International Journal of Otorhinolaryngology and Head and Neck Surgery
Jayakumar V et al. Int J Otorhinolaryngol Head Neck Surg. 2022 Mar;8(3):259-262
http://www.ijorl.com
pISSN 2454-5929 | eISSN 2454-5937

DOI: https://dx.doi.org/10.18203/issn.2454-5929.ijohns20220479

Case Series

Fibrous dysplasia of maxillary sinus-our experience

Varunkumar Jayakumar[1]*, Shubhalaxmi Atmaram Jaiswal[2]

[1]Department of Otorhinolaryngology, Topiwala National Medical College and BYL Nair Charity Hospital, Mumbai, Maharashtra, India
[2]Department of Otorhinolaryngology, Dr. Vaishampayan Memorial Government Medical College, Solapur, Maharashtra, India

Received: 07 January 2022
Revised: 30 January 2022
Accepted: 01 February 2022

***Correspondence:**
Dr. Varunkumar Jayakumar,
E-mail: varun.jvk@gmail.com

ABSTRACT

The fibrous dysplasia is a benign bone disease, of slow growth and unknown etiology. The involvement of the craniofacial skeleton is not uncommon and generally produces facial asymmetries. Presenting case series with fibrous dysplasia occupying the entire maxillary sinus confirmed with the radiological investigations. The patients were clinically examined and with proper explained consent patients were worked up for excision of the involved lesion. Coronal sections of paranasal sinuses was done and diagnosis was confirmed. Patients were posted for recontouring of fibrous dysplasia and achieved cosmetic refigurement. The surgical treatment remains as the main therapeutic approach and the postoperative follow-up is necessary due to this condition recurrent nature.

Keywords: Fibrous dysplasia, Ground-glass appearance of maxilla, Cosmetic surgery of face, Maxillary sinus

INTRODUCTION

Fibrous dysplasia is a benign, chronic, slowly progressive bone disorder of unknown etiology. Histologically it's a fibro-osseous lesion characterized by the replacement of normal bone with a variable amount of fibrous tissue and woven bone which causes progressive expansion and deformity of bones. It comprises 2.5% of all osseous and 7% of all benign bone tumors with prevalence of 1 in 4000 to 10,000 individuals due to mutation of GNAS1 gene. Presenting the case series of patients with clinically, radiologically and histopathologically confirmed fibrous dysplasia of maxillary sinus which is rare and needs prompt diagnosis and management imposing the fact that over correction need not be done.[1]

CASE SERIES

Case 1

A 27 year old female came with complaints of: right cheek swelling since 6 months with no h\o trauma, previous surgery, comorbidities. On examination: ear, nose, and oral cavity: NAD. Right cheek swelling bony hard in consistency, irregularity and non-tender, no e\o warmth with skin induration (Figure 1). Patient was investigated and computed tomography of paranasal sinuses (CT PNS) was done. Right maxilla ground glass appearance with bony thickening and not extending beyond the right maxillary sinus s\o fibrous dysplasia (Figure 2). Patient was worked up for surgical correction for right fibrous dysplasia. Procedure done was right maxilla reconturing.

Patient was under AAP, IV line was started and general anaesthesia was given. Incision was right sublabial incision. Patient postoperative period was insignificant. Patient is under regular follow-up.

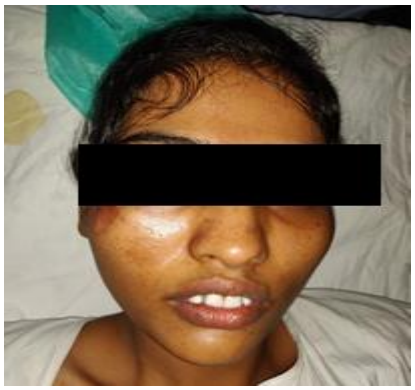

Figure 1: Clinical picture showing right maxillary region swelling.

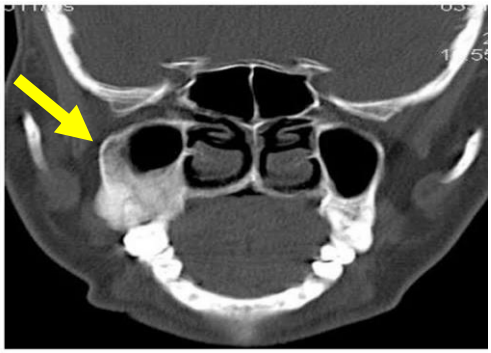

Figure 2: CT PNS showing right maxilla bony thickening.

Case 2

A 12 year old male came with complaints of: right cheek swelling since 1 year with no h\o trauma, previous surgery, and comorbidities. On examination: ear, nose, and oral cavity: NAD. Right cheek swelling bony hard in consistency, irregularity plus non tender, no e\o warmth (Figure 3).

Figure 3: Clinical picture showing right maxillary region swelling.

CT paranasal sinus was done, right maxilla ground glass appearance with walls showing bony thickening with no intraorbital involvement s\o fibrosseous dysplasia. (Figure 4). Patient was worked up for surgical correction for right fibrous dysplasia. Procedure done was right maxilla reconturing. Patient was under AAP, IV line was started and general anaesthesia was given. Incision done was right sublabial incision (Figure 5). Patient postoperative period insignificant. Patient is under regular follow-up.

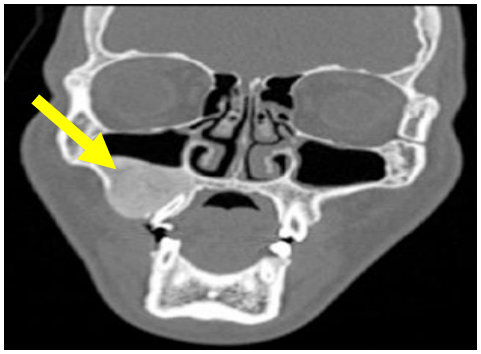

Figure 4: CT PNS showing right maxilla bony thickening.

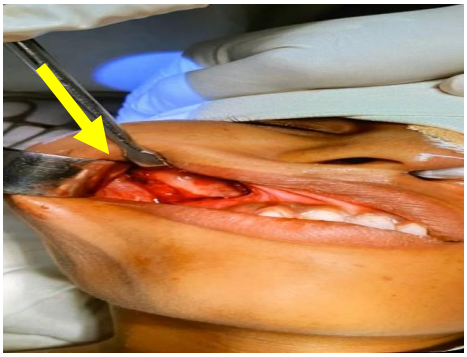

Figure 5: Intra operative picture of right maxilla reconturing.

DISCUSSION

Head and neck region constitute about 25% cases of fibrous dysplasia. Maxilla is the most commonly involved site. Fibrous dysplasia is of 3 types: monostotic (involve one bone), polyostotic (involve multiple bone), McCune-Albright syndrome, presents as a combination of polyostotic FD, skin hyperpigmentation and endocrine dysfunction.

Histology showing fibrous dysplasia with the characteristics thin, irregular (Chinese character like) bony trabeculae and fibrotic marrow space (Figure 6). Treatment plan depends on the extent of involvement, functional disability, danger to function, neurologic symptoms and asthetic consideration. Differentiation should be made between monostotic and polyostotic form of the lesion. The treatment ranges from observation for minor lesions to radical resection. The type of surgery

performed varies from shaving and contouring of the bone to radical surgery. For large cosmetic deformities recontouring and repositioning of bone may be required. Biopsy was done for confirmation and follow up. Radiation therapy is contraindicated because of reported incidence of malignant transformation. Orbital involvement with loss of vision is one of the most severe but relatively uncommon complication of fibrous dysplasia. Fibrous dysplasia is a bone disorder of unknown origin characterized by slow progressive replacement of medullary bone by abnormal proliferative isomorphic fibrous tissue which appears radiolucent on radiographs with classic ground glass appearance.[1] Fibrous dysplasia has 4 different disease patterns: monostotic (70%), polyostotic (30%), craniofacial form and cherubism (rare) (Figure 7).[3,4] The range of skeletal involvement varies from an asymptomatic monostotic lesion to polyostotic involvement resulting in progressive functional deficit and aesthetic problems. The clinical severity depends on time when the mutation of GNAS-1 occurs. In polyostotic form, it is seen if mutation occurs during 6th week of intrauterine life. Multiple bones may get involved. This form commonly involves the skull and facial bones, pelvis, spine and shoulder girdle. Initial symptoms are bone pain and spontaneous fracture of the involved bone. Femur shows shephered's crook deformity.[4] Polyostotic form is again sub-classified into Jaffe's type and Albright syndrome. Both type consists of variable bony involvement with café-au-lait spot. Albright syndrome has additional feature of endocrine disturbances of varying type.[4,5] Polyostotic fibrous dysplasia with soft tissue called Mazabraud syndrome. In monostotic form, if mutation occurs during postnatal life the progeny of that mutated cells are essentially confined to one site resulting in fibrous dysplasia affecting a single bone. About 70% cases are of monostotic form and they involve mainly ribs, femur, tibia and craniofacial bones.[1,2,4] Any bone may be affected, but monostotic forms are never reported as becoming polyostotic.[4] The lesion is asymptomatic and usually discovered incidentally. It causes enlargement and distortion of bone. In craniofacial form, 50-100% of patient with polyostotic disease and 10% patient with monostotic disease have craniofacial involvement.[1,5] Maxilla is more commonly involved than mandible.[1] When maxilla is affected it may involve zygomatic and sphenoid bone. Involvement of frontal, sphenoid, naso-ethmoid and maxillary bone may lead to nasal obstruction, sinus obstruction and sinusitis.[1] Hypertelorism, cranial asymmetry, facial deformity, visual impairment, exophthalmos and blindness may occur due to involvement of orbital and parietal bone.[4] Malignant changes with fibrous dysplasia include osteosarcoma, fibrosarcoma, chondrosarcoma, malignant fibrous histiocytoma and Ewings sarcoma.[1] Association of amelobastoma, cystic degeneration, angiosarcoma, and frontal sinus mucocele have also reported.[1,2] Treatment is primarily surgery. When the only tooth bearing area is involved conservative treatment is bone shaving. Use of calcitonin and pamindronate is also reported for its treatment. Biopsy can be taken to rule out the lesion. Fibrous dysplasia usually get stabilized after puberty.[1]

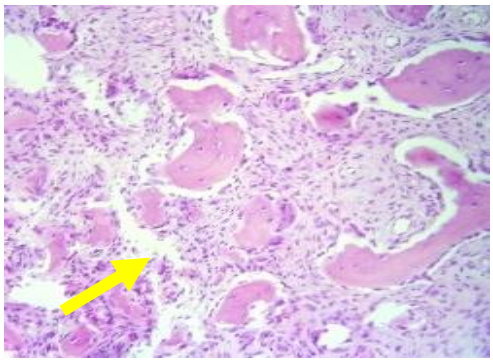

Figure 6: Histology of fibrous dysplasia.

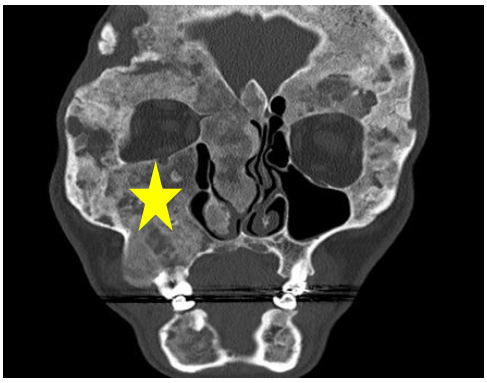

Figure 7: Craniofacial fibrous dysplasia with malignant transformation.

CONCLUSION

Our cases were of monostotic type of fibrous dysplasia involving maxilla, managed by surgery in which the fibrous dysplastic bone was removed by osteotomy. On follow-up after 3 months of surgery: facial asymmetry corrected; there is no diplopia; no watering of left eye; and relieved of nasal obstruction. Histopathological examination (HPE) revealed benign fibro-osseous lesion. The patient and family felt that the surgical procedure resulted in significant improvement of her cosmetic appearance. But 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. Fibrous dysplasia may manifests as monostotic or polyostotic form. Diagnosis of polyostotic form is easier due to extra-skeletal involvement. Monostotic form is common in the jaw. Fibrous dysplasia is a tumor like developmental disorder with minimal chances of malignancies. Aesthetic correction is done by surgeries.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Lumbroso S, Paris F, Sultan C. Activating Gsalpha mutations: analysis of 113 patients with signs of McCune-Albright syndrome-a European Collaborative Study. J Clin Endocrinol Metab. 2004;89(5):2107-13.
2. Watkinson J, Gilbert RW. Stell and Maran's Textbook Of Head and Neck Surgery And Oncology. 5th edition. 2012.
3. Bailey BJ, Johnson JT. Head and Neck Surgery. Fourth edition. Lippincott Williams & Wilkins. 2005.
4. Cummings CW, Haughey BH. Otolaryngology Head and Neck Surgery. Fourth edition. 2005.
5. Smith AG, Zavaleta A. Osteoma, ossifying fibroma, and fibrous dysplasia of facial and cranial bones. AMA Arch Pathol. 1952;54(6):507-27.

Cite this article as: Jayakumar V, Jaiswal SA. Fibrous dysplasia of maxillary sinus-our experience. Int J Otorhinolaryngol Head Neck Surg 2022;8:259-62.