



CASE REPORT OF A RARE CASE OF MAXILLARY AMELOBLASTOMA DEVENDIRAN R

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Abstract : we present a rare case of MAXILLARY AMELOBLASTOMA , a rare tumour of odontogenic origin with an incidence of 1 in 1 lakh population. we describe the clinical features and management with relevant clinical photos.

Keyword : ameloblastoma, odontogenic tumour

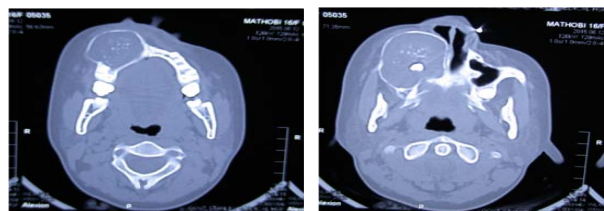
Ameloblastoma : a very rare tumour of odontogenic origin, with an incidence of 1 in 1 lakh population, ameloblastoma can cause slowly progressive deformity of face. This is a case report of 16 yrs old girl with rare maxillary ameloblastoma. This 16 yrs old girl from orisa , came with complaints of right sided cheek swelling for last 2 yrs, insidious onset, slowly progressive in size, no associated with pain / trauma. She also complains the right sided nasal obstruction for last 6 month, progressively increasing obstruction, no aggravating / relieving factors for both. No h/o nasal discharge/ bleed / loss of smell/sneezing/ facial pain/post nasal discharge No h/o difficulty in swallowing / ear complaints On examination, an ill defined bony hard swelling with smooth surface , without signs of tenderness/ warmth seen over right cheek. intra oral examination was normal. On anterior rhinoscopy, inferior turbinate touching midline septum.



Fig. showing anterior view of the patient



Fig . showing intra oral view with no evidence of tumour inside Patient was investigated with CT PNS which showed : Well defined radiolucent lytic lesion of right maxilla occupying maxillary sinus, with radiodense tooth like structure inside the lesion. Advised biopsy and clinical correlataion.



Figures showing axial view PNS showing the well defined unicystic tumour

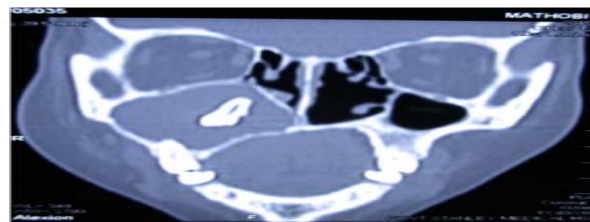
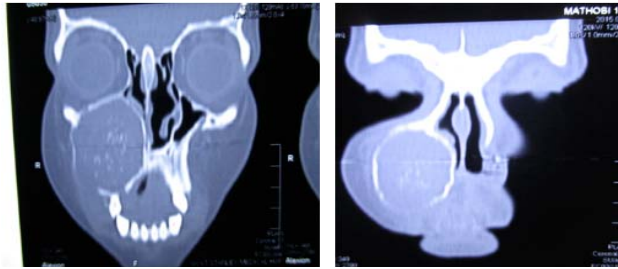
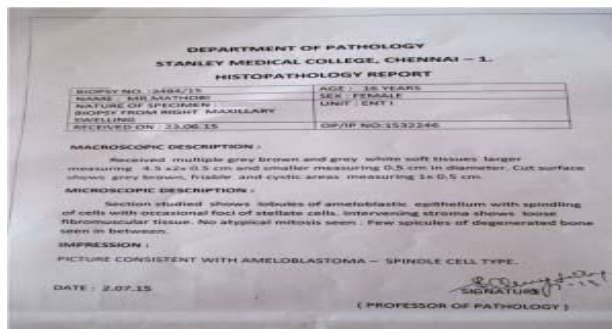


Fig shows unerupted teeth inside the tumour



Figures showing coronal views of the tumour Biopsy was done by Caldwell luc procedure under local anaesthesia and sent for frozen section biopsy. Report came as:



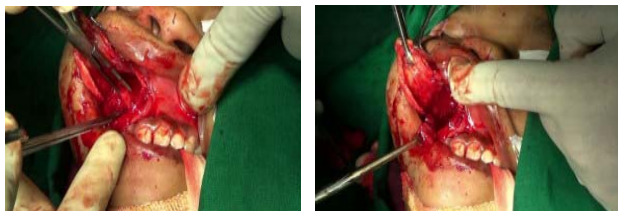
After discussing with the parents of the patient and considering age of that patient we decided to do enucleation and curettage through Caldwell luc approach and endoscopic medial maxillectomy under general anesthesia.



Fig shows Caldwell luc incision



Fig. thinned anterior wall of maxilla incised and retracted laterally as a flap and anterior cystic wall is being cut



Figures show removal of unerupted floating teeth

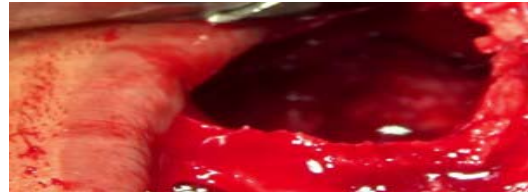


Fig . shows inside of the maxillary sinus with cyst wall

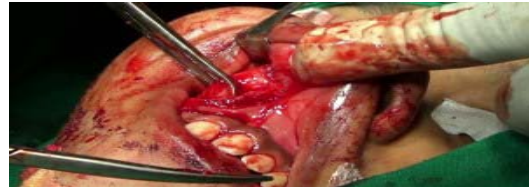


Fig. cyst wall being removed from the medial wall of maxillary sinus



Fig. cyst wall being completely stripped from the maxillary sinus



Fig. cyst wall being cut and released completely By using endoscope intra nasally, inferior turbinectomy and wide middle meatal antrostomy done

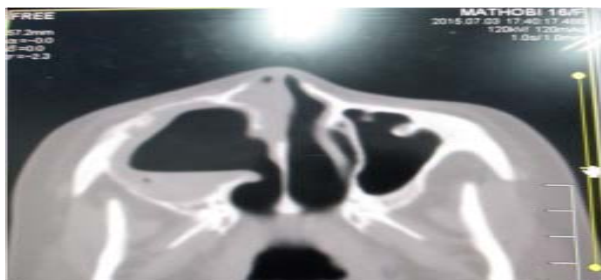


Fig. shows post 3rd post operative day status with swelling reduced



Figure shows the patient on her 10 th post operative day without any obvious deformity

Post operative CT SCANS:



Figures showing axial cuts of post operative status



Figures showing post operative coronal view of ct scans

Discussion :

Ameloblastoma is the second most common benign odontogenic tumour. This rare tumour constitute 1-3 % of all cysts and tumours around the jaw. This locally aggressive benign tumour, has the malignant potential with high recurrence rate following surgery. This was previously known as adamantinoma of jaw. This tumor originates from the odontogenic epithelium which forms the future enamel of teeth. Mostly occurs in mandible (80%, angle of mandible the commonest site), maxilla (20%, posterior maxilla in molar region more common). Approximately 20% cases are associated with dentigerous cysts and unerupted teeth.

Histology :

There are 3 clinical variants of ameloblastoma .

- 1.unicystic
- 2.multicystic
- 3.solid or peripheral type

Histopathology will show cells that have the tendency to move the nucleus away from the basement membrane. This process is referred to as "Reverse Polarization". The follicular type will have outer arrangement of columnar or palisaded ameloblast like cells and inner zone of triangular shaped cells resembling stellate reticulum in bell stage. The central cells sometimes degenerate to form central microcysts. The plexiform type has epithelium that proliferates in a "Fish Net Pattern". The plexiform ameloblastoma shows epithelium proliferating in a 'cord like fashion', hence the name 'plexiform'. There are layers of cells in between the proliferating epithelium with a well-formed desmosomal junctions, simulating spindle cell layers.

Histological variants:

Based on histopathology studies, they are divided into 6 variant types:

- 1.follicular (30%)
- 2.plexiform (30%)
- 3.acanthomatous (very rare)
- 4.granular
- 5.desmoplastic
- 6.basilar

Epidemiology:

Annual incidence 1in one lakh. Affects male and female equally. Mostly occurs in 3 or 4 th decade.

Clinical features:

Mostly asymptomatic .early lesions are incidentally found by radiological studies for other reasons. In symptomatic patients, it will be a slow growing painless hard lesion involving mandible or maxilla. Sometimes it may associated with pain and paresthesia of involved regions. very rarely it can csuse malocclusion of teeth , nasal / ocular complaints. On examination , it will be hard , non tender .sometimes on palpation there will be egg shell crackling , a diagnostically important feature.

Investigations:

CT scan :

Classically this will be seen as a multiloculated (80%), expansile lytic radiolucent lesion with well defined borders with no matrix calcifications with thinning of bony cortices. Sometimes unerupted teeth may be seen inside the cyst. Radiographically, unicystic variant is divided into three subtypes

1. Simple : no nodule inside the cyst (best prognosis)
2. Luminal : single nodule projecting into the cyst
3. Mural : multiple nodules in the wall of the cyst (rare type)

MRI scan :

There will be mixed solid and cystic patterns with thick irregular wall often papillary structures projecting into the

lesion .

Treatment :

This is the more controversial part of these tumors. Though radio and chemotherapy have been tried, surgical en bloc resection is the most accepted treatment modality. Enucleation and local curettage can be done but associated with high incidence of recurrence (45%). Treatment algorithm mainly depends on the following factors.

1. Size of the lesion
2. Anatomical Location
3. Histological type.

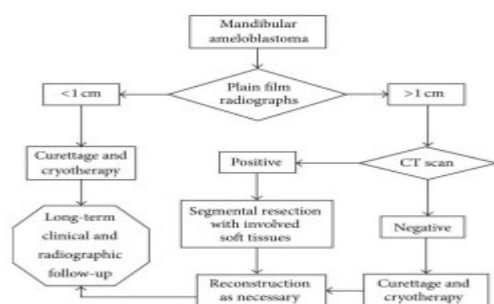


Fig. treatment algorithm of mandibular ameloblastoma (According to a retrospective study done in Northern California for both primary management and treatment of recurrences for mandibular ameloblastoma)

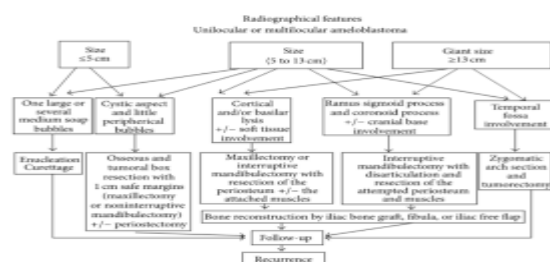


Fig. Pitie Salpeterie hospital- therapeutic algorithm for ameloblastomas

Group I confined to maxilla without involvement of the orbital floor	Partial maxillectomy
Group II involving orbital floor but not involving premaxillary area	Total maxillectomy
Group III involving orbital contents	Total maxillectomy + orbital exenteration
Group IV involving skull base	Total maxillectomy + orbital exenteration + skull base resection

Fig. treatment algorithm of maxillary ameloblastoma . Regular follow for upto 10 years may be necessary as there is a chance late recurrence.

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