

Craniofacial fibrous dysplasia — A Morbid presentation

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Abstract

Fibrous dysplasia is grouped under fibro-osseous lesions with developmental anomaly of bone forming mesenchyme that manifests as a defect in osteoblastic differentiation and maturation. This paper describes a case of 28yrs old female who presented with swelling on right side of face, nostril and intraoral swelling on right half of hard palate since 1-year. The diagnosis was based on clinico-radiological and histopathological investigations. The appropriate management of patient included surgical modality along with placement of obturator to fill the defect. Follow up was done and till date there is no recurrence.

Keywords: Fibrous dysplasia, Monostotic, Polyostotic, Cemento-ossifying fibroma (COF).

Introduction

Fibrous dysplasia is a skeletal developmental anomaly of bone forming mesenchyme that manifests as a defect in osteoblastic differentiation and maturation.^{1,2} It presents with three forms namely monostotic, polyostotic and craniofacial forms. Monostotic form affects one bone whereas polyostotic shows involvement of the multiple skull and facial bones.^{2,3} Gender predilection is equal. Monostotic usually occurs in second to third decade of life, whereas polyostotic variant occurs before 10 years of age.³ The clinical problems are embraced with pathological fractures, serious complications associated with cosmetic and functional problems.^{1,2} Laboratory investigations comprise elevated serum alkaline phosphate levels. Malignant transformation is rare though it may be hastened with radiation therapy.³

Case Report

A 28 years old female patient reported with swelling on right side of face and nostril which was gradually increasing in size since one year (Figure-1). She had difficulty in breathing, with postural changes like work

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Figure-1: Extraoral swelling on right side of face.

and sleep since one week. Watery discharge was seen from right nostril. She also gave history of blurred vision and parasthesia of involved site with associated change in phonation. She had a similar complaint 9 years back, for which she was operated and since one year the swelling had recurred for which she visited our hospital.

On extra oral examination, a solitary diffuse swelling was seen on right side of the face measuring approximately 5×3 cm, extending medially from bridge of the nose and laterally to level of the right lateral canthus of the eye. A linear scar was seen on the medial aspect of the swelling. On palpation, swelling was bony hard and tender, with no local rise in temperature.

Intraoral examination revealed missing 15, Root canal



Figure-2: Intra-oral swelling on right palate.

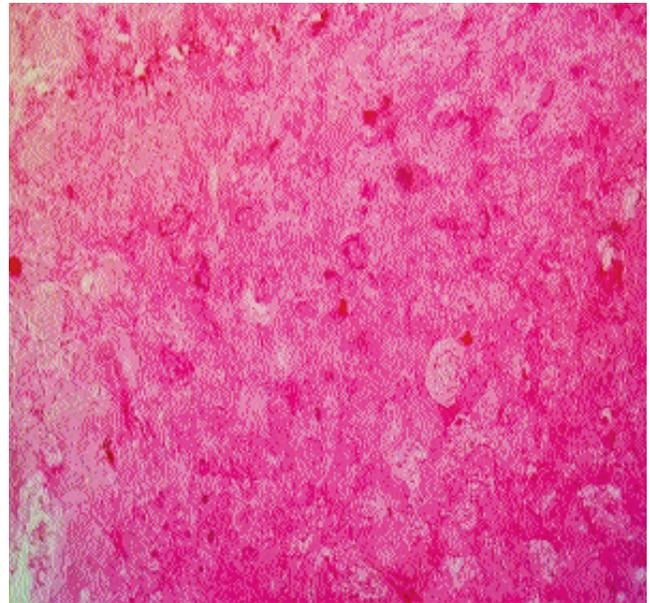


Figure-5: Histopathological picture.

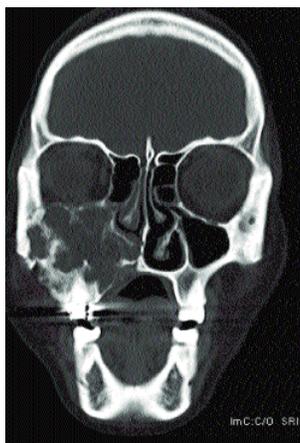


Figure-3: Coronal view of CT showing mixed radiolucent radiopaque multilocular lesion occupying maxillary sinus, inferior orbital rim and lateral wall of nasal floor.

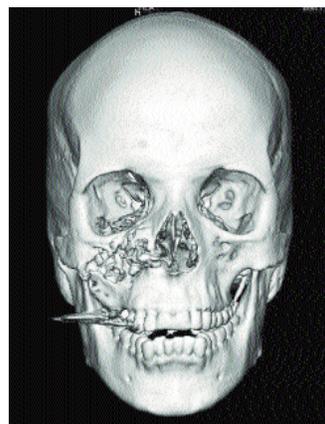


Figure-4: 3D CT depicting lesion extension.



Figure-6: Osteotomy of right maxilla.

treated 13,14,16,17, and a solitary well-defined swelling on right side of the hard palate measuring approximately 2x1cm extending antero-posteriorly from mesial half of 16 to maxillary tuberosity, medio-laterally extending from mid palatine raphe to attached gingiva of 16,17. The mucosa overlying the swelling revealed bluish hue (Figure-2). On palpation swelling was bony hard and

tender. Hence forth pertaining to all the clinical findings provisional diagnosis of fibro-osseous lesion was given.

Para-nasal sinus view (PNS) revealed radiodense area occupying entire right maxillary sinus extending till infra

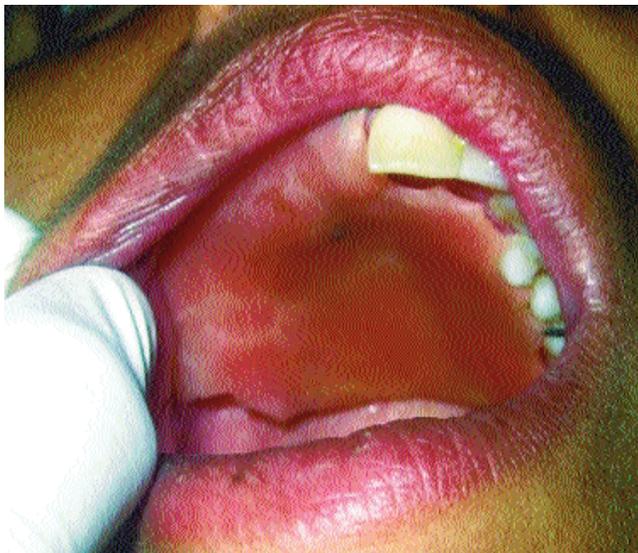


Figure-7: Post-operative photograph with removable denture acting as obturator.

orbital rim and right half of the nasal cavity. The computed tomography of the face and sinuses revealed a mixed radiolucent and radiopaque multilocular expansile lesion with patchy calcifications involving right maxillary alveolus, maxillary sinus bulging into nasal cavity and right orbit causing erosion and involving orbit. 3D CT also depicted the same extension (Figure-3 & 4).

Incisional biopsy was done from right maxilla and histopathology report revealed a delicate connective tissue stroma containing fibroblasts and irregular trabeculae along with delicate collagen fibers and extravasated blood cells. A diagnosis of fibrous dysplasia was given (Figure-5).

The lesion was surgically excised by using a crevicular incision from 11 to distal of 18 and a mucoperiosteal flap was raised. Osteotomy was done through buccal cortex till the base of pyriform fossa and mid-palatal split was given with right hemi-maxillectomy. After surgery the oro-antral communication was closed with surgical obturator (Figure-6&7). The specimen was submitted for histopathological examination and it was confirmed as fibrous dysplasia.

The follow up was done and till date there was no recurrence.

Discussion

Fibrous dysplasia is a developmental tumour like condition that is characterized by replacement of normal bone by excessive proliferation of cellular fibrous connective tissue intermixed with irregular bony

trabeculae.¹ The exact etiology of fibrous dysplasia is uncertain although it may be caused due to non-inherited condition by mutation in GNAS1 gene (guanine nucleotide binding protein) or abnormalities in AMPc which may result in increased proliferation of melanocytes causing cafeu-lait spots and hyperfunction of effected endocrine organs.²

Males and females are equally affected, although the mean age of occurrence was 27 years in 69 patients as reported by Zimmerman.⁴ In the present case patient was female and 28 years old.

Traditionally fibrous dysplasia may affect only one bone or multiple bones associated with cutaneous and endocrine abnormalities. It may be monostotic, polystotic and craniofacial,^{1,2} where monostotic accounts 70-80% and polystotic 20-30%. Usually mandibular lesions are truly monostotic, where maxillary lesions often involves adjacent bones like zygoma, sphenoid and occipital, hence forth they are not strictly monostotic but they fall into designation of craniofacial forms.¹ The other subtypes of polystotic fibrous dysplasia are café-u-lait pigmented skin lesions with endocrinopathies which is called Jaffe Lichtenstein syndrome⁵ and McCune Albright syndrome.^{2,4} Accordingly our case was craniofacial type with no associated endocrinopathy.

The site distribution for fibrous dysplasia focuses more on the maxilla than mandible, When a survey of fibroosseous lesions was done in 52 patients by Abdulai et al out of 16 patients 12 cases had fibrous dysplasia, in maxilla and 4 in mandible.⁶ In the present case maxilla was involved.

The disease is usually asymptomatic however it may be associated with pathological fractures, obstruction of para-nasal sinuses and loss of visual accuracy due to compression of optic nerve.⁷ In our case, the patient had facial asymmetry, blurred vision of right eye and watery discharge due to obstruction of sinus with associated symptoms like anosmia and headache.

The radiographic appearance of fibrous dysplasia is extremely variable and unique. It includes three basic patterns namely:^{2,3}

Type-1: The lesion is generally a rather small uni-ocular radiolucency or large multi-ocular radiolucency, both with a rather well circumscribed border containing a network of fine trabeculae.

Type-2: The lesion is more opaque and typically mottled.

Type-3: It is quite opaque with delicate trabeculae giving ground glass appearance or peau-d orange.

Accordingly our case was surmised into type 1. The differential diagnosis of fibrous dysplasia precludes malignant sarcoma, metastatic osteoblastic lesions and benign lesions like Ossifying fibroma, Paget's disease, aneurysmal bone cyst, ameloblastoma, osteochondroma and hyperparathyroidism.

Histopathology of fibrous dysplasia shows high disparity. It may constitute essentially fibrous component with proliferating fibroblasts. Irregular trabeculae of bone may be scattered or some of them may show a Chinese character pattern.²

The predilection between fibrous lesions like ossifying fibroma, cementossifying fibroma and fibrous dysplasia is controversial, clinically it is impractical to arrive at a correct diagnosis. Generally the boundaries of COF (Cemento Ossifying fibroma) are better defined as, they have soft tissue capsule and cortex, but fibrous dysplasia blends with the surrounding bone.⁸

According to literature when 17 cases of fibrous dysplasia were evaluated in Jamaica by Ogunsalu, 2 cases had recurrence at age of 34 years.⁹ In our case as the patient had similar complaint nine years back, pertaining to her history it implicated as recurrence. However necessary investigations were done to arrive at an accurate diagnosis.

Treatment of fibrous dysplasia is always a dispute. However isolated clinical treatment is surgery though it is disagreed in literature due to functional and cosmetic deficits. The other treatment modalities include bisphosphonates as reported other authors^{10,11} following benefits like reduction in pain and inflammatory symptoms with increased osseous density and recalcification of osteolytic lesion. However the present case was treated with surgery and obturator was placed

to close the Oro-antral communication.

Conclusion

Fibro-osseous lesions are diverse group of lesions which include fibrous dysplasia a benign tumour like condition. It is important for the dentist as it causes cosmetic and functional problems. Many treatment modalities have been recommended though still unsatisfactory, due to recurrence. Henceforth more advanced conservative treatment modalities are yet to emerge. Awareness and scrupulous knowledge of dentist is essential in such cases for prompt diagnosis and immaculate treatment, all of which will enhance the probability of existence.

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