CASE REPORT

INFANTILE HEMANGIOENDOTHELIOMA OF LIVER: CASE REPORT
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HOW TO CITE THIS ARTICLE:

ABSTRACT: Infantile Hemangioendothelioma of the liver is an uncommon tumor. Two subtypes have been described. Type1 has recognizable but irregularly dilated vascular spaces in a fibromyxomatous stroma. Type 2 is described as having a more aggressive histology. It is marked by poorly formed and anatomising channels with papillae lined by atypical endothelial cells and is potentially malignant. Clinical presentation of IHHE is variable: parents or doctors can palpate hepatomegaly or abdominal mass, some hematologic abnormalities may be seen in laboratory investigation, or lesion may be asymptomatic and discovered incidentally. We report a case of 45 day old female child who presented with abdominal distension. CT abdomen showed hypoattenuating mass in the left lobe of the liver along with specks of calcification. The mass was excised and had large areas of gritty calcification simulating a teratoma. However histopathological diagnosis of infantile hemangioendothelioma type I of liver was made. Hemangioendothelioma of the liver has been reported as most common benign symptomatic vascular tumor of the liver in infancy.

KEYWORDS: Hemangioendothelioma, Liver, Vascular Tumor, Infant, Teratoma.

INTRODUCTION: Infantile hemangioendothelioma is the third most common hepatic tumor in children forming 12% of the hepatic tumors. Two sub types have been described based on tumor size and vascularity. Solitary lesions are more common than multiple lesions. The present case is a 45 day old female child who presented with abdominal distension and solitary mass in left hypochondrium.

CASE REPORT: 45 days old female patient presented with distension of abdomen, respiratory distress and regurgitation of milk. Antenatal history was uneventful. Birth history was normal. Parents noticed distension of abdomen with dilated veins at the age of 21 days. The mass was noted in left hypochondrium which gradually increased in size.

Palpation revealed a non-pulsatile mass in the left hypochondrium. USG abdomen showed a large heterogeneous mass in the left renal area 89x5mm size with specks of hyperechogenecity, left kidney was not separately seen and diagnosis of nephroblastoma was suggested. CECT abdomen revealed large peripherally enhancing mass 84x52x87mm mass arising from left lobe of liver extending from left hypochondrium to left iliac fossa with specks of hyperdense areas suggestive of calcifications were noted [Figure 1]. A differential diagnosis of hepatoblastoma, teratoma, neuroblastoma or exophytic hemangioendothelioma of the liver were given on radiological examination. All routine investigations were done. Peripheral blood smear shows normocytic normochromic picture with normal white cell count but Platelet count was 40,000 cells/mm suggesting thrombocytopenia. Liver Function Tests were within normal limits. Serum AFP levels and catecholamine levels were normal.

Wide range of differential diagnosis was given including malignancy by clinical and radiological examination. Surgical exploration was performed and well encapsulated mass was
excised from the left lobe of the liver and sent for Histopathological examination. Gross examination revealed 8x8.5cm well encapsulated heterogeneous mass with solid and cystic areas along with calcified foci. Tumor was looking grossly like a teratoma. [Figure 2]. Microscopy revealed vascular channels lined by continuous layer of plump endothelial cells in supporting fibrous stroma [Figure 3]. Foci of fibrous scarring, specks of calcification and areas of extra medullary hematopoiesis were noted [Figure 4]. Foci of organized haemorrhage were also seen. Mitotic activity was found to be low. Histopathological features were in favour of infantile hemangioendothelioma of the liver type-1.

**DISCUSSION:** Infantile hemangioendothelioma is a rare tumor found almost exclusively in children before 6 months of age. It is a highly cellular hemangioma that may appear as solitary or multiple nodules that may be associated with hemangiomas of other sites particularly skin. A female predominance has been noted as is the present case. Tumor may be asymptomatic & discovered incidentally or with hepatomegaly or as a palpable upper abdominal mass. In present case the child presented with distension of abdomen with prominent vessels.

Hematologic abnormalities like leukocytosis, anemia and thrombocytosis or thrombocytopenia may be seen.\(^3\) The peripheral smear revealed thrombocytopenia in our case.

On imaging they appear as complex mostly solid hepatic lesion with variable hypo and hyper echoic texture.\(^4\) In our case ultrasound abdomen showed a large heterogeneous mass with specks of hyperechogenicity. CECT abdomen revealed large peripherally enhancing mass arising from left lobe of liver with hyperdense areas suggesting calcific foci [Figure 1].

Grossly the tumor shows variegated appearance, grey tan and gelatinous appearance with cystic and necrotic areas.\(^5\) In our case, the tumor was well circumscribed, encapsulated mass with solid and cystic areas [Figure 2].

In 1971 Dehner and Ishak have proposed classifying infantile hemangioendothelioma into two histologic types: Type I IHHE also referred to as benign hemangioendothelioma or hepatic infantile hemangioma is used to describe the more common variant of the two and is formed by capillary sized vessels lined by a single layer of somewhat plump but bland endothelial cells with rare mitotic figures.

Type II IHHE contains vessels with pleomorphic endothelial cells and was often considered to be the same lesions as angiosarcoma.\(^3\) Type II IHHE has also been referred to as epithelioid angiosarcoma and kaposiform angiosarcoma in literature.

Nord et al.,\(^6\) have reported such a case affected by type II IHHE with cutaneous hemangiomas. Microscopy of present case correlated with type I IHHE. The diagnosis of infantile hemangioendothelioma should be considered when the patient is younger than six months of age by specific histological and radiological findings. Most important differential diagnosis to infantile hemangioendothelioma is hepatoblastoma which is a malignant embryonic tumor seen in infants and children.\(^2,7\) Other benign lesions that have to be differentiated from infantile hemangioendothelioma histologically are mesenchymal hamartomas.

Patients with infantile hemangioendothelioma have usually excellent prognosis and spontaneous regression is known to occur after first year of life. And children may die of associated complications such as severe heart failure or rupture.\(^5\) Treatment is determined on the basis of tumor size and severity of symptoms like CHF, AV shunting or coagulopathy. Surgical resection is indicated if life threatening symptoms are present or if the mass cannot be distinguished from a malignant tumor radiologically.\(^4\)
CONCLUSION: We have reported a case of infant with infantile hemangioendothelioma presenting with abdominal distension. Multiple specks of calcification were noted in the CT scan and a wide differential diagnosis was given including malignancy. Therefore understanding of the clinical presentation and detailed histopathology of types of infantile hemangioendothelioma of liver is essential for diagnosis.

ACKNOWLEDGEMENT: Authors acknowledge the immense help received from the scholars whose articles are cited and included in references of this manuscript. The authors are also grateful to authors/editors/publishers of all those articles, journals and books from where the literature for this article has been reviewed and discussed.

REFERENCES:

Abbreviations: IHHE – Infantile Hepatic Hemangioendothelioma

Fig. 1: CT scan showing heterogeneous mass with specks of hyperechogenecity
Fig. 2: Photograph showing single grey tan well circumscribed nodular mass with variegated appearance. Cut section showing calcified areas simulating teratoma.

![Fig. 2](image)

Fig. 3: Micrograph showing vascular spaces lined by endothelial cells with well-preserved bile ductules.

![Fig. 3](image)

Fig. 4: Micrograph showing areas of calcifications.

![Fig. 4](image)
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FINANCIAL OR OTHER COMPETING INTERESTS: None

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Date of Submission: 31/07/2015.
Date of Peer Review: 01/08/2015.
Date of Acceptance: 28/08/2015.
Date of Publishing: 03/09/2015.