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FIBROUS DYSPLASIA OF MAXILLARY SINUS : A CASE REPORT

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ABSTRACT

The Fibrous Dysplasia Is A Benign Bone Disease , Of Slow Growth And Unknown Etiology . The Involvement Of The Craniofacial Skeleton Is Not Uncommo And Generally , Produces Facial Asymmetries . There Are Three Type Of Disease : Monostotic Fibrous Dysplasia , That Involves Only One Bone And Polyostotic Involving Multiple Bones And Mccune – Albright Syndrome . The Surgical Treatment Remains As The Main Therapeutic Approach And The Postoperative Follow-up Is Necessary Due To Its Recurrent Nature.

KEYWORDS

Fibrous Dysplasia , CT Scan , paranasl sinus.

INTRODUCTION :

Fibrous dysplasia is a tumor-like lesion of the bone. It is self-limiting , is not encapsulated and is characterized by replacement of normal bone with cellular fibrous connective tissue , which contains irregular trabecules of immature , nonlamellar , metaplastic bone . The etiology is unknown , but different hypotheses have been discussed. The disease may develop at an early age , progress actively during childhood and stabilize in adulthood. In the majority of cases , fibrous dysplasia is diagnosed before the second decade of life , rarely after the fifth. This disease is much more frequent in white populations than black and the monostotic form is found more often in females than in males.[1]

Clinically, there type of fibrous dysplasia are differentiated:
Monostotic; Polyostotic and McCune – Albright syndrome, which presents as a combination of polyostotic fibrous dysplasia, skin hyperpigmentation and endocrine dysfunction. It occurs in one of 30-40 cases of fibrous dysplasia.[1]

The diagnosis suspicion is made by clinical and radiological methods which need anatomopathological confirmation [3]. The typical radiological exam shows a characteristic aspect of “opaque glass” involved by a dense cortical tissue [4]. The definitive treatment is the le surgical excision. The clinical follow-up of the patient is essential for the early diagnosis of a recurrence. We describe one case of maxillary bone fibrous dysplasia that, in spite of its benign nature, caused facial deformity.

CASE REPORT:

A 45 years old man was referred to department of ENT HNS OPD with complaints of swelling over the left cheek since childhood causing facial asymmetry. The swelling was insidious in onset and gradually progressive. The swelling was initially small in size and gradually increased to Size of about 5x6 cm. The swelling was associated deficit of visual accuracy and unilateral nasal obstruction . There was no associated pain over the swelling and not associated with loosening of teeth. There was no history of difficulty in mouth opening. Although he had no history of trauma over the cheek and no history of dental infections or tooth extractions in the past.

The clinical exam, good general state, the evaluation of organic system didn't reveal alterations. He presented with a painless mass in the left maxilla and left zygoma with normal subjacent oral skin and mucosa.

On extra oral examination diffused swelling of 5x6cm was present on left side of the face extending superi-inferiorly 0.5cm below the canthotragus line; to line joining corner of mouth to tragus & anteroposteriorly 1cm away from the corner of the mouth to 1.5 cm in front of the tragus. On palpation the consistency was bony hard , nontender & no local rise of temperature. Intra orally swelling was present extending buccally from first premolar to about 7mm beyond the tuberosity. Refer Appendix [Fig 1(a)(b)]

The computed tomography of the face sinuses showed a hyperdense , heterogeneous , extensive mass aspect of an 'opaque glass' involving the maxilla ,zygomatic bone and left maxillary sinus.Refer Appendix [Fig 2(a)(b)] Histological study revealed a compatible picture with fibrous dysplasia .

Serological investigation including serum calcium ,serum phosphorus, and alkalin phosphatase(ALP) were within normal range.

DISCUSSION :

Fibrous Dysplasia is a benigne,chronic,slowly progressive bone disorder of unknown etiology that is characterized by the replacement of normal bone with a variable amount of fibrous tissue and woven bone [11].

It's classified as for the number of affected bones and the presence or not of extra-skeleton abnormalities. The monostotic form affects only one bone and corresponds to 70-80% of the FD cases. The polyostotic form, in which several bones are affected, may be divided into three subtypes: craniofacial, in which only the craniofacial complex are involved including the jaw and the maxilla; Lichtenstein-Jaffe , in which in addition to the several skeleton bones involvement there are coffee-with-milk pigmentations; Albright's Syndrome, characterized by the affection of several bones, coffee-with-milk pigmentations in the skin and endocrine affection with a remark for the early adolescence in girls. The polyostotic form corresponds to 20-30% of the cases [6]. The craniofacial bones are more affected in the polyostotic form (50-100%) than in the monostotic form (20%) [3].

Clinical manifestations of fibrous dysplasia include bony deformity , pathologic fracture, and cranial never palsies . The disease starts early in life, usually in childhood; the monostotic from may become quiescent at puberty, whereas the polyostotic from can continue to progress. Sarcomatous transformation can occur, with an estimated incidence of 0.4%.[85]

The CT is the choice exam for the study of the lesion[1], analysis of their extension and surgical preparation [5,8]. Basically, three radiographic standards in the cranium fibrous dysplasia and facial bones are described: pategoid, that alternates the radiodense and radiotransparent areas; sclerotic, homogeneously dense; cystic standard, with spherical or ovoid radiolucid area surrounded by dense limits[9]. In the case reported, the lesion tomographic images presented a hyperdense standard intermixed by imprecise limits hypodense areas, which resulted in the classical aspect of “opaque glass”. The FD definitive diagnosis is made by the correlation of clinical, radiological and anatomopathological findings [10].

The microscopic evaluation shows a collagen matrix stroma with fibroblasts in a entangled standard with osseous trabeculate similar to the “Chinese writing” [9].

The main factors that guide the FD approach are the presence and the intensity of the symptoms, the tumor location and the patient's age. The simple presence of the lesion does not justify surgical intervention. The main indications for surgical treatment of FD are the presence of significant clinical symptoms and the control of large aesthetic deformities [2, 7].

The clinical and radiological follow-up by CT is essential in the

patients with FD because of the moderate recurrence rate of the lesion[1] which may reach 37% according to some authors [7].

The surgical treatment remains as the main therapeutic approach and the postoperative follow-up is necessary due to its recurrent nature.[Fig 3,4]

CONCLUSION :

The Fibrous Dysplasia involving the faciomaxillary region should always be considered as differential diagnosis for any patient presenting with swellings involving facial and cranial bones. It is significant for the otorhinolaryngology because it may affect facial and cranial bones and may cause deformities and dysfunctions. Moreover CT Scane is the investigation of chonce. The invovlment of paranasal sinuses is rare. For obtaining the definite diagnosis, treatment and further management of fibrous dysplasia is mandatory to be carried out imaging studies, histological and laboratory tests. Fibrous Dysplasia is a tumor like development disorder with minimal chances of malignancies .Aesthetic correction is done by surgeries.

APPENDIX:

Fig 1(a)

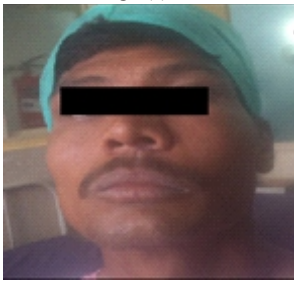


Fig 1(b)

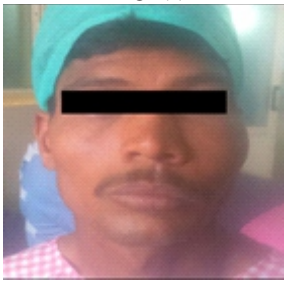


Fig 2(a)

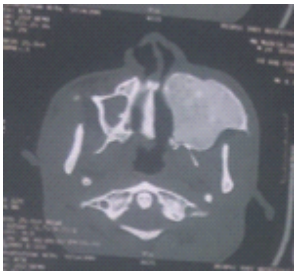


Fig 2(b)

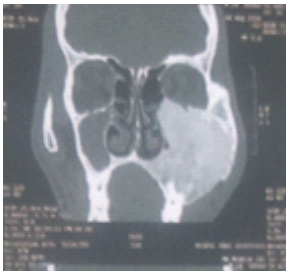


Fig 3

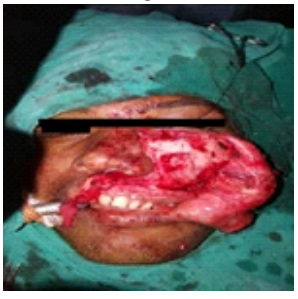
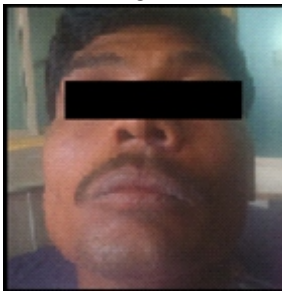


Fig 4



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