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GI IMAGE

Primary Castleman's Disease of the Liver

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Abstract Castleman's disease is a rare lymphoproliferative disorder that may affect the liver as part of mutlicentric disease or, rarely, as unicentric hepatic disease. Primary hepatic disease is difficult to diagnose but demonstrates diffusion restriction on magnetic resonance scan and can be treated curatively with hepatic resection.

Keywords Castleman's disease · Hepatectomy · Diffusion restricted magnetic resonance

Case Presentation

A 64-year-old female was noted to have an arterially enhancing segment IVa liver lesion on follow-up CT for previous aortic aneurysm repair (Fig. 1a). A subsequent ultrasound demonstrated an indeterminant 14-mm hypoechoic focus in a steatotic liver with areas of focal fatty sparing (Fig. 1b), and the lesion was further evaluated with MRI of the liver. This confirmed the presence of a hypervascular segment IVa lesion on T_2 scans with appearances suggesting a hypervascular metastasis (Fig. 1c). The lesion was noted to diffusion-restrict on contrast MRI, indicating a highly cellular mass suspicious for a hypervascular primary tumour or a metastasis (Fig. 1d). Of note, there was no past history of neuroendocrine carcinoma or melanoma. Tumour markers including CEA, CA19-9, CA 125, chromogranin A and alpha fetoprotein were all within the normal range.

On the basis of these findings, the patient's radiology was reviewed in a multidisciplinary meeting, and the lesion was resected with a non-anatomical liver resection. Intra-operatively, the lesion was not palpable; however, it did appear as a hypoechoic mass on the ultrasound. There was no evidence of extrahepatic lymphadenopathy. Histopathology demonstrated a 14-mm lesion with expansion of the portal tracts by a lymphoid infiltrate and hyalinised vessels consistent with the hyaline vascular variant of Castleman's disease (Fig. 2). Following resection, the patient was discharged on day 4 and remains well with follow-up imaging demonstrating no new liver lesions or lymphadenopathy.

Discussion

Castleman's disease is a rare lymphoproliferative disorder.[1] Most commonly seen in the neck and mediastinum [1] although it may also occur in the abdomen, axilla and retroperitoneum. Primary hepatic Castleman's disease is very rare with less than 20 cases reported.[1] Castleman's disease may present as a localised unicentric form and the multifocal form, and it can also be classified histologically as plasma cell, hyaline variant and mixed plasmablastic.[2]

The hyaline variant occurs in 90 % of cases and is also usually asymptomatic and histologically demonstrates increased numbers of small, hyalinised blood vessels within and between follicles.[1] Small lymphocytes of the mantle zones are arranged in concentric circles around the germinal

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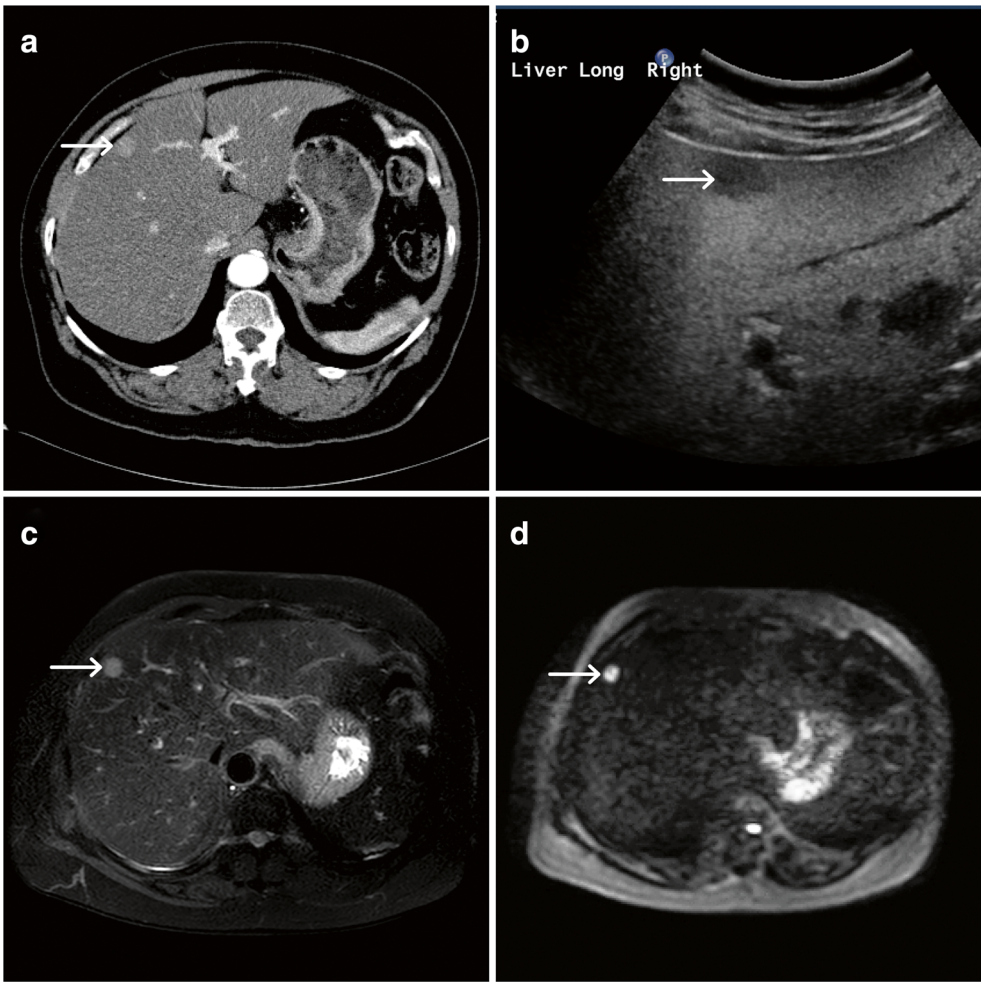

Fig. 1 **a** Portal venous CT scan (*left*) demonstrating a homogenous hypervascular lesion in segment IVa (*arrow*). **b** Trans-abdominal ultrasound confirming the presence of a solid hypodense lesion in segment 4 (*arrow*) of a steatotic liver. **c** T_2 weighted MRI showing a hypervascular mass in segment IVa (*arrow*). **d** Diffusion restricted MRI demonstrating diffusion restriction (*arrow*)

centre [2] (onion skinning), and the follicles may be radially penetrated by a hyalinised blood vessel giving a lollipop appearance.[2]

The plasma cell variant which is commonly seen in multicentric forms is more aggressive,[1] and is usually idiopathic but may be associated with HHV-8 and presents with generalised lymphadenopathy, anaemia, hepatosplenomegaly and chills.[1] The management of the unicentric form is resection, as in this case, while multicentric disease is routinely managed with immunosuppression.[1]

This case demonstrates the importance of diffusion restriction as an MRI finding indicative of malignancy. Diffusion restriction assesses the Brownian motion of water molecules within a magnetic field and, when diffusion restriction is present, suggests a highly cellular soft tissue lesion.[3] This finding is important in assessing both primary and metastatic hepatic malignancies. In this case, the presence of diffusion restriction within the incidentally noted and asymptomatic, hypervascular segment IVa mass was a key determinant in the final multidisciplinary recommendation for resection and

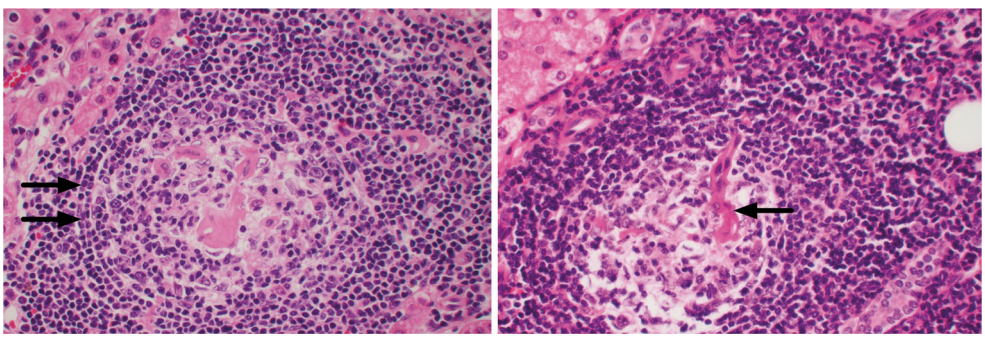

Fig. 2 The hyaline vascular variant of hepatic Castleman's disease demonstrating onion-skinning (*left*: *arrows*), and radial penetration of a hyalinised blood vessel into a follicle (*right*: *arrow*). Haematoxylin and eosin ×500 magnification

ensured that this rare primary hepatic condition was definitely treated.

Authors' Contributions All four listed authors contributed to the conception, analysis and drafting of this manuscript.

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