Symptomatic Infantile Hepatic Hemangioendothelioma Successfully Treated with Steroid

Malay Kumar Dasgupta, Sabyasachi Das, Chaitali Patra, Shatanik Sarkar
Department of Paediatric Medicine, Radha Gobinda Kar Medical College and Hospital, Kolkata, West Bengal, India

ABSTRACT

A case of hepatic hemangioendothelioma presenting as congestive cardiac failure in a neonate is being reported which was managed successfully with oral prednisolone, resulting in improvement of symptoms and regression of tumor within 3 months.

Key words:
Congestive cardiac failure, congestive cardiac failure, hemangioendothelioma, liver tumor, prednisolone

INTRODUCTION

Benign infantile hemangioendothelioma (IHH) is a rare neonatal benign vascular tumor usually located in the liver. Most of the lesions are diagnosed in the 1st few months of life. They may undergo spontaneous regression but can sometimes be life-threatening due to congestive cardiac failure (CCF) and/or consumptive thrombocytopenia and coagulopathy. While in asymptomatic cases, observation is the recommended course, for patients with symptoms optimum management strategy is still controversial. We report a sick neonate with early heart failure caused by infantile hepatic hemangioendothelioma (IHH) which has well been controlled with prednisolone therapy.

CASE REPORT

One day old male infant, after an uncomplicated pregnancy and delivery, was referred from the periphery with a presenting feature of poor feeding and respiratory distress. There was no antenatal ultrasonography report available. Examination revealed an irritable child, weighing 2.7 kg, in respiratory distress with relative ratio about 120/min, hazard ratio 180/min, and a high-volume pulse. There was cardiomegaly, a grade 3/6 ejection systolic murmur in the tricuspid area and a left ventricular S_3. There were fine crackles over both lungs at the basal region. Liver was soft, 4 cm below the costal margin at the right mid clavicular line. A large firm mass, with irregular surface was palpable over left hypochondrium, epigastric and left lumbar region measuring about 8 × 5 cm, not bimanually palpable.

The complete hemogram were normal. There was evidence of prerenal azotemia (blood urea 47 mg/dL and creatinine 0.94 mg/dL). Serum bilirubin, liver enzymes, and serum alpha fetoprotein (AFP) (1.5 mcg/mL) were essentially normal.

The chest X-ray showed cardiomegaly and pulmonary venous congestion. The electrocardiography was normal except sinus tachycardia. Echocardiography showed left and right ventricular dilatation, with normal valves and intact septae. There was no patent ductus arteriosus or coarctation. Ejection fraction (EF) was 58% along with mild tricuspid regurgitation and trivial mitral regurgitation.

Ultrasonography of the abdomen showed a well-defined lump measuring 8.5 × 5.2 cm, having marked internal vascularity in the upper abdomen, separated from spleen and kidney. Liver was enlarged with hugely dilated left hepatic vein (LHV). Intrahepatic biliary radicals and common bile duct were also dilated [Figure 1].

Then computed tomography (CT) scan of the abdomen showed features suggestive of hemangioendothelioma of liver arising from the left lobe exophytically. Focal calcifications were present; LHV was dilated and there was narrowing of aorta below the level of the celiac artery [Figure 2].

Cardiac failure was managed with fluid restriction, oxygen, diuretics, and inotropic support for 2 days. But there

Address for correspondence:
Dr. Sabyasachi Das,
Gopalpur (Charaktola), Post- Sarkarpool,
Kolkata - 700 143, West Bengal, India.
E-mail: dasbabai83@gmail.com

Access this article online

Quick Response Code:

Website: www.jcnonweb.com

DOI: 10.4103/2249-4847.123099

was no clinical improvement noted. Then after studying different literature, oral prednisolone was started in a dose of 4 mg/kg/day in two divided doses. There was a dramatic improvement in general physical condition of the child. Prednisolone was tapered to 4 mg/kg/day on alternate days after 2 weeks, 2 mg/kg on alternate days by 6 weeks and 1 mg/kg on alternate days at 3 months. At 3 months follow-up, the child was gaining weight steadily, size of the mass had decreased considerably, and echocardiography showed normal EF (65%). Follow-up USG showed a decrease in vascularity and size of the hemangioendothelioma (3.2 × 2.1), but LHV was still dilated.

DISCUSSION

Hepatic tumors in children are relatively uncommon (about 2-3% of all pediatric tumors).[1,2] However, IHH is the third most common hepatic tumor in children (12% of all childhood hepatic tumors), the most common benign vascular tumor of the liver in infancy, and the most common symptomatic liver tumor during the first 6 months of life.[3-4] Approximately 85% of affected patients present by 6 months of age,[4-6] and in about 45-50% of cases these patients also have cutaneous hemangiomas.[3,4-6] The tumor has a 2:1 female predilection.[3,5,6] The lesions may be single or multiple, and calcifications are seen at histopathologic analysis in 50% of cases.[4] IHHs are usually benign, but malignant sarcomas have been reported to arise in existing hemangioendotheliomas.[3,4,7] Most tumors continue to grow during the 1st year of life and then spontaneously regress, probably due to thrombosis and scar formation.[3,4,6,7]

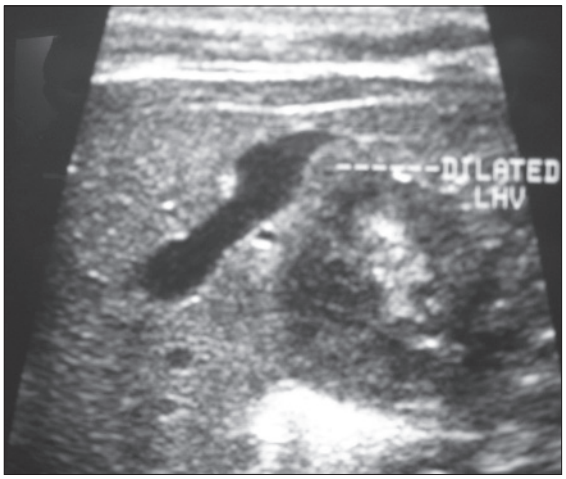

Figure 1: USG of abdomen showed enlarged liver with hugely dilated LHV. IHBR and CBD were also dilated

Clinical manifestations of IHH are variable and mainly depend upon the tumor size and location[2,3] and include hepatomegaly (83%), an abdominal mass (66%), skin hemangioma (65%), anorexia, vomiting (25%), and failure to thrive.[8]

In most of the cases, IHH remains asymptomatic and detected in ultrasound of abdomen by chance. But, sometimes, it can cause severe symptoms such as abdominal distension, gross hepatomegaly, severe arteriovenous (AV) shunting with CCF, anemia, thrombocytopenia (Kasabach-Merritt syndrome), consumptive coagulopathy, intraabdominal hemorrhage, and so on. Rarely, biliary obstruction with jaundice, vomiting, and gastric outlet obstruction[8] has been reported.

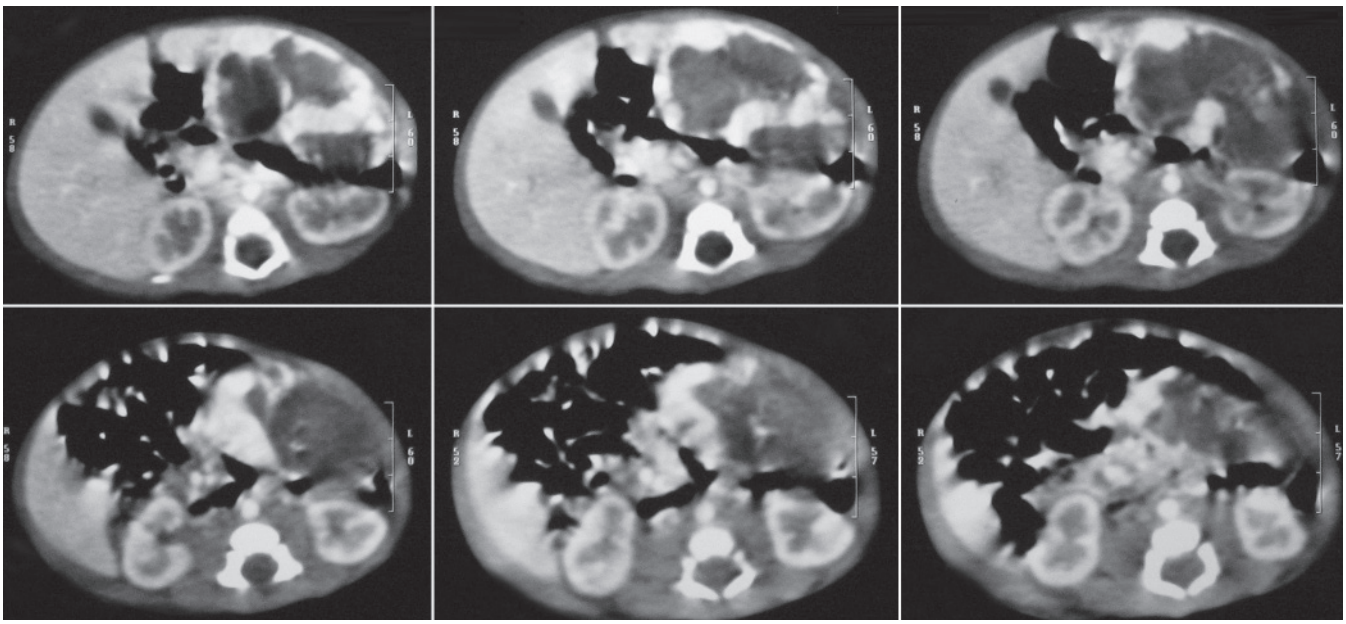

Figure 2: CT scan of abdomen showed Haemangioendothelioma of liver arising from left lobe exophytically. Focal calcifications were present, LHV was dilated and there was narrowing of aorta below the level of celiac artery

Radiological evaluation is useful for patient diagnosis, with sonography often being the initial diagnostic modality. On sonography, IHH is characterized by discrete, hypoechoic lesions (either solitary or multiple) within the liver that may have calcifications or shunting on Doppler evaluation. A more definitive diagnosis requires either a contrast-enhanced CT (CECT) or an magnetic resonance imaging (MRI). While CECT shows a hypodense area which enhances with contrast, MRI may identify IHH as low-signal lesions on T1 and high-signal lesions on T2-weighted images. According to clinical presentations, radiological evaluations and liver biopsy, the major differential diagnoses should include that most cases of IHH are asymptomatic with spontaneous regression. But cases with heart failure experience a high mortality rate (up to 70%).[8] hepatoblastoma, mesenchymal hamartoma, and hepatocellular adenoma.

Serum AFP may be high in case of IHH, though not up to the level seen in hepatoblastoma. In case, a malignant liver mass cannot be excluded beyond doubt, it may be reasonable to proceed with an open biopsy.

Fok TF[8] reported that most cases of IHH are asymptomatic with spontaneous regression. But cases with heart failure experience a high mortality rate (up to 70%).[8] Rapid AV shunting along with dilatation of hepatic arteries and recirculation of blood back to right heart may lead to overt heart failure (58%).[8]

There are confusions regarding the management strategies. Expectant treatment with serial USG to follow the size of the lesion can be done safely in children without any symptoms. Supportive medical care includes control of CCF, administration of blood products to correct anaemia, and coagulopathy. Systemic corticosteroids have become the mainstay in the treatment of hemangioendotheliomas.[9] They can be continued for 2-3 weeks and tapered slowly over 2-3 months. Yet, their mechanism of action is not well-understood. But it is thought that it may hasten involution by inhibiting proliferation of endothelial and smooth muscle cells. Daily doses of 2-3 mg/kg of prednisolone are usually given and some investigations have recommended higher doses like 5 mg/kg/day. This treatment results in dramatic shrinkage of the hemangioma in one third, but another third may show no response. Lesions unresponsive to steroids can be treated with alpha-interferon. Invasive measures like hepatic artery ligation or embolization[8] may be tried with rapid onset of severe symptoms. Small solitary tumours may be best treated by complete resection. In our experience, it is emphasized that prednisolone can be used to treat the patient with IHH complicated with early decompensated heart failure. Early aggressive treatment in symptomatic patient may produce a favorable outcome.

REFERENCES

1. Warvi WN. Primary neoplasms of the liver. Arch Path 1944;37:367-82.
2. Pariente D. The liver, biliary tract and spleen. In: Carty H, Shaw D, Brunelle F, Kendall B, editors. Imaging Children. Vol. 1. New York: Churchill Livingstone; 1994. p. 485-560.
3. Zenge JP, Fenton L, Lovell MA, Grover TR. Case report: Infantile hemangioendothelioma. Curr Opin Pediatr 2002;14:99-102.
4. Mortele' KJ, Vanzieleghem B, Mortele' B, Benoit Y, Ros PR. Solitary hepatic infantile hemangioendothelioma: Dynamic gadolinium-enhanced MR imaging findings. Eur Radiol 2002;12:862-5.
5. Ingram JD, Yerushalmi B, Connell J, Karrer FM, Tyson RW, Sokol RJ. Hepatoblastoma in a neonate: A hypervascular presentation mimicking hemangioendothelioma. Pediatr Radiol 2000;30:794-7.
6. Buonomo C, Taylor GA, Share JC, Kirks DR. Abnormalities of the hepatobiliary system. In: Kirks DR, Griscom NT, editors. Practical Pediatric Imaging: Diagnostic Radiology of Infants and Children. 3rd ed. Philadelphia: Lippincott-Raven; 1998. p. 954-79.
7. Sty JR, Wells RG, Starshak RJ, Gregg DC. The hepatobiliary system. In: Sty JR, Wells RG, Starshak RJ, Gregg DC, editors. Diagnostic Imaging of Infants and Children. Vol. 1. Gaithersburg: Aspen; 1992. p. 247-93.
8. Fok TF, Chan MS, Metreweli C, Ng PC, Yeung CK, Li AK. Hepatic haemangioendothelioma presenting with early heart failure in a newborn: Treatment with hepatic artery embolization and interferon. Acta Paediatr 1996;85:1373-5.
9. Cerar A, Dolenc-Strazar Z, Bartenjev D. Infantile hemangioendothelioma of the liver in a neonate. Immunohistochemical observations. Am J Surg Pathol 1996;20:871-6.

How to cite this article: Dasgupta MK, Das S, Patra C, Sarkar S. Symptomatic infantile hepatic hemangioendothelioma successfully treated with steroid. J Clin Neonatol 2013;2:187-9.

Source of Support: Nil, **Conflict of Interest:** None declared.