

A peculiar case of diffuse hemangiomas of the left hepatic lobe in an asymptomatic adult patient: case report and literature review

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Abstract. – We report a rare case of diffuse hepatic hemangiomas (DHH) of the left hepatic lobe coexistent with giant hepatic hemangioma and without extra-hepatic involvement in an asymptomatic adult patient.

Liver hemangiomas are the most common benign liver tumors. However, DHH without extra-hepatic involvement has rarely been reported in adults. Furthermore, giant hepatic hemangioma coexistent with DHH is even uncommon, although an association between hemangiomas and giant hepatic hemangiomas may be supposed.

In this peculiar case, we observed an exclusive and widespread involvement of the left hepatic lobe with a sharp boundary between normal and altered liver parenchyma running along Cantlie's line.

Key Words:

Hemangiomas, Giant hepatic hemangioma, Liver hemangiomas.

Clinical Presentation

An asymptomatic 48-year-old man was referred to our hospital due to slight elevation of alanine aminotransferase (63 UI/L; upper normal value: 41 UI/L) detected on a blood routine test. Routine blood and liver function tests were normal, apart from mild hypercholesterolemia. Serum tumor markers were all within the normal range. Hepatitis B and C virus serum markers were negative. The previous clinical history was uneventful; the patient was a smoker and did not have a prior history of prolonged intake of drugs.

Imaging Findings

Abdominal ultrasound (US) showed a large heterogeneous exophytic mass of the left lo-

be of the liver, approximately 11 cm in size, with well-defined margins and absence of detectable high-velocity flow signals on color Doppler. Also, left hepatic lobe revealed a heterogeneous echo pattern with multiple small hypoechoic nodules on a hyperechoic background (Figure 1).

No lesions were found in the right hepatic lobe.

Abdominal MRI was performed for further evaluation. The exophytic mass of the left hepatic lobe showed low signal intensity on T1-weighted images and high intensity on T2-weighted images with restricted diffusion; dynamic MR axial images revealed a discontinuous centripetal filling appearance with some remaining unfilled portions. In the adjacent liver parenchyma of the left hepatic lobe, several nodular and coalescent lesions, mainly smaller than 1 cm in size, with low T1-weighted signal intensity (SI) and high T2-weighted SI, were depicted. Fat-saturated T1-weighted contrast-enhanced axial MR images showed early discontinuous enhancement of the nodules with uniform late retention of contrast. MR confirmed the exclusive involvement of the left hepatic lobe and absence of focal lesions in the right lobe with a sharp boundary between normal and altered liver parenchyma running along Cantlie's line. There was no evidence of extra-hepatic masses or lymphadenopathy (Figures 2 and 3). These findings were highly suggestive of a giant hepatic hemangioma with coexistent DHH of the left hepatic lobe.

Because of the lack of symptomatology left hepatectomy was ruled out. At 6 months follow-up clinical conditions and radiological findings (US and MRI) were unchanged.

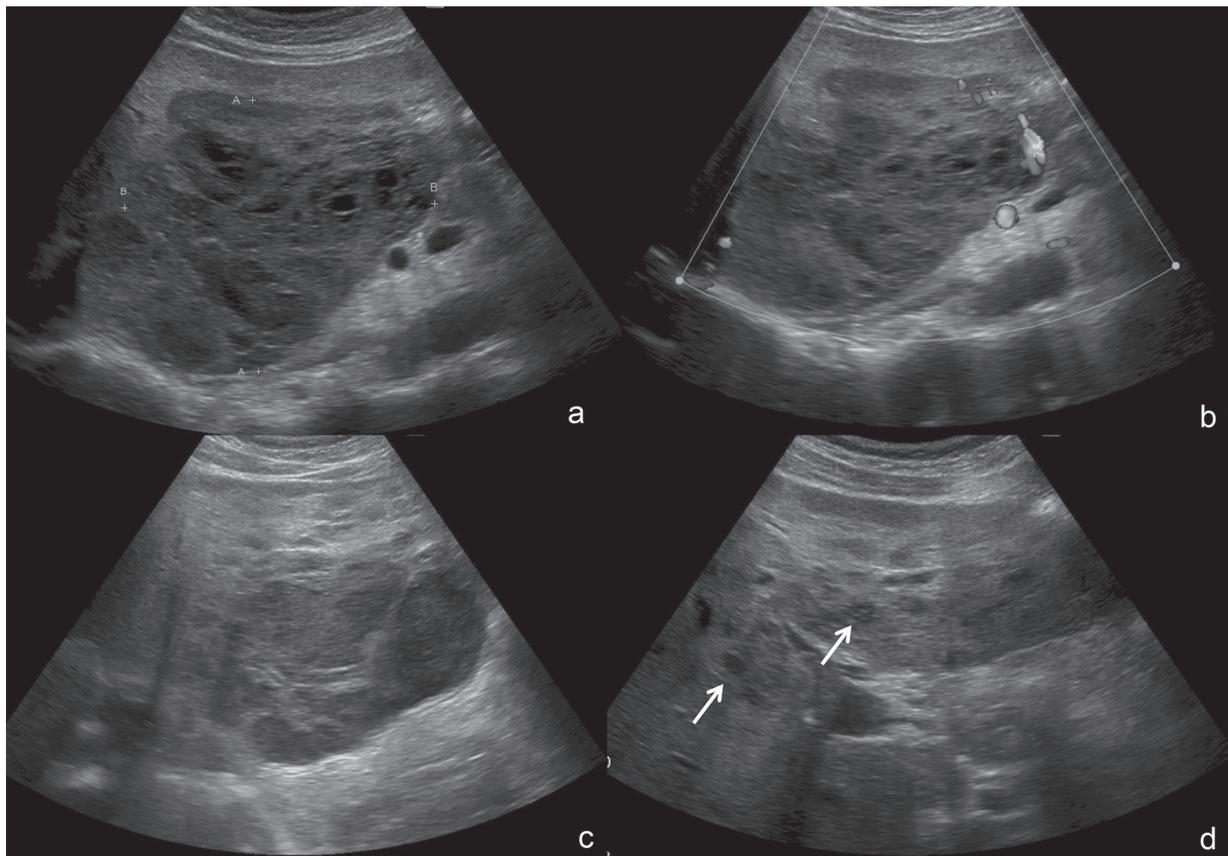


Figure 1. *a*, The abdominal US shows a large heterogeneous exophytic mass of the left hepatic lobe, approximately 11 cm in size, with sharp edges. *b*, Color Doppler does not show flow signals in the central portion of the lesion whereas peripheral scattered color spots are depicted. *c*, US examination shows a heterogeneous echo pattern of the liver parenchyma next to the large exophytic mass. *d*, In particular, US reveals multiple small hypoechoic nodules (yellow arrows) with well or poorly defined margins on a hyperechoic background in the segments II, III and IV.

Written informed consent was obtained from the patient for publication of this case report, including accompanying images.

Discussion

DHH is a rare benign condition characterized by diffuse replacement of liver parenchyma by hemangiomas. It can occur in all age groups, but it is most frequently detected in neonates in whom the entire liver is usually involved, thus acting as an intrahepatic complex arteriovenous shunting leading to high-output cardiac failure and significant mortality. Isolated DHH without extra-hepatic lesions is extremely rare in adults. The etiology and clinical course are not completely understood because of its rarity. It has been reported in patients

with hereditary hemorrhagic telangiectasia (HHT) or associated with hemangiomas of the skin and involvement of at least two visceral organs. However, on physical examination, our patient showed no hemangiomas on the skin, and radiological evaluation allowed ruling out HHT or hemangiomas in other visceral organs. Although previous reports^{1,3} have emphasized the role of prolonged steroid therapy in the development of hepatic cavernous hemangiomas and metoclopramide administration in a patient with DHH⁴, no history of steroid or metoclopramide use was documented in our patient. Even though some cases of long-term adult survival of diffuse neonatal hemangiomatosis have been reported^{5,6}, the prognosis of DHH without extra-hepatic involvement is still unclear and is probably related to the amount of hepatic involvement⁷. Accordingly, different prognoses

have been reported in the literature: one patient developed hepato-renal syndrome and finally died⁸, in another patient, severe arteriovenous shunting and cholestasis resolved spontaneously⁴. Lehmann et al⁹ described a case of a patient with diffuse hemangiomatosis of the left hepatic lobe who developed progressive tumor growth in the remaining liver parenchyma after left hepatectomy. Moreover, only a few papers reported the imaging findings of DHH. In particular, the diffuse (non-nodular) and the nodular patterns of DHH have been described. On US the liver parenchyma affected by DHH appears as a homogeneous hyperechoic area with poorly defined margins (diffuse pattern) or multiple – discrete or coalescent – small heterogeneous nodules usually lower than 5-10

mm in size (multinodular pattern). On MRI, the diffuse non-nodular DHH shows a heterogeneous enhancement during the arterial phase that becomes more homogeneous during portal and delayed phase imaging. The multinodular type exhibits small discrete and coalescent nodules with early homogeneous enhancement during the arterial phase, followed by uniform late retention of contrast, as found in our patient¹⁰. A significant association between giant hepatic hemangioma and hepatic hemangiomatosis has been described by Jhaveri et al¹¹. They found hemangiomatosis adjacent to a giant hemangioma larger than 8 cm in 18 of 41 patients (44%). In most cases, hemangiomatosis involved the adjacent margin of the giant hemangioma, without interposed normal liver tissue, as in our

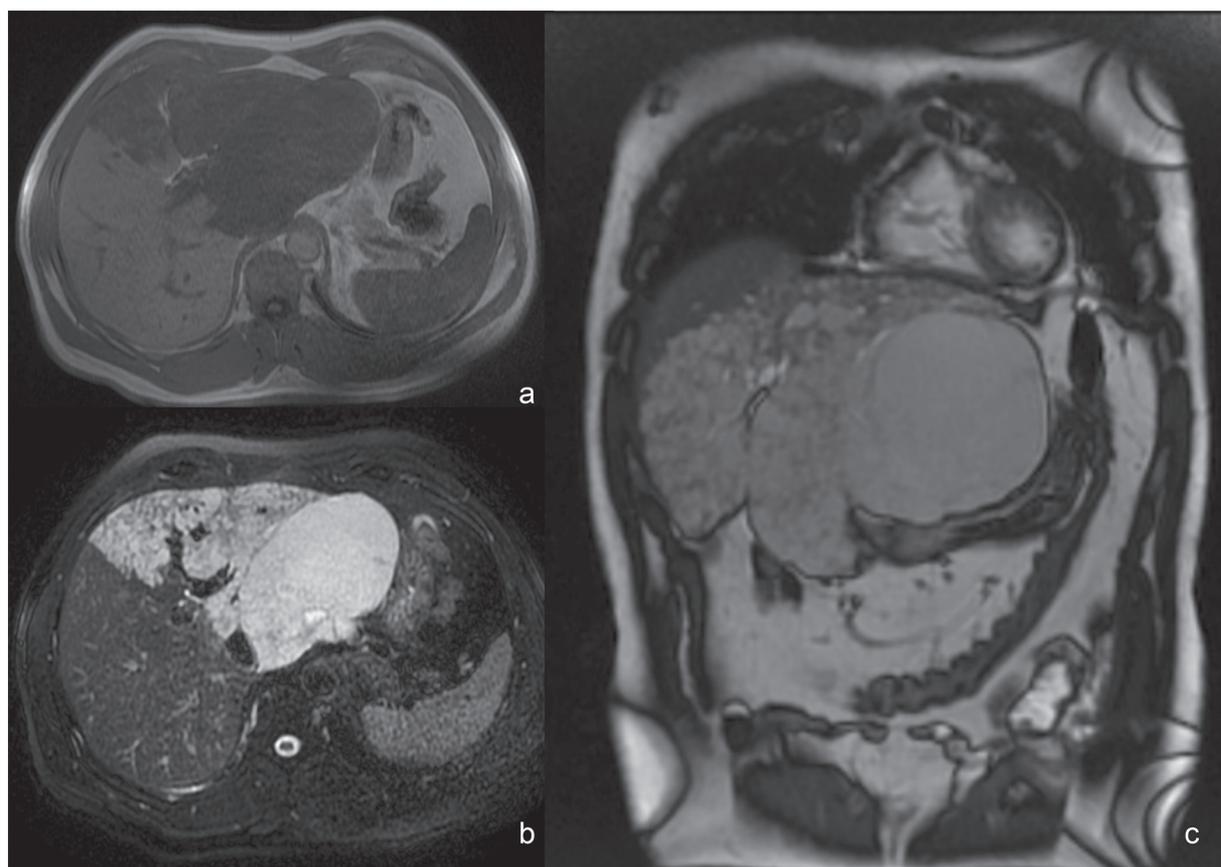


Figure 2. Pre-contrast MR images. *a*, Axial T1-weighted image. *b*, Axial fat-saturated T2-weighted image. *c*, Coronal FIESTA image. Pre-contrast MR images confirm the presence of a large exophytic mass in the left hepatic lobe showing homogeneous low signal intensity on T1-weighted images (a) and high homogenous intensity on T2-weighted images (b and c). In the adjacent liver parenchyma of the left hepatic lobe, several nodular and coalescent lesions with low T1-weighted signal intensity (a) and high T2-weighted signal intensity (b and c) are depicted. No normal liver tissue separates the exophytic mass and the adjacent altered liver parenchyma whereas a sharp boundary between normal liver parenchyma of the right hepatic lobe and altered parenchyma of left lobe is detected.

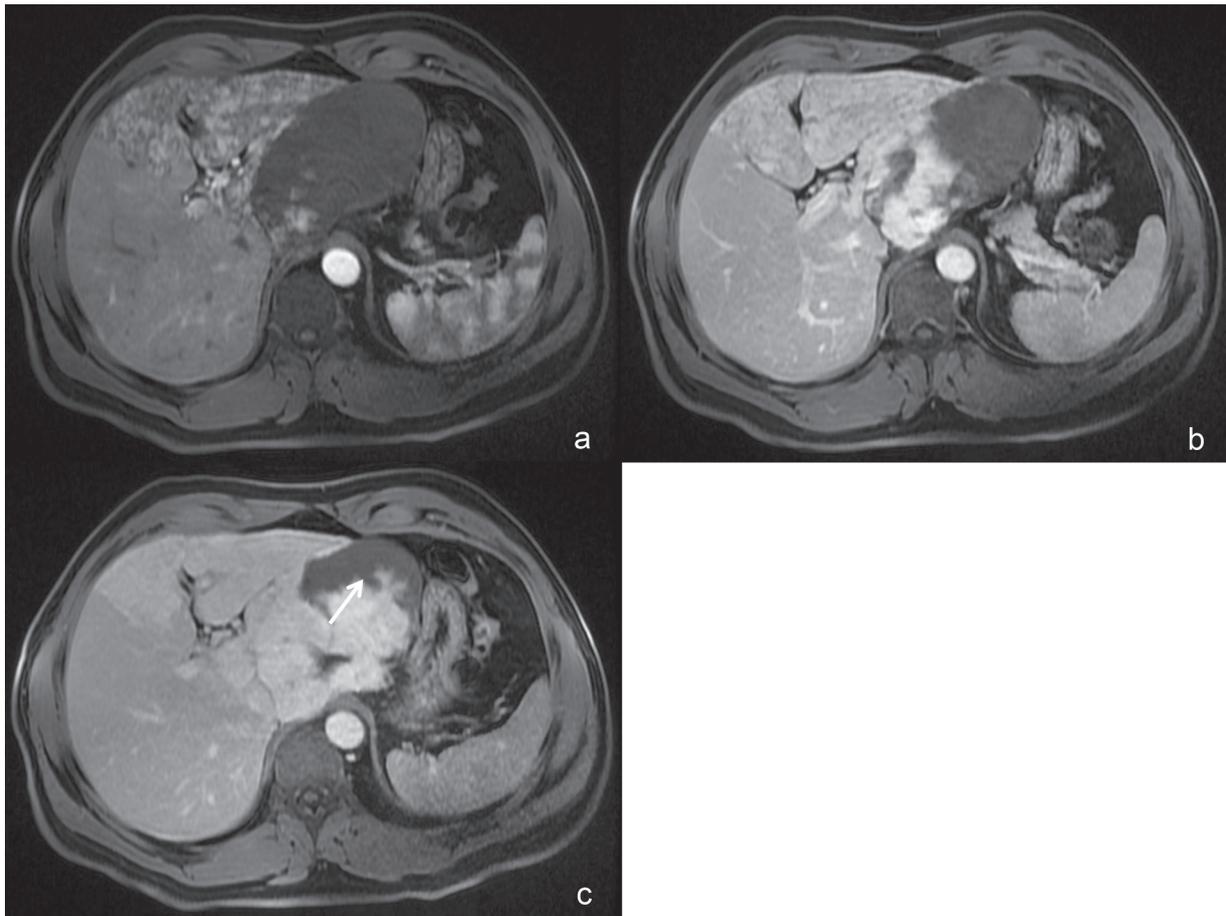


Figure 3. Post-contrast MR images. *a*, Axial post-contrast MR image-arterial phase. *b*, Axial post-contrast MR image - venous phase. *c*, Axial post-contrast MR image - delayed phase. Post-contrast MR images show a discontinuous centripetal filling appearance (a, b) of the large exophytic mass with a remaining unfilled portion of the delayed phase (c-yellow arrow). The coalescent nodules of the adjacent left lobe demonstrated dishomogeneous enhancement on the arterial phase (a), with gradual fill-in and a more homogeneous appearance on the venous and delayed phases (b, c).

patient. The presence of this association and the amount and distribution of normal residual liver parenchyma must be communicated to physicians involved in patient's care because the management strategy of patients with giant cavernous hemangiomas can be modified by the presence of DHH. In symptomatic cases, with selective involvement of liver lobes, hepatectomy should be considered.

Learning Points

- DHH without extra-hepatic involvement is an extremely rare condition in adults.
- The diagnosis of DHH concomitant or not with giant hepatic hemangioma in adults can be suggested by distinctive MRI findings.
- MRI also seems to be the preferable imaging technique for long-term follow-up.

Conflict of Interest

The authors declare no conflicts of interest.

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