POLYOSTOTIC FIBROUS DYSPLASIA

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FIBROUS DYSPLASIA is a congenital anomaly in the development of bone causing tumour-like malformation. It was known variously as fibrous-osteodystrophy, osteo-dystrophia fibrosa, etc. till 1938 when Lichtenstein described it as fibrous-dysplasia. One or several bones may be involved. Involvement of one bone or multiple bones is known as monostotic or polyostotic (the condition being unilateral) respectively. Young (1948) has described the anterior and middle cranial fossa involvement among the skull lesions.

Albright has stated that in several forms usually occurring in childhood, extra-skeletal anomalies such as pigmentations of skin, premature sex development in females, primitive growths and hyperthyroidism occur. It is often associated with skin pigmentations and endocrinal disturbances.

Aetiology of it is unknown. It is believed that normal mature bone on physiological lysis is replaced by a solid mass of proliferating fibrous cells under the periosteum; subsequently bone gets expanded and deformed while the cortex is thinned out. This deranged development maintains a slow pace through many years before clinical abnormalities are identifiable.

Case Report

M.S., 20-year-old male, presented in *December 1965*, complaining of swelling of right cheek, right frontal, right molar and right mandibular region: 12 years. The swellings were gradually increasing in size. There was no pain.

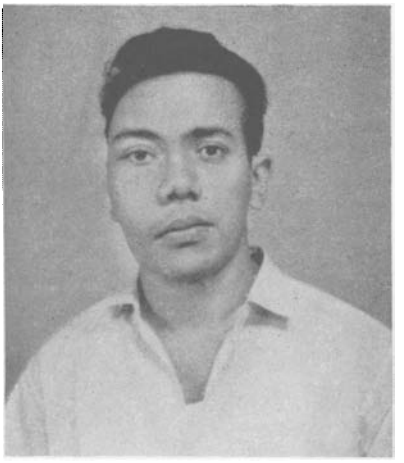


FIG. 1.
Photograph of slide.

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Clinical examination: showed smooth, non-tender, hard swelling of right maxilla, right frontal, right malar extending to right, and right mandible which was diffusely thickened also showed swelling (Fig. 1).

Investigations: routine blood and urine exam: revealed nothing significant. Blood calcium and alkaline phosphatase were within normal limits.

X-ray: of the skeleton showed changes in several bones consistent with fibrous dysplasia. The bones involved were right frontal, right malar, right maxilla and right mandible, right iliac crest, lumbar vertebrae (Harris *et al.*, 1962; Jaffe, 1958) and ribs (Figs. 2, 3, 4 and 5). There was no evidence of thyroid enlargement suggesting parathyroid tumour.

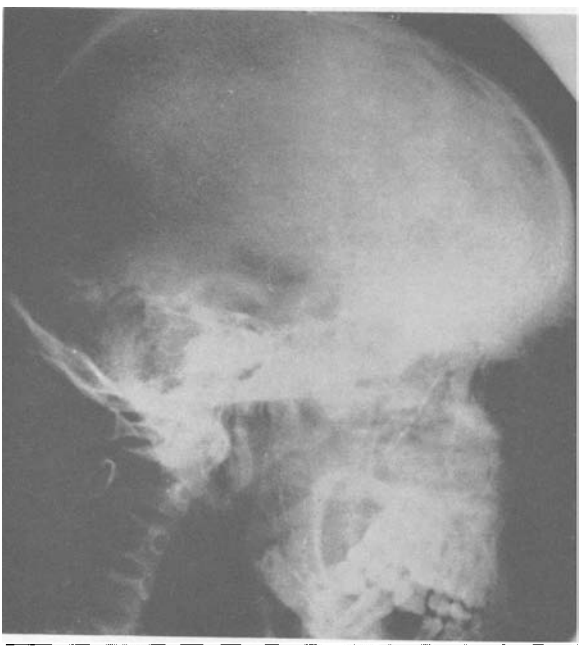


FIG. 2.
X-ray of the skull showing dense shadow on right frontal and right malar region.

Biopsy: was taken from the right maxilla and histopathological examination revealed fibrous dysplasia (bands of fibrous tissue admixed with irregularly arranged trabeculae of bone) (Fig. 6).

Comments

Fibrous dysplasia is a rare disease, particularly in this part of India. The involvement of the lumbar spine is quite rare. Harris *et al.* (1962) has reported the following incidence of bones involved in polyostotic cases:

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Base of skull	50 per cent.	Radius	43 per cent.
Occipital	53 per cent.	Ulna	33 per cent.
Cervical spine	7 per cent.		



FIG. 3.
X-ray lumbar region showing dense shadow of lumbar spine.

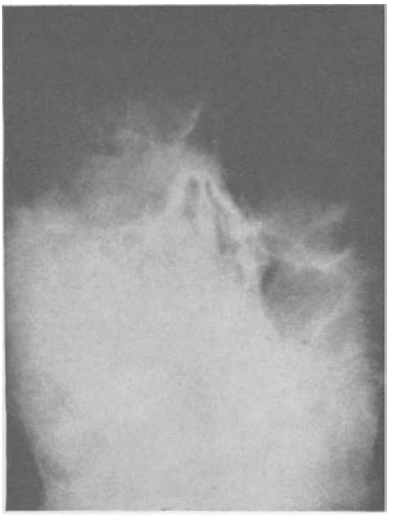


FIG. 4.
X-ray: dense shadow of right maxillary region.



FIG. 5.
X-ray of the pelvic region showing dense shadow of right iliac crest.

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The case under report was unilateral mainly. No pigmentation or endocrina abnormalities were seen in this case (contrary to opinion of Lichtenstein anc Jaffe (1942).

Summary

A case of fibrous dysplasia of polyostotic variety involving lumbar spines, frontal, malar, maxilla and mandible, iliac crest is presented and briefly discussed.

Acknowledgment

We take this opportunity to thank Dr. L. H. Hiranandani, Chief, E.N.T. Department, and Superintendent, Bombay Hospital, Bombay, for his permission to publish this case.



FIG. 6.

Biopsy report: fibrous dysplasia. Fibrous tissue with irregularly arranged trabeculae of bone.

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