GORHAM'S DISEASE
A case report

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Summary

A rare case of Gorham's syndrome is presented in a 6 months old female child. This case report is the first of its kind in Indian literature.

Haemangiomas or vascular birthmarks are the most common cutaneous malformations seen in children. Under the term vascular naevi are included the naevus flammeus, capillary haemangioma, cavernous haemangioma, naevus anaemicus, haemangioma circumscriptum, and vascular naevi associated with other disorders viz, Sturge-Weber's syndrome, Maffucci's syndrome, blue rubber bleb naevus, Von Hippel Lindau syndrome, Kasabach-Merritt syndrome, Klippel-Trenaunay-Weber syndrome, haemangioma with retrolental fibroma, sudoriparous angioma, various angiokeratomas, and Gorham's disease.

Cutaneous haemangiomas of the face or distal part of the extremities may lead to hypoplasia of the underlying osseous tissue. Osseous haemangioma may result in local fibrous substitution of the affected part of the skeleton. In exceptional cases cutaneous haemangioma may lead to osteolysis of the underlying bone, and this is called Gorham's disease or disappearing bone disease[1]. On account of its rarity, we are reporting a case of Gorham's disease in a 6 month old female child. To our knowledge, this is the first time this disease is being reported in Indian literature.

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Case report

A female child aged 6 months was admitted to the skin ward for a swelling with occasional bleeding on the skull. History revealed that the child had a small swelling on the parieto-occipital region from birth. The swelling gradually increased in size and used to bleed occasionally. Though the diagnosis of cavernous haemangioma was evident, the child was admitted to the ward on 5-2-'80 for investigations to detect associated disorder if any.

Local examination revealed a 7.5 cms. × 7.5 cms. round, circumscribed, elevated, partially compressible tumour on the parieto-occipital area (fig. 1). The tumour was reddish-brown in colour and showed bleeding spots and ulceration at places. There was no evidence of any other naevus on the integument.

Systemic examination did not reveal any anomaly.

Family history was non-contributory. Blood for total and differential counts, HB, ESR, bleeding time, clotting time, platelet count; routine examination of urine and stool; X-ray of the chest and barium meal examination of GI tract were within normal limits. X-ray of the skull showed

Fig. 1 Showing a cavernous haemangioma on the parieto-occipital area of the skull.

osteolysis of the parietal bone underlying the cavernous haemangioma and soft tissue swelling (fig. 2).

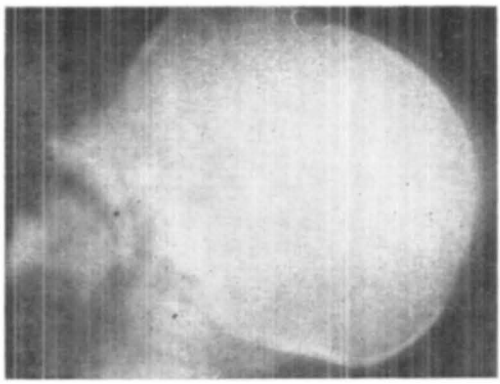

Fig. 2 Showing the soft tissue swelling with osteolysis of the parietal bone.

Discussion

Cutaneous haemangiomas occurring with osteolysis or 'disappearing bones' have been reported by Frost and Calpan[2]. Gorham described the disease as osteolysis of multiple areas of the skeletal system, although usually a single bone is involved[3]. The disorder usually begins in childhood. The disease has been progressive in a few instances but is eventually self limiting[4]. There is partial or complete replacement of the bone with fibrous tissue. Cutaneous haemangioma may be the initial indication of the disease. Deaths have occurred from haemorrhage into serious cavities.

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