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Case Report

Fibrous Dysplasia of Maxilla in a 10 Year Old Boy

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Summary:

We present the case of a surgically treated 10-year old boy with a diagnosis of fibrous dysplasia of maxilla. When first seen in the outpatient clinic of our department he had presented with pain and swelling over right cheek. After initial radiological diagnostics by computed tomography scans (CT-scans) the patient was treated operatively by radical excision of the tumour. The radiographs showed no invasion into adjacent tissue or bone.

Wide local excision was done by Caldwell Luc approach. The histopathology showed greyish white, smooth,shiny,irregularspecimen- diagnosed as fibrous dysplasia. Six monthly followup shows no recurrence at present.

Keywords: *Fibrous Dysplasia, Caldwell Luc, McCune-Albright syndrome, Chondroblastoma*

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Introduction:

Fibrous dysplasia is a benign fibro-osseous bone disease characterized by the replacement of bone with cellular fibrous tissue with foci of ossification.[1,2]

It is a hamartomatous condition related to mutations in the gene (GNAS I) encoding the alpha-subunit of stimulatory G-protein (Gs-alpha) that result in the increased production of cyclic adenosine monophosphate (cAMP), affecting the proliferation and differentiation of preosteoblasts.[1] In over 80% of cases, Fibrous Dysplasia affects only one bone and this form is called monostotic Fibrous Dysplasia, but multiple bones may also be involved -polyostotic Fibrous Dysplasia.[1]

Fibrous Dysplasia is more frequent in the maxilla than in the mandible.

Majority of monostotic lesions are asymptomatic and found incidentally on radiographic imaging.[3] In reality, the polyostotic form manifests earlier, usually in children younger than 10 years of age; has a more serious prognosis.[4] Three percent of polyostotic cases are associated with cafe-au-lait spots and a hyper functional endocrine state characteristic of McCune-Albright syndrome.[5]

The diagnosis of Fibrous Dysplasia is usually based on clinical, radiographic, and histopathologic features.[6] Clinically, the most common presentation is swelling[3,7]; other manifestations include localized pain, deformity, fractures, and reduced vision or hearing.[3]

Case description:

10-year-old male presented to outpatient department with complaints of swelling over right cheek region associated with dull aching pain while chewing food over a period of 6 months which progressed quickly to current size in the past 2 months.

On local examination, there was a diffuse swelling over right cheek region which was firm to hard in consistency, non-fluctuant, non-tender, fixed to underlying surface. The border of swelling was smooth and regular.

Oropharyngeal examination was normal.

Fine needle aspiration cytology of the swelling was done and reported as Giant cell reparative granuloma of right maxilla.

Extrasinus mass involving infratemporal fossa

Mass abutting internal maxillary artery

MRI Face was done which stated well defined, lobulated, heterogeneously enhancing mass lesion from right maxilla along its right posterior alveolar margin and posterolateral wall of right maxillary sinus extending into right masticator space. Extra sinus mass which is involving the infratemporal fossa– either odontogenic lesion or ameloblastoma

CT PNS with contrast was done which shows a well-defined expansile lesion of approx. size 3.9*3*3.7cminvolving posterolateral wall of right maxillarysinus.It showed presence of multiple septa within, forming multiple compartments.These compartments are filled with isodensefluid/ soft tissue. No evidence of obviouscalcification or invasion into adjacent muscles or bone.Features suggestive of Chondroblastoma/Fibrous dysplasia.

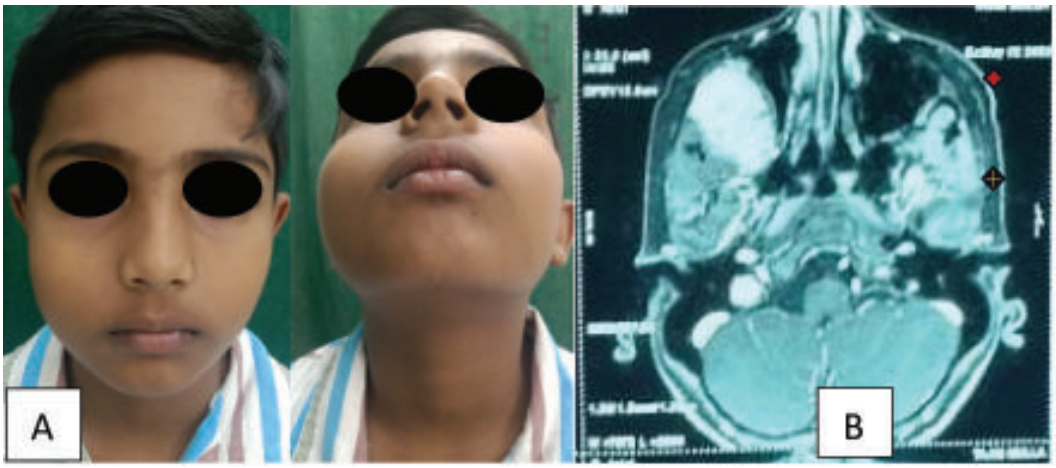


Figure 1 *A) Preoperative clinical picture of patientB)MRI face showing mass in right maxillary region*

Extended Caldwell Luc operation was planned.

The maxillary vestibular incision was given from the right upper canine to the upper second molar of the right side, the flap of mucosa and periosteum was elevated and dissected to expose the anterior wall of sinus where the mass was found. Underlying bone was drilled.

Mass was removed in piecemeal and sent for histopathological examination. The cavity was packed with antibiotic soaked wick which was removed on 2nd postoperative day and there was postoperative swelling which subsided over a period of 5 days.

Histopathological report showed irregular, greyish white, shiny soft to firm, gritty to cut (cartilage) tissue. Histological findings were consistent with Fibrous dysplasia of maxilla.

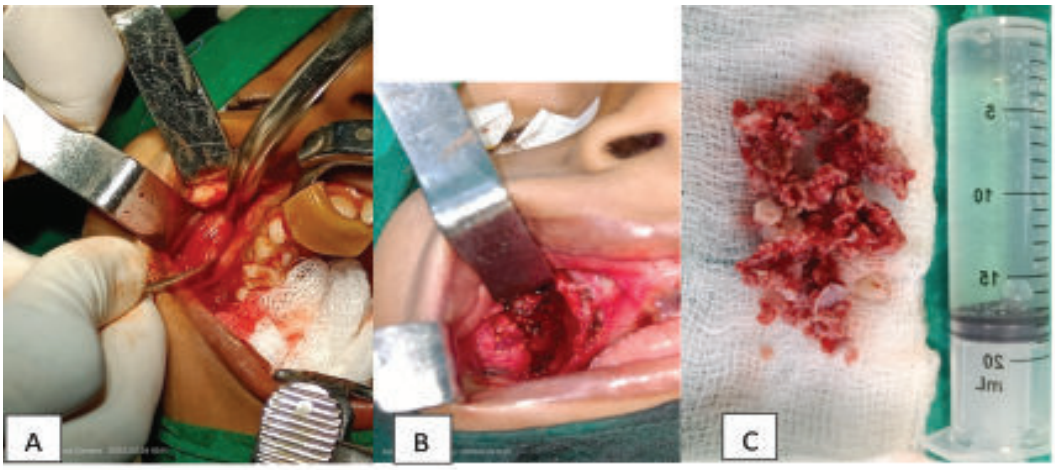
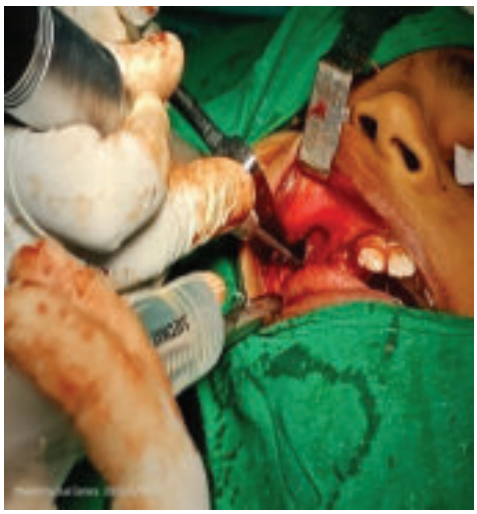


Figure 2

A) Caldwell luc approach; subperiosteal flap being raised
B) Lesion seen above the plane of maxilla.
C) Lesion removed in piecemeal.

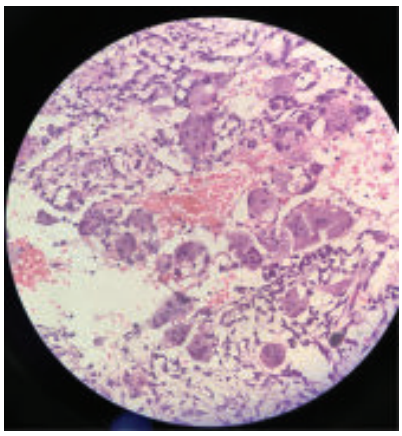


D) Drilling of the underlying bone after removing the mass

Differential diagnosis includes:

1) Chondroblastoma
2) Fibrous Dysplasia
3) Osteoid osteoma

Histopathological slide here



Discussion:

Fibrous dysplasia is a common, benign condition that differs from other fibro-osseous lesions.[8] Before the biopsy or surgical specimen is available, a diagnosis of craniomaxillofacial fibrous dysplasia (CFD) can be made with considerable accuracy based on the patient's medical history and radiographic features. More frequently has an impact on the maxilla than the mandible .[9]

Sudden increase in the level of alkaline phosphatase is one of the symptoms for malignant transformation and for that reason its amount should be periodically observed.[10]

As a result, its levels in these patients should be routinely checked. As a result of the patient's elevated ALP levels, an evaluation of her ALP should be done every six months [11]

There are several treatments, including observation, medication, surgery, and aggressive excision and reconstruction. Treatment with bisphosphonates was made possible by understanding the disease's pathophysiology. They prevent osteoclastic activity from limiting bone degradation. [12]

Even though there are no universally recognized standards for treating fibrous dysplasia, surgical therapy is still the backbone of care [13]. It aims to restore normal facial aesthetics.

Indications of surgery are- pain, vision loss, facial deformity .

As compared to frontoethmoid bone lesion in Otorhinolaryngology, involvement of maxilla is rare.

However, there is a 15-20% chance of recurrence, particularly throughout the growth phase[14].

Conclusion:

Conservative management is usually the mainstay of therapy in asymptomatic cases of fibrous dysplasia. In patients fulfilling criteria for surgical management, craniofacial reconstruction offers a viable option in the surgeon's armamentarium, providing good functional and cosmetic outcomes.

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