HAEMANGIOMATOSIS OF THE SKELETON

Report of a Case

RANBIR SINGH, D. S. GREWAL, A. K. BANNERJEE and V. P. BANSAL, CHANDIGARH, INDIA

From the Departments of Orthopaedic Surgery and Pathology,
Postgraduate Institute of Medical Education and Research, Chandigarh

Haemangioma of bone is uncommon. The commonest site is the spinal column, and after that the skull. Topfer (1928) reported that vertebral haemangiomata were found at necropsy in 11·9 per cent of 2,154 cadavers. Jacobs and Kimmelstiel (1953) estimated that the total number of extravertebral haemangiomata of bone reported, including their own two cases, was forty-one. Twenty cases involved the skull and the rest the bones of the extremities. They did not report any case of generalised haemangiomatosis. Ritchie and Zeier (1956) reported a case of generalised haemangiomatosis with haemangioma of the spleen. The present case report is similar but there was no clinical evidence of involvement of the spleen.

CASE REPORT

A girl aged eleven years presented in June 1969 with a fracture of the mid-shaft of the right humerus caused by a trivial fall. Radiographs showed a cystic lesion around the fracture site and many further cystic lesions elsewhere in the skeleton (Fig. 1). The fracture was immobilised and she was admitted for investigations. She did not complain of pain in any other bone or joint. There was nothing of note in the family history.

On examination the child was of average build. No skin or subcutaneous nodules could be palpated; she had no neurofibromata or café-au-lait spots. The liver and spleen were not palpable.

A radiological skeletal survey showed cystic lesions in most of the bones. They ranged in size from about two millimetres to five centimetres. Some cysts were unilocular, others multilocular. Multiple cysts gave the skull a moth-eaten appearance with thinning of the overlying tables. A similar lesion was seen in the ramus of the mandible. In the ribs, multiple expansile cystic lesions were seen. A number of similar lesions were seen in both scapulae. The long bones of both upper and lower extremities were similarly affected. The pelvic girdle was riddled with multiple punched out areas (Figs. 2 to 5).

Serum calcium, phosphorus, alkaline phosphatase and proteins were within normal limits. Urine analysis was normal.

Biopsy of the left ulna revealed a cyst with a thin papery wall; a few septa were seen dividing the cyst into loculi. Clear fluid was present inside the cyst. The histology was inconclusive. A month later further biopsy was undertaken, the fifth rib being excised. The central part of the rib was expanded in a fusiform manner for a distance of about 2·5 centimetres. This part contained a cyst, the wall of which was no more than a thin shell of bone that could easily be cut with scissors. The cyst was loculated by thin bony septa. The cyst contained brown semisolid material which was thought to be clotted blood.

Microscopically, the decalcified paraffin sections showed large spaces lined by endothelium and containing blood. There were specific areas with collections of small endothelium-lined spaces (Fig. 6). The bony trabeculae were thin, but there was evidence of new bone formation as shown by the irregular lines of calcification in relation to the proliferating vascular channels. These appearances together with the radiological findings were consistent with the diagnosis of haemangioma.