Pediatric Oncall Journal (July - September)
Volume 15, Issue 3 : 85-86
DOI: https://doi.org/10.7199/ped.oncall.2018.33

PEDIATRIC ONCALL
CHILD HEALTH CARE

TEACHING FILES (GRAND ROUNDS)

INFANTILE HEMANGIOENDOTHELIOMA - HOW TO TREAT?

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KEYWORDS
IHE, steroids

ARTICLE HISTORY
Received 05 July 2017
Accepted 01 August 2018

Case Report

A 1 months old boy presented with progressive abdominal distension since 1 month of age and breathlessness for a day. There was no jaundice or pallor. On examination, weight was 3.86 kg. His vital parameters were normal and systemic examination revealed an approximately 4 x 3.5cms sized lump palpable in epigastric region which was not separate from the liver. There were dilated veins were present over abdomen. On investigations, ultrasound (USG) of abdomen showed multiple mass lesions in liver suspected to be hemangioendothelioma with a hypoechoic lesion seen in left liver lobe. Baseline bilirubin was 2.8 mg/dl (direct =1.2 mg/dl), SGOT was 46 IU/L, SGPT was 42 IU/L, total proteins were 7.9 gm/dl, albumin was 3.0 gm/dl, alkaline phosphatase was 479 IU/L. Hemogram showed hemoglobin of 12.5gm%, white cell count of 18,000 cells/cumm and platelets 427000 cells/cumm. Serum alpha feto protein was normal. MRI abdomen showed an enlarged liver with smooth margins and multiple lobulated lesions in right and left lobes, the largest being in left lobe measuring 4.1x3.5x5cm and right lobe being 2.8x2.3x2.9cm. The lesions on post contrast showed peripheral enhancement with multiple internal enhancing septa within. It also showed prominent vessels in anterior abdominal wall with increased caliber of celiac axis and marginally reduced caliber of infrarenal aorta confirming multiple lobe liver epitheloid hemangioendothelioma.

How to treat this child?

Discussion

Patient was started on IV methylprednisolone 15 mg/kg/day for 3 days and 10mg/kg/day for next 4 days and changed to oral prednisone 4mg/kg/day for 20 days followed by gradual tapering of the steroids in next 2 months. At 2.5 months of age, ultrasound showed decreased nodes in liver with lesion size being 1.5x1.3cm in right lobe and 0.9 x0.7cm in left lobe.

Infantile hemangioendothelioma (IHE) is rare because of its low incidence rate of 1/20000. (1) Hemangioendothelioma accounts for 12% of all childhood hepatic tumours and the most common vascular tumor in children. Hemangioendothelioma is predominant in female with the male to female ratio being 1:1.3 to 2. (2) Presenting feature usually is as an abdominal mass but could be hepatomegaly, skin hemangioma, thrombocytopenia, high output cardiac failure, hemolytic anemia and peritoneal bleeding. (3) Though it is a histologically benign tumor it has severe complications like congestive cardiac failure (15%) and liver failure (2%) in infants. (1) A non-complicated tumor may spontaneously regress, but most fatalities occur in patients whose initial presentation is intractable heart failure. (1)

In India incidence rate of infantile hemangioendothelioma is about 1/20000. (1) Eighty six percent of IHE usually presents within first 6 months of life with 1/3rd of them in the first month. (1)Pathologically infantile hemangioendothelioma is a mesenchymal tumor composed of a connecting network of predominantly endothelium lined small-diameter vascular channels. (4) To start with USG is performed which may show single or multiple hypoechoic lesions in the liver. A more definite diagnosis requires either a CT or MRI. Serum alpha feto-protein should be done in all cases and biopsy to be performed in case of a suspected malignancy. (5) Among the treatment modalities for solitary lesions steroid therapy, radiotherapy and hepatic artery ligation can be used along with embolization techniques whereas for large lesions liver transplant is the mainstay of treatment. (6) Symptomatic treatment like digitalis and diuretics and blood products for congestive cardiac failure and anemia accompanied by steroid therapy for regression of lesions is the first step of treatment. (7) 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. (8)The response to steroids can be achieved within 1 to 3 weeks, and the success rate varies from 20% to 70%. (9, 10) One- third of cases may prove unresponsive to steroids and can be treated with interferon alfa-2a. (4) Successful treatment with cyclophosphamide after failure of steroid therapy was also reported. (4) Our patient showed a remarkable response to steroid therapy.

Compliance with Ethical Standards

Funding: None
Conflict of Interest: None

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