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Surgical and biological considerations of extensive Maxillary fibrous dysplasia

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ABSTRACT

Objective:Fibrous dysplasia (FD) is a skeletal disorder including the maxilla and the mandible; nevertheless, its effects on dental tissues and the implications for dental care remain unclear. The aim of this study was to characterize the dental features associated with FD and the reaction of affected bones to routine dental therapy additionally the relation with endocrine disturbances ***Study design:***Thirty-two patients with FD underwent endocrine testing as part of the diagnosis of FD/McCune-Albright syndrome, maxillofacial evaluation and surgical shaving was performed. Any dental anomalies were recorded, and the associations between endocrinopathies and dental anomalies were analyzed statistically by means of the paired *t* test. ***Results:***Eighty-four percent had FD in the maxilla and/or mandible; endocrine dysfunction; and/or renal phosphate wasting. The Malocclusion (81%) and other prevalent dental anomalies (41%) included tooth rotation, oligodontia, and taurodontism. The expansion of the maxilla or mandible by FD did not distort the dental arch curvature, and routine dental therapies such as extractions, restorations, and orthodontic treatment did not exacerbate FD lesions. ***Conclusion:*** Maxillomandibular FD was associated with higher rates of malocclusion than were present in healthy patients. Furthermore, patients with FD did not require special dental management and were able to undergo routine dental care without an exacerbation of FD lesions,

Key words: Surgical ,biological, maxillary and fibrous dysplasia

Introduction

Fibrous dysplasia (FD) of bone is characterized by the replacement of normal bone and marrow by fibrous tissue, within which irregular trabeculae of woven bone are haphazardly distributed, FD may affect a single bone (called *monostotic MFD*) or multiple bones (called p*olyostotic FD* [PFD] and may be associated with endocrinopathy (Shenker *et al.,* 2013)

FD can occur as part of McCune-Albright syndrome (MAS), a rare multisystem disease that was first described as the triad of PFD, skin hyperpigmentation (*cafe´-au-lait),* and precocious puberty (Moran *et al.,* 2012). MAS is also associated with other endocrine disorders of the pituitary, thyroid, and adrenal glands, (Smith, 2011; Ladenson, 1992; AKoutras,1997and Grant,1999) .

Hypophosphatemia and renal phosphate wasting are commonly observed in patients with FD/MAS, (Kushner *et al.,* 2001). Two separate literature reviews of the years 1926 through 1995 revealed only 158 published cases.(MLevine, 1999 and Schwindinger and Levine,1996) FD in association with *cafe´-au-lait* skin hyperpigmentation but no endocrinopathyis known as Jaffe-Lichtenstein syndrome.(Damm *et al.,* 2002)

FD is considered a disease of cells of the mesenchymal stem cell/ osteoblastic lineage in which excess cyclic adenosine mono phosphate impairs the ability of the stem ce1lto differentiate into a mature functioning osteoblastic. (Gejman *et al.,* 1991) However, it is not known how—or even whether excess cyclic adenosine monophosphate affects the developing tooth, either directly or indirectly.

Craniofacial bones, including the maxilla and the mandible, are commonly affected by FD, often causing disfigurement. However, despite the frequency of craniofacial involvement, the dental features of FD have been poorly characterized, mainly in isolated case reports with sparse information about the effects of FD on dental tissues. (DeSanctis *et al.,* 1999; Shenker *et al.,* 2011 and Olander and Hammarstrom, 1985) 12,13,14) The development, eruption, and shedding of primary teeth followed by the development and eruption of permanent teeth are sequential events that may be altered by metabolic dysfunction with in dental tissues or the presence of bony pathosis within the jaws (Gejman *et al.,* 1991).

It remains unclear whether the presence of FD in the jaws has any effect on tooth development and function. The typical appearance of patients with FD of the Maxillomandibular bones is that of facial asymmetry, often associated with palatal asymmetry. FD is often associated with characteristic *cafe´-au-lait* skin *A* pigmentation and rarely with *cafe´-au-lait* pigmentation of the oral mucosa . Panoramic radiographic imaging of the jaws and computed tomography have revealed aground-glass trabeculation that may progress to mixed radiolucent/radiopaque lesions and thinning of the cortical margin. (Shenker *et al.,* 2011)

Craniofacial FD may present with a variety of manifestations depending on the area of bony involvement. Lesions may involve the maxilla, mandible, zygoma, calvarium, sphenoid, temporal or orbital bones.

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Symptoms develop secondary to slowly progressive bony expansion. Craniofacial bone involvement may produce asymmetry and cosmetic deformity. Massive cranial bone involvement has resulted in lion-like faces termed "leontiasis ossea," or maxillary involvement termed "cherubism." FD of the maxilla may also result in nasal obstruction, epiphora, and malocclusion. Visual disturbance can occur secondary to proptosis, extraocular muscle dysfunction, optic nerve or chiasm compression when FD affects the bony orbit. (DeSanctis *et al.*, 1997)

Fibrous dysplasia has often been confused with osseous lesions of hyperparathyroidism (Damm *et al.*, 2002) indicating similar histopathologic features (DeSanctis *et al.*, 1997). parathyroid hormone (PTH) is a peptide hormone produced in the parathyroid gland which acts directly on bone to increase bone resorption and mobilize calcium to control the concentration of calcium in the extracellular fluid; this function is effected through activation of a mechanism that transfers calcium from bone and from glomerular filtrate to the extracellular fluid compartment (AKoutras,1997). Parathyroid hormone-related peptide (PTHrP) was originally identified in tumors associated with hormonal hypocalcaemia of malignancy (Grant, 1999).

Calcitonin (CT) could be classically considered to be a physiologic antagonist of PTH and PTHrP; it lowers the plasma calcium concentration by inhibiting bone resorption, with a stimulation of the renal calcium excretion (Grant, 1999).

Although the ideal technique for the treatment of fibrous dysplasia of the maxilla is still an issue of controversy, The current work attempted to study the surgical intervention ,dental feature and endocrine disturbances as an approach the treatment of fibrous dysplasia of the maxilla, as well as to find out suitability of this approach for treatment of this cases . The present study, have hypotheses that the surgical intervention could be of value as a suitable technique for treatment of fibrous dysplasia of the maxilla

Aim of the work:

The aim of this study was to characterize the dental features associated with FD and the reaction of affected bones to routine dental therapy additionally relation with endocrine disturbances

Material And Methods

Thirty-six patients diagnosed with FD/MAS gave written informed consent and were subsequently enrolled in this study. FD was diagnosed by a combination of the results from clinical history, physical examination, radiographic analyses, and lesion bone biopsy. Testing of the pituitary, thyroid, parathyroid, adrenal, gonadal, adrenal functions was performed.

The maxillofacial evaluation included extra oral and intraoral examinations, panoramic ,CT (as showing in fig 1), intraoral radiographs, and the preparation of maxilla mandible study casts to assess the therapy-related bony changes such as the growth/enlargement of FD lesions or the development of malocclusion in the maxilla or mandible to assess the therapy-related bony changes such as the growth/enlargement of FD lesions or the development of malocclusion in the maxilla or mandible.

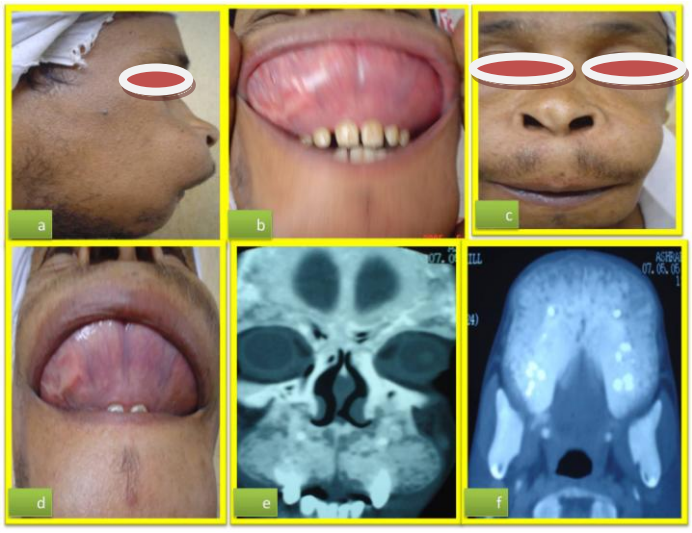

Fig. 1: lateral ,intraoral and frontal photograph preoperatively showing the clinical extent of the lesion with obliteration of nasolabial fold ,(a, b ,c, and d).Coronal and axial section of Computed tomography of the maxillary fibrous dysplasia prior to the first operation showing ‘‘peau d’orange (orange peel)’’ appearance was noted in the maxilla and mandible also th bilateral maxillary sinus was mostly replaced with the lesion.(e and f)

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The CT scan can also evaluate the optic canal, superior and inferior orbital fissure, internal auditory canal, or skull base foramina for evidence of compression.Technetium-99m radionuclide bone scan will demonstrate increased uptake in other sites of skeletal involvement and can be used to determine when the quiescent phase has been reached.

As the priority was to relieve the obstructive symptoms, a conservative bone shaving surgery was performed through intraoral or a modified Weber Ferguson approach as showing in (fig 2)

The patients was regularly followed up once in every 3 months till 1 year and every 6 months since then, and a secondary surgical procedure to further enhance the cosmetic appearance and improve the facial symmetry was refuted by the patient

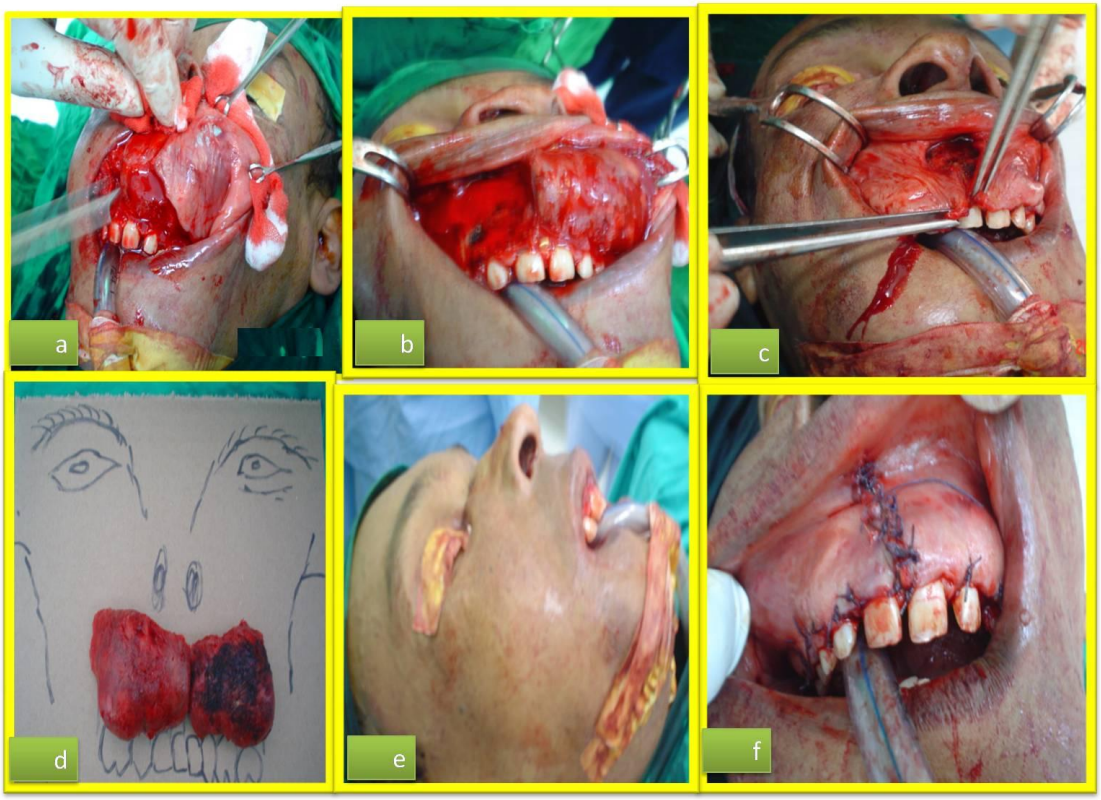

Fig. 2: Intraoperative photograph showing the incision ,shaving the lesion, approximation of the flaps and finally orientation of lesion for pathologist as tags ,(a, b, c, and d).Immediate postoperative photograph showing closure of the mucoperiosteal flap (e and f)

Conservative surgical treatment as in fig 2,is regarded by many as the treatment of choice. This involve decompression of cranial nerves at their respective foramina, contouring of bone to restore symmetry of facial skeleton, canaloplasty to relieve external audit auditory canal stenosis and minimal resection to relieve anosmia, and nasal and sinus ostia obstruction. When the extent of the lesion is such that total excision would result in greater deformity and functional loss than the disease itself.

Endocrinology data and therapeutic consequence of calcitonin administration in association with surgical interventions. Surgical intervention performed after the local bone calcification by a calcitonin treatment, because of alleviation of vigorous hemorrhage by the bone remodeling. .Surgical therapy considered based on the residual cosmetic and functional problems. Some patients with minimal deformity may not require surgical intervention.

In addition, if surgical therapy is postponed until after complications arise (visual or hearing impairment, cranial nerve dysfunction), the deficit maybe permanent and irreversible or make the surgery more technically difficult When optic canal compression occurs and visual acuity is compromised, the risk of damage to the optic nerve during decompression becomes more likely and visual acuity may not return to normal

Statistical analysis:

We analyzed associations between degree of endocrine disturbance and criteria of clinical examination using the paired *t* test, the results of which were calculated with the SAS statistical software package

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Results:

After undergoing full radiographic testing and technetium (99T) full-body bone scanning, 4 of the36 original patients evaluated (11%) were excluded because they did not have an FD lesion in any craniofacial bone.

The 32 remaining patients with craniofacial FD are the focus of this report. There were 11 males and 21 females, ranging from 4 to 50 years of age (mean, 21.1 years old). Twenty-three patients had PFD, and 19 of these 23 were diagnosed with MAS on the basis of PFD in association with *cafe´-au-lait* skin pigmentation and endocrinopathy or renal phosphate wasting, or both; 14 of these had a combination of 2 or more endocrinopathy and renal phosphate wasting .

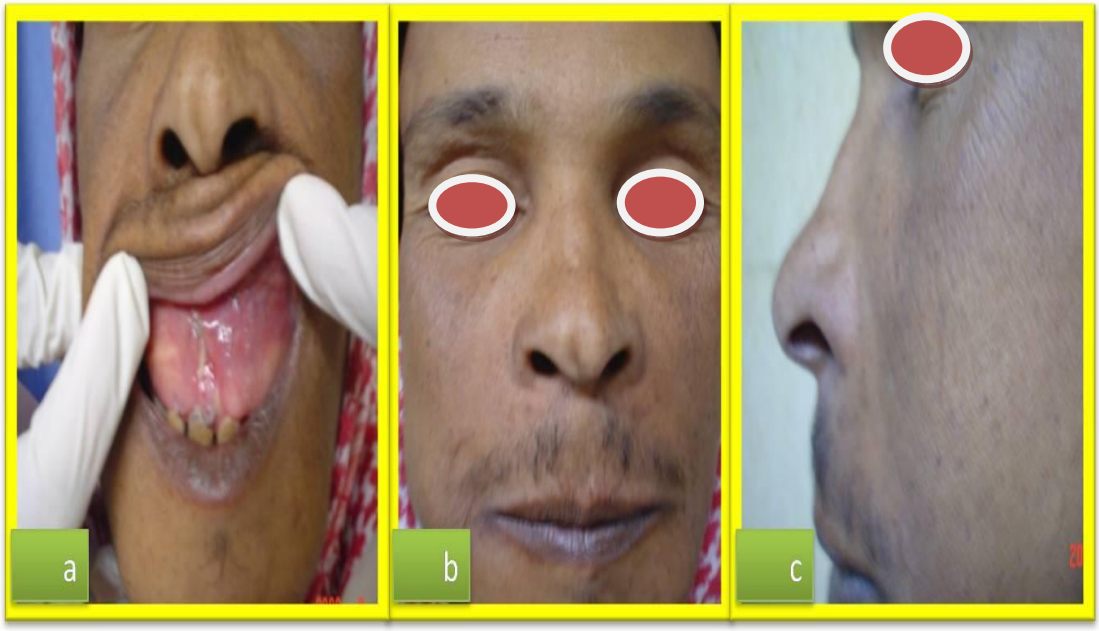

Fig. 3: 1 month postoperatively showing no swelling, a. 6 month postoperatively showing improving the nasolabial fold ,b, 12 month postoperatively showing improving the nasolabial angle

Precocious puberty, followed by hyperthyroidism and renal phosphate wasting ,were the most common abnormalities. FD was present in the maxilla, mandible, or both in 27 patients (84%) and was frequently associated with malocclusion and palatal, maxillary, or mandibles asymmetry, ranging from mild to severe ,severely affected mandibles; nevertheless, the vast majority of these patients did not report any paresthesia or dysesthesia.

Two patients reported unilateral jaw paresthesia. However, both patients had a history of the surgical removal of FD from the associated nerve sites, so the paresthesia was judged to be a result of postoperative changes Another patient reported temporomandibular joint pain and clicking, but there were no degenerative or bony changes in the condylar head or glenoid fossa and the temporomandibular range of motion was within normal limits. The cosmetic of patents was improved as showing in (Fig 3).

Interestingly, there were no changes in maxillary or mandible, FD after dental restorations (20 cases), tooth extractions (6 cases, the removal of odontoma (1 case), the removal of a maxillary cyst (1 case) Furthermore, there were no observed incidents or reports of prolonged bleeding, pain, or swelling; occlusal changes; or infection after these procedures.

The dental anomalies included rotation, oligodontia, displacement, enamel hypoplasia and hypomineralization, taurodontism, retained deciduous teeth, and attrition .Malocclusion was the most common abnormality, but it did not correlate with the concomitant presence of a tooth anomaly and an FD lesion, observed in the jaws of 9 patients (28%) as showing in table 1

There was no statistically significant correlation between any specific endocrine dysfunction or renal phosphate wasting and the DFT scores or tooth anomaly; nor was there a statistically significant relationship between renal phosphate wasting and enamel hypoplasia, hypomineralization, or between such wasting and attrition.

In addition, the 15 impacted third molars that were extracted were normal radio graphically; the first 4 were decalcified and, when examined histological, were found to be normal. The remaining 11 were not examined histological. However, the prevalence of oligodontia and retained deciduous teeth suggest that mutation may

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have prevented the formation of permanent successors .Because of the gross radiopacity and sclerotic nature of gnathic FD, it was difficult to delineate the outline of the mandible canal in panoramic radiographs of severely

Table I: Demographics of clinical findings in patients with fibrous dysplasia:

Variables	*Age groups (years)*		
	5-17	*17-50*	*Totals*
Precocious puberty 11	56 7	58 18	56
Hyperthyroidism 8	40 5	42 13	41
Phosphaturia 9	45 4	33 13	41
GH excess 5	25 1	8 6	19
FD in maxilla or mandible (no.	(%) 17	85 10	83 27
FD in maxilla and mandible	(%) 6	30 4	33 10
Malocclusion	(%) 15	75 11	92 26

Discussion

This work represents the largest group of FD/MAS patients to date in whom the resultant dental characteristics have been described, allowing us to make a number of important observations. First, in patients with craniofacial FD, the maxilla or mandible—or both—is involved 84% of the time.

Clinical clues to the presence of FD in the mandible or maxilla are facial or palatal asymmetry, typical *cafe´- au-lait* spots, and a history of endocrine disorders associated with MAS, especially precocious puberty.

Conversely, if FD is noted in the maxilla or mandible, associated endocrine disease should be suspected and the appropriate referrals should be made.

We were also able to show that FD is frequently associated with dental anomalies, including rotation, oligodontia, displacement, enamel hypoplasia and hypomineralization, taurodontism, and others. The etiology of these anomalies is not known, but it is possible that they are the result of activating mutations in tooth development either directly (enamel hypomineralization and hypoplasia, oligodontia, attrition), or indirectly because of the proximity of abnormal bone (malocclusion, rotation, displacement, retention of deciduous teeth).

Taurodontism, a condition visible radio graphically in ultirooted teeth and characterized by enlargement of the pulp chamber caused by apical displacement of the bifurcation or trifurcation of the roots, has been described in many syndromes, including patients with growth hormone excess,21-26 but never in FD/MAS.

The incidence of taurodontism observed in this cohort was 9% (3/32) (data not shown). An earlier study associated taurodontism with multiple missing teeth and reported a 34.8% prevalence of taurodontism in subjects with oligodontia compared with 7.5% in a control group.27 We did not observe an association between oligodontia and taurodontism in our patients. However, the patients with taurodontism were also diagnosed with 1 or more endocrinopathies, including growth hormone excess, precocious puberty, secondary hyperparathyroidism and hyperthyroidism, or renal phosphate wasting.

It is possible that associated abnormalities may account for this (especially the propensity for renal phosphate wasting to cause enamel hypoplasia or hypomineralization), but there was no statistically significant relationship between any given endocrinopathy and renal phosphate wasting or between a specific anomaly and an increased caries index score. This may be because of the small number of patients that could be classified into each endocrinopathy or renal phosphate wasting subgroup.

Despite the prevalence of malocclusion, 28% of patients had tooth anomaly in the FD bone. The malpositioned teeth were rotated within the socket or inclined in the mesiodistal direction however, no buccolingual displacement or mobility was observed. In essence, the curvilinear pattern of the dental arch was preserved .Of particular importance to the clinician is the lack of complications associated with the routine dental care of patients with FD. We noted no abnormal response or complications in association with dental restorations or extractions.

Conclusions:

Maxillomandibular FD was associated with higher rates of malocclusion than were present in healthy patients. Furthermore, patients with FD did not require special dental management and were able to undergo routine dental care without an exacerbation of FD lesions,

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