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Hepatic angio-sarcoma: An unusual source of intra-hepatic bleeding☆

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ABSTRACT

INTRODUCTION: Hepatic angio-sarcoma represents an uncommon malignant tumor of the liver with a poor prognosis and a high rate of bleeding complications.
PRESENTATION OF CASE: We report a case of hepatic angio-sarcoma with a multi-nodular pattern complicated by intra-hepatic bleeding. The diagnosis was performed by computed tomography (CT). Angiographic procedure was unsuccessfully attempted as a treatment option. Autoptic examination confirmed the vascular nature of the malignant tumor.
DISCUSSION: Hepatic angio-sarcoma represents the most common malignant mesenchymal tumor of the liver. The diagnosis is provided by the histological examination and by specific endothelial markers. However, CT examination allows to recognize the disease and to detect intra-abdominal bleeding occurring in one-fourth of cases. Surgical resection represents the only definitive treatment of hepatic angio-sarcoma. In case of haemoperitoneum, trans-catheter arterial embolization represents the primary procedure used to stop the acute arterial bleeding.
CONCLUSION: CT represents the reference technique for the diagnosis of hepatic angio-sarcoma and allows to recognize the intra-abdominal bleeding which represents its most common complication. This condition always requires an immediate therapeutic approach.

1. Introduction

Hepatic angio-sarcoma accounts for 2% of primary hepatic tumors and represents the most common malignant mesenchymal tumor of the liver. It commonly affects patients aged between 60 and 70 years with a higher incidence in males (male:female, 4:1).[1]

Hepatic angio-sarcoma can be induced by different toxics and carcinogenic substances. Moreover, some diseases such as haemochromatosis and von Recklinghausen's neurofibromatosis are related to hepatic angio-sarcoma. Nevertheless, in 58-75% of cases no clear exposure is reported and the origin of the disease remains uncertain.[1]

Computed tomography (CT) can accurately identify its features and complications with the possibility of differential diagnosis with other similar hepatic lesions.

We report an uncommon case of hepatic angio-sarcoma presenting with intra-hepatic bleeding in an acute abdominal setting.

2. Presentation of case

A 65-year-old man, with professional exposure to vinyl chloride monomer, was admitted to our hospital due to general feelings of illness and severe acute abdominal pain.

A CT examination was mandatory because of the signs of an hypo-volemic shock and a severe acute abdominal pain.

The patient was scanned from lung apices to the pubic symphysis, before and after intravenous injection of 1.5 mL/kg of Iopamidol (Iomeron 400; Bracco, Milan; Italy) at 3.5 mL/s through the ante-cubital vein with an automatic power injector.

A 320-row CT scanner was used (detector collimation 0.5 mm, increment 0.5, 120/250 kVp/mAs). CT acquisition was performed with a biphasic technique during the arterial and portal venous phases after the intravenous injection of contrast material. An additional delayed phase was obtained from the diaphragmatic dome to the pubic symphysis.

All CT data were transferred to a workstation (HP XW 8600) equipped with dedicated software (Vitrea FX 2.1, Vital Images, Minneapolis, Minnesota, US) for image reconstructions.

The unhenanced CT scans of the abdomen revealed hemoperitoneum and hepatomegaly. On enhanced scans, the hepatic parenchyma appeared inhomogeneous because of the presence of multiple and diffuse nodular lesions with a maximum diameter of 20 mm × 20 mm (Fig. 1a and b). Besides, on the arterial phase, active bleeding at the level of V and VI hepatic segments was detected (Fig. 1c). CT findings were suggestive for hepatic lesion rupture with hemoperitoneum. For this reason, trans-arterial embolization was

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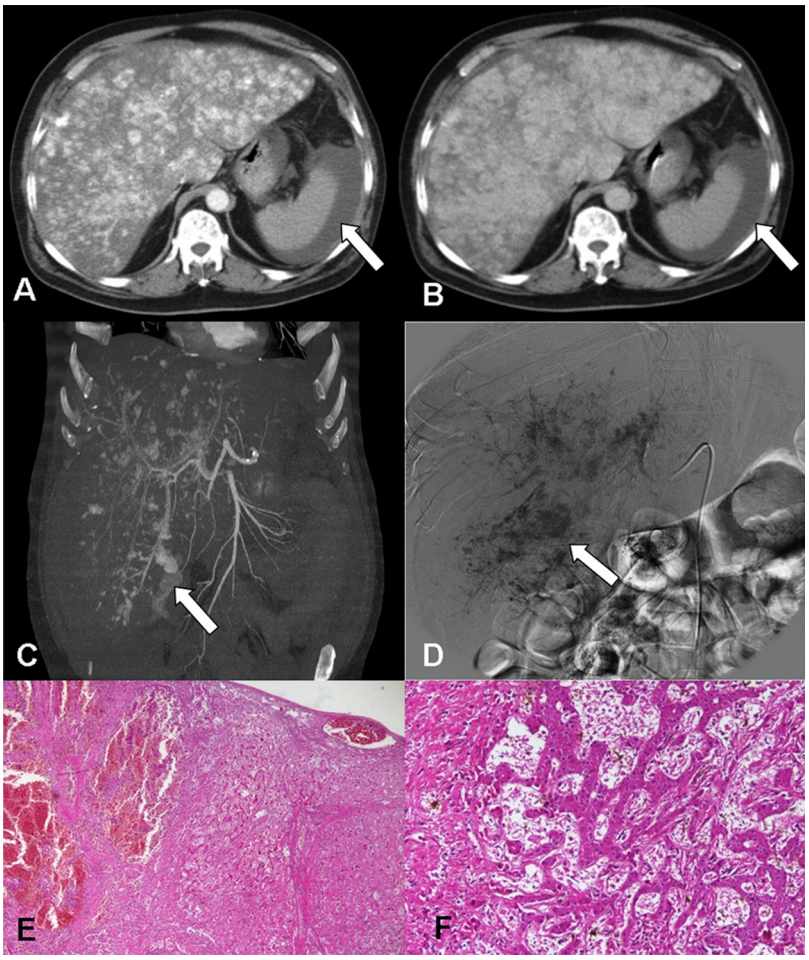

Fig. 1. 65-year-old man affected by hepatic angiosarcoma. (A and B) Transverse CT scans show multiple diffuse intrahepatic hyperdense nodules both in the arterial (a) and portal venous (b) phases. Ascites is also associated (arrows). (C) Maximum Intensity Projection (MIP) CT reconstruction on the coronal plane shows intrahepatic bleeding (arrow) at the level of V and VI segments. (D) The angiographic examination confirms the spreading of contrast medium at the level of V and VI hepatic segments (arrow). (E and F) The histopathological specimen confirmed the diagnosis of hepatic angiosarcoma as neoplastic proliferation with sinusoid growth configuring cavernous spaces covered by atypical cells (hematoxylin–eosin stain, 100× magnification).

performed by using the right femoral artery as vascular access. A 5 French catheter was positioned at the origin of the celiac trunk and superior mesenteric artery. The contrast material was injected at 25 mL/s with an automatic power injector. Images were acquired by means of a digital angiograph (Integris Allura, Philips Medical Systems, Best, The Netherlands). Angiographic examination confirmed active bleeding at the level of V and VI hepatic segments (Fig. 1d). The right hepatic artery embolization was performed by using gelatin sponge. However, even if the interventional procedure interrupted the bleeding from the hepatic artery branches, the patient died the day after, because of the hypo-volemic shock complications.

The autoptic examination diagnosed diffuse and invasive hepatic angio-sarcoma with wide hemorrhagic area at the level of V and VI hepatic segments and haemoperitoneum. Macroscopically, the liver had nodular external surface with blood filled cavities and spongy areas surrounded by normal parenchyma. Microscopically, neoplastic proliferation with sinusoid growth configuring cavernous spaces covered by atypical cells was detected (Fig. 1e and f).

Tumor cells were intensely positive for CD-31 and CD-34 endothelial markers.

3. Discussion

The clinical presentation of hepatic angio-sarcoma is unspecific and includes abdominal pain, weakness and weight loss. However, Huang et al. reported that a substantial number of patients were asymptomatic and were identified during routine health examinations, highlighting the importance of periodical health check-ups.[2]

Hepatomegaly, ascites and jaundice are common findings. Liver function tests are usually abnormal but there is no liver function test or set of tests specific for the tumor. Intra-abdominal bleeding is also typical and occurs in one-fourth of all cases. This complication is probably related to the high incidence of clotting abnormalities and the vascular nature of the neoplasm.[3]

At the time of presentation, most patients have metastases. The most common sites of metastases are the lung and spleen.[4]

The diagnosis of hepatic angio-sarcoma is provided by the histological examination. Endothelial markers such as factor-VIII-related antigen, vimentin, CD 31, and CD 34 are expressed by tumor cells.[5]

However, fatal hemorrhagic complications have been reported during percutaneous biopsy of hepatic angio-sarcoma. For this reason, radiologists should know the imaging findings of this tumor in order to facilitate a safe and accurate diagnostic work-up.[6]

Ultrasound shows single or multiple masses, whose features depend on the different levels of necrosis and hemorrhage.

CT examination allows to highlight the variable features of hepatic angio-sarcoma appearing as multiple nodules, a single large mass, a large mass with smaller satellite nodules or as diffuse infiltrating micro-nodular lesions. Multi-nodular and diffuse micro-nodular patterns represent the most common presentation.

Also our experience reports a case of multi-nodular pattern which can be added to the limited series reported in the medical literature.[7] Generally, no intra-patient variability in tumor appearances has been observed.[8] Magnetic resonance imaging (MRI) allows to depict the heterogeneous composition of hepatic angiosarcomas which may contain areas of necrosis, recent and old hemorrhage, fibrosis and hyalinization.[5,9]

Positron emission tomography (PET) with 2-deoxy-2-[18F] fluoro-D-glucose (FDG) may provide useful information for differentiating hepatic cavernous hemangiomas with irregular shape from angio-sarcomas which show high FDG uptake.[7]

Surgical resection represents the only definitive treatment of hepatic angio-sarcoma. However, even in cases of complete tumor resection, recurrence is common. Some cases of long-term survival after partial hepatectomy are reported.

In cases of hepatic mass rupture with hemoperitoneum, occurring in 15–27% of patients, trans-catheter arterial embolization has been demonstrated to be the primary procedure used to stop the acute arterial bleeding.[10] In fact, our case underwent this kind of treatment.

Major differential diagnoses include hepatocellular carcinoma, hepatic artero-venous malformations (AVM) in hereditary hemorrhagic telangiectasia (HHT), hepatic metastases and haemangioma.[7,10–12]

4. Conclusion

Hepatic angio-sarcoma is an uncommon tumor of the liver with a poor prognosis. CT represents the reference technique for the diagnosis and allows to recognize the intra-abdominal bleeding which represents its most common complication. This condition always requires an immediate therapeutic approach.

Conflicts of interest

The authors declare that there is no conflicts of interest.

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Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Authors' contributions

Study design was done by Amato Antonio Stabile Ianora, Marco Moschetta, Michele Telegrafo, Arnaldo Scardapane.

Data collections were carried out by Marco Moschetta, Michele Telegrafo, Arnaldo Scardapane, Amato Antonio Stabile Ianora, Fabio Fucilli.

Data analysis was done by Amato Antonio Stabile Ianora, Marco Moschetta, Michele Telegrafo, Fabio Fucilli, and Arnaldo Scardapane.Manuscript writing was done by Amato Antonio Stabile Ianora, Marco Moschetta, Michele Telegrafo, Fabio Fucilli.

References

1. Egea Valenzuela J, López Poveda MJ, Pérez Fuenzalida FJ, Garre Sánchez C, Martínez Barba E, Carballo Alvarez F. Hepatic angiosarcoma. Presentation of two cases. *Rev Esp Enferm Dig* 2009;**101**(6), 430–34, 434–37.
2. Huang NC, Wann SR, Chang HT, Lin SL, Wang JS, Guo HR. Arsenic, vinyl chloride, viral hepatitis, and hepatic angiosarcoma: a hospital-based study and review of literature in Taiwan. *BMC Gastroenterol* 2011;**11**:142.
3. Locker GY, Doroshow JH, Zwelling LA, Chabner BA. The clinical features of hepatic angiosarcoma: a report of four cases and a review of the English literature. *Medicine (Baltimore)* 1979;**58**(1):48–64.
4. Koyama T, Fletcher JG, Johnson CD, Kuo MS, Notohara K, Burgart LJ. Primary hepatic angiosarcoma: findings at CT and MR imaging. *Radiology* 2002;**222**(3):667–73.
5. Bruegel M, Muenzel D, Waldt S, Specht K, Rummeny EJ. Hepatic angiosarcoma: cross-sectional imaging findings in seven patients with emphasis on dynamic contrast-enhanced and diffusion-weighted MRI. *Abdom Imaging* 2012.
6. Drinković I, Brkljacić B. Two cases of lethal complications following ultrasound-guided percutaneous fine-needle biopsy of the liver. *Cardiovasc Intervent Radiol* 1996;**19**(5):360–3.
7. Okano A, Sonoyama H, Masano Y, Taniguchi T, Ohana M, Kusumi F, et al. The natural history of a hepatic angiosarcoma that was difficult to differentiate from cavernous hemangioma. *Intern Med* 2012;**51**(20):2899–904.
8. Peterson MS, Baron RL, Rankin SC. Hepatic angiosarcoma: findings on multiphasic contrast-enhanced helical CT do not mimic hepatic hemangioma. *Am J Roentgenol* 2000;**175**(1):165–70.
9. Yang KF, Leow VM, Hasnan MN, Manisekar KS. Primary hepatic angiosarcoma: difficulty in clinical, radiological, and pathological diagnosis. *Med J Malaysia* 2012;**67**(1):127–8.
10. Park YS, Kim JH, Kim KW, Lee IS, Yoon HK, Ko GY, et al. Primary hepatic angiosarcoma: imaging findings and palliative treatment with transcatheter arterial chemoembolization or embolization. *Clin Radiol* 2009;**64**(8):779–85.
11. Scardapane A, Ficco M, Sabbà C, Lorusso F, Moschetta M, Maggialetti N. Hepatic nodular regenerative lesions in patients with hereditary haemorrhagic telangiectasia: computed tomography and magnetic resonance findings. *Radiol Med* 2013;**118**:1–13.
12. Scardapane A, Stabile Ianora A, Sabbà C, Moschetta M, Suppressa P, Castorani L, et al. Dynamic 4D MR angiography versus multislice CT angiography in the evaluation of vascular hepatic involvement in hereditary haemorrhagic telangiectasia. *Radiol Med* 2012;**117**:29–45.