Fibrous Dysplasia of Right Upper Jaw – Uncommon Presentation

Sumanta Kumar Dutta, Clinical Assistant, **Utpalendu Das**, Clinical Tutor,

T. K. Hazra, Assistant Professor

IPGME & R and S.S.K.M. Hospital, Calcutta-700020

30 years, male patient presented with huge nasal mass for 25 years and nasal obstruction for 15 years. On examination, a mass of 10" x 8" size found involving the right side of face, nose, cheek, orbit and palate. The right eye was proptosed eccentrically and was completely blind. Histopathological examination confirmed it as fibrous dysplasia.

CASE REPORT

A 30 year old muslim man attended ORL OPD at SSKM Hospital, Calcutta with history of progressive swelling of right side of the face for 25 years and nasal obstruction for 15 years.

Fig.1

Fibrous dysplasia of right upper jaw

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 attend the present size. Initially the swelling was painless.

The patient experienced lightheadedness about 4 months back as the mass was hollowed out by discharging necrotic material off and on and progressive loss of vision in Right eye which become totally blind at the time of presentation.

Fig.2

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

There was no history of epistaxis, deafness or tinnitus or anosmia.

Examination revealed a huge mass involving right side of face, nose, cheek, orbit and palate. It was 10" x 8" in size with irregular surface and covering skin was shiny here and there. Right eye was proptosed eccentrically with no vision and left eye was normal. There was an opening over the swelling. It was immobile and bony hard.

Anterior rhinoscopy showed both the nasal

cavities were full of mass and the nasal septum was pushed to the opposite side. Oropharynx showed bulged hard palate (more on right side) without any scar mark, ulceration or loosening of teeth. Other systemic examination was unremarkable.

Investigation

Routine investigation (Blood/urine) and Serum Ca^{++} and alkaline phosphatase were normal.

Fig.3

Lateral view of skull X-ray showing marked deformity of facial bones

Radiological investigation (X-ray and cT scan) revealed marked deformity of facio-maxillary and frontal bones and the underlying sinuses. The bones were thick and irregular with inward projection of right frontal bone—rest of the cranial bones and brain appeared normal.

HP study of the 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 difficult⁵. 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 Organization⁶ 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 likely diagnosis is fibrous dysplasia. 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.

References

1. Hammer J E, Scofield H H and Cornyn J (1968) : Benign fibro-osseous jaw lesions of periodontal membrane origin – an analysis of 249 cases. *Cancer*, 22 : 861.

2. Kridel RWH, Miller R H and Greenberg S D (1983) : Ossifying fibroma : diagnostic clarification. *Otolaryngologic Head Neck Surgery*, 91 : 568.
3. Michaels L (1989) : Head and Neck tumours. In *Recent Advances in Histopathology* ed Peter P Anthony and R N M MacSween. Churchill Livingstone, Edinburgh. p. 112.
4. Miller R H, Sturgis E M and Sutton C L (1996) : Neoplasms of the Nose and Paranasal sinuses. In *Otorhinolaryngology Head and Neck Surgery* ed John Jacob Ballenger and James B Snow, Jr. (15th ed). Williams and Wilkins. p. 202.
5. Pindborg J J and Kramer I R H (1971) : World Health Organization, Geneva.
6. Shanmugaratnam K (1978) : Histological typing of upper respiratory tract tumours. *International histological classification of tumours*. No. 19. World Health Organization, Geneva.
7. Waldron C A (1973) : Intraosseous fibrous swelling of jaws,, In Bergsma D (ed) *Birth defects, Atlas and Compendium*. The Williams and Wilkins Co. Baltimore.

Congratulation !

Dr. V. P. Sood was invited to deliver guest lecture on the occasion of annual E.N.T. Conference Trivandrum branch of Kerala State of A.O.I. on 27th July 1997 at Trivandrum.