Rare disease

Sporadic exophytic hepatic angiomyolipoma

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Summary

Angiomyolipomas are rare mesenchymal tumours consisting of smooth muscle, blood vessels and adipocytes in variable proportions. These tumours are more usually seen in kidneys (incidence 0.3–3%), and the extrarenal location is rare. Since its initial description by Ishak in 1976, only about 200 cases of hepatic angiomyolipomas have been reported. We describe the case of a 59-year-old woman, without tuberous sclerosis, with a 6 cm tumour in hepatic segments II and III, gallbladder with parietal calcification and lithiasis. She was proposed to bisegmentectomy (II and III) and cholecystectomy, which were performed without complications. The pathological examination confirmed the diagnosis of hepatic angiomyolipoma (positive for HMB45 and focally positive for HHF35) and chronic cholecystitis. Although typically benign, the emergence of cases with malignant behaviour—recurrence or distant metastasis—led to a change in attitude towards these lesions, from clinical surveillance to surgical excision, with few exceptions.

BACKGROUND

Hepatic angiomyolipoma (HAML) is a rare hepatic mesenchymal tumour that belongs to a family of tumour called 'PEComa'.^{1–3} Angiomyolipomas are more usually seen in kidneys (incidence 0.3–3%),⁴ and the extrarenal location is extremely rare.⁵

Since its first description by Ishak,⁶ approximately 200 cases of HAML have been reported in the literature,⁷ with an increasing frequency in recent years due to the development and popularity of diagnostic imaging techniques.⁸

Histologically, HAMLs are composed by a heterogeneous mixture of blood vessels, smooth muscle and adipose cells,⁹ of varying proportions, leading to various radiological features.¹⁰ Consequently, preoperative diagnosis is challenging, especially with regard to the distinction from hepatocellular carcinoma.¹¹

CASE PRESENTATION

We present the case of a 59-year-old Portuguese woman with diabetes mellitus, obesity (body mass index=35) and

Figure 1 Abdominal CT: arrow, gallbladder with parietal calcification; brackets, 6 cm tumour in liver segments II and III.

Figure 2 Intraoperative photos. 1, gallbladder; 2, hepatic angiomyolipoma and 3, liver surface.

osteoporosis, without tuberous sclerosis. She had been referred by her physician because of a right hypochondrium discomfort, with an ultrasound revealing vesicular lithiasis and a 6 cm heterogeneous hepatic tumour in segments II and III.

A CT was performed diagnosing vesicular lithiasis with calcification of the gallbladder wall, and a 6 cm well-defined heterogeneous hepatic tumour consistent with HAML in segments II and III (figure 1). Analytically, tumour markers and hepatitis virus were negative.

She was proposed to bisegmentectomy II and III and cholecystectomy, which were performed with no complications (figures 2 and 3), and was discharged on the sixth postoperative day.

Histopathological examination confirmed the diagnosis of HAML (positive for HMB45 and focally positive for HHF35) and chronic cholecystitis with bile crystals on the gallbladder wall (figures 4 and 5).

OUTCOME AND FOLLOW-UP

Follow-up without evidence of disease (3 years).

Figure 3 Photograph of the liver resected.

DISCUSSION

The aetiology and histogenesis of HAML remains unknown, but it is known that they are more frequent in women, and may arise sporadically or in the context of tuberous sclerosis.¹¹

It is also known that preoperative imaging diagnostic rate has been reported to be less than 50%. Even the post-operative pathological diagnosis has been mistaken as hepatocellular carcinoma. In the past, this tumour had been considered an entirely benign and slow-growing lesion without risk of malignant transformation. However, since 2000, several reports have revealed that this kind of tumour can be malignant with evidence of recurrence or distant metastasis.^{12 13} Therefore, the proper treatment of HAML has become controversial. Retrospective studies suggest that conservative management with close follow-up is only advised in asymptomatic patients with good compliance and non-hepatitis

Figure 4 Neoplasm composed by adipose tissue and epithelioid cells with eosinophilic cytoplasm and large pleomorphic nuclei, with prominent nucleoli.

Figure 5 Cytoplasmic immunoreactivity of epithelioid cells with HMB45; focal positivity with desmin and HHF35.

virus carrier, with tumour size inferior than 5 cm and with a fine-needle aspiration biopsy proving HAML.⁷

In this case, the CT scan already suggested HAML, distinguishing it from other hepatic tumours. Having that suspicion, a decision between clinical surveillance and surgical excision had to be made. The tumour size was greater than 5 cm, lowering the accuracy of a biopsy, and increasing the risk of malignancy. The patient complaints could be affordable to the gallbladder lithiasis, but could also be secondary to growing of the tumour. Hepatitis virus were negative, lowering the risk of hepatocellular carcinoma. Since cholecystectomy had to be performed, and the removal of the tumour consisted in a relatively low morbidity procedure (segments II and III bisegmentectomy), both were promptly proposed to the patient, who agreed to be submitted to both procedures. Biopsy was not performed because its result would not change the decision already taken. The anatomic-pathologist examination confirmed the diagnosis of HAML (positive for HMB45 and focally positive for HHF35) and chronic cholecystitis with sequestration of bile crystals on the gallbladder wall.

Learning points

- ▶ Hepatic angiomyolipomas are rare and of unknown aetiology.
- ▶ Preoperative imaging diagnostic rate is low.
- ▶ Although typically benign, the emergence of cases with malignant behaviour led to a change in attitude towards these lesions, from the clinical surveillance to the surgical excision as treatment of choice, with few exceptions.

Competing interests None.

Patient consent Obtained.

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