

FIBROUS DYSPLASIA IN THE MAXILLO-MANDIBULAR REGION – CASE REPORT

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ABSTRACT

Craniofacial fibrous dysplasia is 1 of 3 types of fibrous dysplasia that can affect the bones of the craniofacial complex, including the mandible and maxilla.

Fibrous dysplasia is a skeletal developmental disorder of the bone-forming mesenchyme that manifests as a defect in osteoblastic differentiation and maturation.

It is a lesion of unknown etiology, uncertain pathogenesis, and diverse histopathology.

Fibrous dysplasia represents about 2, 5% of all bone tumors and over 7% of all benign tumours.

The aim of this article is to represent a rare case of bilateral fibrous dysplasia of the upper and lower jaws, in combination with Intellectual disability (previously called mental retardation). The clinical diagnostic approach including imaging studies: Orthopantomogram (OPG) and 3D tomography is described. Histological examination was also essential for obtaining a definitive diagnosis.

Key words: fibrous dysplasia, Lichtenstein-Jaffe's disease, Maxillofacial, McCune-Albright's disease, osteodystrophia fibrosa, osteitis fibrosa disseminata, monostotic form, polyostotic form, craniofacial form, cherubism, 3D tomography, Intellectual disability.

INTRODUCTION

Fibrous dysplasia (FD) is a bone development anomaly characterized by hamartoma proliferation of fibrous tissue within the medullary bone, with secondary bony metaplasia, producing immature, newly formed and weakly calcified bone, without maturation of the osteoblast which appears radiolucent on radiographs, with the classically described ground-glass appearance.(1).

In 1937, McCune and Bruch first suggested that among all of the abnormalities of bone formation, this disorder should have its own place as a distinct clinical entity. The following year, Lichtenstein introduced the term "fibrous dysplasia"

Reed's definition states that fibrous dysplasia is an arrest of bone maturation, woven bone with ossification resulting from metaplasia of a nonspecific fibro osseous type. (3). The etiology of this abnormal growth process is

related to a mutation in the gene that encodes the subunit of a stimulatory G protein (G α) located on chromosome 20.1,2 As a consequence of this mutation, there is a substitution of the cysteine or the histidine-amino acids of the genomic DNA in the osteoblastic cells-by another amino acid, arginine.3 Consequently, the osteoblastic cells will elaborate a fibrous tissue in the bone marrow instead of normal bone. (8, 9, 10).

It is a benign bone disorder of an unknown etiology, uncertain pathogenesis and diverse histopathology (2).

Fibrous dysplasia represents about 2, 5% of all bone tumors and over 7% of all benign tumours

Cranial or facial bones are affected approximately in 30% of the patients. (3, 4). The average age of the patients with FD is 25, 8 years (from 5 to 67) without sex preference(46, 7% male) and usually manifests before the 3rd decade of life. (4, 5)

Fibrous dysplasia is described in terms of three major types: monostotic, involving a single bone; polyostotic, having multiple lesions involving multiple bones; and McCune Albright syndrome, a polyostotic form of fibrous dysplasia that also involves endocrine abnormalities.

The monostotic form of fibrous dysplasia is the most common, comprising 70% of cases, most likely to quiesce at puberty. A typical monostotic lesion, usually presented unilateral, will involve the femur, tibia or ribs, with 25% occurring in the bones of the skull. Affection of the craniofacial bone is observed with 10% of the patients suffering from monostotic FD (6, 7).

Twenty-five percent of fibrous dysplasia involves two or more bones. These lesions may be localized to one region of the body or they may be disseminated, involving virtually every bone. There is a female predilection in polyostotic fibrous dysplasia, and up to 50% may involve bones in the head and neck. These lesions are more likely to continue to progress even after puberty.

Deformity is progressive and by mass effect there may be impingement on other structures and functional impairment.

These lesions tend to be structurally weak and are therefore prone to pathologic fracture. Alkaline phosphatase

may be elevated in up to 30% of patients with polyostotic fibrous dysplasia, and a dramatic rise may herald malignant degeneration.

Malignant degeneration occurs in less than 1% of cases of fibrous dysplasia. Malignancies are almost exclusively osteosarcoma. For unknown reasons, monostotic and craniofacial lesions have the greatest potential for malignant degeneration. Pain, rapid growth of a lesion and a dramatic elevation of alkaline phosphatase may herald malignant transformation.

CASE REPORT

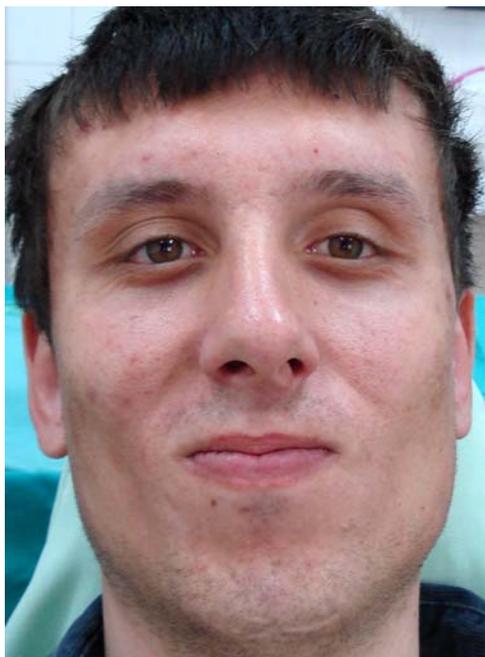
A 28-year-old male presented at the Oral surgery department, Medical university, Plovdiv six years ago with complaining of mild pain in the lower jaw without precise localization.

The past medical history includes skin disorder - rosacea and medium Intellectual disability manifesting with emotional-volitional instability prolonged neurotic decompensation.

There was no family history with similar findings.

The general physical examination revealed a moderately built patient with satisfactory vital signs and mental retardation.

Extraorally no facial deformity was ascertained.



Picture 1.

Oral examination revealed the presence of bilateral symmetrical expansion in the distal part of the alveolar ridge of lower and upper jaws.



Picture 2.



Picture 3.

The covering and surrounding mucosa was normal in color, without any clinical manifestation of inflammation or ulceration.

On palpation affected areas was painless, with hard consistence and plane surface.

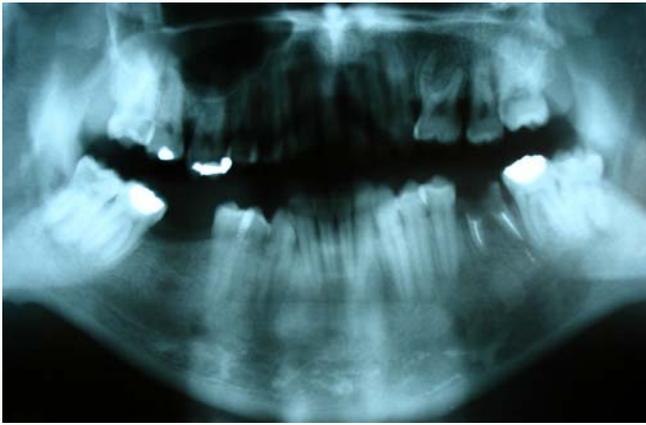
The initial diagnosis based on the clinical examination was fibrous dysplasia of the upper and lower jaws.

Further Imaging Studies with X-ray and 3D tomography was undertaken for accurately definition of the bone density and for obtaining the definite diagnosis.

They confirmed lesion confined to the interior of bone with no soft-tissue involvement which was helpful in distinguishing fibrous dysplasia from a malignancy.

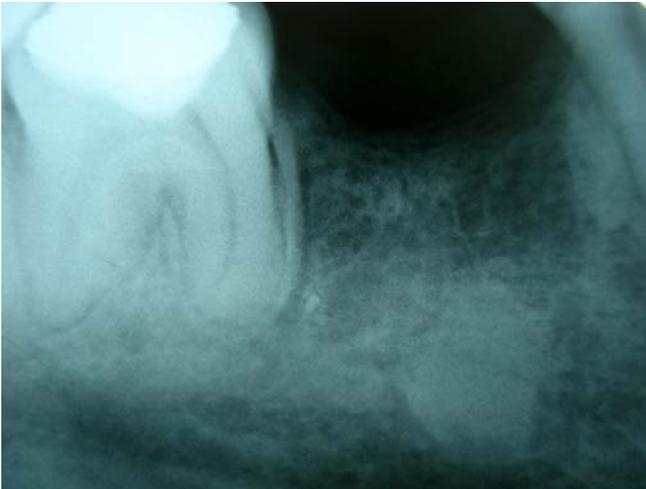
Maxillary and mandibular involvement had a mixed radiolucent and radiopaque pattern.

Orthopantomograph revealed diffusive radiolucent areas in the posterior areas of upper and lower jaws.



Picture 4.

In the area of the lower left first molar above the mandibular canal was observed radio opacity measuring around 10 mm with obliteration of two third of the left maxillary sinus.



Picture 5.

During the extraction of the lower left first molar a bone and soft tissue was taken for histological examination.

The histological findings (№ 12924/ 7. 05. 2007) showed fibrous modification of the bone tissue without soft tissue involvement. This led to the definite diagnosis Fibrous dysplasia.

The patient was advised to visit his dentist regularly for ongoing re-examination and to observe and inform for any change in growth formation and appearance of pain.

In the re-examine periodically is measured the level of alkaline phosphatase.

DISCUSSION

Fibrous dysplasia of the cranium is a rare disorder of unknown etiology in which normal bone is replaced by abnormal fibro-connective tissue proliferation.

In 36, 3% of the cases of FD the clinical beginning is hidden, there is no clear symptoms and obtaining the diagnosis is complicated. The rest of the patients (63, 6%) are with diverse symptoms depending on location, swelling, deformation and presence of pain (8, 11).

Polyostotic fibrous dysplasia affects multitude skeletal bones usually unilateral.

The patient at present is in quiescence and no bone increase is detected during following years.

Sudden increase in the level of alkaline phosphatase is one of the symptoms for malignant transformation and for that reason its amount should be periodically observed.

At present alkaline phosphatase level is normal and maintains a constant level.

The pain in the lower jaw, being the main complaint of the patient, is considered to be caused by compression of the mandible nerve of the enlarged fibrous tissue.

The main aim of the treatment is correction of the functionality in combination with aesthetic effects.

Surgical excision of the affected bone tissue is usually a successful way of treatment. However it leads to a huge functional and aesthetic deficit, as well as long-term post-operative complications.

The conservative therapeutical approach with limited reduction in the size of these lesions is enough to manage the symptoms. Because patients with FD may be at risk of malignant transformation, periodic follow-up is mandatory to detect such transformation. (2, 13). Although the conservative approach in case of FD, affecting the jaws, is widespread, ortognatic surgery is used in many cases in order to restore the occlusion and to correct the dentofacial deformity, caused by the disorder.

Surgical approach aims: stable occlusion, facial aesthetics and evasion of post-operative relapse. The radiotherapy is contra indicated (14, 15).

Biphosphonates are used in cases when an intervention is necessary but the surgery can not be performed (8). Some authors suggest applying calcitonin in combination with surgical treatment. Calcitonin treatment aims local bone calcification which leads to reduction in bleeding during the bone remodeling (16).

Usually the prognosis is good although the bad outcomes occur more frequently among young patients or those with polyostotic forms of the disorder (8). The interval between the diagnosis of FD and the development of malignity is 13,5 years. The risk of malignant

transformation is 0, 5%, if the patient with FD does not undergo treatment (1, 8). The malignant transformation is higher among male with polyostotic FD, craniofacial lesions and monostotic FD. The clinical signs of malignancy development are: pain, immediate swelling and an increase in the alkaline phosphatase. The radiotherapy increased malignant transformations more than 400 times (1, 3).

CONCLUSIONS

Isolated cases of fibrous dysplasia in maxillo-mandibular region are rare and can be difficult to

differentiate from other benign and malignant bone disorders.

The general dental practitioner can be the first to detect such conditions especially when the only affected areas are in maxillo-mandibular region so sufficient knowledge on this condition is important for the proper diagnosis, treatment and prevention of further complications.

For obtaining the definite diagnosis, treatment and further management of fibrous dysplasia is mandatory to be carried out imaging studies, histological and laboratory tests.

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