

## Primary infantile haemangiopericytoma of liver

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### Introduction

Haemangiopericytomas are vascular soft tissue sarcomas arising from the pericytes of Zimmermann cells<sup>1</sup>. They comprise less than 3% of the soft tissue sarcomas<sup>2</sup>.

### Case report

A 3.5 month old, well grown male infant, born of a non-consanguineous marriage, presented with abdominal distension and non-bilious, post feed vomiting since 20 days. Abdominal distension was more in the upper abdomen and was progressive. Examination revealed firm hepatomegaly with a span of 14 cm without splenomegaly. There was no icterus, pallor, lymphadenopathy or bleeding from any site. Birth history was normal and there was no history of previous admissions. Based on the clinical scenario, a differential diagnosis of malignancy and storage disorder were kept in mind.

Routine blood investigations revealed a haemoglobin level of 10g/dl with normal platelet and bilirubin levels. Aspartate transaminase (AST) was 12 IU/L and alanine transaminase (ALT) was 18 IU/L. The prothrombin time (PT), activated partial thromboplastin time (APTT) and international normalised ratio (INR) were normal. Specific investigations such as alpha fetoprotein were done to rule out hepatoblastoma. The value was 237 ng/ml which is within the normal range for age. Ultrasonography (USG) of abdomen showed multiple lesions scattered in the entire liver parenchyma. A computed tomography (CT) scan of abdomen with triple phase study showed hepatomegaly with multifocal lesions diffusely scattered in both lobes of uncertain aetiology (Figure 1).

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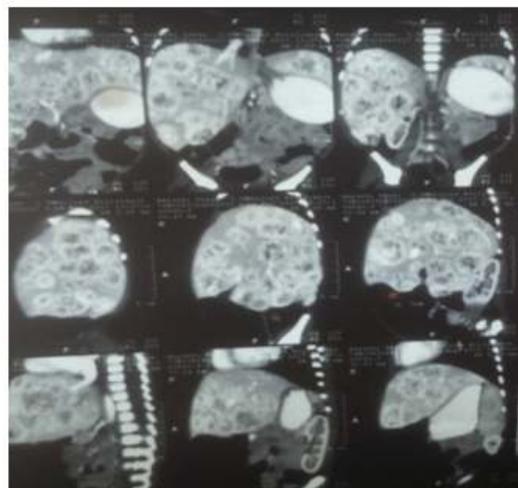
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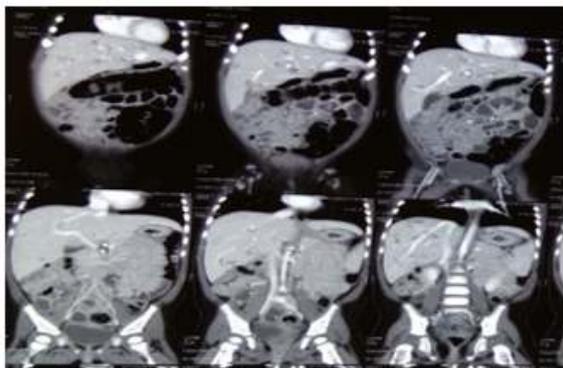
**Figure 1:** CT scan of abdomen showing multiple scattered lesions in the entire liver parenchyma

CT guided biopsy revealed a benign tumour with a rich capillary sized vascular network and an investing proliferation of pericytes around it – endothelial cells expressing CD31, CD 34, ERG Mic 2 and SMA, suggestive of haemangiopericytoma.

The infant was transferred to the paediatric surgery department for further management. Haemangiopericytoma, being a rare tumour in infants, following literature review and discussion amongst the surgical and the radiology teams, the only option for treatment of this baby was liver transplantation, the feasibility and availability of which was very poor. Also, since this was a vascular tumour, and with previous successful outcomes of other liver vascular tumours, the paediatric surgical team took the decision to start high dose oral propranolol, in a dose of 3mg per kg per day. Strict monitoring of pulse rate, blood pressure and finger prick blood sugar was done to identify and rectify the side effects of the high dose propranolol. Since all these parameters were maintained and the baby did not show any side-effects of the drug, the same dose was continued with regular monitoring in the ward and weekly monitoring after discharge. Clinical response was seen within 10 days of starting the drug by a decrease in the episodes of vomiting and also a decrease in the abdominal girth. Baby was discharged about 12 days after starting propranolol and was followed up on an outpatient basis every week for the first 3 months for assessment of response as well as side-effects of the drug and both were satisfactory.

At 3 months, repeat USG of abdomen showed a reduction in the vascularity of the tumour as well as the size and number of lesions. Clinically, the child kept feeding and growing well with no adverse effects of propranolol and the dose was continued as 3mg/kg/day, dosages being modified as per weight.

CT-scan of abdomen done after 6 months showed a significant reduction in the size and vascularity of the lesions (Figure 2).



**Figure 2:** Repeat CT scan of abdomen showing decrease in the size of tumour post propranolol

The child is on regular follow-up and received propranolol therapy for one year following which the drug was gradually tapered and stopped in the last month. USG of liver shows almost complete regression of the lesions. However, repeat CT-scan is planned for 3 months later.

### Discussion

The child presented with isolated hepatomegaly with no other symptoms. Thus, the clinical differential diagnoses in this case were metabolic/storage disorders like tyrosinaemia and primary hepatic malignancy like hepatoblastoma. Hepatic haemangiopericytoma usually occurs as a metastatic lesion, especially in adults, as seen in the study done by Regina *et al*<sup>3</sup>. Microscopically, the tumour consists of tightly packed cells around thin walled, endothelium-lined vascular channels ranging from capillary-sized vessels to large gaping sinusoidal spaces. They closely resemble the adult type except for frequent endovascular proliferation of tumour cells, increased mitotic activity and focal necrosis, features that indicate malignancy in an adult-type haemangiopericytoma. This, however, is generally not the case with the infantile form. Most of these tumours tend to follow a benign clinical course and are curable by local excision<sup>4</sup>. In a case series by Atkinson *et al*<sup>5</sup>, 6 cases of primary haemangiopericytomas were studied of which only one had primary hepatic involvement. The infantile haemangiopericytoma probably shows more benign and less aggressive behaviour than those in children above 1 year of age and neo-adjuvant chemotherapy

has been used in infants with documented response to therapy<sup>6</sup>.

Limited data is available regarding treatment of infantile haemangiopericytoma. It is usually surgical comprising surgical excision of tumour<sup>7</sup>. Preoperative embolisation is preferred to avoid excessive bleeding. Chemotherapy comprising vincristine, cyclophosphamide and methotrexate is advised for malignant residual tumour post-surgical excision<sup>8</sup>. Anti-angiogenic agents are potential promising therapeutic options for treatment of haemangiopericytoma<sup>9,10</sup>. In this patient, due to the high vascularity of the tumour and multiple lesions, propranolol was used and there was significant reduction in the size of the tumour as seen in follow up CT. There are no studies in the literature citing the use of propranolol to reduce the tumour size, and not many studies reported of this tumour in infancy. Thus, timely detection of the tumour as in this case helped in the early medical management of the patient.

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