

Picture Story

A case of diffuse neonatal haemangiomatosis

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A 5 day old baby girl was transferred to the surgical casualty unit of the Lady Ridgeway Hospital (LRH) from Base Hospital, Negombo for management of severe, persistent rectal bleeding since 2nd day of life despite repeated blood transfusions. At the surgical unit at LRH further blood transfusions were given and she underwent exploratory laparotomy. At surgery blood was seen within the small and large intestines but a bleeding site was not detected. The child continued to have profuse rectal bleeding and was transferred to our unit for further management on the 8th day of life.

She is the first child of non-consanguineous healthy parents. She was born at term with a birth weight of 2.7 kg and was exclusively breast fed. Intramuscular vitamin K was given at birth and there were no risk factors for sepsis. There was no family history of bleeding disorder and her bleeding was confined to the gastrointestinal tract (GIT).

On examination, she was pale and afebrile. There were no dysmorphic features. Multiple cutaneous haemangiomas (0.5 to 2 cm in size) were noted over the back of chest and buttocks (Figure 1). Haemangiomas were also present in the tongue. The rest of the examination was normal.

The haemoglobin was 4 g/dl, white cell count 8.5 x 10⁹/l (N 63%, L 34%, M 3%) and the platelet count 92 x 10⁹/l with a normal blood picture. The bleeding time was 4 minutes and the clotting time 5 minutes; the prothrombin time was 14 seconds (control 12 seconds) and the activated partial thromboplastin time was 46 seconds (control 43 sec). The septic screen was negative. Ultrasound scan of abdomen and thorax showed no haemangiomas in solid organs.

A presumptive diagnosis of diffuse neonatal haemangiomatosis (DNH) was made and she was started on high dose steroids (IV hydrocortisone 100 mg/kg/day). The bleeding subsided within 2 weeks.

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Figure 1. Multiple cutaneous haemangiomas

Lower GI endoscopy, performed at the Colombo South Teaching Hospital after the active bleeding had subsided, showed multiple mucosal haemangiomas along the large intestine 5-25 cm from anus (Figure 2). The multiplicity of the lesions precluded cauterization. Thus the diagnosis of DNH was established. The steroid dose was gradually reduced to a low level and finally stopped at 9 months of age.

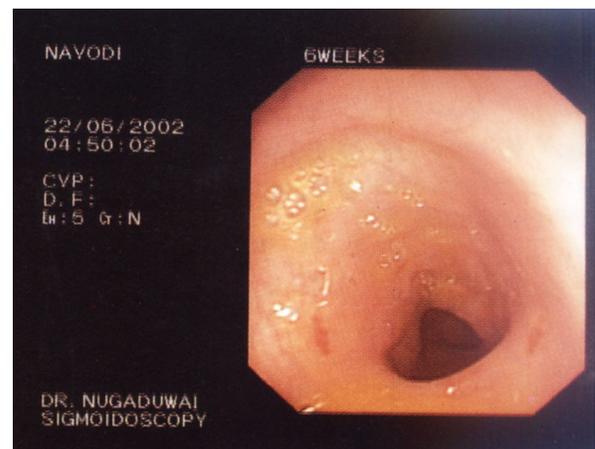


Figure 2. Multiple cutaneous haemangiomas - Lower GI endoscopy

The cutaneous haemangiomas started fading and, apart from one episode of mild bleeding per rectum at 3 months of age, she did not have any significant GIT bleeding. Initially, she had some cushingoid features but later, with low dose steroids, they disappeared. She was given inactivated polio vaccine along with other EPI vaccines. Now, 11 months old, she remains asymptomatic with normal growth and development.

Discussion

DNH is a rare, frequently fatal disease characterized by multiple cutaneous and visceral haemangiomas¹. The skin, liver, lungs, intestines and central nervous system are the most commonly affected organs². Lesions are usually present at birth (70%) or develop within the first weeks of life and may number in the hundreds³. DNH carries a mortality rate of 60-90%, with death usually occurring within the first months of life as a result of high-output cardiac failure caused by arteriovenous shunting or from thrombocytopenia caused by haemorrhage from gastrointestinal or respiratory tract haemangiomas³. DNH must be differentiated from benign neonatal haemangiomas, in which cutaneous lesions are also present at birth but there is no associated visceral involvement or increased mortality⁴. Treatment of DNH includes systemic steroids, radiation therapy, ligation or embolization of feeding vessels of haemangiomas and interferon alfa-2a⁵. It has also been successfully treated with flashlamp pulsed dye laser⁶. DNH is a life-threatening disorder that requires a multimodal, interdisciplinary approach⁷.

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