FAMILIAL OSTEODYSTROPHY OF THE SKULL AND FACE

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For many years intermittent attention has been paid to the aptly named phenomenon of leontiasis ossea. Numerous conditions affecting the appearance of the face have been included but the application of a reasonable definition, such as appears in Dorland's Medical Dictionary, makes a considerable proportion of cases unworthy of the designation. Dorland declares the condition to be a bilateral and symmetrical change in the bones of the face and cranium leading to a lion-like facial expression.

Considering only the contracted group of conditions which have been more accurately named I could find no familial incidence recorded. This prompts me to report briefly the findings in three British Columbian children of one family which has been studied and investigated.

These three children, all girls, were first seen in 1953, aged eight, six and two years. The older children were the more affected and a baby brother one year old appeared normal.

Each girl exhibited the broad face with prominent cheek bones which gives the condition its name and it was for this reason that the parents took them to the family doctor. Clinically there was only mild deformity, difficult to demonstrate photographically (Fig. 1). The prominences lateral to the nose were, however, bony hard. Figure 2 reproduces original lateral views of one child's skull. The film on the left resulted when ordinary technique was used, while on the right the same skull is seen using between two and three times the penetration usually required for a normal adult skull. Figure 3 presents recent more revealing views of the same skull. Thickening of the facial bones, as well as of the entire bony plate of the skull and failure of development of sinuses, can be seen. Examination of the actual radiographs
exhibited more effectively an increase in the size of the maxillae and malar bones, with great deposition of bone in the paranasal regions. The normal osseous structure was replaced by bone having a more or less homogeneous appearance which might be described as granular. The mandible revealed similar structural changes but without any alteration in the size or shape of the bone. The palate bones and nasal septum all revealed changes in the direction of increased density. The skull revealed evidence of thickening of the vault and base,
most marked in the frontal and occipital regions. It was difficult to differentiate diploe from cortex. Considerable thickening of the walls of the orbit and ethmoidal bones was apparent from the postero-anterior radiographs. All three children revealed similar changes although there were varying degrees of involvement of individual regions. The dentition appeared to have developed and erupted normally. The skull of the male child exhibited similar but less advanced change.

A radiological survey of the long bones in each child failed to reveal any generalised osteopetrosis. General physical examination was quite unremarkable and the children were in no way affected mentally. No relative was known to suffer from this condition. Laboratory investigations including blood Wassermann, mineral metabolism and electrolyte studies, phosphatase estimation and complete blood and urine examination were all within normal limits.

A biopsy specimen consisting of the complete thickness of the skull in the parietal region was removed from the eldest child. The cortex was greatly thickened but not unduly hard. Microscopically the picture was of normal bone.

COMMENT

It is difficult to be certain of the diagnosis in these cases. There is no basis for suspecting an infective origin, such as has been attributed to the local sclerosing leontiasis of the facial bones. The single biopsy was not consistent with fibrous dysplasia, but the radiographic appearances were characteristic of one manifestation of this disease in the face and skull. It is unusual for the distribution of such lesions to produce a leonine facies and to warrant the designation leontiasis ossea.

Further examinations will be necessary to follow the progress of this disease, which may be expected to cease at the time of, or before, the normal cessation of skeletal growth.

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REFERENCES