A diagnostic pitfall in hepatic mesenchymal hamartoma mimicking hepatoblastoma: A case report

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Case Report

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Abstract

Background: Hepatic mesenchymal hamartoma (HMH) is an uncommon benign tumor in children. While mesenchymal hamartomas may be angiomatous and blood vessels may be identified, HMH with a malignant tumor symptom on the contrast-enhanced hepatic computed tomography angiography (CTA) has not been described. Here, we present the first case of HMH mimicking hepatoblastoma on the hepatic CTA from pathological point of view and review the imaging and histological features of this unique lesion.

Case presentation: A 2-year-old female child was found a distention in the right abdomen and was admitted to our hospital. The Hepatic CTA showed that the blood vessels were thickened, the tumor blood vessels were clustered in the tumor. According to the hepatic CT findings, the tumor was considered to be malignant, possibly a hepatoblastoma. Microscopic examination showed a tumor arranged in lobules, composed of loose myxoid mesenchyme surrounding ductal structures, with intervening vascular channels. The Immunohistochemical staining revealed positive CK7 and CD34 for the bile duct elements and the lining endothelial cells of the vascular channels. There were abundant blood vessels around the nodules and the margins of remaining hepatocytes, but few in the central region. The blood vessels are small, thin-walled vessels and presented like capillaries and venules.

Conclusions: A histological diagnosis of Hepatic Mesenchymal Hamartoma was confirmed by the microscopic examinations. This case adds learning points to radiologists when heterogeneous reinforcement on enhanced CT scan was presented in such a large childhood liver tumor.

Background

Hepatic mesenchymal hamartoma (HMH) is an uncommon benign tumor in children, with 80% being detected under the age of 2 years.[1] With regard to this report, HMH was confused with hepatoblastoma on the computed tomography angiography (CTA) of the liver. Mesenchymal hamartomas could be composed of various epithelial and mesenchymal components, including bile duct elements, hepatocytes, myxoid stroma with myofibroblast-like cells, dilated vessels and lymphatics. While mesenchymal hamartomas may be angiomatous and blood vessels may be identified,[2] HMH with a malignant tumor symptom on the contrast-enhanced hepatic CTA has not been described. Here, we present the first case of HMH mimicking hepatoblastoma on the hepatic CTA from pathological point of view and review the imaging and histological features of this unique lesion.

Case Presentation

A 2-year-old female child was found a distention in the right abdomen by her parents inadvertently. She did not present any specific clinical symptoms herself. Ultrasound examination at the local hospital indicated liver cyst. The hepatic CT showed a large space-occupying lesion of the liver, considering hepatoblastoma. The child was then admitted to our hospital on March 29, 2019. The child has no

history of febrile seizures, acute infectious diseases or any other specific disease. The child is the second child of the family, with birth weight 2800 grams and good condition except neonatal jaundice in the neonatal period. At the time of admission, there was no obvious abnormality in the physical examination. Tumor marker testing results showed that the alpha-fetoprotein was slightly elevated to 14.5 ng/ml (normally < 10 ng/ml), and others (carcinoembryonic antigen, ferritin) presented normal value.

The enhanced hepatic computed tomography (CT) scan revealed a large mass in the right lobe of the liver, with the size of 91.9mm × 125.4mm × 176.4 mm (Fig. 1). The boundary was clear. Multiple circular cystic densities were observed in the plain CT scan, and the CT value was 11hu. After enhanced scan, the tumor showed heterogeneous enhancement, with the CT value of 45hu. Low density cystic change enhancement was not significant, CT value of venous stage lesions was 66hu, and CT value of delayed stage lesions was up to 84hu. In summary, according to the CT scan, multiple macular necrosis and cystic degeneration were observed in the lesion, and the contrast enhanced scan showed slightly delayed enhancement. The CTA showed the hepatic artery supplied blood to the right lobe of the liver. The blood vessels were thickened, the tumor blood vessels were clustered in the tumor, showed a signal of vascular agglomeration. The hepatic segment of the inferior vena cava was obviously compressed and tapered, the middle hepatic vein and right hepatic vein were not shown. The left hepatic vein ran naturally, no obvious filling defect was observed. The right branch of the portal vein was compressed and tapered, and the left portal vein ran naturally. The tumor volume was about 1,329.55 cm^3 and the remaining liver volume was about 219.56 cm^3.

According to the hepatic CT findings, the tumor was considered to be malignant, possibly a hepatoblastoma. The surgical resection was then planned immediately. As the tumor was huge in volume and may be ruptured before surgery, an 11-day preoperative preparation is performed. Finally, the tumor was removed by enucleation method, thus saving the rest of the liver. Intraoperative exploration of the abdominal cavity showed the tumor was located in the right lobe and invaded the hilum and middle lobe of the liver, almost occupied the abdomen and pelvic cavity.

The size of the surgical resection was 180.0mm × 150.0mm × 80.0 mm and the actual tumour size was 170.0mm × 120.0mm × 78.0 mm (Fig. 2). The tumor cut surface is soft, fleshy, white-tan, and had areas of hemorrhage, necrosis and cyst formation. The cyst wall was intact and did not invade the surrounding area. Microscopic examination showed a tumor arranged in lobules, composed of loose myxoid mesenchyme surrounding ductal structures, with intervening vascular channels. The vascular size profile is small - capillary to small venules (Fig. 3). The mesenchyme was loose, edematous and mucoid degeneration, with scattered inflammatory cells, lymphocytes, monocytes and neutrophils. Some areas of the junction were composed of infantile regenerated hepatocytes and proliferated blood vessels. The Immunohistochemical staining revealed positive CK7 for the bile duct elements and positive CD34 for the lining endothelial cells of the vascular channels (Fig. 3). Interestingly, there were abundant blood vessels around the nodules and the margins of remaining hepatocytes, but few in the central region. The blood vessels are small, thin-walled vessels and presented like capillaries and venules. Together, a histological diagnosis of Hepatic Mesenchymal Hamartoma was confirmed by the microscopic examinations.

Because of the distribution of abundant blood vessels around the nodules in the tumor, the mass was probably misdiagnosed as a hepatoblastoma on the hepatic CT imaging.

During the follow-up, the ultrasound showed that the size and shape of the liver changes after operation, the internal echo of liver is normal and evenly distributed. The structure of the vascular network was clear, and the intrahepatic bile duct did not expand. The liver function investigations and coagulation profile were within normal limits.

Discussion And Conclusions

The components of hamartoma are complex. Most of them are tumor like malformations formed by abnormal development of normal tissues, and a few of them are mesenchymal tumors.[3] Hepatic mesenchymal hamartoma was frequently cystic. The mesenchymal components were composed mainly of architecturally abnormal biliary structures.[4–7] Therefore, both CK7 and CK19 were positive in most cases which indicates biliary epithelium in the cystic component.[8–10] Usually, there was limited immunoreactivity to CD34 and CD31 markers for hepatic mesenchymal hamartomas.[9] Hamartoma can originate from various components, such as mesenchymal, vascular, neurovascular and so on. CD34 and CD31 expression were positive in the case of vascular hamartoma, which was a very rare benign vascular proliferation.[11] However, in our case, the majority of hamartoma components were mesenchymal and bile duct elements, which indicated positive immunohistochemistry expression of the degenerated bile duct epithelium (positive for CK7, CK19 and P16). The positive expression of CD34 and CD31 (positive CD31 not shown) revealed the distribution of abundant vessels around the tumor nodules, which may explain the misdiagnosis of hepatoblastoma on hepatic CTA findings.

The heterogeneous enhancement of the tumor in CT scan imaging may also indicate the tumor contained a set of arterial and venous vessels. However, bile duct elements and hepatocytes are not presented in vascular hamartoma.[12] The sign of a hyperintense center (liquefactive necrosis) surrounded by a less intense rim of viable tumor in the CT scan may also reveal a possibly malignant signal. The abundant vessels around the tumor nodules through microscopic examination may explain the areas of hemorrhage, necrosis and cyst formation of this uncommon benign liver tumor. This case adds learning points to radiologists when heterogeneous reinforcement on enhanced CT scan was presented in such a large childhood liver tumor. To the best of our knowledge, this is the first case of hepatic mesenchymal hamartoma with abundant vessels and presented a hepatoblastoma-like characteristics on hepatic CT imaging.

The observation of hamartomas which mimic hepatoblastoma on hepatic CTA is interesting and remarkable. Moreover, this report suggests that, hepatic mesenchymal hamartoma could be composed of abundant vessels and should not be misdiagnosed as malignant tumors.

Abbreviations

HMH = hepatic mesenchymal hamartoma, CTA = computed tomography angiography, CT = computed tomography.

Declarations

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Authors' contributions

ZC and LF conceived and wrote the manuscript together, YZ and YY jointly collected clinical data and WJ analyzed the imaging data.GM performed pathological diagnosis and immunohistochemical analysis.All authors have read and approved the final manuscript.

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Availability of data and materials

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Ethics approval and consent to participate

Not applicable.

Consent for publication

Obtained.

Competing interests

The authors declare that they have no competing interests.

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Figures

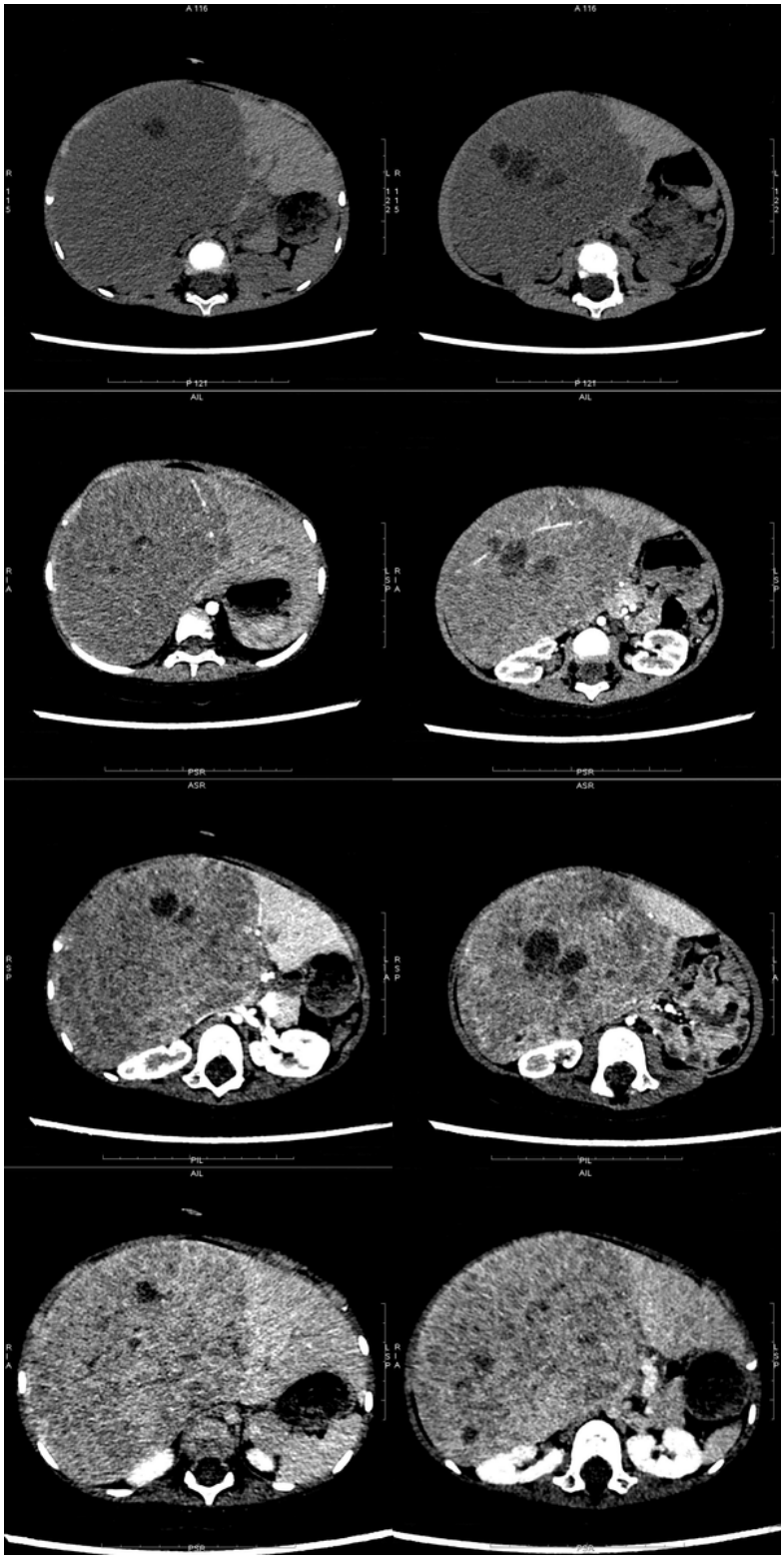

Figure 1

Hepatic contrast computed tomography angiography image and vascular reconstruction. The blood vessels were thickened, the tumor blood vessels were clustered in the tumor.

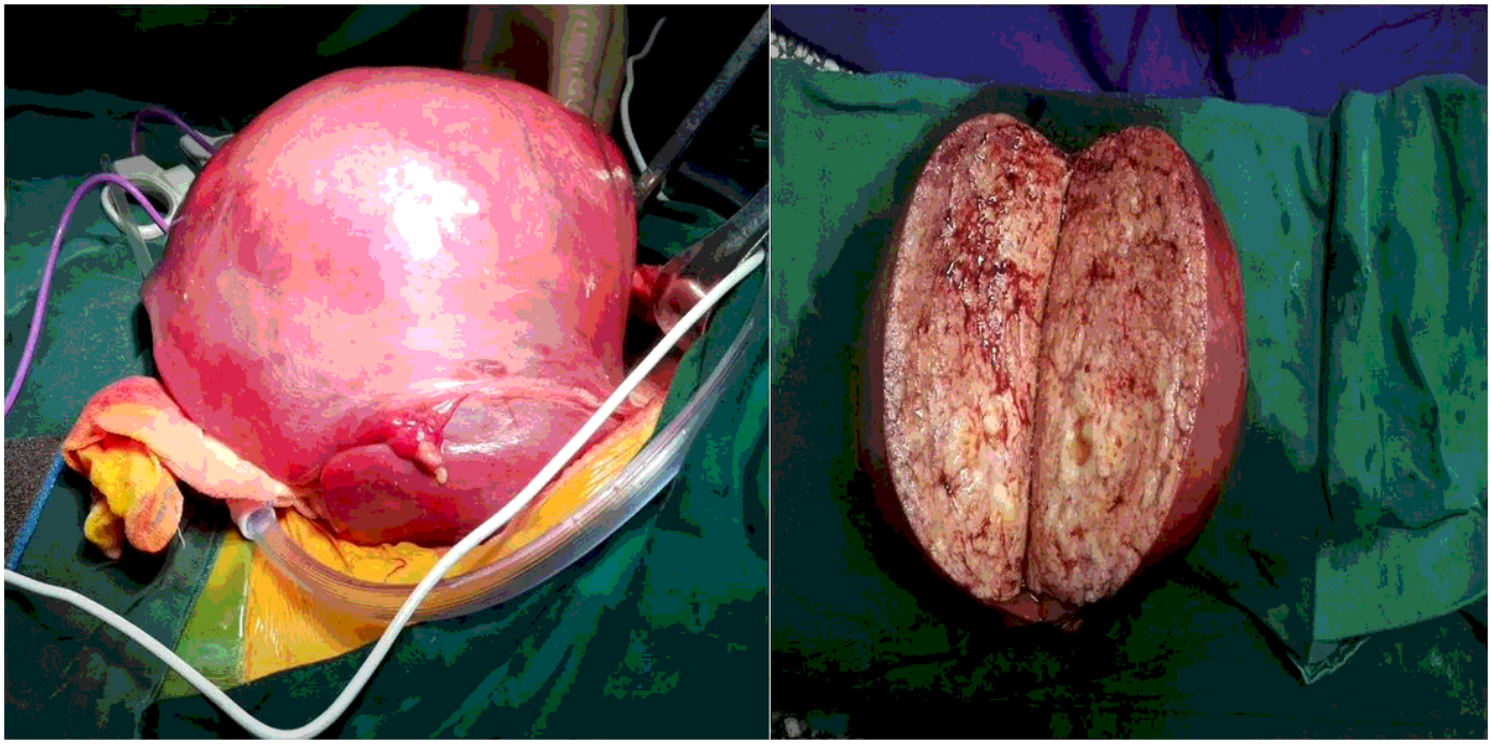

Figure 2

Intraoperative photo of the tumor.

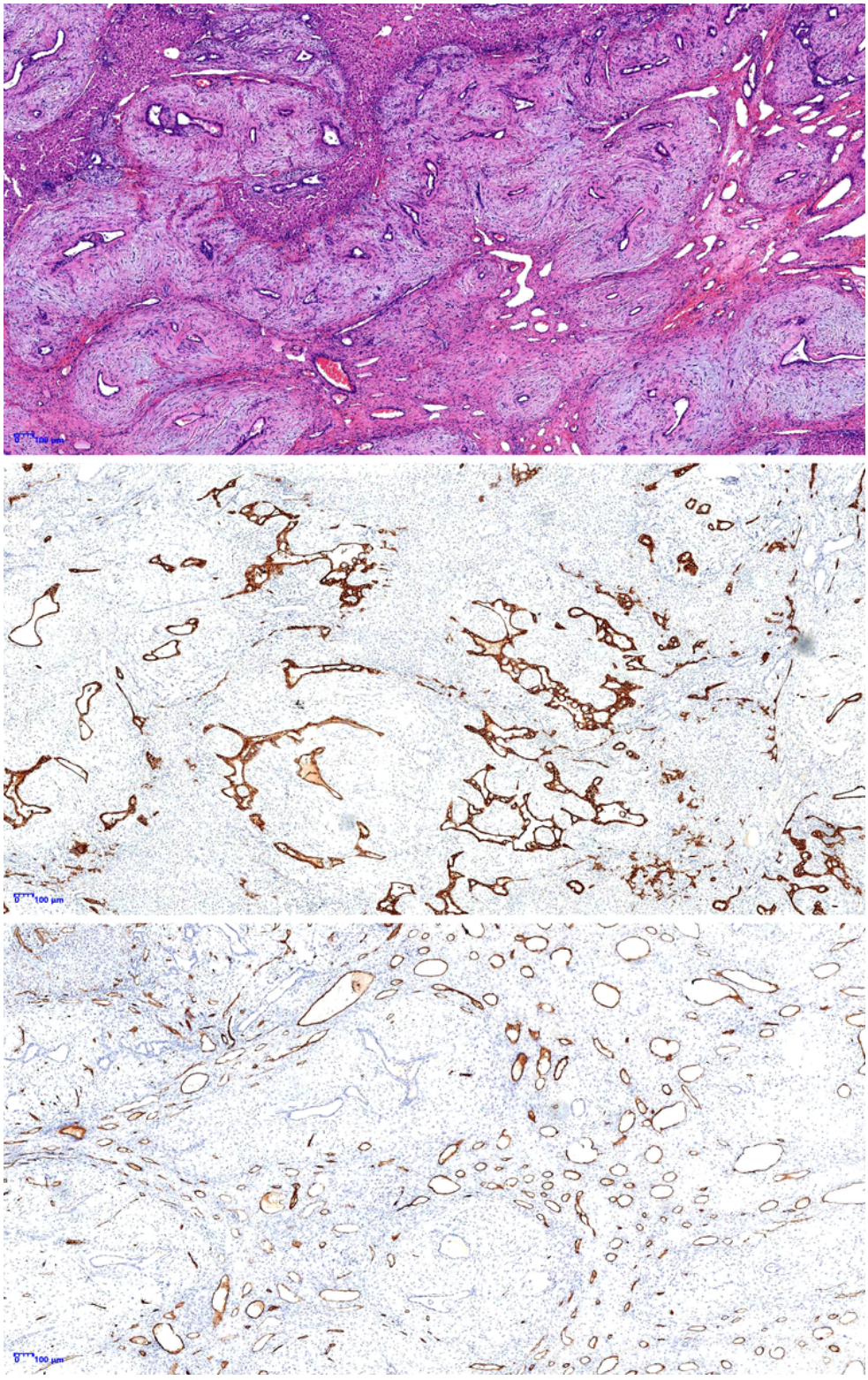

Figure 3

H&E photomicrographs of a hepatic mesenchymal hamartoma. Immunohistochemistry expression of CK7 (middle figure) and CD34 (bottom figure) revealed the distribution of vessels around the tumor nodules. Scale bars are listed within the images. Images were captured at 2.5× (low power) magnification.

Supplementary Files

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