CASE REPORTS

INFANTILE HEPATIC HEMANGIOENDOTHELIOMA: FINDINGS IN THE LIVER WITH SPECTRAL AND COLOR DOPPLER SONOGRAPHY

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SUMMARY: We present a case of infantile hepatic hemangioendothelioma (IHH) in whom abdominal sonography, CT, and later, angiography were performed. Abdominal sonography demonstrated multiple diffuse clearly defined hypoechoic areas in both lobes of the liver. Enlarged vessels around the hypoechoic lesions, and high velocity and disturbed blood flow within the lesions were detected with Doppler sonography. Arteriovenous shunting was also shown in the right lobe of the liver. Although these findings are nonspecific for IHH, in the appropriate clinical setting, Doppler ultrasound (US) findings revealing AV shunts are significant indicators of IHH.

Key Words: Liver Neoplasms, Hemangioendothelioma, Adolescence, Ultrasonics.

INTRODUCTION

Infantile hepatic hemangioendothelioma (IHH) is the most common benign vascular liver tumor encountered in infancy which may be either solitary or multicentric. No genetic transmission is known. The majority of the patients with hepatic hemangioendotheliomas present before the age of six months and there is female predominance varying from 1.3:1 to 2:1 (1). The majority the patients are under six months of age. It is a rare vascular anomaly of the liver that usually presents in infants with hepatomegaly, high-output congestive heart failure and cutaneous hemangiomas. IHH has also been occasionally associated with thrombocytopenia, hemorrhage caused by rupture, anemia, consumptive coagulopathy and obstructive jaundice (2). Although the sonographic, angiographic, tomographic and magnetic resonance abnormalities of the liver in IHH are well known, description of duplex and color Doppler sonographic findings is limited. We present a case in which the diagnosis was established with duplex and color Doppler sonography and correlated well with computed tomography (CT) and angiography.
CASE REPORT

A 6-month-old, previously healthy girl was referred to our hospital for evaluation of right-upper-quadrant mass and hepatomegaly. The liver extended 4 cm below the right costal margin. The lungs and the heart were normal and there was no evidence of high-output congestive heart failure. Also, there were no cutaneous hemangiomas. Liver enzymes (SGOT and SGPT) were slightly elevated. Alpha-fetoprotein (AFP) level was also elevated (410 ng/ml). Blood sample demonstrated normochromic and normocytic anemia and thrombocytopenia. The level of bilirubin was normal.

Plain roentgenogram of the chest was normal. Abdominal sonography demonstrated hepatomegaly and multiple, clearly defined hypoechogenic areas of about 1-2 cm in diameter in both lobes of the liver (Fig. 1A). Bile ducts were not dilated, portal vein branches were normal and hepatic veins were slightly enlarged. Duplex and color Doppler sonography also revealed an enlarged hepatic artery and intrahepatic branches. Several tangled masses of vessels around hypoechogenic lesions were detected and we considered that they represented enlarged arteries (Fig 1B). Spectral analysis demonstrated high velocity and disturbed blood flow in the enlarged arteries. Arteriovenous shunting was shown in the right lobe of the liver that revealed a spectrally broadened, mixed arterial and venous, high-amplitude pattern (Fig 1C).

The CT scan before contrast medium administration revealed a diffuse infiltrative hypodense lesion in the liver. In the lesion, also, there were more hypodense, local, round or oval distinct areas. There were lower density values measured in the lesion than those of surrounding liver (Fig 2A). After intravenous bolus injection of contrast medium, the lesion generally enhanced to a greater degree than the normal liver but more hypodense local areas in the lesion did not enhance (Fig 2B). On delayed CT scan, the lesion became isodense with the surrounding normal liver. CT also revealed narrowing of the aorta, distal to the celiac artery and enlargement of the hepatic artery.

On angiographic examination, the distal segment of the abdominal aorta was significantly narrow compared to the proximal segment (Fig.3A) Celiac root, and hepatic artery were

Fig 1: (A) US image shows multiple clearly defined hypoechogenic lesions of about 1-2 cm in diameter in both lobes of the liver. Some of the lesions tend to join together. (B) Color Doppler US image shows several tangled masses of vessels around hypoechogenic lesions. (C) Duplex Doppler image reveals arteriovenous shunting with a spectrally broadened, mixed arterial and venous, high-amplitude pattern in the vascular lesion located in the right lobe of the liver.
Fig 2: (A) CT scan shows a diffuse infiltrative hypodense lesion in the complete left and partial right lobe of the liver. In the lesion there are more hypodense local areas that correspond to hypoechoic areas in US images. (B) Contrast enhanced CT scan shows that lesion enhances generally. More hypodense areas do not show enhancement.

Fig 3: (A) Significant narrowing of the aorta distal to the wide hepatic artery is observed at the angiographic image. (B) Multiple, small, contrast media collection areas are observed in the complete left and partial right lobe of the liver.

DISCUSSION

There is some argument about the definition and classification of IHH. A variety of names including vascular malformation, capillary hemangioma, cavernous hemangioma, and hemangioendothelioma have been described by different authors. A widely used classification by Dehner and Ishak (3) divides this tumor into hemangioendotheliomas (type 1 and 2) and cavernous hemangiomas. Mulliken and Young (4) suggest that hemangioendotheliomas and cavernous and capillary hemangiomas in infants are probably the same entity in different phases of
evolution. On the other hand Enzinger and Weiss (5) suggest the term hemangioendothelioma for vascular lesions that show a clinical course intermediate between those of hemangioma and angiosarcoma. The classification and terminology of Enzinger and Weiss was used for this report.

There are reported studies that the diagnosis of IHH was established from a synthesis of the clinical presentation, laboratory tests and results of imaging examinations (2,6,7). Since the diagnosis of IHH is more infrequent, liver biopsy was not necessary to confirm the diagnosis and it might result in significant internal bleeding. Although histologically benign, hemangioendotheliomas may grow rapidly. Malignant degeneration has also been reported (8). In the study of Selby et al. (1) the 6-month survival rate based on 71 patients was 70%. Mortality may approximate 70% in complicated cases. Death usually results from high-output cardiac failure secondary to arteriovenous shunting within the tumor, but respiratory compromise, hepatic failure, hemorrhage, and disseminated intravascular coagulation may also be responsible (2). Jaundice, multiple tumor nodules and histological absence of cavernous differentiation are also prognostic indicators of death (1). If the status of the patient is not significantly complicated by AV shunting, IHH eventually undergoes spontaneous resolution (9). Interestingly, serum AFP level was elevated in our case. It was reported by Seo et al. (10) that it occasionally could be elevated in IHH.

Ultrasonography for hepatic masses in infants and children is not specific. Therefore, integrated imaging of hepatic tumors in childhood is necessary. Ultrasonographic appearance of IHH can be defined as a solitary or multicentric, generally hypechoic, sometimes hyperechoic, complex liver mass, with large sonoluent areas throughout the sinusoids. A dilated proximal abdominal aorta, hepatic artery and large draining hepatic veins on ultrasound, as in our case, indicates arteriovenous shunting within the hepatic mass that shows a hemangiomatous tumor (11). Increased blood flow is not pathognomonic for IHH because it can also be seen with hamartomas and malignant liver tumors (9).

One of the liver lesions which must be differentiated from IHH is mesenchymal hamartoma (MH). MH is mainly cystic in appearance on US, with dense echoes scattered throughout the lesion (11). Confusion on the basis of hypervascularity is more likely to occur with hamartoma than with malignant liver tumors (11). Since MH is a highly vascular tumor, unless cystic changes are present, differentiation from IHH may be difficult, but there is very little sinusoid formation and sinusoidal pooling in hamartomas. Therefore, arteriovenous shunting is not generally an expected finding.

In their study of infantile hepatic hemangiomata (that also covers IHH in the same title), Paltiel et al. (6) found different arterial and venous spectral patterns in separate portions of a single tumor with high peak Doppler shifts or similar to those of normal hepatic parenchyma or vessels with little systolic-diastolic variation that shows AV shunts. They found that there was no correlation between the tumor size and peak Doppler shift, and between the presence or absence of CHF and either tumor size or peak Doppler shift. They also stated that there was no specific Doppler finding for infantile hepatic hemangiommas (6).

We believe that arteriovenous shunting is a significant Doppler finding for IHH, particularly in infants with high-output heart failure. Although arteriovenous shunting in the liver could be seen in hepatocellular carcinoma (HCC), hereditary hemorrhagic telangiectasia (HHT), hepatoblastoma, and IHH, all these diseases have different clinical, laboratory and imaging findings. Duplex Doppler and as well as color Doppler can very clearly outline the vascular nature of these diseases. HCC is usually found in children above five years of age (11). In HHT, despite possible arteriovenous fistulas or arteriovenous malformations in the liver, clinically, a triad of telangiectasia, recurrent epistaxis, and a family history of disorder are expected. Hepatoblastoma is generally a hyperechoic solitary lesion but occasionally may be multiple or diffuse and have rarely arteriovenous shunting (11).

Doppler US findings such as high systolic-diastolic velocity in enlarged hepatic arteries, in the vascular structures within or near the liver lesions and arteriovenous shunts are nonspecific for IHH. These findings can overlap with malignant tumors. We suggest that in
addition to appropriate clinical, laboratory and other imaging investigations, Doppler US findings, especially arteriovenous shunts are positive indicators of IHH.

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REFERENCES