Hepatic angiosarcoma: A suggestive pattern of enhancement on dynamic MR imaging

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Primary hepatic tumors can arise from different components of the liver such as hepatocytes, bile duct epithelia, neuroendocrine cells, and mesenchymal cells. Despite its rarity, hepatic angiosarcoma (HA) is the most common malignant mesenchymal tumor of the liver [1]. HA usually develops in the absence of an underlying or pre-existing liver disease and its pathogenesis remains unknown. In most cases, no obvious risk factors can be identified [2].

Radiologically, hepatic angiosarcomas may have variable appearances, so that a definitive diagnosis requires histopathological examination [3]. HA may present as different growth patterns such as multiples nodules, dominant masses or rarely, diffuse infiltrating micronodular tumor [2]. On computed tomography (CT) examination, nodules are predominantly hypoattenuating, potentially mixed with some hypoattenuating areas caused by intratumoral variations like fresh or hemorrhage [4]. However, studies describing the morphologic features or enhancement pattern of HA on MRI are scarce. We report herein the MRI features of hepatic angiosarcoma.

Case report

A sixty-two-year-old Caucasian woman was admitted to our hospital because of weight loss, fatigue and jaundice. She had a previous history of strong alcohol intake and hepatitis B virus infection. Clinical examination showed abdominal distension with ascites, collateral venous circulation and stellar angiomata without encephalopathy or focal neurological abnormalities. The laboratory data at admission included: total bilirubin, 63 mmol/dL (normal: 3–18 mmol/dL); albumin, 35 g/dL, prothrombin time, 65%. The serum alpha fetoprotein level was normal.

Abdominal ultrasonography revealed hepatomegaly with presence of multiple, large heterogeneous nodules within both lobes of the liver. MRI examination was subsequently performed for further characterization of the liver lesions on a 1.5 Tesla closed magnet system (Achieva, Philips Medical Systems, Best, The Netherlands) with a dedicated abdominal phased array coil for signal reception.

MR images demonstrated a multifocal macronodular infiltration of both hepatic lobes. The tumors showed low signal intensity on T1-weighted images and heterogeneous, and became markedly hyperintense on T2-weighted MR images. Some of them, particularly those with small diameter, did not enhance after contrast administration or displayed a rim of mild enhancement. Those with larger diameter showed early central and heterogeneous enhancement. On delayed enhanced MR images, enhancement spread centrifugally until heterogeneous filling was achieved (Fig. 1). Histopathological specimens were obtained for further confirmation using US-guided liver biopsy with a 18-gauge 22mm—penetrating needle on both non tumoral and nodular liver.

Pathological examination revealed malignant proliferation characterised by an intrasinusoidal scaffold-like growth on the surface of residual liver-cell plates (Fig. 2). Tumoral cells showed a spindle shape, an atypical nuclei and a specific strong immunoreactivity to vascular endothelial markers (CD31, CD34) (Fig. 2). Non tumoral liver tissue was involved by the same tumoral process with a peliotic pattern.

The patient had no significant history of exposure to arsenic, radiation, vinyl chloride or Thorotrast. “Metronomic” chemotherapy using cyclophosphamide was started without favourable response. The patient died 5 months after diagnosis with hepatic failure.

Discussion

Probably because of its rarity, relatively little has been written of enhancement pattern of hepatic angiosarcoma HA in the radiology literature. In the present case, MR features were atypical from all of common liver masses. HA presented as multiple macronodules disseminated into both hepatic lobes. The tumors were heterogeneous and strongly hyperintense on T2-weighted MR images. Most of lesions presented with central and heterogeneous enhancement, which spread progressively over time. Koyama et al. already described this specific, central and progressive enhancement within dominant masses of 3 patients with HA who underwent MR examination [2].

Pathologically, HA is composed of malignant spindle cells of endothelial cell derivation that can be arranged in sinusoidal or large cavernous spaces, or form solid nodules or...

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masses [5]. The irregular and progressive enhancement pattern within dominant masses might be related to the gradual slow filling of high volume vascular spaces.

Prior sporadic case reports have suggested that HA might exhibit a progressive centripetal enhancement similar to that of cavernous hemangioma. In our patient, none of the masses showed MR findings that were consistent with hemangiomas as they displayed heterogeneous hypersignal intensity on T2-weighted MR images and heterogeneous centrifugal fill-in pattern with the intensity of enhancing areas different to that of the aorta and blood pool. Other recent reports have demonstrated that enhancement pattern of HA should not be confused with those of cavernous hemangioma. Peterson et al. described in 6 patients with HA varied enhancement patterns on multiphasic contrast-enhanced helical CT [4]. None of them showed features of peripheral nodular enhancement isoattenuating to vessels during the arterial and portal phases which could simulate hepatic hemangioma. However, some hemangiomas may not show typical features on dynamic contrast-enhanced CT and MRI. Centrifugal enhancement pattern of hemangiomas has been rarely described [6,7]. Moreover, giant hemangiomas may show a heterogeneous hyperintensity on T2-weighted images, and for giant hemangiomas (greater than 10 cm), a central or an irregular flame-shaped pattern of enhancement [8,9]. However, the multiplicity and heterogeneity of lesions in HA, the patient history together with clinical examination and liver function tests should permit distinction.

Disclosure of interest

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

References


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