CASE REPORT

Cranio-Facial Fibrous Dysplasia

Parmod Kalsotra, Monica Manhas

Abstract

An unusual case of fibrous dysplasia involving left cranio-facial region in an adult male is reported. The clinical features, radiological findings and the treatment have been discussed.

Key-words

Fibrous dysplasia, Cranio-facial.

Introduction

Fibrous dysplasia is a pathologic condition of bone of unknown aetiology with no apparent familial, hereditary or congenital basis. Lichtenstein first coined the term in 1938 (1) and in 1942 he and Jaffe separated it from other fibro-osseous lesions (2). In fact, there were thirty-three synonyms listed in their paper in 1942, and the term fibrous dysplaisa was not listed in the index medicus until 1967 (3).

Fibrous dysplasia constitutes 2.5% of all bony neoplasms, and 7% of all benign bony tumours (4). Common sites of involvement are femur, tibia, fibula, ribs and facial bones. In the cranio facial region, the maxilla is most frequently involved (5,6). The mandible, ethmoid and sphenoidal regions follow in the order.

Many theories have been suggested regarding the cause of fibrous dysplasia, including the trauma with a nonspecific distrubance in local bone reaction, (7, 8) a congenital anomaly (2) "Perverted" activity of mesenchymal bone-forming cells, (9, 10) and a complex endocrine disturbance with local bone susceptibility. However, the most readily acceptable theory at this time is the abnormal activity of mesenchymal cells.

Fibrous dysplasia is divided into *monostotic and polyostotic* variety (11). It may involve multiple bones associated with cutaneous pigmentation and precocious puberty known as Albright's syndrome (12).

Case Report

A thirty year old male presented with a swelling on the left side of the nose and left cheek and mild proptosis for a period of five years (Fig. 1). There was no history of pain, trauma, epistaxis, loosening of teeth, trismus or diminished vision. Examination of the region revealed a smooth bony-hard swelling involving left maxilla, zygoma

From the Department of ENT, ASCOMS and Hospitals, Sidhra, Jammu, J&K.
Correspondence to : Dr. Parmod Kalsotra, Asstt. Prof. ENT. ASCOMS and Hospitals. Sidhra, Jammu.

and temporal region. The skin over the swelling was normal. The swelling was bulging in the gingivo-buccal sulcus with normal teeth. There was no other swelling in the body and cafe'-au-lait spots were not seen. The throat and nose examination was found to be normal. The routine blood and urine investigation were normal. Plain X-ray of the paranasal sinuses showed radio-opaque shadow over left cheek region. The CT scan (Fig. 2, 3, 4,) showed a radio-dense mass involving left maxillary sinus pushing the orbit from below and causing proptosis. The mass was also involving left zygoma, sphenoid and frontal bone with ground-glass appearance and expansion of bones. There was no evidence of bony destruction or intra-cranial extension. Patient was subjected to partial excision and currettage of the mass under general anaesthesia via sublabial approach.

Post operative period was uneventful. The histopathological examination suggested it to be fibrous dysplasia. At one year follow-up, there was no recurrence of symptoms.

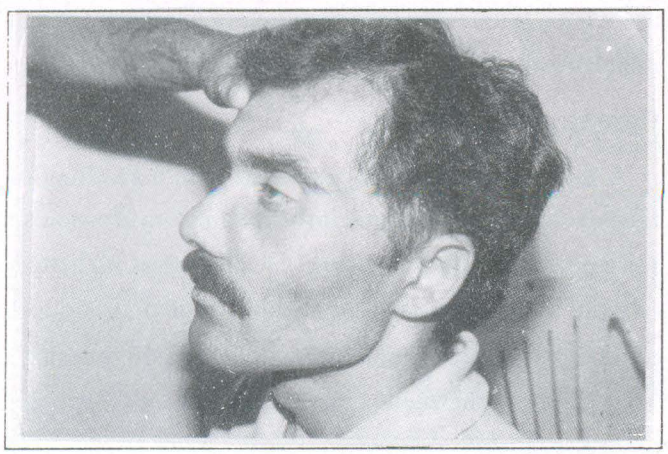

Fig. 1. Lateral view of patient showing swelling involving left cheek, maxilla and zygoma.

Fig 2, 3, 4–NCCT scans show radio-dense mass involving left maxilla, zygoma, pterygoid plate, sphenoid and parietal bones. Note the expansion of bones with no destruction or intracranial extension.

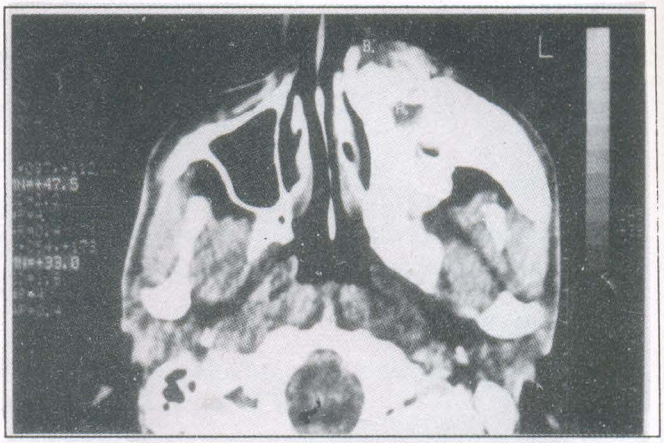

Fig. 2

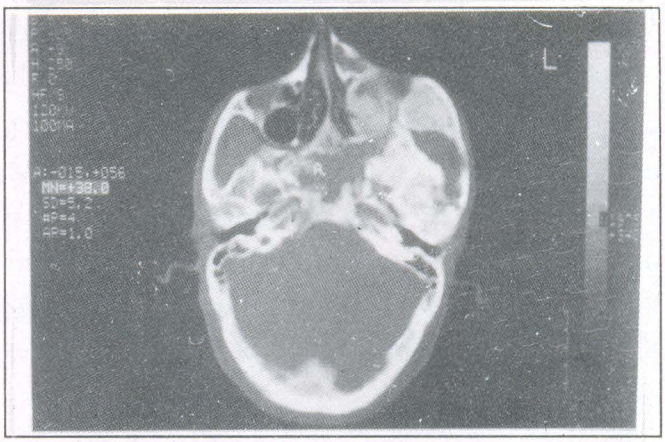

Fig. 3 .

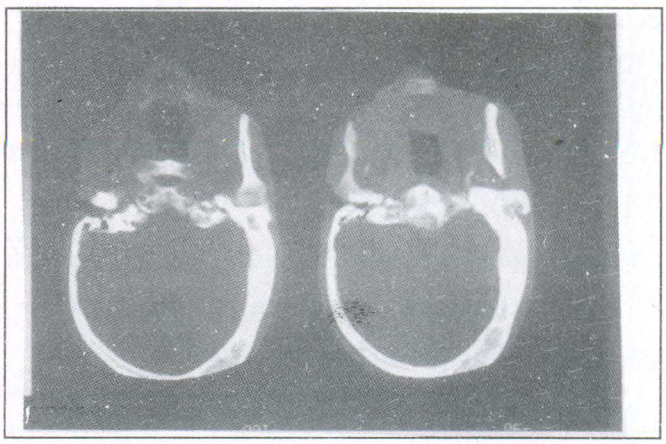

Fig. 4

Discussion

The mere presence of fibrous dysplasia of the craniofacial region is not in itself an indication for treatment. Many small solitary lesions will remain static and asymptomatic for long periods. A marked or progressive deformity, pain or functional disability suggest the need for intervention. Complete surgical resection of the involved area is the treatment of choice, but this results in considerable functional and cosmetic defect demanding extensive reconstruction. Conservative surgery (partial resection and currettage) is the basic surgical promise (11, 13). Also, the disease nearly always burns itself out around the puberty. Radiotherapy is to be avoided because it can induce malignant changes in the fibrous dysplasia (9, 14).

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