

Endoscope assisted sublabial approach for management of Fibrous dysplasia of right zygomatico-maxillary bone: A case report

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Abstract- Fibrous dysplasia (FD) is an uncommon rare skeletal disorder in which normal bone is replaced by abnormal fibro-osseous tissue. It comprises about 2.5% of all bone tumour. Monostotic fibrous dysplasia is more common than the polyostotic form and constitutes about 70-80% of fibrous dysplasia and has no gender predilection and mostly seen in second and third decade of life. Here we present a case report of a 52 years old female presented with swelling over the right cheek for last 4 months with occasional facial pain. On NCCT scan of Nose and PNS showed a well demarcated expansile medullary bony lesion containing unorganized bony trabeculae measuring 34mm (CC)*26mm (AP)*26mm (ML) arising from right zygomatic bone with intralesional calcification causing mass effect and indenting the lateral wall of right maxillary sinus - feature suggestive of Fibrous dysplasia. The patient was treated under general anaesthesia by the surgical contouring and bone shaving of the tumour. Endoscope assisted sublabial approach was made to counter limited visualization and complete excision of Fibrous dysplasia of right zygomatico-maxillary bone which was later confirmed on histopathological examination. The cosmetic challenge from facial deformity due to fibrous dysplasia was overcome by intrabdominal fat graft with no intraoperative and postoperative complication and uneventful postoperative follow-up.

Keywords- Fibrous dysplasia, Maxilla, NCCT Scan of Nose and PNS & 3D reconstruction of face, Endoscopy, intrabdominal fat graft, HPE.

INTRODUCTION:

Fibrous dysplasia (FD) is a sporadic non-neoplastic hamartomatous developmental fibro-osseous lesion of bone characterized by an abnormal mixture of fibrous and osseous elements in the interior of affected bones replacing cancellous bone which comprises 2.5% of all bone tumours and in total about 7% of all benign bone tumours [1]. Monostotic fibrous dysplasia is more common than the polyostotic form constitutes about 70-80% of FD patients and has no gender predisposition and is mostly seen in the second and third decade [1]. Clinically, the monostotic form is characterized by a lytic expanding bone lesion, which may cause pain, deformity, fractures, and nerve entrapment. Maxilla is the most commonly affected bone in craniomaxillofacial region. Maxillary lesions may extend to involve the maxillary sinus, zygoma, sphenoid bone and floor of the orbit and require surgical intervention resulting in an acquired defect of the involved site [2].

Here we aimed to report a case of fibrous dysplasia because of the rarity of the tumor, the practicality of the new surgical approach where use of endoscope to counter the limited visualization inherent with sublabial approach and thus helps in complete excision of tumor and the use of intra-abdominal fat graft that overcomes the facial deformity due to FD which is a cosmetic challenge.

Case Report:

A 52-years old female presented on 16th August 2022 to the Department of Otorhinolaryngology, JNIMS, Imphal, Manipur, India with a complaint of swelling over the right cheek for last 4 months with occasional facial pain. In her past medical history, the patient reported no systemic diseases as well as no syndromic presentation. On clinical examination, mild facial asymmetry associated with facial swelling was observed in the right side of the face. On palpation of the lesion a hard consistency associated with mild tenderness was elicited. There was no lymphadenopathy, skin pigmentation, tachycardia or any other alteration of skin colour (Fig.1). Intraoral examination was unremarkable with normal appearance of oral mucosa. On 0^o endoscopy of nose mild DNS to the left was noticed.

On NCCT Scan of Nose and PNS- shows a well demarcated expansile medullary bony lesion containing unorganized bony trabeculae measuring 34mm (CC)*26mm (AP)*26mm (ML) arising from right zygomatic bone with intralesional

calcification. The lesion is causing mass effect and indenting the lateral wall of right maxillary sinus- feature suggestive of Fibrous dysplasia (Fig. 2a & 2b)

On 3D reconstruction of Face- The anterolateral wall of right sided maxillary bone is comparatively more elevated than left side with absence of sign of erosion. (Fig. 3)



Figure 1-Preoperative pic

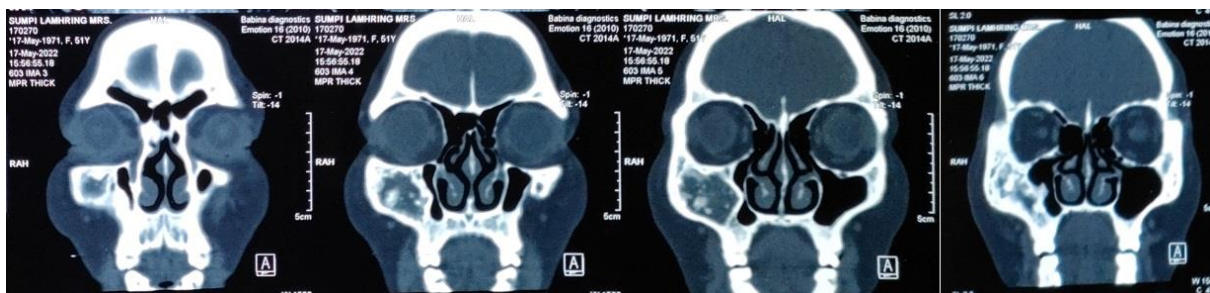


Figure 2a- NCCT Nose and PNS- Coronal view



Figure 2b-NCCT Nose and PNS- Axial view



Figure 3- 3D Reconstruction of Face

Surgical management-After receiving informed consent from the patient, surgical excision of the mass with right facial contouring was planned under general anaesthesia. A 4cm length horizontal sublabial incision was given. After raising the mucosal flap access to affected bone was established. Complete excision of lesion was achieved by combination of hammer and chisel (fig 4a) and micromotor drill (fig 4b). Extension to the zygomatic bone was excised with endoscopy guided drilling of affected bone. Haemostasis was achieved by electrocautery (fig 4c). Intrabdominal pad of fat was harvested (fig 4d) and the cavity was filled. Endoscopic right sided middle meatal antrostomy was performed (fig 4e) and intactness of the walls of maxillary sinus was ascertained.

Figure 4-Surgical contouring and bone shaving of the right maxillary mass with reconstruction.

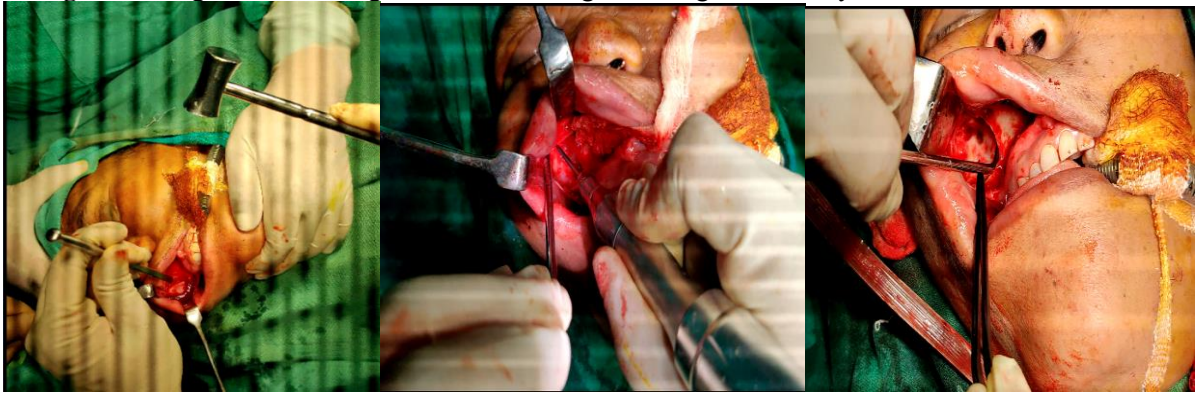


Figure 4a

Figure 4b

Figure 4c



Figure 4d

Figure 4e

The specimen was send for histopathological examination(HPE). Microscopic examination using haematoxylin-eosin stain, shows admixture of fibrous and osseous tissues. Osseous component is composed of woven bone in branching and anastomosing irregular trabeculae with absence of conspicuous rimming(fig 5a). Intervening fibrous tissue reveals bland fibroblastic tissee(fig 5b). There was absence of any cytologic atypia of abnormal mitosis. Hence overall features were suggestive of fibrous dysplasia.

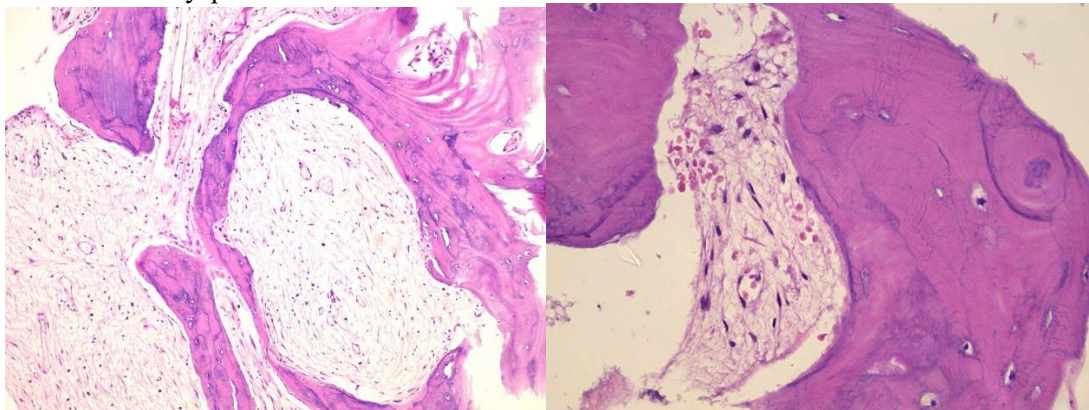


Figure 5a-10X view-Anastomosing strand of bony trabeculae.

Figure 5b- 40X view- No conspicuous osteoblastic rimming.



Figure 6-Post operative pic.

There was no post operative complication. The follow-up was done on 1st wk, 1st month, 3rd month and 6th month with absence of any complaint.

DISCUSSION :

As per the modified classification of fibro-osseous lesions of the craniofacial complex as proposed by Eversole et al, there are mainly 6 pathological process based on which disease conditions and their subtype have been classified [3]. Fibrous dysplasia, the disease condition under the bony dysplasia is a sporadic (non-inherited), non-neoplastic developmental disorder, being subclassified as monostotic form (including craniofacial variant), polyostotic form, polyostotic with endocrinopathy (McCune-Albright syndrome) [4]. It is caused by mutation of somatic cells in fetal (polyostotic FD) or postnatal (monostotic FD) life resulting in alterations in osteoblast growth/differentiation due to faulty expression/signalling of the GNAS1 gene (point mutation on chromosome number 20). These changes inhibit GTPase activity that is normally required to deactivate the G protein, thereby altering signal transduction pathways. The increased amount of cAMP in bone stromal cells leads to unregulated proliferation and differentiation [5].

FD comprises only about 2.5% of all bone tumour, which clearly defines the rarity of these uncommon skeletal disorder. Monostotic fibrous dysplasia is more common than the polyostotic form constitutes about 70-80% of FD patients and has no gender predisposition and is mostly seen in the second and third decade [1]. Although most cases of FD are self-limiting and hamartomatous, some cases of FD do not go into dormancy at the end of adolescence and may be activated or reactivated in adulthood in response to a life event, such as pregnancy. Monostotic FD of the jaws more commonly involves the maxilla than the mandible [6]. Based on the location of the craniofacial FD lesion(s), the symptoms and signs may differ. The monostotic FD present as painless, slow growing facial asymmetry or swelling. The smooth, fusiform enlargement is more marked buccally than palatal affecting the disordered eruption of teeth and with involvement of maxillary sinus and zygomatic process, sphenoid bone, floor of orbit results in exophthalmos and proptosis. The progression of the lesion may cause aesthetic impairment and deformities such as facial asymmetry, sinusitis, orbital dystopia, nasal malfunction [6]. In our patient volume increase in the maxillary region, hard consistency on palpation, expansile facial swelling in right zygomaticomaxillary region without orbital or buccal involvement were noted.

CT is the test of choice for the study of lesion(s), evaluation of its extension, and surgical preparation. There are three general radiographic patterns of CFD including: Ground glass appearance with mixed radio-dense and radiotransparent areas; sclerotic, and cystic patterns [7]. In our patient a well demarcated expansile medullary bony lesion containing unorganized bony trabeculae arising from right zygomatic bone with intralesional calcification was noted. CT scans before the surgical procedure to evaluate the actual size of the lesion, anatomical structures, and their involvements. Furthermore, this modality assists the surgeons to estimate the extent of surgical osteotomy and osteoplasty.

The differential diagnosis includes benign lesions: nonossifying fibroma, eosinophilic granuloma (bevelled margins in skull with typical bulls eye lesion on x-ray), Paget's disease, osteochondroma, giant cell reparative granuloma (no sclerotic margins), aneurysmal bone cyst; simple bone cyst, osteofibrous dysplasia (almost exclusively in the tibia of children <10 years with anterior bowing, monostotic, lesion begins in cortex, spontaneous regression) and malignancies like metastatic osteoblastic lesions [7][8]. In our patient, the initial diagnosis on clinical aspects along with imaging findings was in accordance with fibrous dysplasia of monostotic form which was later confirmed by the histopathological examination post-surgery.

Surgical treatment is recommended whenever clinical symptoms occur and to relieve intractable pain and skeletal deformity. In our patient to achieve complete excision and to prevent the recurrence with maintenance of facial aesthetics, endoscope assisted sublabial approach was employed for excision of tumour and facial contouring along with filling up of the defect with intrabdominal fat graft.

CONCLUSION:

Fibrous dysplasia is a rare tumor [1]. It presents with painless bony swelling, most patients generally report very late, after significant enlargement and involvement of contiguous structures with cosmetic deformity have occurred. Surgery is recommended as the modality of treatment to relieve the intractable pain and the skeletal deformity. Surgical treatment remains the mainstay of therapy and is directed at correcting or preventing functional deficits and achieving normal facial aesthetics which is a cosmetic challenge. Because it is a relapsing tumour, it is critical to remove as much tissue as possible without inflicting mutilation, functional impairments, or lesions of noble structures [7]. Hence use of endoscope assisted sublabial approach for facial contouring with intra-abdominal fat graft overcomes the challenge. The follow-up is of fundamental importance in order to detect relapses.

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