Fibrous Dysplasia of Right Upper Jaw— uncommon Presentation

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Abstract

MR, 30 years, M/M had huge nasal mass for 25 years and nasal obstruction for 15 years. On examination, a huge mass involving right side of face, nose, cheek, orbit and palate of 10" x 8" size was noticed. The right eye was proptosed eccentrically and was completely blind. Histopathological examination diagnosed the case to be fibrous dysplasia.

Case Report

A 30 year old muslim man attended ORL OPD at SSKM hospital, Calcutta with history of

- huge mass over right side of face for 25 years
- nasal obstruction for 15 years.

History of present illness revealed that at the age of 2½ years the patient had slight bulging of right eye. Gradually the eye was pushed out and the swelling increased to attain the present size. Initially the swelling was painless.

The patient experienced lightheadedness 4 months back as the mass was hollowed out by discharging necrotic material off and on.

He had complete nasal obstruction for 5 years.

Fig. I Fibrous dysplasia of right upper jaw.

Vision in the right eye gradually diminished with no residual vision at the time of presentation.

There was no history of epistaxis, deafness or tinnitus.

Examination revealed a huge mass involving right side of face, nose, cheek, orbit and palate. (Fig. 1) It was 10" x 8" in size with irregular surface and unhealthy shiny skin here and there. Right eye was proptosed eccentrically with no vision and left eye was normal. There was an opening over the swelling. The tumour was bony hard in feeling without any mobility.

Anterior rhinoscopy showed both the nasal cavities were full of hard mass and the nasal septum was pushed to the opposite side. Oropharynx showed bulged hard

Fig. II CT scan of nose & paranasal sinuses (Axial cut) shows marked deformity of facio-maxillary and frontal bones.

palate (more on right side) without any scar mark, ulceration or loosening of teeth. Other systemic examination was unremarkable.

Investigation

Serum Ca⁺⁺ and alkaline phosphatase were normal.

Radiological investigation (X-ray and CT scan) revealed marked deformity of facio-maxillary and frontal bones along with underlying sinuses. (Fig. 2)

The bones were thick and irregular with inward projection of right frontal bone-rest of the cranial bones and brain appeared normal.

Biopsy from nasal mass showed trabeculae of woven bone surrounded by fibroblastic tissue-but without any osteoblastic rimming. The diagnosis was stamped to be benign fibro-osseous lesion consistent with fibrous dysplasia.

Discussion

Though fibrous dysplasia, ossifying fibroma and cementifying fibroma are said to be lesions of bone with well defined characteristics, in the nose and paranasal sinuses a decision as to whether a lesion is fibrous dysplasia or ossifying fibroma is often difficult (Pindborg and Kramer, 1971). Again the criteria for the different entities show some overlap and the problem is further compounded by the variability in histological appearances in different parts of the same lesion. These difficulties may be resolved to some extent by employing the umbrella term benign fibro-

osseous lesion for conditions in which we cannot arrive at a more specific diagnosis.

The World Health Organization (Shanmugaratnam, 1978) defined **ossifying fibroma** as consisting of spindle-shaped fibroblastic cells and small islands of woven bone. The lesion is usually encapsulated and the bony trabeculae are rimmed by osteoblasts.

Fibrous dysplasia is defined as a non-encapsulated lesion of fibrous connective tissue containing islands of trabeculae or immature non-lamellar metaplastic bone. Osteoblastic rimming of bone trabeculae is said to be absent or inconspicuous. If there are multiple bony lesions of this type and particularly in the presence of excessive skin pigmentation, the diagnosis of fibrous dysplasia is easily made. The latter features are, however, rarely present in benign fibro-osseous lesions of the nose and paranasal sinuses. But some authors (like Michaels L, 1989) do not ascribe any significance to the presence or absence of osteoblastic rimming.

Miller et al (1996) are of opinion that fibrous dysplasia represents a developmental maturation defect or fibro-osseous metaplasia. It usually occurs in the first decade and is more common in girls than in boys. It involves maxilla more frequently than the mandible (in contrast to ossifying fibroma). The lesion is difficult to remove and contouring is usually the only therapy when it causes significant deformity or impinges on vital structures. The histopathological appearance shows presence of woven bone only with feathery, irregular borders (often described as Chinese characters). One sees random birefringence with polarized light.

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