CASE REPORT

Infantile hepatic haemangioendothelioma: a case report

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Abstract

Infantile haemangioendothelioma is a rare tumour of infancy arising from mesenchymal tissue. The liver is the commonest site for this tumour in children. This is a report of a 3 month old boy who presented with hepatomegaly, hypertension, congestive cardiac failure and protracted diarrhea. Abdominal ultrasonography demonstrated diffuse hepatic nodules involving the whole liver. A diagnosis of infantile hepatic haemangioendothelioma was made on histologic examination of tissue following an open liver biopsy. The patient died 3 days after liver biopsy. We report this case to raise awareness of this rare tumour of infancy in our environment.

Key words: Infantile haemangioendothelioma, hepatic, congestive cardiac failure

Introduction

Primary hepatic tumours are uncommon in children, accounting for only 1-4% of childhood tumours. 1, 2 Haemangiomas are however common in childhood. 3, 4 Hepatic haemangioendothelioma in children presents with a triad of hepatomegaly, cutaneous haemangiomas and cardiac failure. We report a case of infantile hepatic haemangioendothelioma in a 3-month old boy with the classical triad, hypertension and diarrhea.

Case report

A 3-month old boy was admitted into our emergency paediatric unit on account of a rapidly progressive right abdominal swelling that was noticed 6 weeks before presentation. The boy also had recurrent episodes of diarrhea over the same period. There was no cough, no dyspnoea no feeding difficulties, and no haematuria. He had been delivered via a spontaneous vertex delivery at term and weighed 3.69 kg at birth. He was being exclusively breastfed. At the time of presentation, he was pale, afebrile (temperature 37.2oC) and well hydrated. He weighed 5.7 kg (95% expected for age). Three small strawberry-like lesions (0.5-1 by 1-1.5 cm) were noted each on his right forearm, right chest wall and left side of the anterior abdominal wall respectively. The swellings were cystic, non-pedunculated, attached to underlying structures, and non-tender. He was mildly dyspnoeic and tachypneic, with a respiratory rate of 79/minute, but had good air entry into both lungs and no added sounds. The pulse rate was 156/minute, full volume and regular. Blood pressure was 150/90 mmHg. A third heart sound gallop rhythm was audible. The abdomen was markedly distended (girth 49.8cm at the level of the umbilicus), with a small umbilical hernia. It was firm and the skin was shiny with areas of desquamation and distended superficial veins. A markedly enlarged liver (left and right lobes) that extended to the left flank and measured 114 cm below the right costal margin at the mid-clavicular line was palpable. It was firm and had a macronodular surface and a rounded edge, but was non-tender, non-pulsatile, with no audible bruit. A reducible right inguinoscrotal hernia was present. Rectal examination was essentially normal.

The clinical impression was that of an intra-abdominal malignancy, possibly a neuroblastoma with hepatic infiltration, or a hepatoblastoma, with congestive cardiac failure secondary to anaemia and hypertension. Abdominal ultrasonography revealed a huge complex intra-abdominal mass of mixed echogenicity, which had displaced the right kidney to the level of the pelvic brim and distorted the splenic margins. Both kidneys were clearly distinguishable and had well preserved cortico-medullary differentiation. The serum biochemistry results showed mild hyponatraemia, hypocalcaemia, hyperurecaemia and hypoproteinaemia. The initial haematocrit was 21%, while the white cell counts (total and differential) were within normal limits. The initial platelets were normal, but later results showed thrombocytopenia. Intravenous urography showed prompt bilateral contrast excretion but the right kidney had been displaced downwards by the upper abdominal mass. Urinary vinyl mandelic acid (VMA) and alpha-fetoprotein were not done.

On admission, he was treated with intravenous frusemide, 1mg/kg 12 hourly for 48 hours, and oral alpha methyldopa, 5 mg 8 hourly, while the blood pressure was monitored 4 hourly. He was transfused with packed red cells. Alpha methyldopa was replaced with propranolol on
the 14th day of admission due to suboptimal blood pressure control (130/90 mmHg) using the former. A satisfactory BP level of 90/40 mmHg was subsequently attained using the propranolol. Two weeks after admission he developed evidence of bronchopneumonia (fever and coarse crepitations in both lungs) for which antibiotics (ampicillin and cloxacillin) and antimalarial (chloroquine) were administered. Oral allopurinol was also added at this time in anticipation of commencing cytotoxic therapy (hyperuricaemia was already present). After 6 weeks on admission he was considered stable enough for an exploratory laparotomy, which revealed multiple huge hepatic masses involving the left and right lobes of the liver. This precluded any form of partial resection. A wedge biopsy of the liver was therefore taken. All the other intra-abdominal organs were normal. A biopsy of one of the skin lesions (that on the left forearm) was also taken. The patient had a transient fever (38.6°C) on the day of surgery and an episode of bilious vomiting 2 days later. The latter stopped when the naso-gastric tube that had not been draining was removed and re-passed. On the second post-operative day, he bled extensively from the laparotomy site and was febrile again (temperature 39.0°C). The dressings were changed. Early in the 3rd Post-operation day he was found to be dyspnoeic with cold extremities and bilateral basal coarse crepitations. His pulse rate at this time was 128/minute. He died shortly thereafter while efforts were being made to transfuse. Histologic examination of both biopsy specimens later revealed the diagnosis of infantile hepatic haemangioendothelioma.

Discussion

Infantile haemangioendothelioma is uncommon in our environment though haemangiomas are quite common. Infantile hepatic haemangioendothelioma often presents within 6 of life as hepatomegaly and cutaneous haemangiomas are easily noticeable. Most infants present with congestive cardiac failure in addition to the hepatomegaly as in our patient. Cutaneous haemangiomas occur in 45-50% of reported cases. Our patient had cutaneous haemangiomas. The vascular lesions produce systemic haemodynamic disturbance similar to arteriovenous shunts in addition to local intra-abdominal pressure and discomfort produced by the tumour. Cholestatic jaundice, haemolytic anaemia and thrombocytopenia have been noticed in some patients. In addition to congestive cardiac failure, our patient had thrombocytopenia which he developed while on admission. He did not have cholestatic jaundice and neither was the spleen enlarged. Some tumours affect the bowel causing significant gastrointestinal bleeding. Although our patient did not have gastrointestinal bleeding, there was intractable diarrhoea that started at about the same time the abdominal mass was noticed. No organism was cultured from the stool. Vasomotor phenomenon with the release of histamine causing a carcinoid-like syndrome has been reported in an adult with haemangilendothelioma. 

References