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Epithelioid angiomyolipoma of the liver: an incidental diagnosis

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Dear Editor,

We present the case of a 76-year-old woman with a history of dyslipidaemia, hypertension, diverticulosis and stable angina. She was studied after an episode of acute pancreatitis with an acalculous gallbladder, with no symptoms and an unremarkable physical examination. Laboratory tests with tumor markers showed no alterations. An angio-CT scan was requested with the finding of a hypercaptant nodule in the arterial phase in segment V measuring 2.8x2 cm. The study was completed with upper and lower gastrointestinal endoscopy, finding a lipoma in the rectosigmoid junction measuring 2.5 cm and a known diverticulosis. The MRI (Fig.1) showed a nodular image in the medial area of segment V measuring 2.8 cm, with an intermediate signal in T2 and low signal in T1 with diffusion restriction.

A liver biopsy was performed by core needle biopsy, where, despite a limited sample, it suggested the possibility of angiomyolipoma in the presence of a positive HMB45. However, given the scarcity of the sample, it was decided to repeat the biopsy, which on this occasion did not reach the lesion.

Surgical intervention was decided, with excision of the tumor and laparoscopic cholecystectomy. The histopathological study (Fig.1) revealed an epithelioid angiomyolipoma variant, positive for Melanin A, HMB45 and smooth muscle actin.

The patient was discharged on the third postoperative day without any remarkable complications.

Angiomyolipomas are rare neoplasms of mesenchymal origin, derived from perivascular epithelioid cells. In the liver it is a very infrequent neoformation, but in the kidney, it represents 3% of the renal masses[1] and is associated with tuberous sclerosis in up to 20% of patients, hepatic angiomyolipomas are associated with tuberous sclerosis in only 6% of patients[2].

In most cases this is an incidental finding, with abdominal discomfort being the most frequent symptom[3].

Due to their varying composition, particularly their frequent paucity of fat, diagnosis can be challenging. The gold standard for the diagnosis is histologic examination coupled with an immunohistochemical study[2], expressing positivity for HMB-45, Melan-A and smooth muscle actin[4].

The main differential diagnosis of epithelioid angiomyolipoma is hepatocellular carcinoma or hepatocellular adenoma[2].

On an exceptional basis, it may show a malignant behavior including growth, recurrence after surgical resection and metastasis. Malignant transformation is thought to occur mostly in the epithelioid type[3]. The best treatment option remains controversial, many researchers advocate surgical resection over a non-surgical approach consisting of regular follow-up3. Biopsy is indicated when imaging is inconclusive and resection should be considered in case of symptoms, growth in follow up, inconclusive biopsy, atypical epithelioid patterns or high proliferation activity[3].

Follow-up after surgical resection is necessary due to its malignant potential[5].

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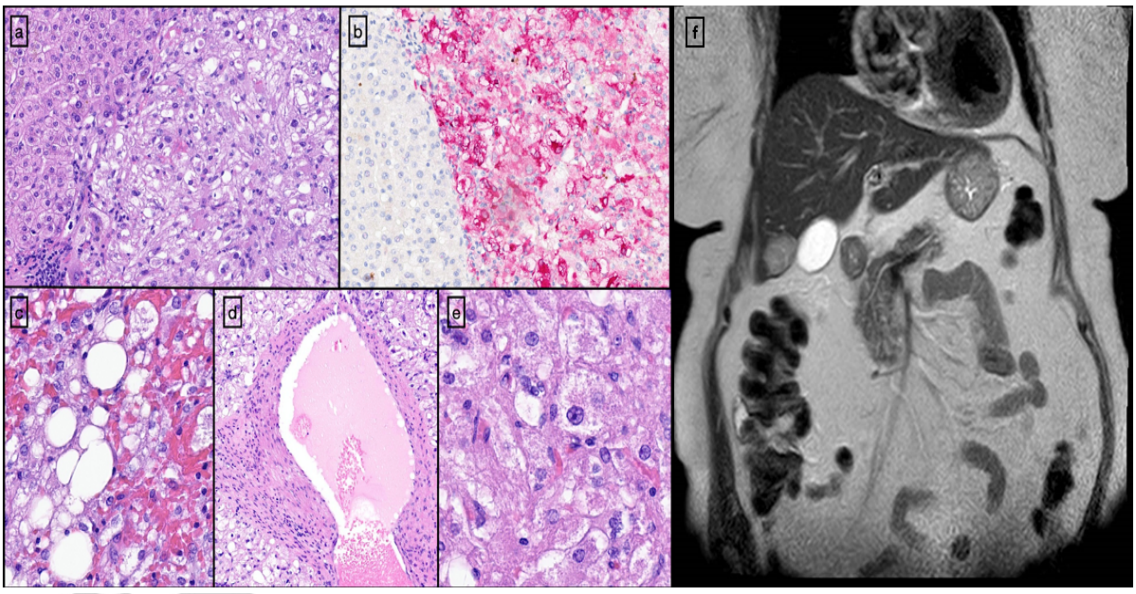

Fig 1 a: The neoplasm was demarcated from the surrounding liver tissues (left) with relative clear boundary. b: HMB-45 stain (melanocytic marker) strongly positive in epithelioid cells (right). c: Fat vacuoles were variably scattered through the tumor. d: Occasional tortuous thick-walled vessels could be identified. See the intimate relationship of the eosinophilic neoplastic cells with a large vessel. e: The myoid cells (specific and diagnostic component in Angiomyolipoma) were predominant epithelioid

in shape with large nuclei, prominent nucleoli and eosinophilic or clear cytoplasm. f: Coronal section MRI: nodular image in the medial area of segment V, 28 mm, with intermediate signal in T2 and low in T1 with diffusion restriction and no signs of aggressiveness.