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Case Report

Craniofacial fibrous dysplasia of the maxilla: A case report

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ABSTRACT

Fibrous dysplasia (FD) is an asymptomatic local alteration of bone in which the typical architecture is replaced by fibrous tissues and nonfunctional trabeculae like osseous structures. The lesion most frequently affects the craniofacial skeleton. The maxilla is affected twice compared to the mandible and occurs more commonly in the posterior area. An unusual fibrous dysplasia involving a female's left craniofacial region is reported. The clinical features, radiological findings and treatment have been discussed. Bisphosphonate therapy may help improve function for treatment; surgery is indicated for biopsy for confirmed diagnosis.

1. Introduction

Fibrous dysplasia is a pathologic condition of bone of unknown aetiology. Lichtenstein first coined the term in 1938[1] and in 1942, he and Jaffe separated it from other fibro-osseous lesions.[2] There are two types of fibrous dysplasia. Monostotic and polyostotic. Polyostotic fibrous dysplasia involves a more significant part of the skeleton and skeletal complications. The aetiology is linked to an activating mutation in the alpha subunit of stimulatory G protein located at 20q13.2-13.3 Few patients with polyostotic variants have endocrinopathies (- Albright syndrome) or myxomas (Mazabraud's syndrome). Surgery is indicated for biopsy retrieval and confirmed diagnosis.

2. Case Report

A 13-year-old female patient residing at Malegaon was reported in the OPD of a tertiary Hospital with a chief complaint of swelling in the upper left side of the face. The patient was all right one year back when she noticed swelling on the left side of the face, which was initially small (approximately of a small betel nut) and gradually increased to the present size (approximately of a large nut). No history of pain was current. Onset was insidious, duration: 1 year. The patient did not elicit any relevant medical or dental history. The patient brushed her teeth twice daily with a dentifrice and a toothbrush. Patients do not report any tissue-abusing habits and consume a mixed diet.

2.1. Extra oral examination

Revealed facial asymmetry, non-palpable lymph nodes and bilaterally synchronous TMJ movements. A single unilateral, diffuse swelling is present on the left middle third of the face. The size was approximately 4cmx5cm with an irregular to roughly spherical shape. It extended superioinferiorly from the left infraorbital margin to the line joining the left corner of the mouth to the left tragus. Mediolaterally from the left ala of the Nose to 1cm from the left tragus. The left globe of the eye appeared to be slightly

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pushed up.

2.2. Intra-oral examination

Figure 1 showed a single, diffuse swelling around the alveolar process in the left maxillary molar and premolar regions, leading to obliteration of the buccal vestibule. Extending anteroposterior from 25 to 27 areas. Superioinferiorly from left buccal vestibule till cervical margin of maxillary teeth. They were mediolaterally obliterating the buccal vestibule. Swelling felt like a smooth expansion of buccal and palatal cortices, which blend smoothly with surrounding areas. Margins were ill-defined, firm in consistency, non-tender, and non-compressible with no discharge or induration.

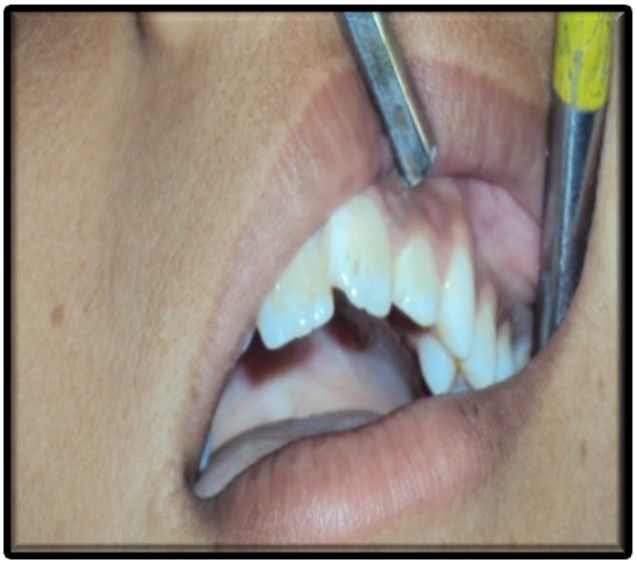

Figure 1: Intra oral examination

A clinical differential diagnosis of a benign oral cavity tumour was given.

2.3. Investigations

2.3.1. OPG

A diffuse homogenous radio-opaity involving a large part of the left maxillary sinus is seen.(Figure 2) It is extending supero-inferiorly from the alveolar crest into the left maxillary sinus. Antero-posteriorly from distal of canine to third molar region. The floor of the maxillary sinus cannot be appreciated.

2.3.2. Occlusal

To check the mediolateral extent of the lesionMaxillary cross-section occlusal radiograph was obtained, which showed a diffuse homogenous radio-opacity extending on the left side's medial and lateral aspects of the alveolar ridge. Medially, it is seen to develop within 5mm of midline

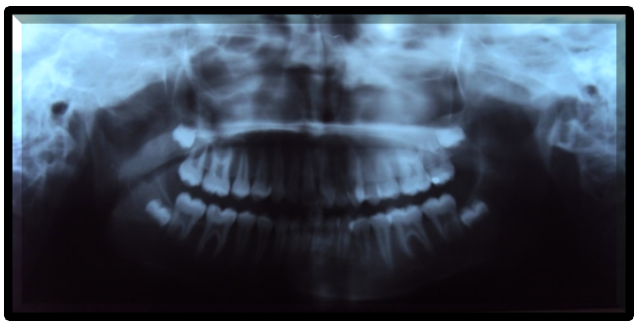

Figure 2: Oral pantograph

laterally. It has caused expansion of the buccal cortex and extends beyond the film.

2.3.3. IOPA

(Figure 3) It showed increased fuzzy trabeculae, giving it a ground glass appearance around the left premolar molar region. Lamina dura was not appreciable. The floor of the maxillary sinus was also not noticeable.

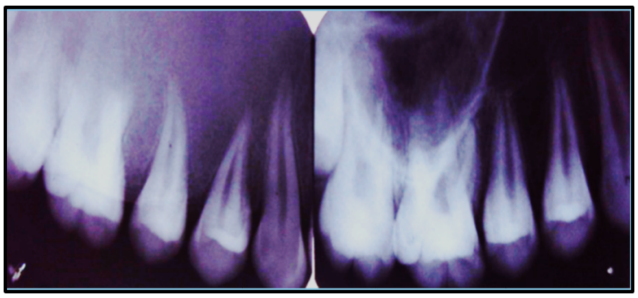

Figure 3: IOPA

A (Figure 4) sagittal view X-ray was taken. It illustrated thinning in the cortical boundaries of the lesion without loss of continuity. (yellow arrows). However, the lamina dura of the left posterior permanent teeth was lost (red arrows).

2.3.4. Axial CT

A hyperdense mass originated from the left maxillary alveolar process. (around roots of premolars and maxillary sinus and filling the anterior $3/4^{th}$ of the left maxillary sinus. The 3D reconstruction revealed It showed uneven enlargement of the left maxillary sinus. Radiographic differential diagnosis included Mono-ostotic fibrous dysplasia, Ossifying fibroma, hyperparathyroidism, ground glass appearance, Paget's disease of bone, and Chronic diffuse sclerosing osteomyelitis.

Fibrous dysplasia was given as a provisional diagnosis based on its distinct clinical and radiologic features. Differential diagnosis includes:

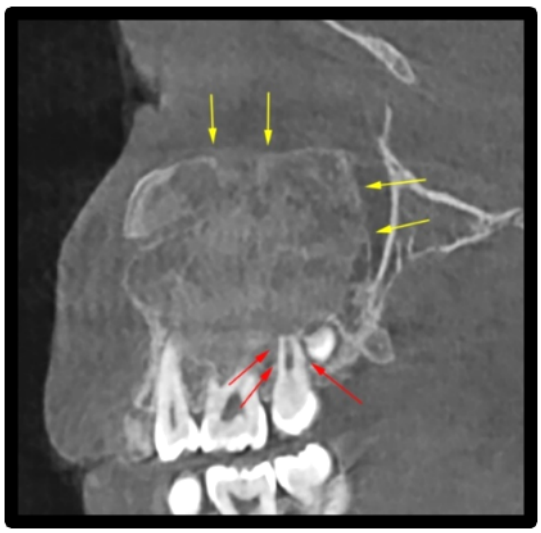

Figure 4: Saggital view x-ray

2.3.5. *Ossifying fibroma*

Ossifying fibroma has a definitive capsule and can be seen most of the time. It exhibits well-demarcated margins, whereas fibrous dysplasia does not. Ossifying fibroma grows centrifugally, producing a ball-like circular lesion that enlarges equally in all directions, expanding the buccal and lingual/palatal cortical plates and, most notably, the inferior cortex of the mandible. The developed inferior cortex is precisely parallel to the margin of the tumour mass above. Fibrous dysplasia causes a linear expansion of the cortex; thus, the expanded cortex cannot be in an exact parallel relationship to the tumour mass. hence, to differentiate between central osteosarcoma and fibrous dysplasia, the distinction may be made based on the lack of a reactive shell, permeative borders, denser mineralisation, and more aggressive changes over time in low-grade central osteosarcoma.

2.3.6. *Hyperparathyroidism*

The lesions show a ground glass appearance on radiology with elevated serum calcium and reduced serum phosphorous. We had done Blood and biochemical investigations for both cases, which showed alkaline phosphates, Serum Calcium and serum Phosphorous, which were within normal range.

2.3.7. *Pagets disease of bone*

2.3.8. *Chronic diffuse sclerosing osteomyelitis*

Histological examination: (Figure 5) An incisional lesion biopsy received six soft tissue bits and one tricky tissue bit. The most significant quiet tissue bit was 0.5x0.6 cm in dimension, brownish-black in colour and friable in consistency. All soft tissue bits were processed together. The hard tissue bit was 0.5x0.5 cm in size, yellowish in colour, and hard in consistency. It was put in 5% HNO3 for decalcification and then processed.

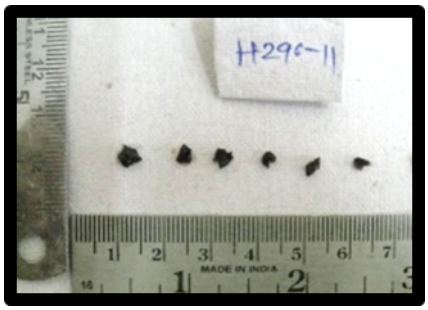

Figure 5: Gross microscopic examination

Microscopic examination: (Figure 6a,b) The fibrous tissue has a monotonous cellularity, and the delicate pattern of bony trabeculae is repeated throughout the entire lesion. Rather than being a haphazard mixture of woven bone lamellar bone and spheroid particles.

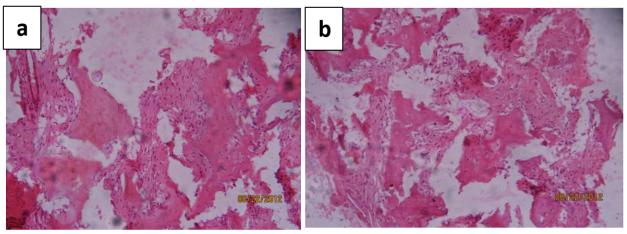

Figure 6: a,b): Microscopic examination

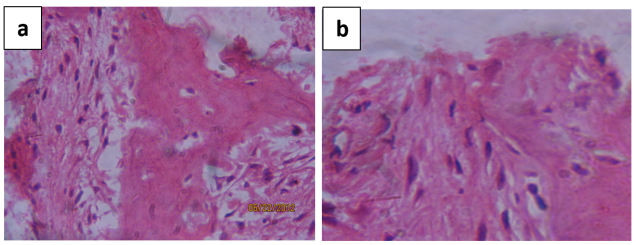

Figure 7: a, b): Photomicrograph of a specimen from a fibrous dysplasia lesion, showing the characteristic pattern of bizarrely contoured, disconnected, dysplastic trabeculae enmeshed in primitive mesenchymal cells (Hematoxylin and Eosinx 100)

Fine branching, irregularly shaped, curvilinear trabeculae of woven bone (immature), not connected with little evidence of osteoblasts rimming, which have been linked to Chinese script writing, were seen.

2.4. *Final diagnosis*

Combining clinical, radiographic, and histopathologic interpretations, a definitive diagnosis of mono-ostotoc fibrous dysplasia of the maxilla was given.

3. Discussion

Fibrous dysplasia is a regional alteration of bone where standard architecture is replaced by fibrous tissue and non-functional trabeculae-like structures. The lesion may be polyostotic or monoostotic, with or without endocrinal disturbances. Authorities have suggested five different types of fibrous dysplasia: monoostotic, polyostotic, polyostotic with pigmentation (Jaffe), polyostotic with endocrine disorders (Mc-cune- Albright syndrome) and Craniofacial fibrous dysplasia.

In 1891, Von Recklinghausen reported three groups of bone disease, one of which most likely contained cases of what is today known as POFD. Subsequently, several issues were described under the term osteitis fibrosa. Generalist Bright et al. reviewed cases of FD with disseminated bone lesions, skin pigmentation, and precocious puberty and gave the term Albright's syndrome. In 1938, Lichtenstein described eight osteoporotic bone lesions without extraskeletal components recognized as polyostotic FD. In 1942, Lichtenstein and Jaffe recognized that a monostotic form of the disease also occurs. Fibrous Dysplasia in the Oral Cavity.[2]

Maxillary involvement: FD commonly affects the maxilla, causing facial asymmetry, dental malocclusion, and potential obliteration of the maxillary sinus.[3,4]

Dental anomalies: FD can lead to dental crowding, spacing, and root displacement. Teeth may show splaying around FD lesions.[4]

Radiographic appearance: FD in the jaws typically presents as a homogeneous, granular radio-opacity with a "ground-glass" appearance.[3]

Histopathology: Microscopic examination reveals fibrous connective tissue with irregular bone trabeculae, often described as having a "Chinese letter" configuration.[5]

Fibrous Dysplasia in the Body: Skeletal involvement: FD can affect any bone but commonly involves long bones, ribs, and craniofacial bones.[6]

Clinical presentation: Symptoms may include bone pain, deformity, fractures, and uneven growth.[7]

Types: FD can be monostotic (single bone) or polyostotic (multiple bones).[8]

Associated conditions: Some cases are associated with McCune-Albright syndrome, which includes endocrine abnormalities and skin pigmentation

Neurological symptoms: FD in the skull can potentially compress nerves, leading to vision or hearing problems in rare cases.

Radiographic features: FD lesions typically appear as expansile, radiolucent, or mixed-density lesions with a "ground-glass" appearance on imaging studies.

Aetiology: Results from a post-zygotic mutation in the GNAS 1gene (guanine nucleotide-binding protein, alpha-stimulating activity polypeptide). Clinical severity depends upon the time the gene is transformed during fetal or postnatal life.[9] If it happens in undifferentiated stem cells, Jaffe-Lichtenstein or McCune Albright syndrome. If it occurs in skeletal progenitor cells, Polyostotic FD occurs, and in post-natal life, Mono ostotic FD occurs. Mandibular lesions are truly monostotic. Maxillary lesions often involve adjacent bones (zygoma, sphenoid, occiput), better called craniofacial FD. The teeth involved remain firm but displaced by bony mass. Painless swelling in the affected area, slow growth. Node is non-tender on palpation. There is rarely a disturbance of function; teeth may be replaced, occlusion interferes with, or eruptions fail.

Some earlier literature on this disease suggested that it represents a permanent maturation arrest in the woven bone stage. It proposed that lesions demonstrating lamellar bone transformation should not be diagnosed as FD. However, it is generally well accepted now that lesions of FD of the jaws, especially the craniofacial type, will mature over time, and lesional tissue may show lamellar bone. Lamellar bone and osteoblastic rimming do not exclude the diagnosis of FD, as would be the case for lesions outside the maxillofacial bone.[10] The lesion's margins and overall pattern may appear in an ideal biopsy, allowing some confidence in a histological diagnosis.

Treatment is pursued only when lesions are cosmetically unacceptable or interfere with sight, breathing, chewing, or speech. Following growth, FD slows after the onset of puberty. So, for smaller lesions, no treatment is needed; for larger disfiguring ones, cosmetic and functional correction and osseous recontouring are required. About 25-50% of patients may experience regrowth after surgery. Some clinicians believe cosmetic surgical osteoplasty will accelerate it from indolent to aggressive.

4. Conclusion

If radiographic findings are not characteristic, a biopsy is required to confirm the diagnosis of fibrous dysplasia. Bisphosphonates have been shown to improve skeletal strength and offer pain relief. Occasionally, surgical intervention is necessary to correct a bony deformity or see a pathologic fracture.

5. Source of Funding

None.

6. Conflict of Interest

None.

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