Infantile hepatic hemangioendothelioma in an 11-year-old child: A case report and review of literature

Abstract

Infantile hepatic hemangioendothelioma (IHH) is a rare benign mesenchymal tumor. As the name implies, it is commonly seen in infancy and has variable clinical presentation and outcome. This tumor can spontaneously regress or can have expansile growth replacing whole of the liver. In very large tumors, there may be extensive arteriovenous shunting within the lesion, resulting congestive heart failure (CHF) in a significant number of patients with poor outcome. Since there is no tumor marker for this tumor, the diagnosis can only be suspected on the basis of clinical features and radiological investigations from among the various mass occupying lesions of the liver in this age group, which includes hepatoblastoma, mesenchymal hamartoma or hepatocellular carcinoma. As a result, the management strategy cannot be defined and in the literature also there are isolated reports with diverse treatment protocols. The medical management includes corticosteroid, interferon, cytotoxic agents and irradiation or even the radical interventions such as hepatic artery ligation, transcatheter hepatic artery embolization. The surgical options are biopsy, complete resection of the lesion or even liver transplantation. We are reporting a case of IHH who was managed with surgical resection and postoperative corticosteroid. However, the patient had recurrence of the tumor and developed CHF, which led to the demise of the patient. The idea of reporting this case is that this patient was 11-year-old child who very unusual for this tumor at this age, and the surgical resection of this lesion should be radical rather than just excision biopsy as was done in the present case.

Key words: Arteriovenous shunting, infantile hepatic hemangioendothelioma, liver

INTRODUCTION

Infantile hepatic hemangioendothelioma (IHH) is a rare mesenchymal tumor of infancy and the reported incidence is estimated to be about 1/20,000.[1,2] The tumor has a 2:1 female preponderance and the clinical course is variable and depends on the tumor size, localization, and its complications. Most tumors continue to grow during the 1st year of life and then spontaneously regress, probably due to thrombosis, and scar formation. More often, however, the tumor is large and manifests as hepatomegaly, abdominal distention, or a palpable upper abdominal mass. There may be extensive arteriovenous shunting within the lesion, resulting congestive heart failure (CHF) in up to 50-60% of patients. Hematologic abnormalities may be seen, including anemia and especially thrombocytopenia caused by trapping of thrombocytes within the hemangioendothelioma with consumptive coagulopathy (Kasabach–Merritt syndrome).[3-5] There is no specific tumor marker for this lesion, so it is difficult for clinicians to make an accurate diagnosis, and one has to consider the differential diagnosis, which include hepatoblastoma and mesenchymal hamartoma. The diagnosis is suspected on the basis of clinical presentation and radiological investigations, which include ultrasonography and computed tomography (CT) scan of the abdomen.[6,7] Because of nonavailability of definitive criteria for specific diagnosis for this lesion so it is difficult to standardize a treatment protocol. There is always a need of surgical biopsy to confirm the diagnosis. The treatment options described in the literature are: Corticosteroid,[8] interferon,[9] Cytotoxic agents and irradiation[10] or even the radical interventions such as hepatic artery ligation,[11] transcatheter hepatic artery embolization,[12] surgical resection or even liver transplantation may be required in few cases.[13,14] Due to rarity of this lesion
the experience of management of this lesion is limited and only few case series are available where also the management is not tailored. We are reporting a rare case of IHH in an 11-year-old male child where the diagnosis could only be confirmed by surgical biopsy, but the patient developed complication of CHF and could not be salvaged. In this report, a review of literature will also be discussed.

CASE REPORT

An 11-year-old male child admitted with lump right side of abdomen for 1-month duration. Lump was 10 × 8 cm occupying the right hypochondrium and right lumbal region. Ultrasonography showed a complex heterogeneous predominantly hypodense septated, well-defined, irregular margnated mass in right lobe of the liver with dimensions 113 × 93.2 mm and volume 746.67 ml [Figure 1]. The septas of mass show vascularity. No evidence of calcification. Hb-8.5 g, total leukocyte count 16,000/cmm (80,17,0201). Platelets 1.5 lac. Renal and liver profiles were normal. HIV, HBsAg hepatitis C virus nonreactive. Contrast-enhanced CT (CECT) scan - a large solid/cystic mass with areas of hemorrhage and fluid filled levels in the right lobe of the liver. It also shows slight peripheral and septal enhancement [Figure 2]. A possibility of undifferentiated embryonal sarcoma/hepatoblastoma/hemangioendothelioma was kept. Preoperative findings a large tumor of size 11 × 10 × 7 cm arising from the right lobe of the liver. Excision biopsy of the tumor was done. Histopathological examination showed a tumor composed of numerous irregular vascular spaces lined by endothelial cells with fibrosis and myxoid changes along with cavernous foci. Areas of extramedullary hematopoiesis are also seen. Features suggestive of infantile hemangioendothelioma [Figure 3]. In the postoperative period, patient was discharged on 7th day and advised to take 2 mg/kg/day. In follow-up ultrasound (US) the lesion 4 × 3 cm (residual) in the right lobe. After 2 months, patient had recurrence of symptoms. Repeat CT scan-the lesion almost filled the right abdominal compartment and leaving only a rim of liver [Figure 4]. Patient developed features of right heart failure and could not be salvaged.

DISCUSSION

Hepatic tumors in children are relatively uncommon (about 2-3% of all pediatric tumors). However, infantile hemangioendothelioma 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. It is rarely reported in older children as present patient was an 11-year-old.

The clinical manifestation of infantile hemangioendothelioma is variable. The tumor may be asymptomatic and discovered...
incidentally. The clinical features of IHH mainly depend on the tumor size and location, and include hepatomegaly (83%), an abdominal mass (66%), skin hemangiomia (65%), anorexia, vomiting (25%), and failure to thrive (25%). The present case was admitted with a large painless lump in the abdomen.

On ultrasonography, infantile hemangioendothelioma appears as a complex, mostly solid hepatic lesion with variable hypo and hyperechoic echotexture. In cases of significant arteriovenous shunting, dilated hepatic vasculature with prominent blood flow at Doppler US is typical. Because of the increased vascular supply to these tumors, a striking decrease in the aortic caliber is often seen distal to the celiac artery origin. This can be taken as sign of significant arteriovenous shunting and can help in reaching a diagnosis. At unenhanced CT, infantile hemangioendothelioma usually manifests as a well-defined mass that is hypoattenuating relative to the normal liver parenchyma. In about 16-40% of cases, the lesion is heterogeneous with central high-attenuation areas due to hemorrhage or calcifications. At CECT, the enhancement pattern may resemble that of an adult giant hemangioma with “nodular” peripheral puddling of contrast material in the early phase, subsequent peripheral pooling, and central enhancement with variable delay. In larger tumors, central enhancement is often lacking due to fibrosis, hemorrhage, or necrosis. Conversely, small lesions, which tend to be multifocal, frequently enhance completely and typically do not demonstrate hemorrhage or necrosis. In the present case, CECT scan revealed only scanty enhancement defining the septas.

At unenhanced magnetic resonance imaging, the lesions have low signal intensity on T1-weighted images and high signal intensity on T2-weighted images. In tumors with arteriovenous shunting and high blood flow, flow voids may be observed on T2-weighted images. Because of the simultaneous presence of hemorrhage, necrosis, and fibrosis, the mass often appears heterogeneous on both T1- and T2-weighted images.

At angiography, a dilated and elongated hepatic artery and early filling of dilated draining hepatic veins may be seen in a hemangioendothelioma with significant arteriovenous shunting. Pooling of contrast material in large vascular spaces may be present.

At histopathologic analysis, infantile hemangioendothelioma manifests as a mesenchymal tumor composed of a connecting network of predominantly small diameter vascular channels lined by endothelial cells. Unlike cavernous hemangiomas, which contain larger blood-filled spaces with endothelial cell lining, infantile hemangioendotheliomas are not obviously vascular tumors at gross examination of the cut surface. Areas of varying degrees of hemorrhage, necrosis, calcification, thrombosis, or fibrosis are often present in large tumors. Two types of infantile hemangioendotheliomas have been identified on the basis of the tumor size and vascularity. Type I lesions are often calcified and consist of multiple small, vascular channels with an immature endothelial cell lining and a fibrous stromal separation containing bile ductules between the channels. Type II lesions have a more disorganized appearing endothelial cell lining and no stromal bile ductules. The present case was categorized as Type I hemangioendothelioma. However, it is not possible to evaluate the prognosis on the basis of the histopathology and Zhang et al. proposed to classify IHH into solitary and multifocal lesions rather into Types I and II.

Serious complications associated with this disease include high cardiac output CHF, consumptive coagulopathy, and thrombocytoopenia (Kasabach-Merritt syndrome). Rapid arteriovenous shunting through vascular channels leads to increased cardiac output. Dilation of hepatic arteries and recirculation of blood back to right heart may lead to overt heart failure (58%). In general, the median age of patients with hepatic hemangioendothelioma presenting with early heart failure in infancy is about 1-month old. Heart failure can cause death in up to 70% of untreated infants without adequate regression of the lesion.

The present case in this report also developed CHF and could not be salvaged, the unusual being the age of this patient, which is not reported in the literature.

Corticosteroid therapy is the first-step to treat complications of IHH. Prednisolone (2-10 mg/kg/day) for an average of 6 weeks or methyl prednisolone pulse therapy may hasten involution by inhibiting proliferation of endothelial and smooth muscle cells. The precise mechanism of action is unclear. When IHH is not responsive to high-dose steroids, interferon alpha-2a or alpha-2b (106 U/m2/day subcutaneous injection, with a therapy duration of about 3-6 months) may be another choice to inhibit endothelial cell proliferation, migration, and angiogenesis. Potential adverse effects include elevation of liver enzymes, bone marrow depression, alopecia, diminished appetite, and psychomotor regression. In addition, radiotherapy carries variable risks of other sequelae (e.g., cirrhosis, hepatosarcoma, and leukemia) so it should not be considered for young infants unless other therapies are contraindicated or unsuccessful. In addition, the experience of using cyclophosphamide for treating IHH complicated with CHF is scarce, and the striking response warrants further investigation. In rapid deterioration of cardiac status or medical therapy failure, one should consider an urgent interruption of the arteriovenous shunt, such as by hepatic artery embolization. Obliteration of the vascular channels to accelerate involution by the use of steel wire, platinum coils, and polyvinyl alcohol has been reported. Furthermore, if the large size and diffuse nature of the tumor render it unsuitable for embolization, operative hepatic artery ligation can be performed.

REFERENCES


How to cite this article: Kadian YS, Singh M, Rattan KN, Gupta V. Infantile hepatic hemangioendothelioma in an 11-year-old child: A case report and review of literature. Onc Gas Hep Rep 2014;3:36-9.

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