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Case Report

Revisiting Midfacial Degloving Approach for Craniofacial Fibrous Dysplasia

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ABSTRACT

Craniofacial Fibrous dysplasia is a tumor like expansile benign lesion which may end up in disfigurement of the face. The skull is involved in 15% with the majority being monostotic. One-third of the cases are located in the maxilla or mandible. Of the skull bones, maxilla is more commonly involved especially in a growing age. Palpable or visible swelling of the involved bone is found initially and later deformity and compression of functionally important structures create the main symptomatology. Surgery is the mainstay treatment. The approaches usually used are lateral rhinotomy, Weber Ferguson's incision along with a medial maxillectomy or total maxillectomy. They usually leave a scar on the face and offer limited access to the opposite side in a same sitting. Here, in the young lady case, a sublabial midfacial degloving approach was taken which gave the excellent access to both the maxilla, and the entire extent of the lesion with no visible post operative scar.

Key words: Fibrous dysplasia, maxilla, midfacial degloving approach

INTRODUCTION

Craniofacial Fibrous dysplasia commonly involves maxilla and mandible and maxilla is more commonly involved especially in a growing age. It is a tumor like expansile benign lesion which may end up in disfigurement of the face. It is usually self limiting, not encapsulated and is characterized by replacement of normal bone with cellular fibrous connective tissue, which contains irregular trabecles of immature, nonlamellar, metaplastic bone. The aetilology is unknown. The disease may progress actively during childhood and stabilize in adulthood. Clinically, three types

are seen; monostotic, polyostotic and McCune -Albright syndrome. The skull is involved in 15% with the majority being monostotic. One-third of the cases are located in the maxilla or mandible. In most of the cases, palpable or visible swelling of the involved bone is found initially and later deformity and compression of functionally important structures create the main symptomatology. The disease has tendency to recur following surgery and in cases of young patients recontouring surgery or, in special situations decompression of the optic nerve are important options. ^[1] Radiation should be avoided as there is a sarcomatous

transformation rate of about 0.4% which increases over 40% following to radiotherapy.^[1] Medicines like alendronate and other biphosphonates can be given in painful lesions but surgery is the mainstay treatment. The approaches usually used are lateral rhinotomy. Weber Ferguson's incision along with a medial maxillectomy or total maxillectomy. They usually leave a scar on the face and offer limited access to the opposite side in a same sitting. Here, in this young lady, a sublabial midfacial degloving approach was taken which gave the excellent access to both the maxilla and the entire extent of the lesion with no visible

post operative scar. This approach is recommended for the lesions and tumors arising in the midfacial region.

CASE REPORT

This 14 years old girl presented with a swelling over the left cheek for the last 6 months. The swelling was insidious in onset, gradually progressive with no aggravating or relieving factors or any other associated symptoms like pain, visual disturbances, loosening of teeth or paraesthesia over the face. The only complaint was the progressive disfigurement of the midface on left side (Pictures A & B of Fig 1).



Fig. 1 (Pre operative and post operative pictures)

The nasal endoscopic examination of the patient showed medial bulging of the lateral wall of the left nasal cavity with difficulty to negotiate the endoscope, although the patient didn't have any complaints of nasal obstruction or anosmia. CT scan showed an expansile lesion with ground glass homogenous matrix seen in the medulla of maxillary bone (left) involving left maxillary alveolar process, hard palate, bones of left maxillary sinus, zygoma and medial wall of left orbit. The left maxillary sinus was compressed and displaced medially. There were no definable edges for the lesion (Picture E of Fig 2). The patient was taken up for surgical debulking with recontouring of the left maxilla by a midfacial degloving approach under general anaesthesia. Sublabial incision was given in the upper gingival and gingivobuccal sulcus 1 cm above the roots of upper maxillary teeth extending from second premolar on the right side to second molar on the left side. The upper lip along with its soft tissue and underlying periosteum reflected upwards to expose the anterior surface of the maxilla (Picture H of Fig 3).

In the nose, intercartilagenous incisions along with marginal incision were given bilaterally and connected with a full transfixation cut to separate the flap from septum, upper lateral cartilage and medial crura of lower lateral cartilage (Picture I of Fig 3). A bipedicled flap laterally was raised on the left side and the lateral nasal mucosa incised at the piriform aperture to get more access on the affected side. Dissection was done in the subperiosteal plane on either side to expose the infra orbital rim superiorly and zygomatic butteress laterally on the affected side. Total degloving of the midfacial region achieved after identification of the infra orbital nerves which were preserved. The lesion was drilled out as a modified Caldwell Luc

approach upto the level of the posterior wall of the maxillary sinus which was displaced superomedially by the lesion. The mucosa of the left maxillary sinus was excised and maxillary ostium widened on the lateral nasal wall. The left maxilla was drilled further at the orbital rim and laterally till zygomatic complex. The maxilla was drilled using cutting burrs and polished with diamond burrs to give a cosmetically normal looking midfacial skeletal contour (Pictures J & K of Fig 3).

The soft tissue wound was closed in layers (Pictures L & M of Fig 4).



Fig 2 (CT scan & Histopathological pictures)



Fig 3 (Surgical steps- Midfacial degloving and drilling of maxilla)



Fig 4 (Wound closure in layers)

The post operative period was uneventful and the patient recovered quickly. There was no cosmetic deformity or scar over the face (Pictures C & D of Fig 1). The diagnosis of fibrous dysplasia was confirmed by histopathological examination (Pictures F & G of Fig 2). Clinically there were no signs of recurrence or facial deformity after 1 year of follow up.

DISCUSSION

The degloving approach involves no external scaring and improves visibility of total operative field. ^[2, 3] By this approach, we can accomplish en bloc dissection of lateral nasal wall, maxilla and is extended to include medial orbital wall, as indicated by the extent of the disease. The Weber Fergusson incision although provides comparable access, it is limited by the unilaterality and a visible midfacial surgical scar. ^[4] A facial degloving procedure was first suggested by Portmann in 1927, but the modern technique had its origin in 1974 with the report of Casson et al. Conley and Pirce first suggested the use of the midfacial degloving procedure for excision of neoplastic diseases in 1979. Later, Maniglia popularized this technique for adequate exposure to bilateral maxilla and lower nasal cavity without cosmetic deformity. ^[5] This approach can also be used in benign fibroosseous conditions, facial fractures, orthognathic surgery, bone grafting and in Juvenile nasopharyngeal angiofibroma. ^[5, 6]

In the last few years, this approach is slowly increasing in popularity in the management of extensive benign lesions of the sinonasal region, lateral sphenopalatine and infra-temporal fossa, ^[2] in selected cases of malignancy in this area and to afford access to the nasopharynx. ^[7] The authors recommend this approach for the above mentioned indications.

CONCLUSION

Midfacial degloving approach is a cosmetically well accepted approach for

benign fibro- osseous lesions, repositioning of facial bone fractures, maxillofacial surgeries and as an approach to nasopharynx for excision of tumours like nasopharyngeal angiofibroma. It leaves no scars on the face and can give access to both sides of midface.

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