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Case Report

Maxillary Tumefaction Revealing a Fibrous Dysplasia: A Case Report

Faten khanfir*[1,3], Abdellatif Chokri*[2,3], Mohamed Tlili[1,3], Mohamed Salah khalfi[1,3], Jamil Selmi[2,3] & Faten Ben Amor[1,3]

[1]Anatomy department, Faculty of Dental Medicine, Monastir, Tunisia
[2]Oral surgical department, Faculty of Dental medicine, Monastir, Tunisia
[3]University Of Monastir, Research Laboratory of Oral Health and Orofacial Rehabilitation LR12ES11, Monastir, 5000, Tunisia

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Abstract: ***Purpose:*** Fibrous dysplasia (FD) is a benign fibro-osseous pathologic condition characterized by the replacement of bone with fibrous tissues. Its diagnosis is based on clinical, radiological and histological findings. ***Objective*:** In this study, we report a case of unilateral FD in the left side of the maxilla, precisely in the palate region, using computed tomography and histological examinations. ***Results***: On examination, expansion of the buccal cortical plate and obliteration of the vestibule were found .The covering and surrounding mucosa was normal in color, showing no clinical manifestation of inflammation or ulceration. Tomography showed a diffuse and heterogeneous thickening of the bony elements, involving the hard palate. As a treatment, we opted for remodeling osteotomy in order to improve the esthetic outcome. ***Conclusion***: knowledge of the morphological changes is important for the diagnosis of bone pathologies. Thus, the dentist must be familiar with the normal morphology of the structures as well as their possible abnormalities.

Keywords: fibrous dysplasia, maxilla, osteotomy, lesion, monostatic, symptoms.

INTRODUCTION:

Fibrous dysplasia (FD) is a benign fibro-osseous pathologic condition characterized by the replacement of bone with fibrous tissues. (Jundt .G, 2005) (Neville *et al.*, 2002).

It is a benign intramedullary fibro-osseous lesion, originally described by Lichtenstein in 1938 and later by Lichtenstein and Jaffe in 1942(Lichtenstein L, 1938).

If FD affects only one bone, it is called monostatic FD, corresponding to 70% to 80% of FD cases. However, multiple bones may also be affected. This form is called polyostatic FD, which corresponds to 20%to 30% of cases (Jundt .G, 2005). This type of FD can be subdivided in three types: craniofacial, in which only the bones of the craniofacial complex are involved, including the mandible and maxilla; Lichtenstein-Jaffe, having similar involvement of multiple bones of the skeleton but it also presents "café-au-lait" spots in the skin; Albright syndrome, which is characterized by the involvement of multiple bones, "café-au-lait" spots in the skin and endocrine impairment, especially during early puberty in girls. In addition to these forms, Jones described a hereditary familial form of localized FD, which is called Cherubism (Jones .WA, 1965).

Thus, our aim through this study was to present a case of FD of the maxilla, at the left side of the palate.

CASE REPORT:

A 46 -year -old female patient presented to the oral surgery department at the dental clinic of Monastir, Tunisia. Her chief complaint was a slowly-growing painless swelling in the upper left jaw posterior teeth region persisting for 4 years. Clinically, the patient presented with facial asymmetry (figure 1). There was no history of trauma, trismus, diminished vision, teeth loosening or epistaxis.

The extraoral examination revealed a well-defined bony hard swelling in the left maxillary region. Obliteration of the nasolabial fold was also noticed.

The intraoral examination showed a swelling that started with the size of a pea and gradually increased to attain the present size, extending from the region of tooth 23 to that of tooth27, with expansion of the buccal cortical plate obliteration of the vestibule.

***Corresponding Author:** Faten Khanfir

The covering and surrounding mucosa was normal in color, without any clinical manifestation of inflammation or ulceration. On palpation, the affected area was painless, with hard consistency and a plane surface.

Based on the clinical examination, the initial diagnosis was fibrous dysplasia of the upper jaw.

Radiological examination: the axial section of CT scan view revealed a heterogeneous, granular radio-opacity of the left maxillary alveolar bone with expansion of the buccal and palatal cortical plate, extending from tooth 23 to tooth 27, and giving a granular or ground-glass aspect.

On The coronal section, we noticed a sinus floor elevation and the presence of active and early lesions, appearing more radiolucent than the mature ones (figure 3).

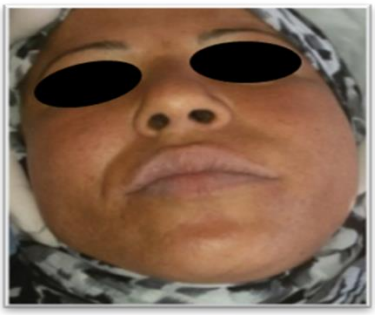


Figure 1

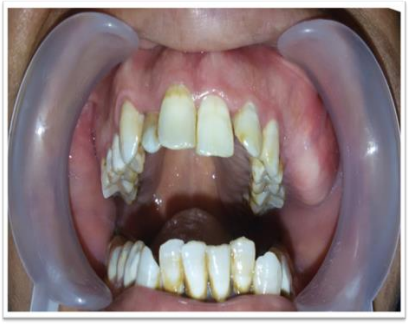


Figure 2

Figure1: facial Photography: Facial asymmetry and obliteration of the left nasolabial fold
Figure2: expansion of the buccal cortical plate and obliteration of the vestibule

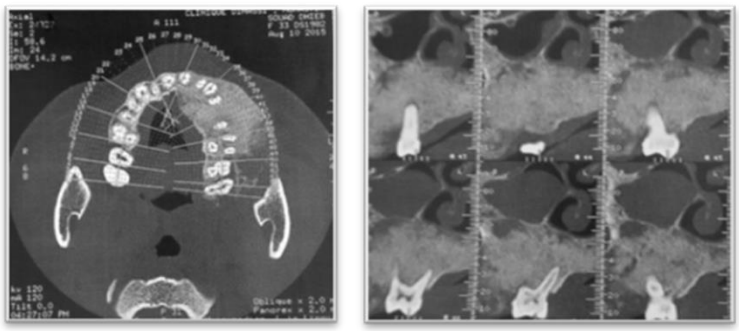
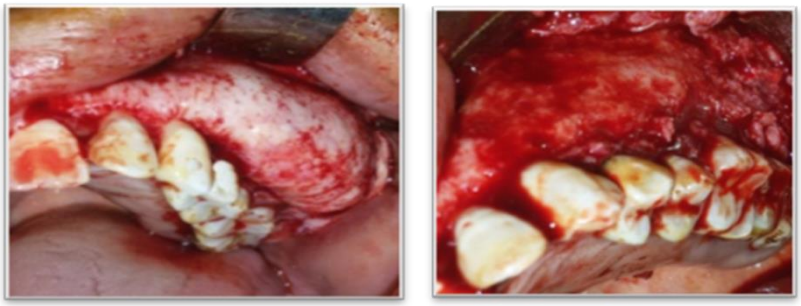


Figure3: (a) (b)

Figure 3: radiological examination a: axial section: heterogeneous, granular radio-opacity of left maxillary alveolar bone;b:coronal section ;b:the presence of active and early lesions which appear more radiolucent than the mature ones

The Surgical Treatment:



(a) (b)

Figure 4: a: Extreme change of the affected bone contour; b: Remodeling osteotomy

The lesion was surgically excised using remodeling osteotomy (figure 4). The histopathology of the excised tissue showed a benign fibro-osseous lesion formed by a hypercellular fibroblastic stroma, consisting of spindle cells arranged in whorls with a storiform pattern and in loose sheets with intervening collagens. Among these, there was an immature woven bone with Chinese letter configuration (figure 5).

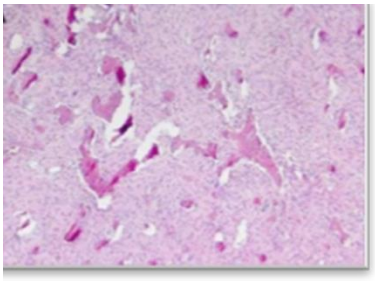


Figure 5

Figure 5: Histopathologic section showing bony trabeculae resembling Chinese letters in the connective tissues

DISCUSSION

Fibrous dysplasia of bone (FD) is a sporadic, uncommon, fibro-osseous skeletal disorder with a broad severity spectrum. Initially, it was identified by Donovan McCune and Fuller Albright.as a distinct entity in association with skin spots and hyper functioning endocrinopathies.

Albright described it as Osteitis fibrosa disseminata since it reflects a similarity with the skeletal disease of hyperparathyroidism, Osteitis and fibrosa cystica (Lee.AT *et al.,* 2007).

Frequency: FD is essentially a disease affecting the young with an incidence ranging from 1:4000to 1:10000. (Jones WA, 1965)The bones of the face and skull are frequently involved .It results in asymmetry and spontaneous fractures. The craniofacial structures are involved in 10%of monostatic type, 50% of mild polyostotic cases, and100% of severe polyostotic cases. The maxilla and mandible are commonly affected with the temporal bone being involved in 18% of cases (Cholakova. R *et al.,* 2010).

Etiology: The exact etiology of FD is still unknown. It has been linked to a mutation in the Gsα gene located at chromosome 20q13.2-13.3. The specific location of mutation is at position 201, which is usually occupied by arginine (R201) and is replaced by either a cysteine (R201C) or a histidine (R201H) (DiCaprio .MR, 2005).

Symptomatology: the maxilla is more often affected than the mandible and the lesions are first detected in the late first and early second decades of life without any gender or racial predilection. (Lan Su .RE *et al.,* 2008) Most lesions are monostatic, asymptomatic, and incidentally- identified. (DiCaprio *et al.,* 2005) The disease is characterized by painless osseous expansion with facial asymmetry.

The craniofacial bones affected by fibrous dysplasia are in the following order: frontal, sphenoid, ethmoid, maxilla, mandible, zygoma, parietal, occipital and temporal (Mahajan .S *et al.,* 2005).

The most important FD symptoms reported by patients are: throbbing or discomfort, diplopia, warmth at the injury site , decreased visual acuity leading even to loss of vision, deafness, dizziness, and nasal obstruction (SANTOS *et al.,* 2010) .Its slow growth can bring several complications, such as facial asymmetry, chronic sinusitis, recurrent headache, nasal obstruction, retention and dental malocclusion. The presence of ulceration and the involvement of cranial nerves are rare but can be found, causing paralysis resulting from the foramina narrowing (Soussa.R *et al.,* 2016).In our case, the patient was referred to our department with a swelling complaint.

Radiology: The density and trabecular pattern of FD lesions is variable. Early lesions may be more radiolucent than mature ones. However, in rare cases, they may appear to have granular internal septa, giving the internal aspect a multilocular appearance. The abnormal trabeculae are usually irregularly- shaped, shorter, thinner, and more numerous than the normal trabeculae. This creates a variable radio-opaque pattern. It may have a granular appearance ("ground-glass" appearance, resembling the small fragments of a shattered windshield), a pattern resembling the surface of an orange (peau d'orange), a wispy arrangement (cotton wool), or an amorphous dense pattern. A distinctive characteristic of FD is the organization of the abnormal trabeculae in a swirling pattern, similar to a fingerprint.

Histologically, a fibro-osseous pattern is typically seen .Yet akin to the imaging features, subtle changes are seen at various stages of the disease's natural history. In the early phase of development, pronounced osteogenesis is observed, presenting thin osteoid anastomosing trabeculae, rimmed by osteoblasts. The stromal fibroblastic element is proliferative and hypercellular although no pleomorphism can be seen. In the ensuing weeks, the trabeculae thicken, yet the osseous collagen pattern remains woven and the trabeculae assume the classic "Chinese letter" characteristic. The fibrous element continues to be hypercellular. In the later stages of the disease, woven bone is replaced by lamellar bone trabeculae. Extensive remodeling may result in a mosaic pattern of resting and reversal lines. In the

present case, the trabeculae showed a c- shaped pattern within a fibrous connective tissue stroma (Neha Vaid *et al.*, 2013).

Differential Diagnosis:

The main differential diagnosis should be made in relation to cemento-ossifying fibroma, presenting the same histological characteristics of fibrous dysplasia. Nevertheless, the first one presents a radiographic image encapsulated with a radiolucent halo delimiting the lesion Contrary to FD characterized by a frosted glass appearance and ill-defined margins (15) (PERDIGÃO, PF *et al.*,2004).

Differential diagnosis can also be made with osteosarcoma, an alteration found in the pelvis, femur, humerus, and skull as well as and facial bones. The radiographic signs would be the extensive soft tissue components, aggressive osteolysis, cortical rupture and extensive periosteal proliferation (JOSÉ, FF *et al.*, 2008).

Furthermore, Paget's disease is characterized by high alkaline phosphatase levels and a bilateral involvement while fibrous dysplasia is predominantly unilateral.

Treatment: Surgery is the primary treatment of choice. It usually consists in a conservative "recontouring" of the tooth-bearing area of the jaw (PERDIGÃO, PF *et al.*, 2004). However, surgical treatment of the cranio-maxillo-facial lesions is controversial. Excision of all the affected bone is usually fruitless since it is impossible to determine the limits of the disease. Yet, it is only indicated if an important function is threatened, when deformity becomes substantial, or complications develop.

CONCLUSION:

Isolated cases of fibrous dysplasia in the maxillomandibular region are rare and can be difficult to differentiate from other benign and malignant bone disorders. For this reason, knowledge of the structural changes is important for the correct diagnosis of bone pathology. Thus, the dental surgeon should be familiar with the normal morphology of the structures as well as their possible abnormalities.

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