Hepatic angiosarcoma with multiple metastases in a young man

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Abstract: Hepatic angiosarcoma, also called Kupffer's sarcoma, is a malignant mesenchymal neoplasm of endothelial cells, represents less than two percent of all primary liver neoplasm. Hepatic angiosarcoma is an infrequent and difficult-to-diagnose disease, mostly discovered by chance. Because of its rapid progression and usually fatal outcome, early diagnosis is necessary and complete surgical resection is the key to improve prognosis, but the neoplasm is often disseminated at the time of diagnosis, making resection impossible. Rare cases of hepatic angiosarcoma have been reported in the literature. Here, we report a case of hepatic angiosarcoma with spleen, lungs, right atrium and spine infiltration. Contrast enhanced abdomen CT and MRI scans revealed multiple nodules in the liver and spleen with rich blood supply, at the same time many metastases were noticed at bilateral lungs, right atrium and spine. The lesions rapidly deteriorated during the 2 months following the exams. The diagnosis of hepatic angiosarcoma was made after an open biopsy.

Keywords: Hepatic angiosarcoma; computed tomography; magnetic resonance imaging

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A 26-year-old male complained continued short breath without sputum for more than 20 days and fever for 1 week. His medical history and laboratory examination were unremarkable, except serology for HBV was positive for more 20 years. After CT and MRI scans with contrast, multiple nodes in the liver and spleen were revealed with rich blood supply. At the same time, many abnormal imaging findings which were considered to be metastases were noticed in bilateral lungs, right atrium and spine (*Figures 1*,*2*). With these findings, fine-needle biopsy was obtained of the hepatic lesions. The histological study showed non-regular dilation of hepatic sinusoids and proliferation of endothelial cells. The patient left the hospital for personal reasons. The followed exams after 2 months showed rapid progression both in the liver and lungs (*Figure 3*). Laboratory tests revealed: total bilirubin 52.7 umol/L, ALT 323 U/L, AST 246 U/L, AFP 2.5 ng/mL. Because fine-needle biopsy before were inconclusive, so recommendation was made to carry out an open abdominal cavity biopsy. Photomicrograph of the resected specimen showed: the tumor cells showed nodular and cord like distribution. Tumor cells were deeply stained, nucleoli and mitotic were seen, cells were positive for endothelial markers CD-31 and CD-34 (*Figure 4*). These were compatible with primary hepatic angiosarcoma. The patient died from multiple organ failure, disseminated intravascular coagulation, and peritoneal bleeding 1 week later.

Disclosure: The authors declare no conflict of interest.

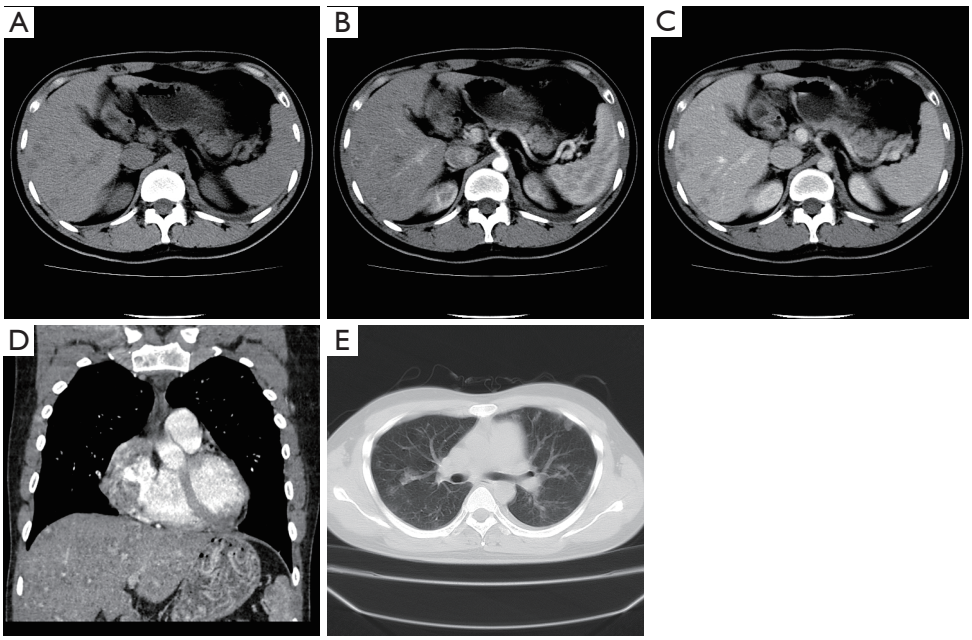

Figure 1 First exam at admission. (A) Unenhanced image depicts multifocal hepatic nodules (≤3 cm); (B) contrast-enhanced early phase image shows slightly irregular enhancement of the nodules, and some lesions without enhancement; (C) contrast-enhanced portal venous phase image shows nodules have almost no enhancement progression; (D) contrast-enhanced image shows abnormal thickness of right atrium; (E) chest axial CT scan (lung window) demonstrates multiple scattered node-like lesions in both lungs.

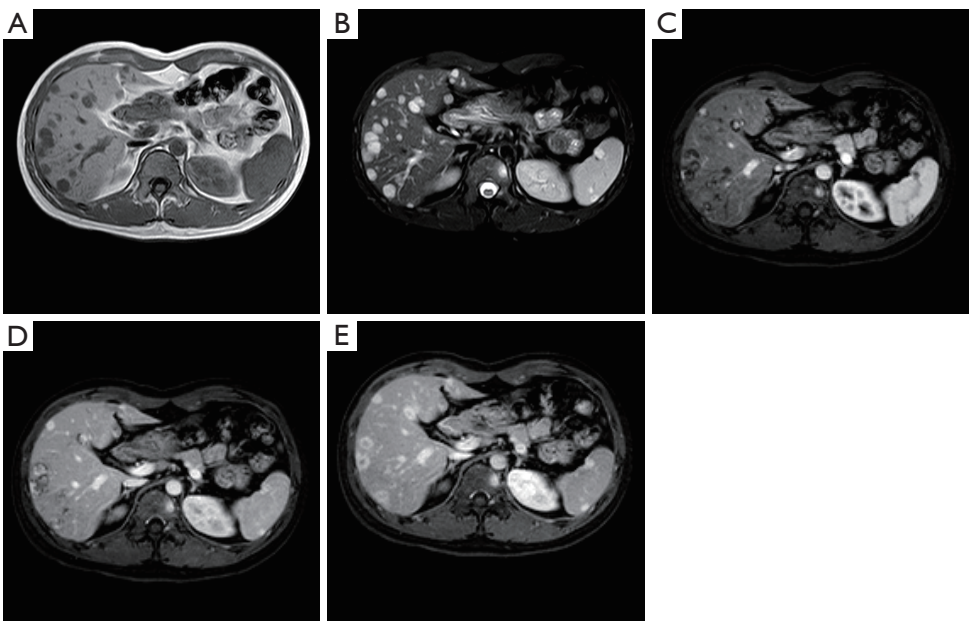

Figure 2 First CT MR exam at admission. (A,B) Abdomen MR images show multiple nodules in the liver with low signal intensity on unenhanced T1-weighted image and high signal intensity on T2-weighted fat suppressed image. At the same slice some node-like lesions were noticed in the spleen and spine; (C) contrast-enhanced early phase image shows variable patterns including patchy peripheral or bizarre shaped intralesional foci of enhancement, peripheral rim enhancement, and small lesions without enhancement; (D,E) the majority of lesions present with varying degrees of progressive enhancement. Small nodules frequently display homogeneous enhancement on delayed phase images due to complete fill-in.

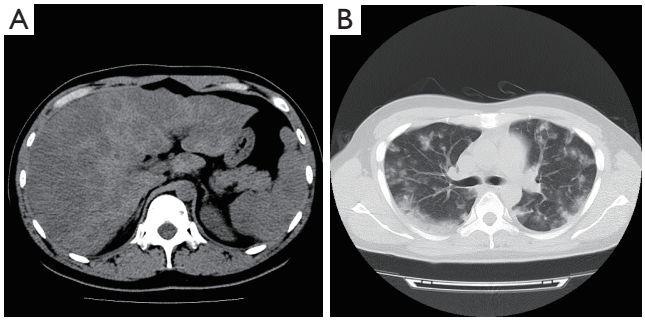

Figure 3 Second exam 2 months later. (A,B) Both abdomen and thorax CT scans shows rapid progression of lesions in the liver, spleen, and lungs.

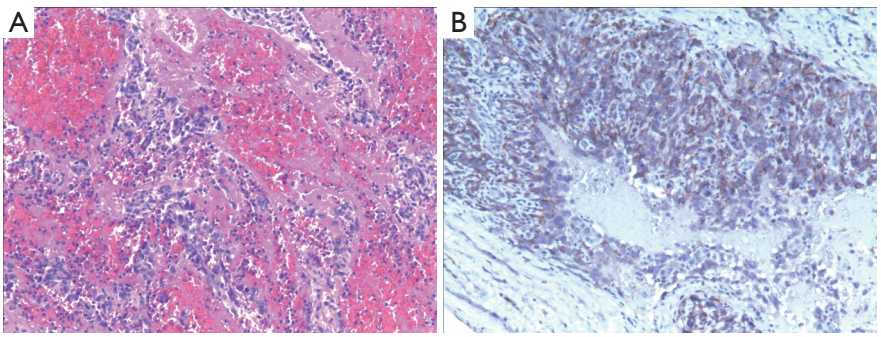

Figure 4 (A) Photomicrograph (×100/HE) reveals that the tumor cells show nodular and cord like distribution. Tumor cells are deeply stained, nucleoli and mitotic are seen; (B) immunohistochemical (×100/CD31) staining shows CD-31 positive.

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