INFANTILE HEMANGIOENDOTHELIOMA - HOW TO TREAT?

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IHE, steroids

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 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 fetoprotein 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.
REFERENCES: