

Case Report

Infantile Hepatic Hemangioendothelioma with Complete Response to Propranolol Monotherapy

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Abstract

Infantile Hepatic Hemangioendothelioma (IHH) is an extremely rare and painless vascular tumor. IHH is usually in benign nature but it can also be life-threatening with a rapid growth tendency. There are approaches in the treatment of IHH, ranging from drug therapy to liver transplantation. Propranolol, which is used in first-line therapy for most patients, is effective alone in the treatment.

Keywords: Infantile hepatic hemangioendothelioma, propranolol, massive hepatomegaly

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In childhood; the most common benign tumors are vascular tumors and most of them are hemangiomas. IHH rates less among vascular tumors. Blood vessels are lined with epithelioid cells that begin to grow abnormally. IHH mostly presents in the first 6 months of life. There is usually a large palpable abdominal mass with hepatomegaly and abdominal distention. Prognosis is good usually but it may cause even death due to rapid growth of the mass, heart failure, arteriovenous shunt or coagulopathy. In this study; We aimed to share the experience of successful treatment with single propranolol in an infant with IHH and to review the current literature on the treatment of IHH.

Case Report

A two-month-old female patient with dyspnea and abdominal bloating was evaluated in the outpatient clinic. Antenatal ultrasonography of the patient, who was a term baby and weighed 3200 grams, was normal. On abdominal

examination, massive hepatomegaly was remarkable. Skin examination revealed a 2x1 cm hemangioma on the forehead. No bleeding tendency or signs of heart failure were observed. Thyroid function tests were normal. In laboratory tests, alpha fetoprotein (AFP) was 1210 ng/ml (normal for AFP age: 323±278), liver function tests and coagulation parameters were normal. Abdominal ultrasound revealed multiple diffuse mass lesions in both lobes of the liver, the largest of which was 4.5x3.7 cm in size, containing heterogeneous areas. Dynamic computed tomography (CT) showed multiple mass lesions in the liver, the largest of which was 5 cm, with a liver size of 100 mm and larger than normal. The diagnosis of IHH was confirmed by trucut liver biopsy. Oral propranolol treatment was started with an initial dose of 1 mg/kg/day for 1 month and was gradually increased to 3 mg/kg/day over two months. On the tenth day of treatment, rapid clinical improvement was observed, respiratory distress resolved, and there was a marked improvement in abdominal bloating. On the 40th day, AFP decreased by 54%

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(565 ng/ml) in the control, while the reduction in lesion diameters by USG was 20%. At the end of 3 months, while the reduction in mass was over 80% with CT (Fig. 1), there was a 90% reduction in AFP level (120 ng/ml). No significant side effects were recorded during the treatment. Currently, patient is 18 months old, under follow-up, AFP: 10 ng/ml (normal for age). On dynamic computed tomography (CT), diffuse and heterogeneous mass lesions were observed in both lobes, the largest of the lesions was 1.1 cm, the liver size was 65 mm and normal, with smooth contours. The patient's written informed consent was obtained for the publication of the case report (clinical details and images) concerning his family.

Discussion

The incidence of IHH in the general population is <0.1/100000. In IHH, diffuse lesions are usually nodular and hepatomegaly develops. They cause abdominal compartment syndrome by compressing the inferior vena cava, diaphragm and surrounding tissues. The mortality rate at this stage is high if not treated. IHH should be differentiated from hepatic hemangioma, hepatocellular carcinoma and metastatic tumors by imaging or biopsy. When we look at the literature, we see that there are many different treatments for IHH. IHH is rare, so studies in large study groups with different treatment options are limited. Different treatment options for IHH include steroids, propranolol, surgical resection, and liver transplantation. Vascular endothelial growth factor (VEGF) is thought to play a role in the growth

of IHH. VEGF inhibitors such as bevacizumab, sorafenib and pazopanib have also been tried in the treatment of IHH.^[1,2] In one study, 4 of the pediatric patients with IHH in the liver/lung died with rapid progression. Complete remission was achieved with 12 complete liver resections.^[3] In another study, propranolol and prednisolone were administered to 9 pediatric patients diagnosed with IHH and good response was obtained.^[4] Liver transplantation was performed in a patient who was also diagnosed with hepatic hemangioendothelioma that was not suitable for resection, and a 4-year survival was observed after transplantation.^[5] In these patients, the need for surgical resection and liver transplantation can be eliminated with propranolol treatment. They can be successfully treated with propranolol alone, avoiding the side effects of steroids. IHH is a potentially fatal condition; the successful results obtained with propranolol is markable to suggest a drug easy to use, cheap and have few side effects, are promising and shows the need for further studies and contributions in the literature.

Disclosures

Informed Consent: Written informed consent was obtained from the parents of the patient for the publication of the case report and the accompanying images.

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Conflict of Interest: None declared.

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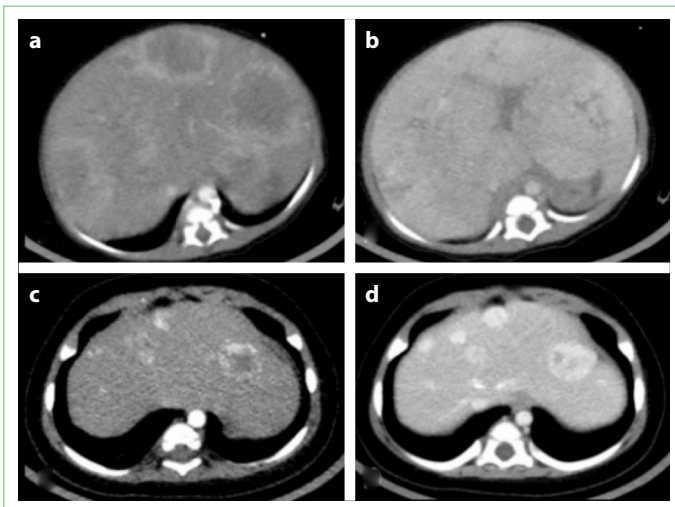


Figure 1. On early arterial phase after intravenous iodine based contrast material administration, axial CT image (a) shows multiple peripherally enhancing solid masses in both lobes of the liver. On late venous phase (b), masses become entirely hyperintense due to filling with contrast material. On early arterial phase (c) and, late phase (d) CT images that obtained after treatment and the diminished sizes of the masses are well depicted.