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**Case Report** **Journal of MAR Ophthalmology (Volume 5 Issue 4)**

# Monostotic Fibrous Dysplasia of the Ethmoid Presenting as a Visual Field Defect: A Case

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***Abstract***

*Fibrous dysplasia is a benign, slowly progressive, skeletal disorder, wherein the normal medullar bone is replaced by the abnormal fibro-osseous tissue. We report an extremely rare case of fibrous dysplasia of ethmoid in a young Arab lady who presented with a positive scotoma in the inferotemporal quadrant in the right eye. Radioimaging and histopathological examination confirmed the diagnosis of monostotic fibrous dysplasia of ethmoid bone. Literature search suggests that this may be the first case of ethmoid fibrous dysplasia presenting as visual field disturbances. Growth hormone supplements administered to the patient during adolescence to improve the general growth could be a probable aetiology for the fibrous dysplasia.*

***Keywords:*** *Fibrous dysplasia, ethmoid, visual field defect, growth hormone.*

## Case Report

A 24 years old Arab lady patient presented to the Department of Ophthalmology with complaints of a positive scotoma in the inferotemporal field of vision in the right eye for the past few weeks. Ophthalmological examination revealed proptosis of 1-2 mm in the right eye on the Hertel Ex ophthalmometer. Visual acuity was 6/6 in both eyes. Pupillary response to direct and consensual light was normal in both eyes. Fundus examination showed solid growth tenting the superonasal retina (in the right eye. The optic nerve head showed well defined margins and normal cup disc ratio. The examination of the left eye was within normal limits.

A magnetic resonance imaging showed enlarged and expansile right anterior and middle ethmoidal cells in the ethmoid, and measured approximately 32 × 18 × 21 mm (L × W × H) that was isointense of T1 weighted and hyperintense of T2 weighted sequences, with no contrast enhancement, suggestive of an ethmoidal mucocele, that was impinging on the globe and medial extraconal space causing proptosis (Figure 1).

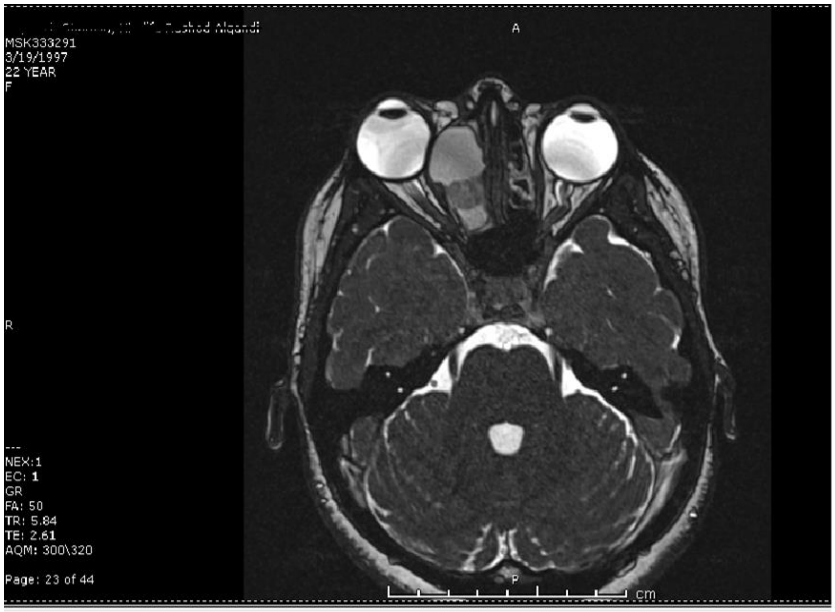


**Figure 1:** MRI showing the initial presentation of right mucocoele causing proptosis and compression of optic nerve

The patient was referred to the ENT department and endoscopic surgical excision of the mass was performed. Histopathological examination was consistent with the diagnosis of fibrous dysplasia of ethmoid bone (Figure 2).

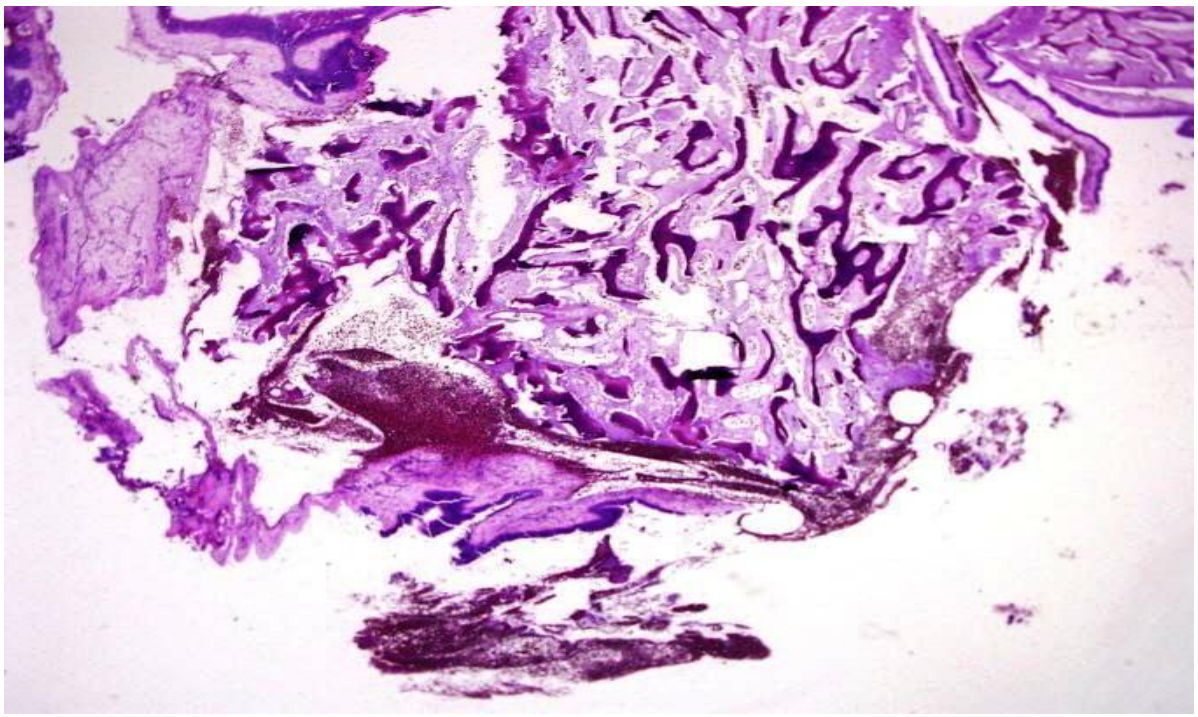

**Figure 2:** Histopathological examination following excision confirming the fibrous dysplasia

Laboratory examinations including serum alkaline phosphatase and serum growth hormone values were within normal limits. The patient gave a history of having taken growth hormone supplements during her adolescence as the general growth was poor during that time. A follow up CT scan after the surgery and a detailed complete clinical and radiological examination did not reveal any other bony lesion in the body and a diagnosis of monostotic fibrous dysplasia of the ethmoid bone was made. The patient was clinically asymptomatic post -surgery and is on regular follow-up.

## Discussion

Fibrous dysplasia is a fibro-osseous lesion and ethmoid bone involvement is rarely reported.**[**1**]** The lesion at the medullary bone expands and involves the nearby cortex; the bone trabeculae are irregularly shaped, and a bone tissue without internal lamellar structures develops. Fibrous dysplasia is of special interest to the ophthalmologist as it can affect the craniofacial bones either as a monostotic fibrous dysplasia or as a part of the McCune-Albright syndrome.**[**2**]** Patients may commonly present with symptoms of atypical pain in the head and face as well as sinusitis symptoms. Encroachment on the paranasal sinuses, orbit, and foramina of the skull may lead to optic neuropathy and visual loss, proptosis, diplopia, and epiphora.**[**3**]** Literature search showed that our case was the first of its kind where the patient presented with symptoms of only visual field disturbances. Magnetic resonance imaging (MRI) provides excellent definition of ocular and soft-tissue involvement and allows assessment of adjacent neurovascular structures. T2-weighted images in patients with FD reveal low signal from the involved bones and very high signal from the cystic component (or mucoceles).**[**4**]**

Another interesting aspect of our case was the probable iatrogenic cause of fibrous dysplasia. The patient had received growth hormone supplements during her adolescence to stimulate growth. The effects of growth hormone on metabolism of skeletal tissues are complex. Increased bone remodeling is noted in both acromegaly as well as exogenous administration of growth hormone.5 The exact mechanism by which growth hormone stimulates fibrous dysplasia of bone is not known but may be related to increased proliferation of bone marrow stromal cells. Impaired differentiation and increased proliferation of these stem cells is a primary effect of the activation of Gαs, driving the formation and expansion of fibrous dysplasia lesions.**[**5**]** Growth hormone excess is one of the endocrinopathies in the McCune Albright Syndrome which was positively correlated with expansion of lesion in FD and with pain prevalence at FD sites in adults. While our patient had an isolated ethmoid dysplasia with no systemic involvement, exposure to growth hormone supplements during adolescence could be a probable cause for its presentation. Careful monitoring of the skeletal growth after growth hormone

supplements is important for early diagnosis of fibrous dysplasia. Differential diagnosis of fibrous dysplasia should be considered in patients with a history of growth hormone supplements presenting before the age 30 with a chronic retro-orbital pain or visual field disturbances.

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