Monostotic Fibrous Dysplasia of the Ethmoid Sinus : A Report of An Unusual case.

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Abstract : Fibrous dysplasia(FD) of bone is non neoplastic ,slowly progressive benign lesion that develops over the years. It is seen in long bones and affection of craniofacial bone is the second commonly involved site. However, FD of paranasal sinuses is rare. Involvement of Ethmoid sinus has been seldom documented. The radiological studies i.e. computed tomography (CT)helps in diagnosis, which is confirmed by microscopicl examination. Surgical intervention is the treatment of choice with rewarding results. We ,encountered a case of FD of ethmoid sinus extending into nasal cavity, leading to nasal obstruction.

Introduction : Fibrous dysplasia (FD) is a genetic sporadic disease of bone that may affect single (monostotic: MFD), or multiple bones (polyostotic: PFD). However, FD occurring in multiple adjacent craniofacial bones is regarded as monostotic (craniofacial FD)[1] WHO. It most frequently occurs in the long bone, and craniofacial bone is the second common site of involvement.[2] However, fibrous dysplasia of paranasal sinuses is rare.[3] There is paucity of reports of FD involving Ethmoid Sinus. [4,5,6]

Case Report : A female aged 17 years presented with complaints of gradual nasal obstruction for last 6 months, since last one month she had also experienced difficulty in breathing. The lesion was insidious in onset with gradual progression. On physical examination ,the patient had no facial deformity, nor eye movement limitation. There was no history of epistaxis. No history of bleeding, pain or ophthalmic problem. On examination the cervical lymph nodes were not palpable. On anterior rhinoscopy mass was found in left nasal cavity with displacement of nasal septum to opposite side. On posterior rhinoscopy the same lesion was visualized on left choanae.

Computed tomography (CT) scans on axial section [Figure 1A] and coronal view [Figure 1B] of paranasal sinuses revealed lobulated soft tissue density lesion occupying entire ethmoid sinus with extension into maxillary sinus and occupying left nasal cavity.It measured 6.4 x 4.8 x 4.6 cm (AP X ML X SI). Left ethmoid trabeculae and turbinates could not be identified separately .

Mild scalloping of medial wall of orbit was seen. However,visualized orbital structures and lamina papiracea appeaeared normal. Routine hematological investigations were non-contributory. Intraoperatively, Weber Fergusor incision was taken, flap refected and maxillary antrum accessed. The lesion in nasal cavity ,affected medial part of maxillary antrum and total mass lesion in ethmoid was removed. The patient made uneventful recovery was discharged on 6th day of operation.

Histopathological examination revealed the lesional tissue composed of trabeculae of immature bone in the form of curved, irregular shapes like “Chinese” letters and characteristic fishhook configurations. Most of these were not rimmed by osteoblasts. These were separated by collagen rich fibroblastic stroma.(Figure2).Based on histopathological and radiological findings final diagnosis of Fibrous Dysplasia was offered. Follw-up after 6 months showed total relief from difficulty in breathing and there was no evidence of recurrence.

Discussion : Fibrous dysplasia (FD) is a non-neoplastic lesion in which normal bone and marrow shows replacement by fibrous tissue and haphazardly distributed woven bone. Patients may present with affection of one bone (monostotic FD; MFD), multiple bones (polyostotic FD; PFD) or they may have features of McCune-Albright syndrome (MAS), which is the triad of PFD, café-au-lait cutaneous spots and endocrine disorders, including among others like precocious puberty [7]. Our case was in the category of monostotic FD of ethmoid sinus ,as there were no skin lesions nor any evidence of endocrinopathies.[1] Microscopically the close differential diagnosis, is ossifying fibroma which has islands of osteoid rimmed by osteoblasts form lamellar bone, and the stroma reveals a interlacing bundles and whorled arrangement of the collagen rich fibrous stroma these features are were absent in in our case.[8]

The patients of cranio-facial FD may show , facial deformity and sign and symptoms due to distortion of adjacent parts such as optic nerve, eye , nasal airway,

cranial nerve VII, middle ear ossicles, and teeth with gradual and insidious progression.[9]

Present case did not reveal any of these. The radiological and clinical findings correlated well in this case. However ,it must noted that the lesion shows characteristic "Ground Glass"appearance on CT scans on bone window.[1]

Fibrous dysplasia is by and large slowly progressive ,benign disorder however few cases of aggressive FD and very rarely development of sarcoma has been documented in the literature.[1,10]

The experience of this suggests FD must be entertained in the differential diagnosis for children with nasal obstruction coupled with typical imaging findings.

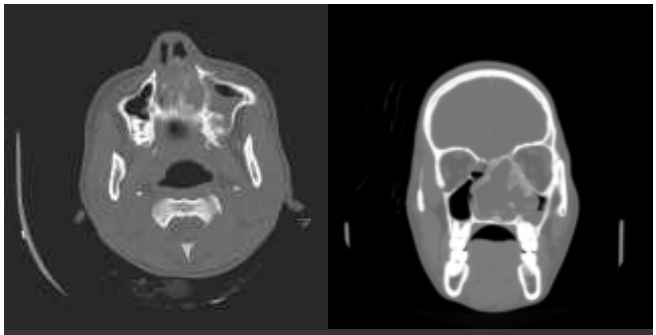

Figure 1a CT Scan axial view

Figure 1b

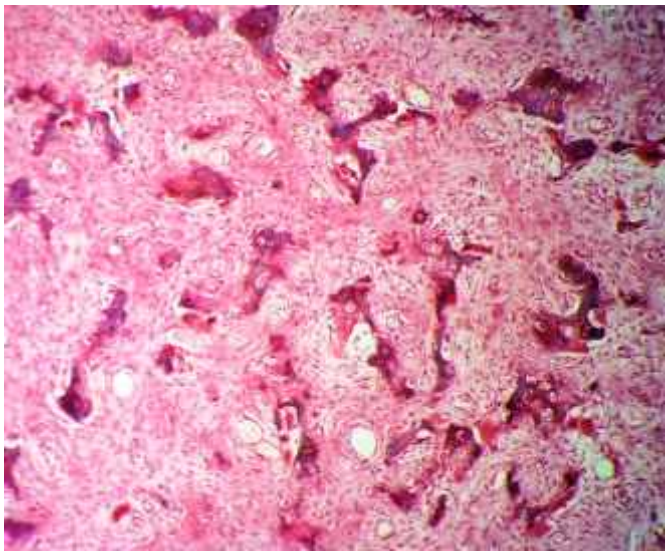

Figure 2: Photomicrograph showing trabeculae of woven bone, Chinese letters and characteristic fishhook configurations without osteoblastic rimming embedded in collagen rich fibrous stroma.

(H&E, x100)

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