

A Rare Cause of Hepatomegaly: Infantile Hepatic Hemangiomendothelioma

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ABSTRACT

Infantile hepatic hemangiomendothelioma, a bening vascular neoplasia derived from endothelial cells, is the most common newborn tumor located in the liver. The tumor has female predominancy. Mostly, it is associated with a good prognosis and may show spontaneous regression by the time the infant is 1 year of age. Although most of the patients are asymptomatic, abdominal mass, hepatomegaly, jaundice, nausea, and vomiting can be encountered among common clinical findings. Herein, a 1.5-month-old boy with abdominal distention was referred to our gastroenterology unit for hepatomegaly. Non-invasive diagnostic modalities revealed infantile hepatic hemangioendothelioma.

Keywords: Infantile hemangiomendothelioma, liver, childhood

INTRODUCTION

Infantile hepatic hemangiendothelioma (IHH) is the third most common hepatic tumor in children with a prevalence of 1% and an incidence of 0.4–1.9/1,000,000/year. It is the most common benign vascular neoplasm that originates from endothelial cells during infancy. Besides the liver, it can be seen in tissues such as the lung, spleen, brain, ovaries, heart, central nervous system, and bone (1-4). Of the cases, 85% are children younger than 6 months and 40%–50% have hemangiomas in their skin (5-8). It is reported to be seen more commonly in girls than in boys at a rate of 2/1-3/1 (4, 6). Hepatic involvement of hemangioendothelioma may be in the nodular or diffuse type, and histopathologic calcification may be seen in 50% of cases. While nodular hepatic involvement is seen in the early phase of the disease, most patients have multiple hepatic involvement. In later stages, extensive involvement is observed with the enlargement of the nodules (8). Infantile hemangioendothelioma is usually benign and can spontaneously become smaller up to 1 year of age; however, it is reported that sarcomas are sometimes observed (4, 8). Among the hepatic vascular neoplasms, infantile hemangioendothelioma is the one that most commonly causes symptoms (9). Clinical findings occur when the child is 47 days old on average (1-365 days), and more than 80% of the patients are diagnosed in the first 3 months of life. Although most cases are asymptomatic, clinical findings vary depending on the tumor size and location. Symptoms such as hepatomegaly (83%), abdominal mass (66%), hemangiomas in the skin (65%), loss of appetite, vomiting (25%), heart failure, anemia, thrombocytopenia, coagulopathy, and jaundice may be seen (5, 10, 11). Visualization methods are very useful in the diagnosis. While single or multiple hypoechoic calcified lesions in the liver are observed in ultrasonography, computerized tomography, magnetic resonance

imaging, and liver biopsy, which is an invasive method, are helpful in diagnosis (12-14). Histologically, this tumor, which is in a benign appearance, causes severe life-threatening complications such as heart failure at a rate of 70% (10, 14).

Here we report the case of a 1.5-month-old male infant who was brought with a diagnosis of infantile hemangioendothelioma and with a complaint of abdominal swelling and decreased weight gain and was handled through clinical and radiological methods, along with the review of literature.

CASE REPORT

A 1.5-month-old male patient was brought to the pediatric polyclinic with the complaint of abdominal swelling. It was found in his history that his parents were unrelated, he was born mature and weighing 3500 g through spontaneous normal vaginal delivery after a followed unproblematic pregnancy, and he did not have any features in the postnatal period; abdomen swelling was noticed on the 20th postnatal day, and despite the fact that they consulted various doctors, no diagnosis could be made and they visited to our clinic because the abdomen swelling increased gradually. On physical examination, the general condition of the patient who weighed 3800 g (10-25 p) and was 54 cm (25-50 p) tall was moderate and he had a pale appearance; a 1×1 cm purple hemangioma that did not fade with compression and had a cauliflower appearance was detected on the left thoracic part of the vertebra. In the cardiovascular system, the cardiac apex beat was 140/min and rhythmic, and a 2/6 pansystolic murmur was detected at the apex. Respiratory sounds were bilateral and natural, and the respiratory rate was 44/min. His abdomen was distended, and the liver was palpated as 4 cm in the midclavicular line. The spleen was not palpable.

In the laboratory tests, the Hgb level was found to be 8.8 g/dL, Hct level was 24%, white blood cell count was 7500/mm³, and PLT count was 195,000/mm³. His AST level was 120 U/L, ALT level was 18 U/L, ALP level was 170 U/L, GGT level was 10 U/L, total protein level was 5 g/dL albumin level was 3.7 g/dL, iron level was 42 ng/ dL, iron-binding capacity was 200 ng/DL, ferritin level was 65 ng/ mL, IgA level was 190 mg/dL, IgG level was 1040 mg/dL, IgM level was 170 IU/mL, prothrombin activity was 51%, and INR was 1.7. In the peripheral spread, erythrocytes were hypochromic and platelet clusters were observed, while atypical cell and hemolysis findings were not observed. Cardiac functions were evaluated as normal in the echocardiography of the patient. In the ultrasonography of the abdomen, in both lobes of the liver, a 24×17×14 mm smooth-contoured hypoechoic solid nodule was observed in segment 2 and an 18×18 mm smooth-contoured hypoechoic solid nodule was observed in segment 4. Peripheral and intralesional flows were normal in Doppler ultrasonography. In the abdominal MRI examination, solid lesions that were in the left lobe of the liver, the largest of which was 2 cm in diameter and showed a diffuse contrast material involvement, were evaluated as multiple hemangiendomas and infantile hemangioendotheliomas (Figure 1a, b). Hepatitis markers and TORCH IgM were negative, and neuron-specific enolase (NSE) and vanilmandelic acid (VMA) levels in the blood and urine were normal. Liver biopsy was not performed because the patient had low prothrombin activity and the lesion was found consistent with infantile hemangioendotelyoma through radiological imaging methods. The patient was transferred to the oncology polyclinic for further follow-up and treatment. The patient, who is still clinically asymptomatic, is under observation without any treatment from the oncology clinic. Verbal consent was received from the mother of the patient.

DISCUSSION

Childhood liver tumors constitute 2%–3% of all childhood tumors. Infantile hemangioendothelioma is the third most common benign vascular tumor, occurring at a rate of 12% among childhood liver tumors (4, 8). Although most patients are asymptomatic, the clinic occurs in the first 6 months in approximately 85% of them (4). Although the clinical findings change depending on the tumor location and size, jaundice, hepatomegaly, and abdominal distension are frequently encountered (2, 5). Although it is reported to be more common in girls, our patient was a 1.5-monthold male who was brought with the complaints of abdominal distention and hepatomegaly, which are among the frequent clinical findings. As in our case, anemia is a common hematological abnormality in thrombocytopenia.

While liver biopsy or angiography is used for the diagnosis, diagnosis is now possible with non-invasive techniques by taking advantage of typical contrasting features. Imaging techniques such as USG, color Doppler USG, CT, MRI, and angiography are used for the diagnosis (12-14). Ultrasonography is generally regarded as the first step. Ultrasonographic imaging of infantile "hemangioendothelioma" frequently reveals well-circumscribed hypoechoic lesions and abnormal large vascular structures in the liver, infarct, hemorrhage, calcified focus, and fibrous tissue (12). In our case, as shown in the literature, while smooth-contoured hypoechoic solid nodules were observed by ultrasonography in the liver, the MRI examination showed that these lesions were



Figure 1. a, b. (a) It is apparently observed as hyperintense on T2weighted MRI. (b) In the coronal image, it is observed as common heterogeneous hyperdense lesions from which the hepatic artery originates.

multiple hemangiomas showing diffuse contrast material involvement. The patient was diagnosed as having infantile hemangioendothelioma with non-invasive imaging methods without the need for liver needle biopsy.

While asymptomatic benign infantile hemangioendothelioma usually undergoes spontaneous regression of 70% without treatment within 1 year, it may sometimes require treatment in symptomatic patients. In symptomatic cases, although the treatment options vary according to the patient, medical or surgical options are the final choice. The indications for treatment of infantile hemangioendothelioma are coronary failure, difficulty in breathing, coagulopathy, abdominal compartment syndrome, and deterioration of hepatic function test results (4). Because our patient, who we followed for 9 months clinically, remained asymptomatic, no medical treatment was applied.

CONCLUSION

Here we emphasized that infantile hemangioendothelioma, which is a benign tumor of the liver with a fairly good prognosis, should be kept in mind and can be diagnosed by ultrasonography, CT, or MRI. Further, invasive procedures can be avoided using radiological imaging methods.

Informed Consent: Verbal informed consent was obtained from patients' parents who participated in this case.

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