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MODIFIED MANAGEMENT IN THE TREATMENT OF CRANIO-FACIAL FIBROUS DYSPLASIA:A CASE REPORT

Dental Science

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KEYWORDS:

Introduction:
Fibrous dysplasia (FD) of bone is a genetic, non-inherited disease caused by activating mutations of the GNAS1 gene which encodes the α-subunit of the stimulatory G-protein Gs. It may present as an isolated skeletal lesion (monostotic form) or involve multiple skeletal sites (polyostotic form) in addition to single or multiple endocrinoapthies.[1] McCune-Albright syndrome (MAS) is a condition which includes a classical triad of polyostotic fibrous dysplasia, Café au lait macules and endocrinopathies.[2] Whereas, Mazabraud syndrome is the term used to note the combination of FD of bone with myxomas of the skeletal muscle.[1]

Cranio-facial FD is an aggressive lesion with rapid enlargement of the involved bones which may involve the adjacent vital structures such as the optic nerve, globe, and auditory canal including nasal airway resulting in functional deficits.[2]For this reason some authors suggest an aggressive resection of the lesion. Weber-fergusson's (WF's) approach is most commonly used for the resection of the tumours involving midface.[3]The present paper focusses on a modified Weber-fergusson's incision to resect FD involving the maxilla,zygomatic bone and floor of the orbit.

Case report:
A 20 year old male patient reported to the clinic with a chief complaint of swelling on the right side of mid-facial region since 10 years.

On extra oral examination a diffuse swelling was noticed on the right side of the face, extending superiorly from the infra-orbital region to inferiorly up to 2cm above the level of upperlip. Medially, the lesion was extending from the right philtrum of the nose to 1cm away from the outer canthus of the eye. There was a raise in the pupillary level of the right eye and obliteration of the naso-labial fold. [Figure 1]

Fig 1: Preoperative View

On intraoral examination a diffuse swelling was noticed on the right side of the maxilla extending from mesial surface of the right canine to distal surface of third molar. Based on the above findings a provisional diagnosis of craniofacial fibrous dysplasia was made. [Fig 2]

Fig2: Intraoral examination

A computerized tomography scan was taken which showed diffuse radioopacity on the right side involving the maxilla, maxillary sinus, zygomatic bone and infra-orbital rim and outer canthus of the orbit.[Fig 3].

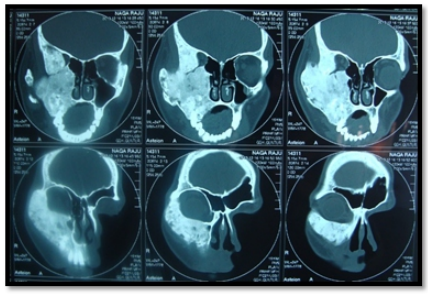

Fig 3 : Computerized tomography scan

Before starting of the treatment all the preventive treatments were done (Oral prophylaxis and Restorations). An incisional biopsy was performed by opening a window on the right side of the maxilla and was submitted for histopathological examination. Histopathological examination revealed numerous bony trabeculae arranged in curvilinear pattern interspersed with uniformly spaced fibroblasts. The trabeculae showed numerous lacunae containing osteocytes with absence of osteoblastic rimming. The trabeculae appeared to be separated from the connective tisuuestroma and were blended with the surrounding fibrous tissue at the periphery. Based on the above findings a final diagnosis of fibrous dysplasia was made. The patient was then explained about the surgical excision procedure and informed consent was taken.

A modified Weber-Fergusson's (lateral canthal extension) incision was planned for the surgery by extending the incision beginning from the upper lip in the philtrum to transversely through the nasolabial groove to beyond the nasal ala and then superiorly through the nasofacial groove to the lower eyelid. Once the medial canthus of the eye was reached the incision was extended laterally inferior to the lower eyelid. (Dieffenbach's modification of WF's approach) Further, the incision was extended 2cm (not more than 3cm) beyond the external canthus of the eye, to prevent the injury to the temporal branch of the facial nerve. [Fig 4]

Fig 4

After the incision was made surgical resection of the lesion was done and debulking of the involved bones was done. [Fig 5]

Fig 5: Surgical resection of the lesion

The globe of the eye was retracted with help of globe retractor and surgical recountouring of the orbital floor was done. [Fig 6] Orbital contouring is done to correct the globe position of right eye and make it equal to pupillary level. Floor is contoured so as not to extend to medial side, to prevent injury injury to lacrimal apparatus and medical canthon tendon.

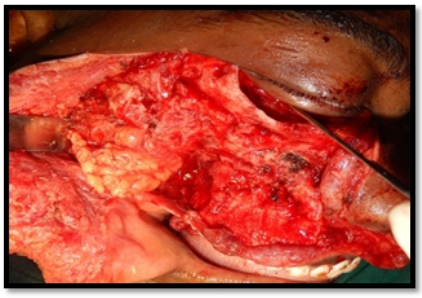

Fig :6 Surgical recountouring of the orbital floor

The excised specimen was submitted for histopathological examination and diagnosis was confirmed as fibrous dysplasia. The patient was followed up post-operatively, and no recurrence was noticed till date.

Discussion:

Fibrous dysplasia was first reported by Von Recklinghausen about a century ago who named it as ***osteitisfibrosageneralisata***. Jaffe and Lichtenstein first introduced the term fibrous dysplasia.[4] FD accounts for 2.5% of all bone tumours and 7.5% of the benign neoplasms. The three subtypes of FD are monostotic, polyostotic and craniofacial. The term craniofacial fibrous dysplasia (CFD) is used to describe FD were the lesions are confined to contiguous bones of the craniofacial skeleton.[5]

Clinically CFD presents as an asymptomatic swelling of the involved bone, typically contiguous involvement of maxillofacial and cranial bones. The lesions are usually diagnosed in children and young adults.[6]Themonostotic form has equal gender distribution, but polyostotic forms are common in females. Maxilla and paranasal sinuses are affected commonly than mandible.[7]

Radiographically, they present as a poorly defined lesion that merges with the adjacent bone. Early lesions may be radiolucent, but they become increasingly radiopaque and typically show a diffuse radiopacity or 'ground glass' appearance.[7]

Although, the use of bisphosphonates such as alendronate, pamidronate or zoledronic acid reduces the pain and growth rate of the lesion;[2] surgical approach is the gold standard for the reduction of the swelling and symptoms associated with CFD.

The options for surgical management of FD include either en bloc resection or contour excision with or without bone grafting. Various approaches to the midfacial region can be used for the resection of the tumours. Among them Weber Fergusson's approach and Midfacedegloving incision are commonly used.[8,9]

The classical Weber-Fergusson's approach may be modified depending on the extent and nature of the tumour. The incision may be modified to Dieffenbach's incision or Lynch incision or Borle's extension.[3,8,9,10] In the present paper, a different modification of Weber-Fergusson's incision was followed. After reaching the external canthus of the eye the incision was extended further 2cm beyond, to prevent damage to the temporal branch of the facial nerve.

Conclusion:

We have modified the approach for wide exposure of the lesion and orbital floor contouring for correction papillary level, and provided the current understanding of the biologic and clinical characteristics of FD and recommendations for the clinical management in the craniofacial region. Most importantly, each patient may present with variable symptoms and clinical findings, thus the care of these patients must be customized to their needs and sites of involvement.

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