Case Report

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Mass Abdomen in an Infant: A Pandora Box

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Abstract

Background: Infantile hepatic hemangioendothelioma/hemangioma (IHH) is the most common type of benign tumor of Liver, contributing to 12% of all childhood liver tumors. (1) It is a neonate's common hepatic mesenchymal tumor, 90% being less than six months old. (1) The tumor is slightly more predominant in females. It may be asymptomatic or can cause serious complications like cardiac failure or coagulopathy. There is no treatment protocol established yet and increasing number of cases have been reported. There is a varied treatment options like pharmacological or interventional on the basis of the presentation. IHH is classified as a benign vascular tumor with spontaneous regression in some patients.

Case presentation: Here is a 6 month old infant presented with Infantile colic and incidentally diagnosed to have IHH. Imaging like Ultrasonography and MRI confirmed the diagnosis. The child had an asymptomatic presentation and regressed with follow up.

Conclusion: Increasing awareness about the condition needed for proper follow up. Early identification and prompt treatment is necessary to avoid fatal complications.

Keywords: Mass abdomen, Infancy, Vascular, Liver.

INTRODUCTION

Infantile hemangio endothelioma/haemangioma (IHH) is a rare benign vascular tumor of the liver. [(1)] IHH is the third most common hepatic tumor in children (12% of all Childhood hepatic tumors). It is the most common benign vascular tumor of the liver in infancy and the most common symptomatic liver tumor during the first six months of life.[(2,3)]

The tumor has slightly more female predominance.[(4)] The lesions may be single or multiple and have calcification on 50% of cases on histopathological analysis.

In most of the cases, IHH remains asymptomatic and detected in ultrasound of abdomen incidentally. But sometimes, it may cause abdominal pain, hepatomegaly, severe arteriosclerosis venous shunting with CCF, anaemia, thrombocytopenia (Kasabach Meritt syndrome), consumptive coagulopathy, Intraabdominal haemorrhage. Rarely, biliary obstruction with Jaundice, vomiting and gastric outlet obstruction has been reported.[(4)]

We report 1.5 month old child with Infantile Hepatic Haemangioendothelioma which was accidentally detected on ultrasound abdomen while evaluating for colic.

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CASE REPORT

1.5-month-old male infant, first born to non consanguineous parents, had uneventful Antenatal and postnatal period in our hospital, came with history of excessive crying and distention of abdomen noticed by parents for the past 3 days. After detailed history taking and thorough clinical examination, child was treated with anti colic medication. Child had persistent symptoms and was subjected to imaging studies. Ultrasound abdomen showed small well defined oval hypo echoic lesion in liver measuring 20*18*8.7mm in left lobe of liver (segment II) showing peripheral vascular rind with a small echogenic focus of 2mm, suggestive of high flow vascular neoplasm, hemangioendothelioma. CT Abdomen confirmed the findings. Child was referred to paediatric surgeon and was advised to follow up with annual ultrasound abdomen.

Child is asymptomatic with normal Growth, developmentally appropriate and vaccinated upto date and on regular follow up. Recent Ultrasound abdomen showed small hypo echoic focal lesion in the left lobe of Liver in subcapsular area having peripheral vascularity suggestive of high flow vascular neoplasm- Haemangioendothelioma.

DISCUSSION

Infantile hepatic hemangioendothelioma (IHH)is the most common vascular tumor of liver in children constituting 12% of all childhood hepatic tumors. About 85% become symptomatic in first 6 months of life and it is the most common symptomatic tumor. There is a female preponderance with a ratio of 1.3 to 2:1.[7]

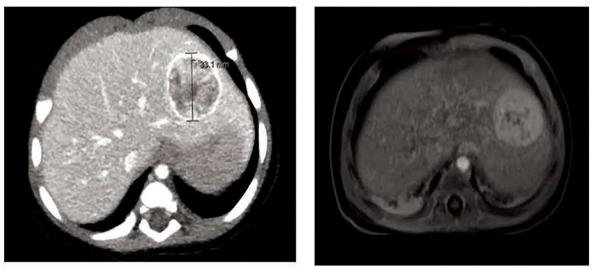
The most common presenting symptom is abdominal mass. Other presentations include hepatomegaly, high-output cardiac failure, skin hemangioma, thrombocytopenia or peritoneal bleeding. Asymptomatic lesions undergo spontaneous regression within a year, symptomatic lesions may cause heart failure or death.

Serum Alpha fetoprotein is an important tumor marker for the diagnosis and prognostication of pediatric hepatic masses. It may be used for prenatal diagnosis. It may be diagnosed in ultrasonogram.

This tumor can be incidentally found in the fetus during antenatal ultrasound. It can also be found in children who underwent abdominal computed tomography for other indications. Other presentations may be coagulopathy, heart failure or jaundice.[6,7]

IHH is divided into two types according to histology.[8] Type I IHH is composed of vascular channels lined with benign endothelial cells displaying small nuclei. Type II IHH demonstrates vascular channels with pleomorphic endothelial cells with hyperchromatic nuclei and mitosis. Type II IHH was once classified as a high grade tumor due to its aggressive histopathological characteristics than type I.[8]

Radiological evaluation is useful for patient diagnosis with sonography often being the initial diagnostic modality. On sonography, IHH is characterised by discrete, hypo echoic lesions (either solitary or multiple) within the liver that may have calcification or shunting on Doppler evaluation. A more definitive diagnosis requires a contrast enhanced CT (CECT) or a magnetic resonance imaging (MRI). While CECT shows a hypodense area which enhances with contrast. MRI may identify IHH as a low signal lesion on T1 and high signal lesion on T2 weighted images. Most cases of IHH are asymptomatic with spontaneous regression. But cases with heart failure experience a higher mortality rate (upto 70%).[5,8] (Fig. 1)



(A) (B)

Fig. 1: Showing CT/MRI image showing enhanced localised tumor seen within liver

Courtesy (image): Diagnostics 2021, 11(2), 333; https://doi.org/10.3390/diagnostics11020333

The differential diagnosis of Infantile Hepatic Haemangioendotheliomaare cavernous haemangioma, angiosarcoma, hepatoblastoma and mesenchymal hematoma.[9]

CONCLUSION

Infantile hepatic haemangioendothelioma may be an incidental diagnosis. They may be asymptomatic or symptomatic. Not all children will be symptomatic. The diagnosis is based on the symptoms and operability. Awareness is important for prompt diagnosis and management.

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