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CASE REPORT

A CASE REPORT ON CRANIO FACIAL FIBROUS DYSPLASIA INVOLVING LEFT ORBIT

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A R T I C L E I N F O

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A B S T R A C T

Purpose: Craniofacial fibrous dysplasia is a benign slowly progressive disease in which normal craniofacial bones are replaced by immature fibro-osseous tissue. This case evaluates the need for early surgical intervention in such patients for its management and to stop further progression.

Methods and Materials: A 35yrs year old man with normal visual acuity and fundus finding presented with axial proptosis of left eye. On further evaluation by MDCT a diagnosis of craniofacial fibrous dysplasia involving left orbit was made. His rest ocular functions were normal including normal audiometric finding.

Result: Left frontal craniotomy followed by medial maxillectomy and ethmoidectomy done on left front medial region with front medial osteotomy and debulking surgery. The specimen was further diagnosed as Fibrous Dysplasia involving bone. In periodic follow-up there was regression of proptosis. Initially there was diplopia in lateral gaze of left eye but subsequent follow-up resolved the complaint.

Conclusion: Early surgical intervention is needed to stop further progression. Periodic follow-up is recommended to monitor for recurrence and malignant transformation.

INTRODUCTION

Fibrous Dysplasia is a benign slowly progressive disease in which normal craniofacial bones are replaced by immature fibro-osseous tissue. It is of following types Monostotic – Involving single skeletal site and is the most frequent form. Polyostotic – Multiple sites involve, here severe bone deformity is seen. McCune Albright Syndrome – Polyostotic fibrous dysplasia with endocrine and skin changes seen. Most common sites – craniofacial skeleton, ribcage, femur and tibia.Craniofacial fibrous dysplasia is one of the types of fibrous dysplasia and is characterised, as the name suggests, by involvement of the skull and facial bones. Monostotic forms accounts for approximately 75% of all cases with 20-30% are of the Polyostotic variant. 23% of all fibrous dysplasia involves the craniofacial skeleton. 33% recurrence rate and 0.2-2.5% Malignant transformation. Craniofacial dysplasia may be seen in over half of polyostotic patients and 10-35% of monostotics. Young adult patients are most frequently affected. Genetically there is activation somatic missense mutation of GNAS1 gene on chromosome 20 occurs. This leads to signaling of G protein, Gs-alpha triggers replacement of normal bone by immature bone marrow stromal cells in a fibrous matrix.

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SUBJECT AND METHODS

Case Report

A 35 yr old man coming with a chief complain of unilateral Axial Proptosis in Left eye for 3 yrs, which was gradual in onset and progressive in nature. There was no history of pain. On ocular examination left eye was 5mm more proptosed as compared to right eye with 15 degree Exotropia. Vision was normal with Extra-ocular movement was full free and painless in both eyes.

Investigations

Fundus finding was normal with normal field testing and O.C.T to rule out optic nerve involvement.

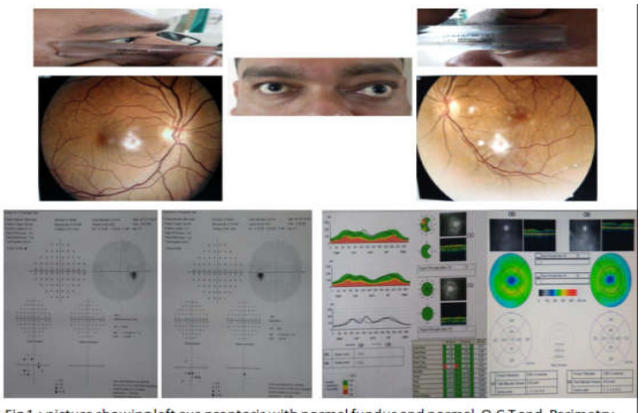

Fig 1 : picture showing left eye proptosis with normal fundus and normal O.C.T and Perimetry

M.R.I study reveals an expancile mass involving the left frontal and ethmoidal sinus measuring 5.5*4.3*4.8 cms with hperosteosis of wall possibly of fibrous dysplasia C.T reports displays fibrous dysplasia involving left side of frontal and ethmoidal sinus with no involvement of brain parenchyma.

Ultra sound whole abdomen with Audiometry and serum alkaline phosphatase test was done to rule out any secondary metastasis or spread.

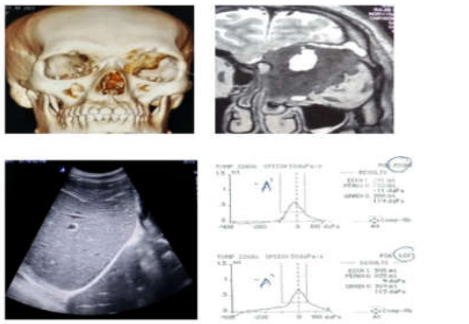

Fig 2 : showing CT and MRI scan diagnosing fibrous dysplasia with normal USG whole abdomen and Audiometry

So a provisional diagnosis of Fibrous Dysplasia Involving Left Frontal, Ethmoidal And Maxillary Sinus Sparing Optic Nerve was made.

Management

The patient has undergone Left frontal Craniotomy followed by Medial Maxillectomy and Ethmodectomy and finally Frontomedial Osteotomy.

Post-op day 1 and 2 shows some exposure. And the histopathological reports confirms the diagnosis of fibrous dysplasia.

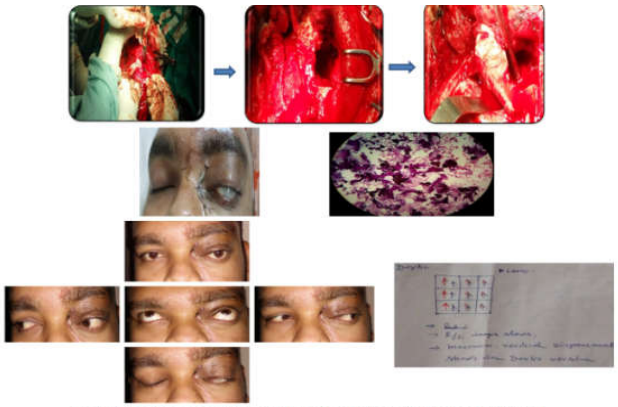

Fig 3 : Operative procedure with first day post-op with histopathological picture with Second week Post-op and Left eye diplopia under lateral gaze

First two weeks follow-up patient had incomplete closure of left lower lid which was managed by artificial tear drops and topical antibiotics. He also had diplopia in lateral gaze due to left lateral rectus underaction. But on subsequent follow-up diplopia was reduced and closure was complete with full and free extraocular muscle movement.

RESULTS

Post operative results were good and the visual acuity was maintained at 6/6 with Occular movement being full. Left eye axial proptosis was reduced to 3mm. But 7 months post CT shows that Few fibrodysplastic growth noted on left medial canthus. So regular follow-up were done to see any progression of disease and the patient was encouraged to maintain bone density through diet and exercise.

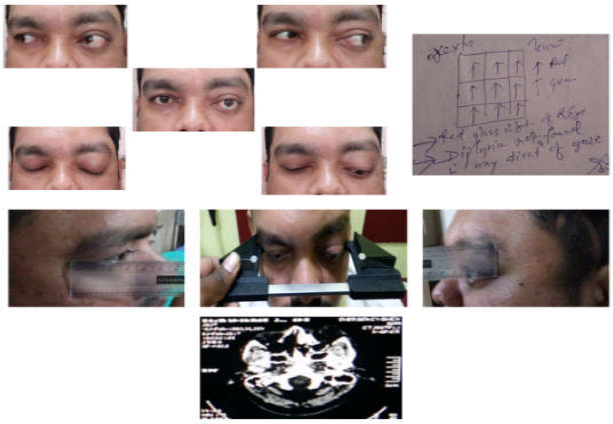

Fig 4 : seven months post-op with no diplopia and CT showing few fibrodysplastic growth

DISCUSSION

Fibrous dysplasia is a benign disorder of bone. It can involve any bone, but most commonly affects the long bones of the extremities or the craniofacial skeleton. Early surgical intervention is needed to stop the further progression. In this case after diagnosis early surgical intervention was done but seven month follow-up showed presence of fibrodysplastic growth. For this periodic follow-up is needed to monitor for recurrence and malignant transformation.

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Monostotic Fibrous Dysplasia: A Case Report

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