Case report

Imaging findings of multiple infantile hepatic hemangioma associated with cardiac insufficiency

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Background: **Infantile hepatic hemangioma (IHH) as a benign liver tumor in infancy and childhood is commonly associated with high output cardiac failure. The present study aims to describe the imaging findings in a patient who was diagnosed as having multiple IHH with congestive cardiac insufficiency.**

Methods: **The imaging findings and clinical manifestations of the patient with multiple IHH associated with cardiac insufficiency were retrospectively reviewed.**

Results: **Ultrasonography showed multiple intrahepatic lesions with mixed echoes and markedly expanded hepatic veins and the inferior vena cava of the patient. Echocardiography revealed right heart insufficiency and pulmonary hypertension. Contrast-enhanced MRI showed early mild enhancement of lesions and more obvious delayed enhancement. The patient died after combined therapy of surgery and hormone.**

Conclusions: **The imaging findings of multiple IHH associated with cardiac insufficiency are typical and diagnostic. Early imaging assessment may facilitate the diagnosis and treatment of the disease.**

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Introduction

Infantile hepatic hemangioma (IHH) is a benign liver tumor commonly seen in infants and newborns.[1] Although it is histologically benign, the tumor has proliferative potentially and may become malignant or develop into malignant angiosarcoma.[2]

IHH is often accompanied by vascular malformations of the skin; its pathological changes are similar to those of the hepatic artery to the hepatic vein or hepatic artery to portal vein arteriovenous fistula. Patients with a large degree of arteriovenous shunting may experience congestive heart failure,[3] and the prognosis of the disease is poor when complications are present and the mortality rate can be as high as 90%.[4] Reports[5,6] have suggested that large IHH is usually fatal to the infant with compromised congestive heart failure which is often an end-stage event. We report a patient with multiple IHH associated with congestive heart failure who was treated at our hospital.

Case report

A 59-day-old female infant with multiple cutaneous hemangiomata was admitted to our hospital because of abdominal distention and anemia. Her alanine aminotransferase level was elevated to 278 U/L and the serum alpha-fetoprotein level was up to 22 734 ng/mL. Her hemoglobin level was 97.00 g/L. The other laboratory tests were not remarkable. Chest X-ray revealed an enlarged heart shadow, and ultrasonography showed multiple intrahepatic lesions with low-attenuation masses (Fig. 1A). The hepatic vein and inferior vena cava (IVC) were markedly expanded (Fig. 1B), and color doppler ultrasound revealed dilated draining veins in the tumor with a high-velocity flow (Fig. 1C&D). Echocardiography demonstrated right heart enlargement and hypertrophy, and pulmonary hypertension (120 mmHg) (Fig. 1E&F). Contrast-enhanced MRI showed mild enhancement of lesions and more obviously delayed enhancement (Fig. 2). The patient was subjected to surgery plus hormone therapy, but she died of congestive heart failure and pulmonary hypertension. Pathological examination revealed IHH

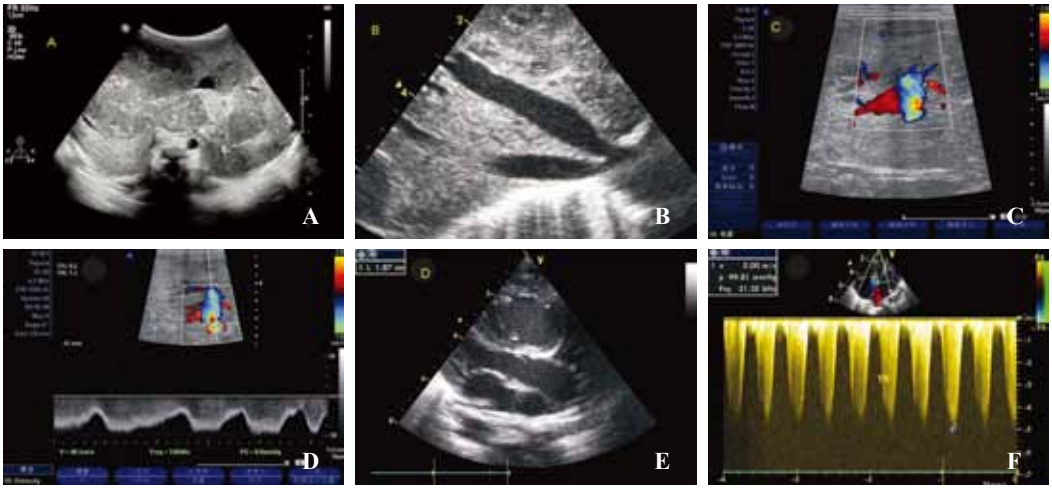

Fig. 1. A: Ultrasonograpy revealing multiple intrahepatic lesions with low-attenuation masses; **B:** Ultrasound image revealing the hepatic vein and the inferior vena cava were markedly expanded; **C&D:** Color doppler ultrasound inflow image of the dilated draining veins in the tumor demonstrates a high-velocity flow; **E&F:** echocardiography study of the enlarged right heart and pulmonary hypertension. RV: right ventricle; AAO: ascending aorta; LVOT: left ventricular outflow tract; LA: left atrium; TR: tricuspid regurgitation.

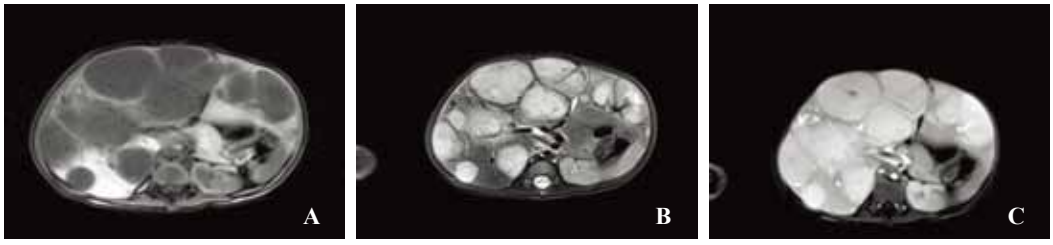

Fig. 2. MRI characteristics of the patient with infantile hepatic hemangioma. **A:** Axial T1-weighted image of liver showing multiple well-defined spherical masses; **B:** Axial T2-weighted image of liver showing multiple well-defined spherical strongly hyperintense masses; **C:** Axial gadolinium-enhanced T1-weighted image of liver showing homogeneous enhancement of hepatic masses.

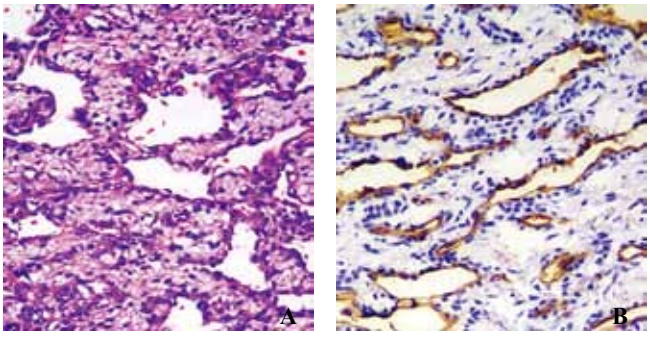

Fig. 3. The pathological results of the patient with infantile hepatic hemangioma. **A:** anastomosing vessels with atypical endothelium; **B:** immunohistochemistry with CD34.

in the right liver lobe: anastomosis of vessels to the atypical endothelium and CD34 immunohistochemistry positive (Fig. 3).

Discussion

Percutaneous biopsy is contraindicated for IHH because of the risk of massive hemorrhage. The diagnosis of IHH relies on clinical findings and imaging studies. IHH associated with cardiac insufficiency has more severe clinical symptoms. In this report, the patient had manifestations of abdominal distention, anemia, and multiple cutaneous hemangiomas at distant sites.

A precise imaging of IHH is essential.[7] Imaging examinations performed to diagnose multiple IHH with congestive heart failure include ultrasonography, echocardiography and MRI. Each of these imaging modalities reveals characteristic findings in this condition. In our patient, ultrasonography revealed multiple intrahepatic lesions with mixed echo patterns, and that the hepatic vein and IVC were markedly expanded. Color doppler ultrasonography showed dilated draining veins in the tumor.[8] Echocardiography showed right ventricular enlargement and hypertrophy, and pulmonary hypertension. MRI revealed multiple foci with early nodular enhancement, and delayed scan showed gradual enhancement from the periphery to the center and more obviously delayed enhancement.[6]

Multiple IHH associated with congestive heart failure is characterized by an early age of onset, and the disease condition is severe. Because pulmonary hypertension may be a manifestation of IHH, pediatric

patients with unexplained pulmonary hypertension should undergo ultrasound examination. When multiple IHH is detected by ultrasonography for the first time, the hepatic vein and IVC should be observed carefully, and echocardiography should be performed to provide detailed information about the disease. Therefore, for infants with severe and complex conditions, early or even bedside ultrasonography and echocardiography should be performed so as to detect multiple IHH complicated by congestive heart failure early, the disease condition should be assessed to facilitate the diagnosis and treatment.

Characteristic MR findings in this disease may be useful in differentiating this disease from other space-occupying lesions of the liver to make a correct diagnosis and a prognostic evaluation. Therefore, this disease should not be misdiagnosed as liver cancer or metastatic liver tumor, so that unnecessary surgery or radiotherapy and chemotherapy can be avoided. Metastatic liver lesions of hepatoblastoma and neuroblastoma are often diffuse or multiple, their images are often similar to those of IHH; alpha-fetoprotein is significantly elevated in hepatoblastoma. In hepatoblastoma, ultrasonography shows hyperechoic tumors separated by hypoechoic areas or areas of cystic necrosis. In 40%-50% of cases, scattered or clustered calcified foci may be seen.[9] Thus, it is difficult to differentiate IHH from hepatoblastoma and neuroblastoma only by ultrasonography. The main characteristics are the enhancement patterns of MRI. Metastatic liver lesions of hepatoblastoma and neuroblastoma are characterized by rapid filling and excretion, which are markedly different from the enhancement characteristics of IHH, rapid filling and slow excretion. In addition, metastatic liver lesions of hepatoblastoma and neuroblastoma are not accompanied by expansion of the hepatic vein and the IVC, enlarged right heart, or pulmonary hypertension. Therefore, echocardiography is conducive to the differentiation of IHH. Additionally, liver metastasis of neuroblastoma is derived from a primary lesion, and the level of 24-hour urinary vanillyl mandelic acid is elevated, which may assist in differential diagnosis.[3,9]

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Competing interest: All authors disclosed no competing interest.

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