healthy liver. These include HCC, which is rare but not exceptional, hypervascular metastases from primary endocrine, renal or thyroid tumors and hepatocholangiocarcinoma. Benign lesions include adenomas, focal nodular hyperplasia (FNH), rapid flow cavernous hemangiomas or more rarely epithelioid hemangioendothelioma.

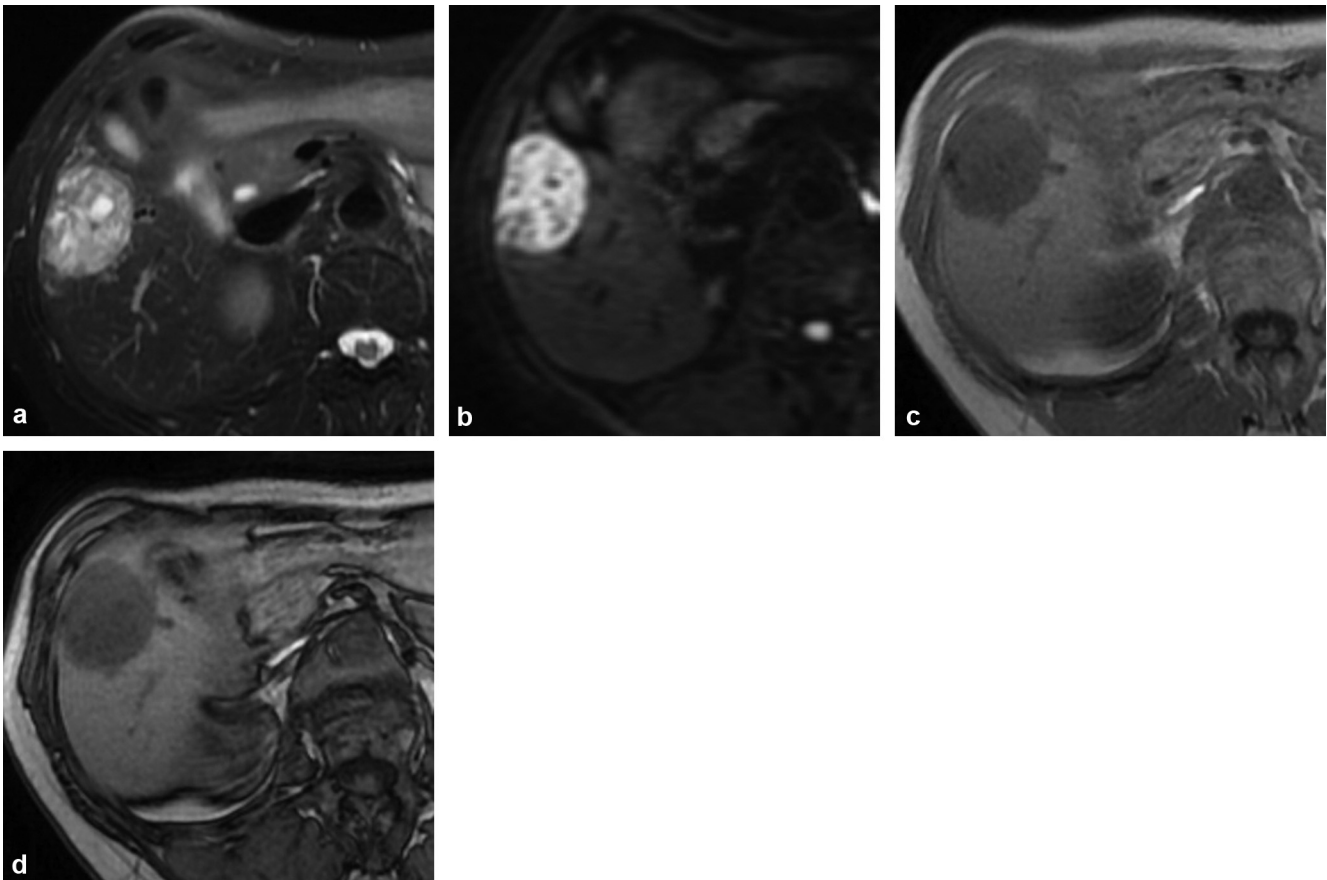
In our case, some of the lesions, particular FNH, angioma and endocrine tumor metastases could easily be excluded on imaging. The other diagnoses could not be formally excluded and a percutaneous liver biopsy was needed.

Because of the small amount of tissue sample and investigation for cells with morphological features similar to those of lymphocytes, the diagnosis of hepatic angiomyolipoma is difficult from a liver biopsy alone. In our case, the diagnosis of HCC was made initially.

From a histological point of view, hepatic angiomyolipoma is a mesenchymal tumor consisting of adipose tissue, smooth muscle, and blood vessels that occasionally have dystrophic walls. Hepatic angiomyolipoma is usually asymptomatic and can be associated with Bourneville tuberous sclerosis in 6 to 10% of the cases [1].

Hepatic angiomyolipoma belongs to the group of PEComas (perivascular epithelioid cell tumors), which is a newly identified group that contains tumors with different histological presentation but with a common histochemical feature of co-expression of the melanin markers HMB 45, Melan A and smooth muscle associated with negative epithelial cell markers too [2].

Large variations in the proportion of each of the three components may be present, thus, defining different histological subtypes. The most common form is that of the mixed angiomyolipoma although angiomyolipoma with a high fat component (containing more than 70% fat), a high smooth muscle component (containing less than 10% fat) or with an angiomatous component are seen. Focal areas of hemopoiesis within the tumor, which were not observed in our case, are common and are a clue to the diagnosis [3].



**Figure 2.** a: fast spin echo (FSE) T2 axial section; b: diffusion-weighted axial section; c: in phase T1-weighted image in the axial planesection by MRI; d: out of phase T1-weighted MR image in the axial plane.

These different histological subtypes are defined by variable proportions of each component and result in a broad spectrum of radiological appearances and diagnostic difficulties with imaging [4].

Histologically, the main difficulty is due to the smooth muscle and fatty components, which are a cause of erroneous diagnosis of a hepatocyte tumor (particularly HCC) from cytology, biopsy specimen (as in our case) and sometimes even from a surgical specimen. These two components, derived from epithelioid cells, primary mesenchymal cells, are able to differentiate [3].

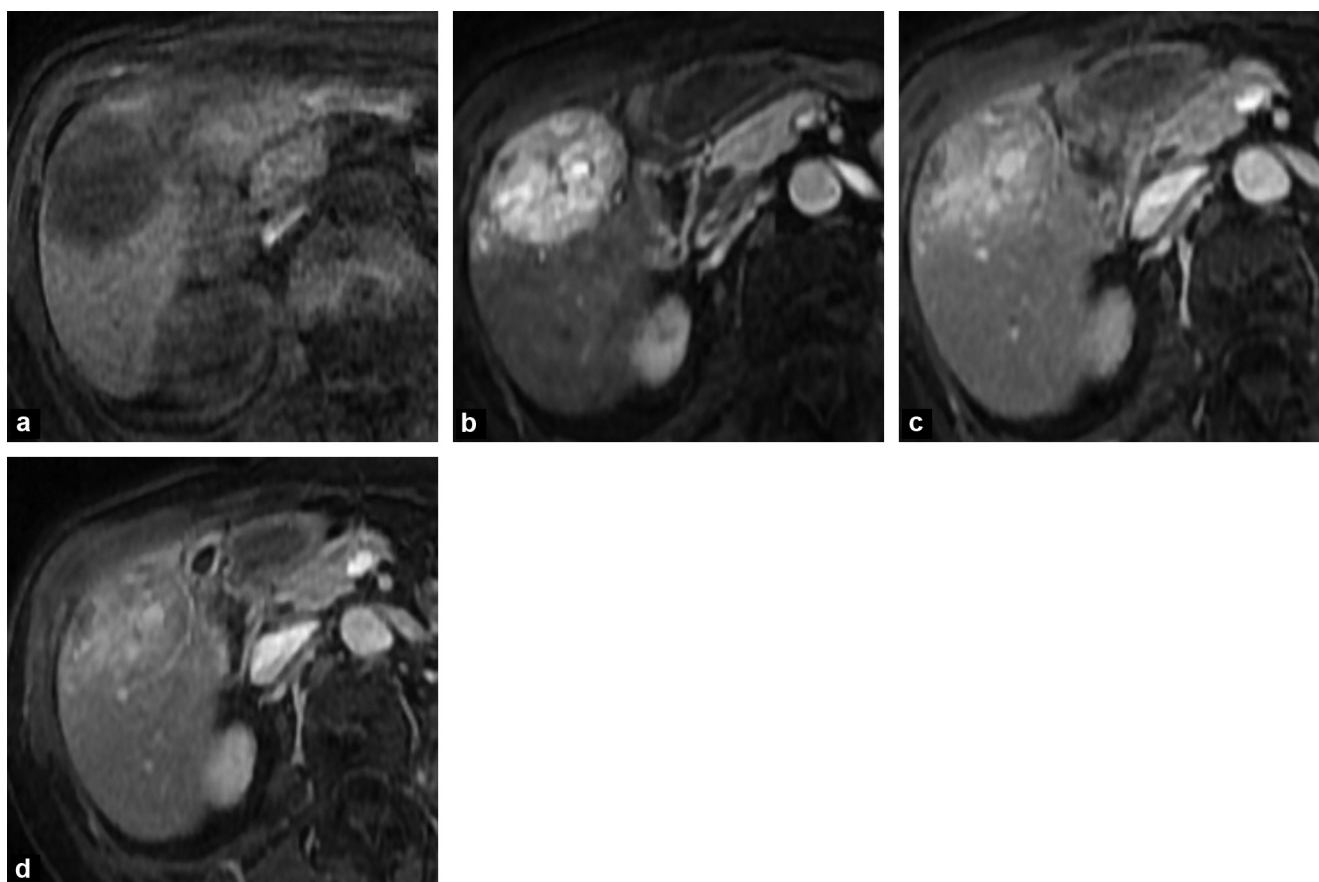
In hepatic angiomyolipoma, the smooth muscle cells are very often epithelioid in appearance with irregular nuclei and can be confused with malignant hepatocytes. The fatty component in the lesion is also difficult to identify as the adipocytes can be confused with steatotic hepatocytes (macrovascular steatosis). All of the histological variations, therefore, lead to confusion and any hypervascularized liver tumor with an unusual epithelioid fatty appearance on histology should be subject to immunohistochemical analysis with measurement of hepatocyte markers (anti-hepatocyte), adipocyte (PS 100) and PEComas (HMB45, Melan A, angiomyolipome [AML]) markers.

When the angiomyolipoma contains a large fatty component (which may range from 10 to 90%), it may be identified by imaging. It is better seen on T1-weighted MR sequences as

an area of increased signal intensity. The same area shows a marked drop in signal intensity when fat suppression is used because of abnormally high accumulation of triglycerides in the hepatocyte cellular cytoplasm and the drop in signal intensity seen on the "out of phase" sequence is due to the presence of water and triglycerides in the same voxels [5]. This contingent should be differentiated from a steatotic contingent of cells seen in the far more common hepatocyte tumors, adenoma, focal nodular hyperplasia and HCC.

The presence of an adipocyte contingent is not specific for this diagnosis although it limits the range of diagnoses. Lipomas are made up of adipocytes although these are not vascularized; these are also very rare. The third diagnosis to consider is HCC with a fatty contingent.

The outcome of these lesions is poorly understood because of their rarity. Whilst no cases of degeneration of a benign hepatic angiomyolipoma have been reported in the literature, only a few cases of a malignant angiomyolipoma have been described [4,6]. The treatment of this lesion is still controversial. Some authors propose surgical resection when the following criteria are met: symptomatic patients, size more than 6 cm, extra-hepatic growth with risk of rupture, increase in size or lack of histological diagnostic certainty [4]. Angiomyolipomas require close follow-up after diagnosis. To date, only one recurrence has been reported 6 years after resection of the initial lesion.



**Figure 3.** a: axial section MRI, liver acquisition with volume acceleration (LAVA) without injection; b: axial section MRI, gadolinium injection arterial phase: hypervascular lesion; c: axial section MRI with gadolinium injection portal phase: no washout; d: axial phase section MRI, gadolinium injection late phase: no washout.

## Conclusion

Hepatic angiomyolipoma is a rare liver tumor of which diagnosis is made difficult by the large variations in the different components, leading to large variations in imaging presentation. The adipocyte component must always be searched for using T1-weighted MR sequences, using fat-saturated sequences. Histologically, the tumor is difficult to diagnose with cellular atypia, particularly in smooth muscle cells, which may suggest a diagnosis of a hepatocyte tumor. If a hypervascular hepatic lesion containing an atypical fatty contingent is found, immunohistochemistry with hepatocyte (anti-hepatocyte), adipocyte (PS100) and a PEComas (HMB45, Melan A, AML) markers is essential to make the diagnosis, to avoid diagnostic errors and confirm the diagnosis of a PEComa.

## Disclosure of interest

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

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