Fibrous Dysplasia: Cause of Nasolacrimal Duct Obstruction

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Abstract
Introduction: Fibrous dysplasia (FD) is a slow growing, benign, idiopathic skeletal disorder. The most common site of involvement is ribs followed by craniofacial bones in which the incidence is 25% to 30%
Case Report: We report a case of 35 years old female patient with history of swelling left cheek for 10 years with complaints of nasal obstruction, epiphora with facial asymmetry involving left maxillary sinus. The final diagnosis came out to be FD with NLD (nasolacrimal duct obstruction) confirmed by radiological and histopathological examination. The lesion was removed with lateral rhinotomy approach followed by dacryocystorhinostomy for NLD obstruction.

Keywords: Fibrous Dysplasia, Epiphora, NLD, Lateral rhinotomy.

Introduction
FD is an idiopathic skeletal disorder in which trabecular bone is replaced and distorted by fibro-osseous tissue.1,5 FD lesions are in three forms mono-ostotic 75-80% and poly-ostotic FD where multiple bone involvement is seen in 30%.2,3 The third is McCune-Albright syndrome seen with café-au-lait pigmented skin lesions with multiple endocrinopathies.2,3 FD is commonly a benign lesion with irregularly distributed spicules of bones lying in cellular fibrous stoma. FD involves maxilla two times more than mandible.1,1 The most common site of involvement is ribs followed by craniofacial bones in which the incidence is 25% to 30%.14 The differential diagnosis of FD are Paget disease, ossifying fibroma, and osteosarcoma which are usually confused either clinically or radiologically with fibrous dysplasia.1,5 Common clinical features involves facial asymmetry, paraesthesia, sinus obstruction. Ocular proptosis, visual disturbance, and epiphora are also some rare presentations. Chronic dacyrocystitis caused due to obstruction of NLD presents as Purulent epiphora.7 we report a case of Fibrous dysplasia with NLD.

Case Report
35year old female with 10 year history of swelling left side of face and 6 months history of purulent epiphora affecting the right eye reported to the department of ENT of a tertiary care centre. Initially the swelling was small, pea size but it gradually increased to present size. There was no history of trauma, paraesthesia and no difficulty in...
chewing food. On examination there was a diffuse swelling 4×5 cm, hard, non tender extending from 1 cm below the infra-orbital margin superiorly and mediolaterally from left side of nasal septum up to the ramus of the mandible. (Image 1) Obliteration of naso-labial fold was present on the left side. On anterior rhinoscopy there was a soft tissue mass in the left nasal cavity. Vision and ocular motility were normal and blockage was seen after inferior canalicular syringing that is regurgitation test was positive. Complete hemogram was within normal limits. Serological investigations were normal including serum phosphorus, serum calcium and alkaline phosphatase. X-ray paranasal sinuses showed homogenous radio-opaque ground glass appearance on left maxillary sinus. (Image 2) High resolution computed tomography of nose and paranasal sinuses showed an ill-defined lobulated mass of size 5×5×4.7 cm seen arising from left maxilla which had completely obliterated the left maxillary sinus upto the lateral nasal wall obliterating the left osteo-meatal complex. (Image 3) Obliteration of NLD was seen. Histopathology showed fragments of variable thickened lamellar bony trabeculae separated by fibro-connective tissue with scattered osteoclasts suggestive of fibrous dysplasia. The lesion was removed by lateral rhinotomy approach followed by dacryocystorhinostomy. Post-op period was uneventful, follow-up was done after two weeks and patient was relieved of her symptoms of epiphora.

Discussion
Fibrous dysplasia is a slow growing benign disease in which medullary bone is distorted and replaced by fibrous tissue proliferation most commonly seen in first and second decade of life. Out of the three forms i.e. mono-ostotic, poly-ostotic and McCune-Albright syndrome the mono-ostotic involvement is more common in the cranio-facial region. GNAS1 (guanine nucleotide binding protein, alpha stimulating activity polypeptide gene (20q13.2) post natal mutation is considered as the causative factor for fibrous dysplasia. Activation of undifferentiated mesenchymal cell cause arresting of bone development in woven phase converting into fibro-osseous tissue. Most common presentation of FD of maxilla is facial asymmetry, dental problems, headache, and paresthesia. Epiphora is an uncommon presentation due to
NLD or sac obstruction. Radiological pattern of cranio-facial FD has been described by Fries into three types. The first is pagetoid or ground glass with alternate areas of radiodensity and radiolucency with bone expansion usually seen in patients having symptoms for more than 15 years. The second type shows sclerotic pattern with bone expansion with homogenous radiodensity. The third type shows cyst like pattern with sclerotic border. Mostly the clinical and radiological findings are enough to diagnose a case of FD even without biopsy. Small asymptomatic lesion of cranio-maxillary facial skeleton are managed conservatively. Medical treatment includes bisphosphonates because they control bone erosion by inhibiting osteoclastic action but there is no uniformly accepted protocol for treatment, so the mainstay of therapy is surgical treatment. Surgical contouring or remodelling procedure to achieve acceptable aesthetics are the part of conservative surgery. Proximity of the lesion to the vital structures determines the extent of surgery. Endonasal endoscopic surgery can be done for limited disease in nasal and sinonasal areas. Treatment for NLD obstruction is dacryocystorhinostomy by endonasal endoscopic or external approach. In case of insufficient exposure external approach is suggested. In our patient dacryocystitis occurred from stenosis of NLD. Lesion was removed by lateral rhinotomy approach followed by dacryocystorhinostomy.

**Conclusion**

Fibrous dysplasia of maxilla is a benign disease which may result in nasolacrimal duct obstruction. In these cases conservative treatment like surgical contouring will never suffice but complete resection with dacryocystorhinostomy should be the approach.

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**References**
