Improving aesthetics in fibrous dysplasia maxilla: Case report

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Abstract
An intriguing case of the polyostotic fibrous dysplasia of the craniofacial region undertaken for osteoplastic facial remodeling through hemi mid facial degloving approach for facial asymmetry shall be discussed.

Keywords: Maxilla, expansile, osteolytic, chiselling, osseous

Introduction
Fibrous dysplasia of craniofacial region necessitates multilevel surgical intervention ranging from mere chiselling to extensive drilling of the exuberant osseous tissue. The facial contour is likely to be distorted with obliteration of the hollow of the canine fossa due to osteoid proliferation. Thereby the zygomatic and the nasomaxillary buttresses lie at the same plane. Sometimes a bony bulge is noted beneath the body of zygoma. The frontal and the helicopter profile/ view is ideal to appreciate the facial deformity sculpturing to restore the canine fossa of the maxilla in the pre adolescent population and an osteoplastic osteotomy of the maxilla anterolateral wall in post teens is the likely recourse a middle meatus antrostomy or a medial maxillectomy, re-establishes the mucociliary drainage of the effected sinus in ethmoidal-sphenoidal lesions, optic nerve and orbital extensive drill work serves to decompress and manage proptosis and visual impairment as the case may be. An intriguing case of the fibrous dysplasia of the craniofacial region shall be discussed.

Case report
28-year-old male patient reported to the Department of otorhinolaryngology and head and neck surgery with the chief complaint of painless swelling on the right side of face since childhood gradually increasing in size to attain the present size. There was no history of trauma, paresthesia, and difficulty in chewing food, and it was not associated with any other symptoms. A diffuse swelling (5x 4 cm approximately) was seen, superiorly starting from 1 cm below the infraorbital margin, medially obliterating the right nasolabial fold, to ramus of mandible on the right side laterally, which was bony hard and non tender in nature. (figure-1)
The skin over the swelling was normal. The swelling involving the right maxillary alveolar bone extended from upper incisors to the last molar with expansion of palate obliterating the buccal vestibule. Overlying mucosa appeared normal, firm, and was nontender. Complete haemogram showed normal parameters patient had primary mitral valve disease - severe mitral regurgitation. With mild aortic and tricuspid regurgitation. The computed tomography of the patient reported multiple expansile lesions with ground glass matrix and lucent area seen involving frontal bone, temporal bone, and body and greater wing of sphenoid, ethmoids, petromastoid part of right zygomatic bone, right side hard palate, maxilla and walls of maxillary sinus and right temporal bone, pterygoid plates, crista galli on right side with obliteration and partial opacification of cavities of involved sinus. Consistent with polyostotic fibrous dysplasia. (Figure-2, 3)

1x2 cm osteoplasty was undertaken and outer cortex was removed using chisel leaving 1 cm superiorly below infraorbital rim and above the alveolar ridge. Intramaxillary bone which was cancellous and friable was curreted out and it was communicated with nasal cavity. (Figure- 5)

A trans nasal endoscopic middle meatus antrostomy was widened to complete a medial maxillectomy thereby facilitating drainage of maxillary sinus into the nasal cavity (Figure-6).

The residual mucosa over the haller cell region and infraorbital nerve area was retained to facilitate mucociliary clearance. Postop period was uneventful any expected swelling due to intra op retraction induced edema was countered by placing a rollerguaze bolster over the new canine fossa area which was removed on the fifth post op day. Head elevation and steroids were given along with aerobic and anaerobic coverage. (Figure-7)
The histopathological report of the patient was consistent with fibro-osseous lesion. (Figure-8)

Discussion

Fibrous dysplasia is a fibro-osseous disorder of mesenchymal origin with exuberant proliferation of the fibrous matrix with interwoven osseous tissue. Lichtenstein in 1938 and Lichtenstein and Jaffe in 1942 described fibrous dysplasia as benign intramedullary fibro-osseous lesion [1] while Reed defined it as disorder of arrest of bone maturation with presence of woven bone with ossification [2]. WHO too classified Fibrous dysplasia as developmental in origin with etiology being mutation in the Gsα gene located at chromosome 20q13.2-13.3 [3]. The specific location of the mutation is at position 201, which is usually occupied by arginine (R201) and is replaced by either a cysteine (R201C) or a histidine (R201H) [3].

Three entities of fibrous dysplasia (FD) have been described: monostotic FD, where only one bone is involved; polyostotic FD, where multiple bones are involved; McCune-Albright syndrome, where FD is associated with café au lait spots and multiple endocrinopathies (rare) [4, 5].

Primarily a disease of young population fibrous dysplasia has incidence of 1:4000 to 1:10,000 [6]. Craniofacial structures are involved in 10% of monostotic type, 50% of mild polyostotic cases, and 100% of severe polyostotic cases. Maxilla and mandible are commonly affected with the temporal bone involved in 18% of cases [6]. Craniofacial fibrous dysplasia in the wings of the sphenoid may compress the optic nerve with consequent effect on vision during expansile growth phase of the facia skeleton more so in prepubertal individuals though in our case facial asymmetry was the main presenting complaint midface involvement can progress to nasal obstruction, restricted movement of the eye. Mastication is affected due to malocclusion of alveolar ridges and palatal expansion. Predominantly sclerotic (34 to 38% of cases), Predominantly lytic or cystlike (11 to 22% of cases), and mixed (40 to 55% of cases) are the three radiographic patterns described in fibrous dysplasia [7]. A reason for the difference in appearance between maxillofacial FD and FD of long bones is that the former occurs in skeleton derived from membranous bone [8]. Solitary unilocular cyst, non-ossifying fibroma, eosinophilic granuloma, cholesteatoma, meningioma, Paget's disease, osteochondroma, ossifying fibroma, giant cell reparative granuloma, exostoses, aneurysmal bone cyst, cystic fibrous osteitis; and malignancies, such as: Sarcoma and metastatic osteoblastic lesions are the various differential entities [9]. Skeletal deformities require a surgical approach which includes both conservative and radical procedures. Conservative shaving or osseous contouring has been recommended by some authors who maintained that periodic contouring could be performed until a static phase was reached. Radical approach permits the complete removal of the lesion followed by immediate reconstruction [10].

The Michael Moure, Lateral Rhinotomy, Weber-Ferguson, midfacial degloving approaches can be undertaken for surgical access of fibrous dysplasia in the midface [11].

We performed osteoplastic radical procedure for facial asymmetry with removal of osteoplastic flap in the middle so that soft tissues of the cheek prolapse and fill the hollow of the new canine fossa. Moreover drillwork was done on the naso maxillary buttress and zygomaticomolar buttress to smoothen the prominence in these regions. Malignant changes with FD include osteosarcoma, fibrosarcoma, chondrosarcoma, and Ewing's sarcoma [12].

Chemotherapy is ineffective as a curative modality while radiotherapy is contraindicated due to likelihood of late malignant transformation [13]. A regular follow-up is essential to detect early relapses or malignant transformation.

Conclusion

Surgical intervention limited contouring or radical osteoplastic flap procedure is undertaken in fibrous dysplasia to improve the facial aesthetics and sometimes functional impairments as the case may be.

References

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