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RESEARCH ARTICLE

CRANIOFACIAL FIBROUS DYSPLASIA: A CASE REPORT.

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Abstract

Fibrous dysplasia is a developmental tumor like condition that is characterized by replacement of normal bone by excessive proliferation of cellular fibrous connective tissue intermixed with irregular bony trabeculae. caused by sporadic, congenital mutations in the cAMP regulating protein, Gsa. Craniofacial form is a type of fibrous dysplasia, occurs in association with monostotic or polyostotic form can also occur in an isolated pattern. Here a case of craniofacial fibrous dysplasia has reported.

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Introduction:-

Fibrous dysplasia was first reported by Von Recklinghausen in 1891 & he coined the term Osteitis Fibrosa Generalisata^[1,2]. In 1938, Lichtenstein and Jaffe first introduced the term Fibrous Dysplasia^[1,3]. Fibrous dysplasia (FD) is an uncommon developmental bone disease of benign origin. It leads to expansion & replacement of medullary bone by disorganized fibroosseous tissue^[4]. It comprises 2.5% of all osseous & 7% of all benign bone tumors^[5]. In general they affect 1 in 4,000 to 10,000 individuals^[3]. Male to female ratio is equal^[1,5]. Most frequently found in metaphysodiaphyseal region of long bones i.e. in rib (28%), followed by femur (23%), tibia, craniofacial bone (10-25%)^[6]. Sarcomatous transformation is rare^[5]. As earlier stated Fibrous dysplasia is a developmental tumor like sporadic condition that results from a post zygotic mutation in GNAS1 (Guanine Nucleotidebinding Protein, α -stimulating activity polypeptide 1) gene^[1,6]. GNAS1 gene codes for G protein which stimulates cAMP production in affected tissue; which result in (1) Endocrinal disturbances (2) café-au-lait spots (3) Aberrant activity during osteoblasts differentiation, which results in normal medullary bone to be replaced by fibrous tissue^[6].

Case report:-



Figure 1:- Extraoral photograph showing solitary swelling present in the right middle third of the face.



Figure 2:- Intraoral photograph showing expansion in right maxilla.



Figure 3:- Intraoral photograph showing right palatal expansion.

A 11 year old male patient reported to our department with complaint of swelling in the right side of the face noticed since 3 months. Initially swelling was small and gradually increased to present size. On extra oral examination diffused swelling of about 3*4 cm was present on right side of the face extending superioinferiorly 1 cm below the cantho tragus line; to line joining corner of mouth to tragus and anteroposteriorly 1 cm away from the corner of the mouth to 1 cm in front of right tragus. On palpation the consistency was bony hard, nontender, and no local rise in temperature. Intraorally swelling was present extending buccally from right upper deciduous canine to beyond right tuberosity. Labiopalatal dimension at premolar region was about 2 cm. Buccal and palatal cortical plates were expanded. Overlying mucosa appeared normal, firm, and was non-tender. Laboratory investigations were normal except for a raised serum alkaline phosphatase level which was 309IU/L. Radiological investigations includes maxillary occlusal view, orthopantomogram(OPG), and CT Scan and bone scan.

Maxillary occlusal view and OPG shows gross radiopacity in the maxillary bone from first premolar region to posterior to tuberosity region, which gives ground glass appearance.

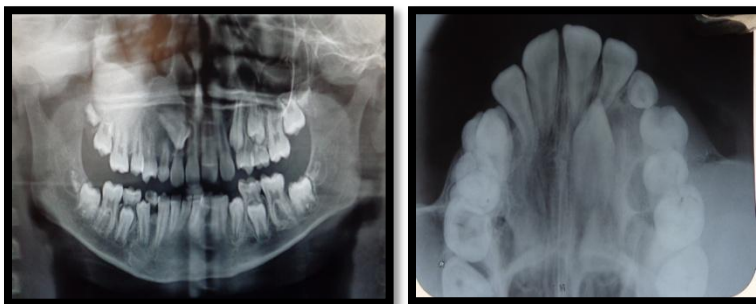


Figure 3:- OPG shows diffuse radiopacity in right maxilla extending from right premolar region to upper third molar further till the infraorbital region.

Figure 4:- maxillary occlusal view shows diffuse radiopacity with buccopalatal extension from first premolar to maxillary tuberosity.

CT image shows a radio opaque lesion occupies most of the facial bones in the mid and upper facial skeleton on the right side, which is significantly expanded. The maxillary sinus and nasal airway on the right is obliterated



Figure 5: pre-operative coronal CT.



Figure 6: pre-operative 3D CT.



Figure 7-surgical operation of the case

Discussion:-

Fibrous dysplasia is a bone disorder characterized by slow progressive replacement of medullary bone by abnormal proliferative isomorphic fibrous tissue with classic ground glass appearance^[4]. Fibrous dysplasia has 4 different disease patterns Monostotic (70%), Polyostotic (30%), Craniofacial form and Cherubism(rare)^[3,6]. The range of skeletal involvement varies from an asymptomatic to progressive functional deficit & aesthetic problems. The clinical severity depends on time when the mutation of GNAS-1 occurs.

Monostotic Form: If mutation occurs during postnatal life the progeny of that mutated cells are essentially confined to one site resulting in fibrous dysplasia affecting a single bone. About 70% cases are of monostotic form and they involve mainly ribs, femur, tibia & craniofacial bones^[4,5,6].

Polyostotic Form: It is seen if mutation occurs during 6th week of intrauterine life. Multiple bones may get involved. This form commonly involves the skull & facial bones, pelvis, spine & shoulder girdle. Femur shows shepherd's crook deformity^[6]. Polyostotic form is again sub-classified into Jaffe's type & Albright syndrome. Polyostotic fibrous dysplasia with soft tissue called mazabraud syndrome^[7].

Craniofacial Form: 50%-100% of patient with polyostotic disease & 10% patient with monostotic disease have craniofacial involvement^[1,4]. Maxilla is more commonly involved than mandible^[4]. When maxilla is affected it may involve zygomatic & sphenoid bone. Involvement of frontal, sphenoid, naso-ethmoid & maxillary bone may lead to nasal obstruction, sinus obstruction & sinusitis^[4]. Hypertelorism, cranial asymmetry, facial deformity, visual impairment, exophthalmos and blindness may occur due to involvement of orbital and parietal bone^[6]. Malignant changes with fibrous dysplasia include Osteosarcoma, Fibrosarcoma, Chondrosarcoma, Malignant fibrous histiocytoma & Ewings sarcoma^[4]. Association of Ameloblastoma, Cystic degeneration, Angiosarcoma^[8], Frontal sinus mucocele^[9] have also reported. Treatment is primarily surgery. When the only tooth bearing area is involved conservative treatment is bone shaving. Use of Calcitonin & Pamidronate is also reported for its treatment^[10]. Biopsy can be taken to rule out the lesion. Fibrous dysplasia usually get stabilized after puberty.

Cherubism: It is a special variant of fibrous dysplasia is also known as familial fibrous dysplasia. It is an autosomal dominant disorder of variable penetrance, with onset in childhood and is typically more severe in males^[11].

Conclusion:-

Fibrous dysplasia is a difficult disease to manage. The sporadic mosaic nature of the disease means that it is an uncommon disease with a variable expression. Hence managing the outcome of this disease is challenging. In the future, we can look to new medical treatments that are on the horizon, and should work for the initiation of international collaborative studies to better define optimal surgical approaches to the treatment of this challenging disease.

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