

# RARE PATHOLOGIES OF MAXILLOFACIAL REGION

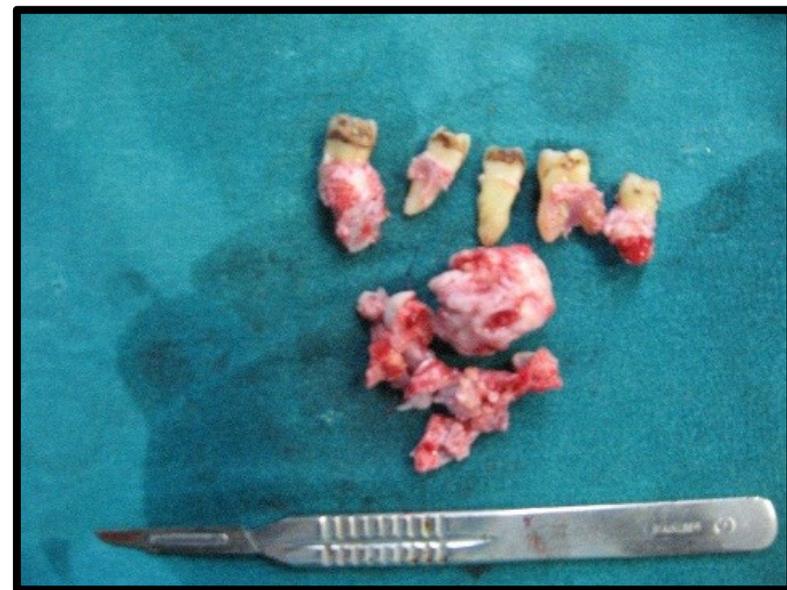
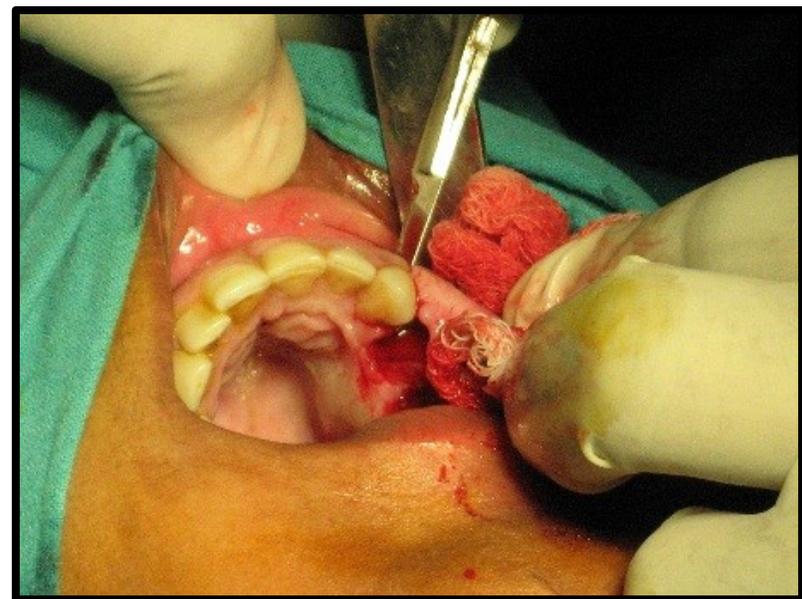
**Dr. RAMAKANT DANDRIYAL (HOD & PROF.)**

# CEMENTO-OSSIFYING FIBROMA

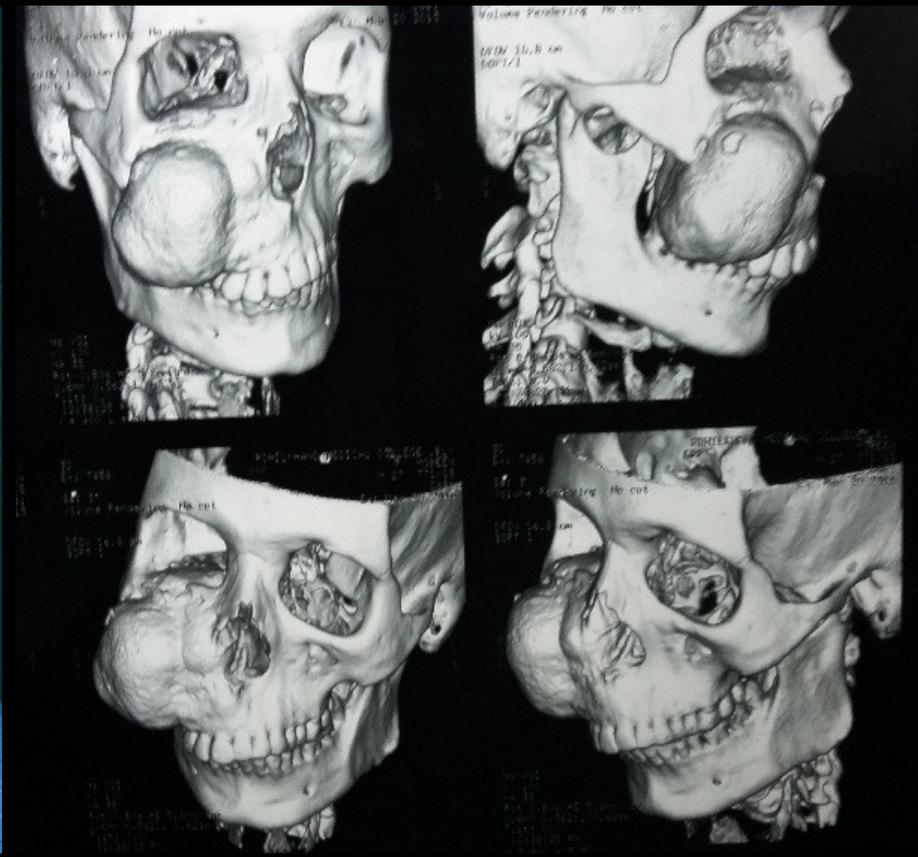
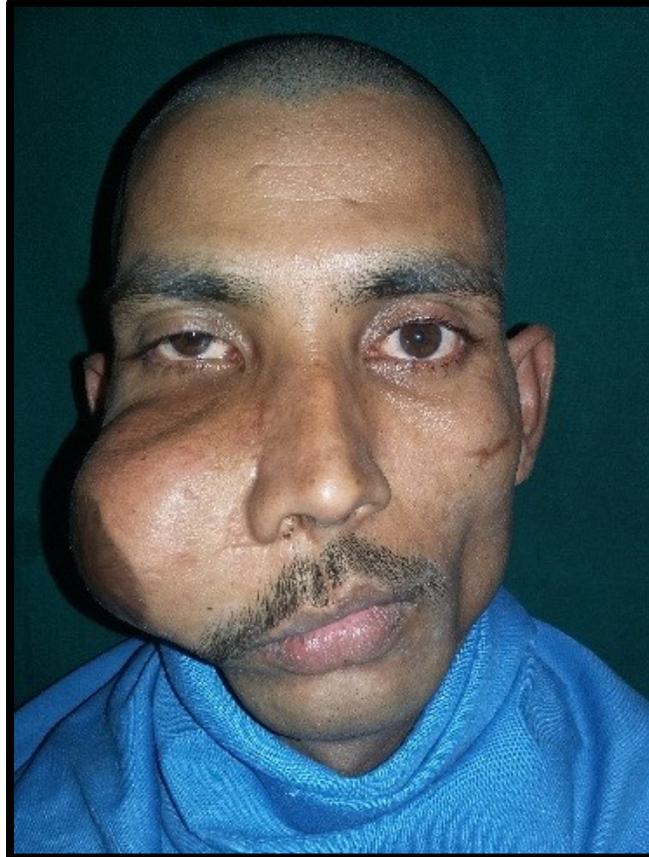
- Cemento-ossifying fibroma is a benign fibro-osseous tumor belonging to the same category as fibrous dysplasia and cement-ossifying dysplasia.
- It is most commonly seen between the third and fourth decades of life.
- is more frequent in women than in men .
- The most common location is the mandible, with 70-90% of all cases.
- It is a non-odontogenic tumors derived from the mesenchymal blast cells of the periodontal ligament, with a potential to for fibrous tissue, cement and bone, or a combination of such elements
- Clinically, these tumors manifest as a slow-growing intrabony mass that is normally well delimited and asymptomatic – though over time the lesion may become large enough to cause facial deformation.
- Two basic patterns have been defined: one characterized by the presence of a unilocular or multilocular radio transparent image, and another showing mixed density due to a variable internal amount of radiopaque material
- Histologically, these tumors are composed of well vascularized fibrocellular tissue with the capacity to form immature bone trabeculae and cementoid formations.
- Treatment comprises surgical resection of the lesion with enucleation and curettage of the bone bed.

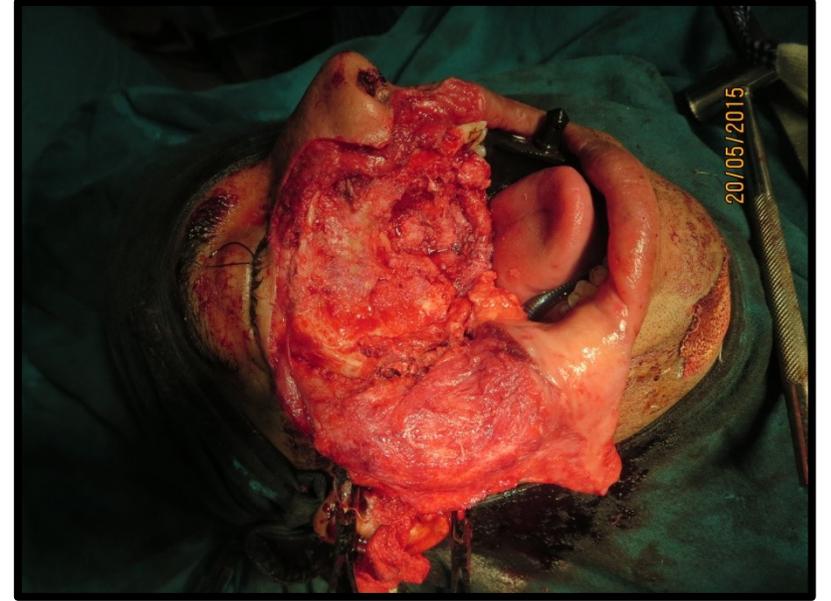
# CASE REPORT-1 COF





# CASE REPORT-2 COF







# CHONDROSARCOMA

- Chondrosarcoma is a malignant tumour wherein the tumour cells form cartilage.
- It usually involves the pelvic girdle, chest wall and scapula.
- It does not involve the jaws commonly and if it does, it affects the maxilla rather than the mandible.
- In a series at Mayo clinic chondrosarcoma involved the jaw in only 3% of cases (Dahlin and Unni, 1986)
- The molar region of mandible is affected more compared to ramus, condyle, coronoid process or symphysis. (Fred et al., 1991 and Takayuki et al., 2006).
- Chondrosarcoma are slow growing tumour with a tendency for local recurrence after surgery.
- However with recurrence they exhibit rapid and aggressive growth.
- Chondrosarcomas of head and neck present without pain.
- Male to female ratio of 2:1.(Brad et al.,2004)
- Age ranges- 30 to 50 years of life(Fred et al. 1991).



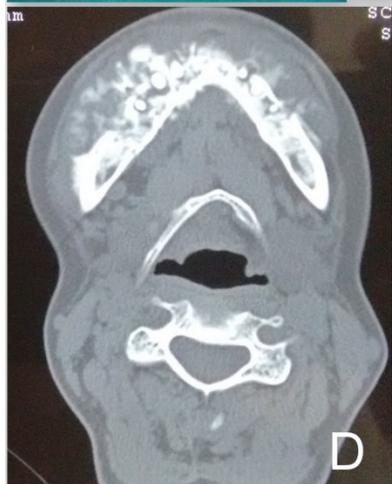
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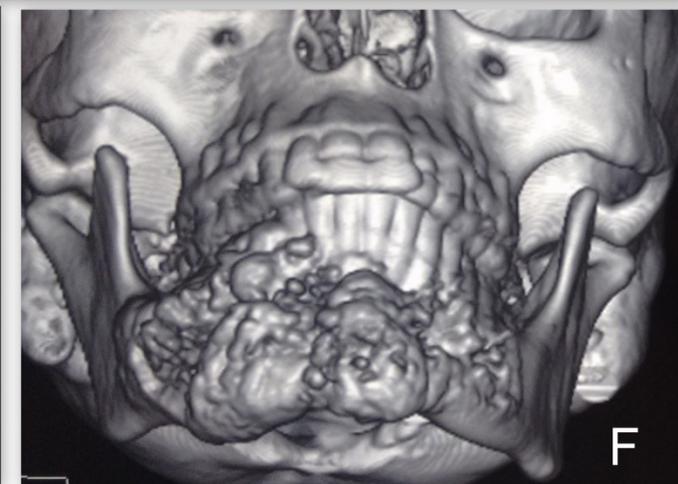
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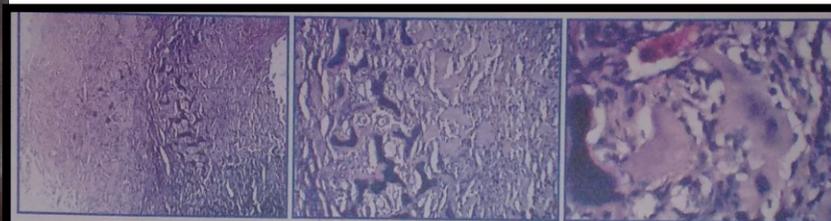


Fig. 1

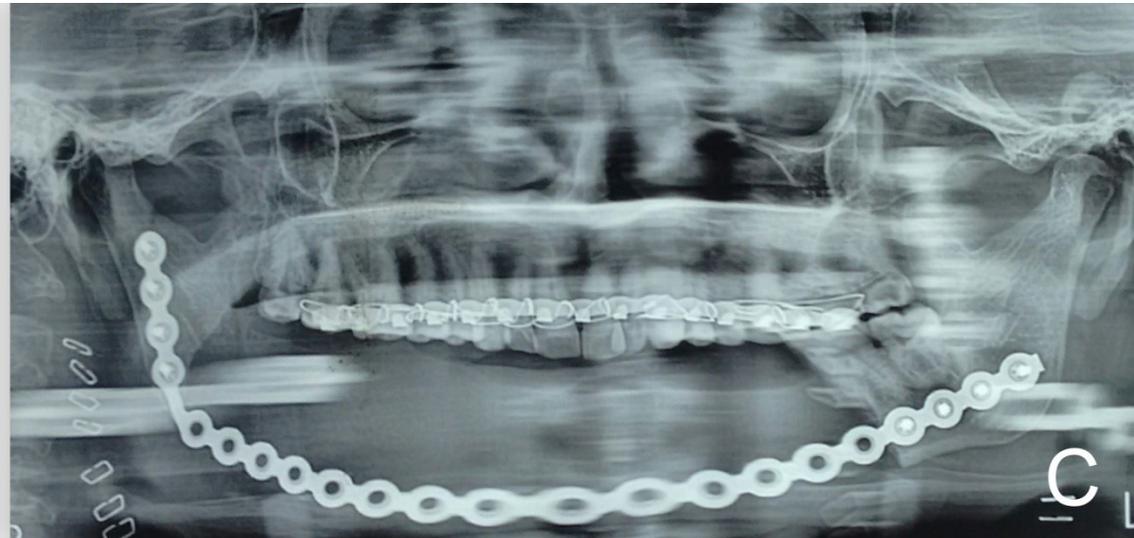
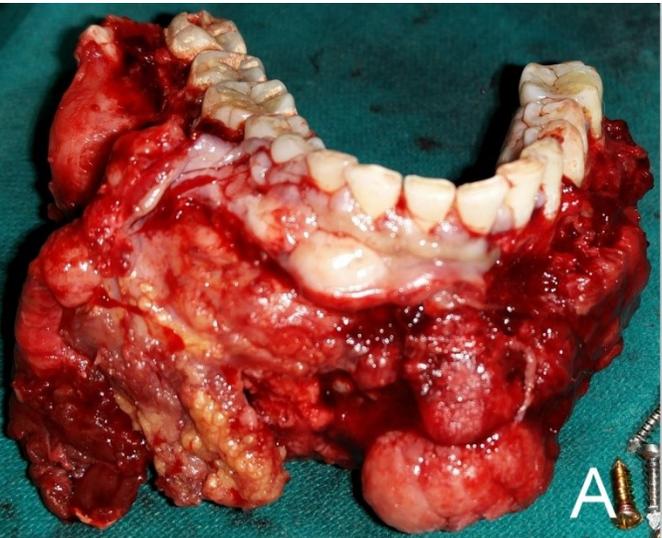
Fig 2

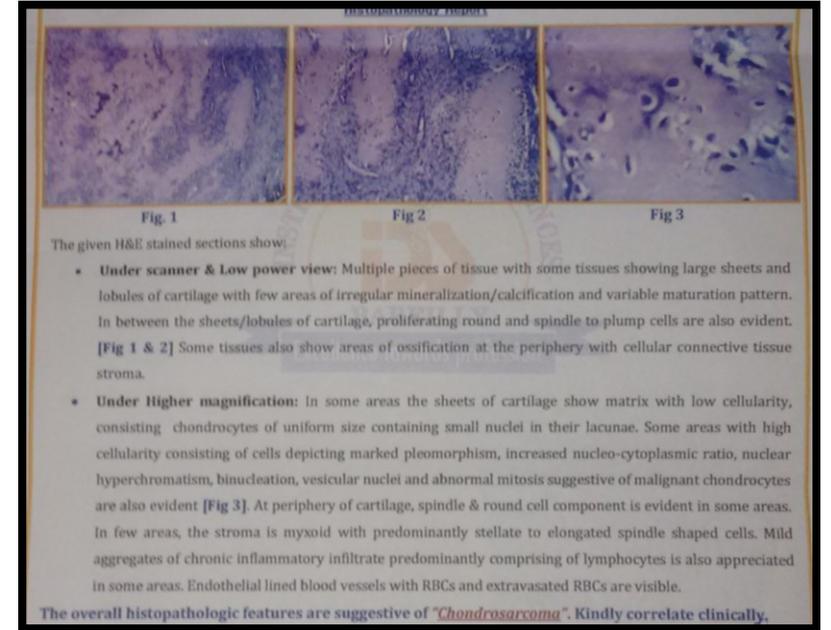
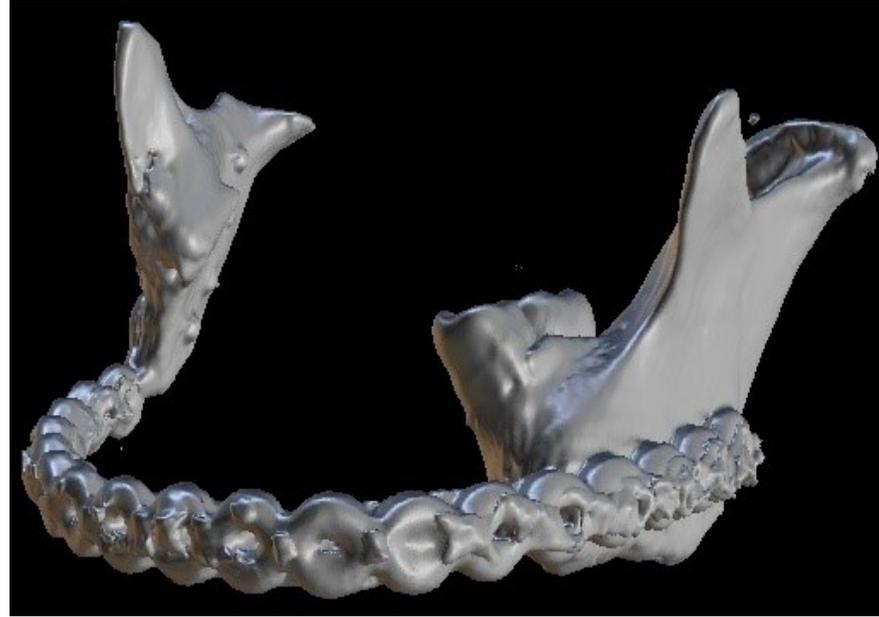
Fig 3

The given H&E stained sections show:

- **Under scanner and Low power view:** two pieces of tissue with irregular fragments of osseous tissue (in different stages of maturation) embedded in **fibro-cellular connective tissue stroma**. [Fig 1 & 2]
- **Under Higher magnification:** Some osseous fragments show peripheral rimming of osteoblasts and centrally entrapped osteocytes. Some of the bony fragments exhibit patchy basophilia representing mineralization. The connective tissue stroma is hypercellular and comprised of bundles of collagen fibres with predominantly plump shaped fibroblasts. Endothelial lined blood vessels with RBCs are also seen. [Fig 3]

The overall histopathologic features are suggestive of "**Ossifying Fibroma**". Kindly correlate clinically.





The given H&E stained sections show:

- **Under scanner & Low power view:** Multiple pieces of tissue with some tissues showing large sheets and lobules of cartilage with few areas of irregular mineralization/calcification and variable maturation pattern. In between the sheets/lobules of cartilage, proliferating round and spindle to plump cells are also evident. [Fig 1 & 2] Some tissues also show areas of ossification at the periphery with cellular connective tissue stroma.
- **Under Higher magnification:** In some areas the sheets of cartilage show matrix with low cellularity, consisting chondrocytes of uniform size containing small nuclei in their lacunae. Some areas with high cellularity consisting of cells depicting marked pleomorphism, increased nucleo-cytoplasmic ratio, nuclear hyperchromatism, binucleation, vesicular nuclei and abnormal mitosis suggestive of malignant chondrocytes are also evident [Fig 3]. At periphery of cartilage, spindle & round cell component is evident in some areas. In few areas, the stroma is myxoid with predominantly stellate to elongated spindle shaped cells. Mild aggregates of chronic inflammatory infiltrate predominantly comprising of lymphocytes is also appreciated in some areas. Endothelial lined blood vessels with RBCs and extravasated RBCs are visible.

The overall histopathologic features are suggestive of "**Chondrosarcoma**". Kindly correlate clinically.

# OSTEOCHONDROMA

Osteochondroma (OC) is defined as an osteo-cartilagenous exostosis with cartilage capped exophytic lesion that arises from the bone cortex.

It is one of the most common benign tumor of the axial skeleton, but is rarely associated with the facial bones.

It has been described in the head, cranial base, jaw, maxillary sinuses, condyle, ramus, body, coronoid process and symphyseal mandibular region.

The embryonic development of the temporo mandibular joint (TMJ), by the endochondral ossification, makes this area the most frequent facial site for OC.

It represents approximately 35% to 50% of all benign tumors, and 8% to 15% of all primary bone tumors.

They are easily identified as they lead to facial asymmetry and malocclusion.

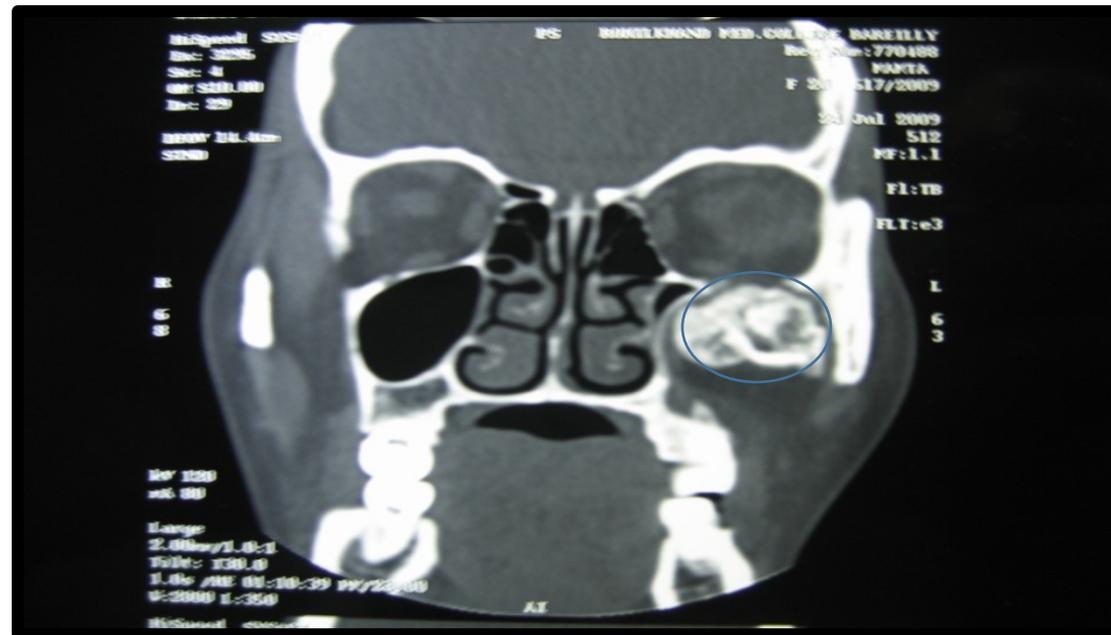
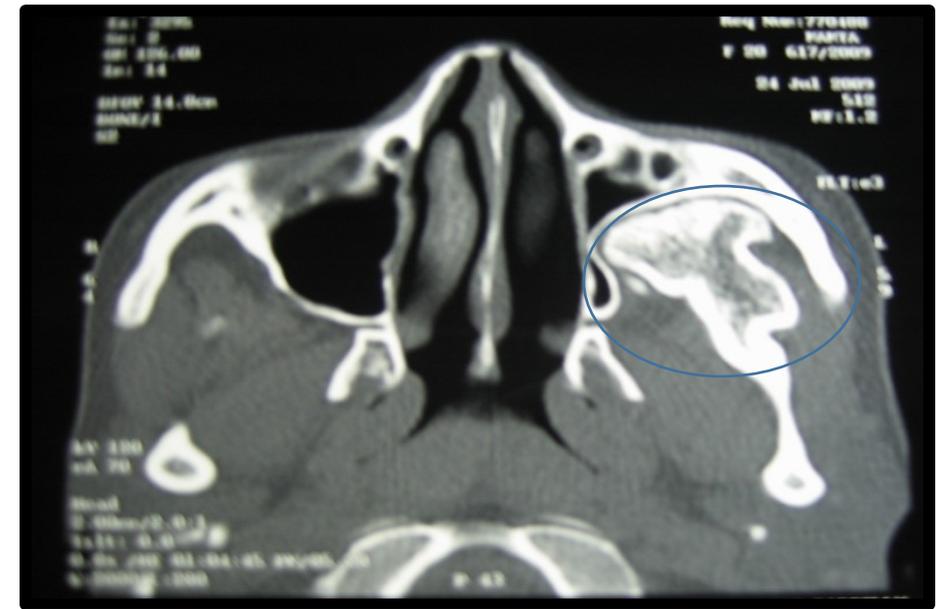
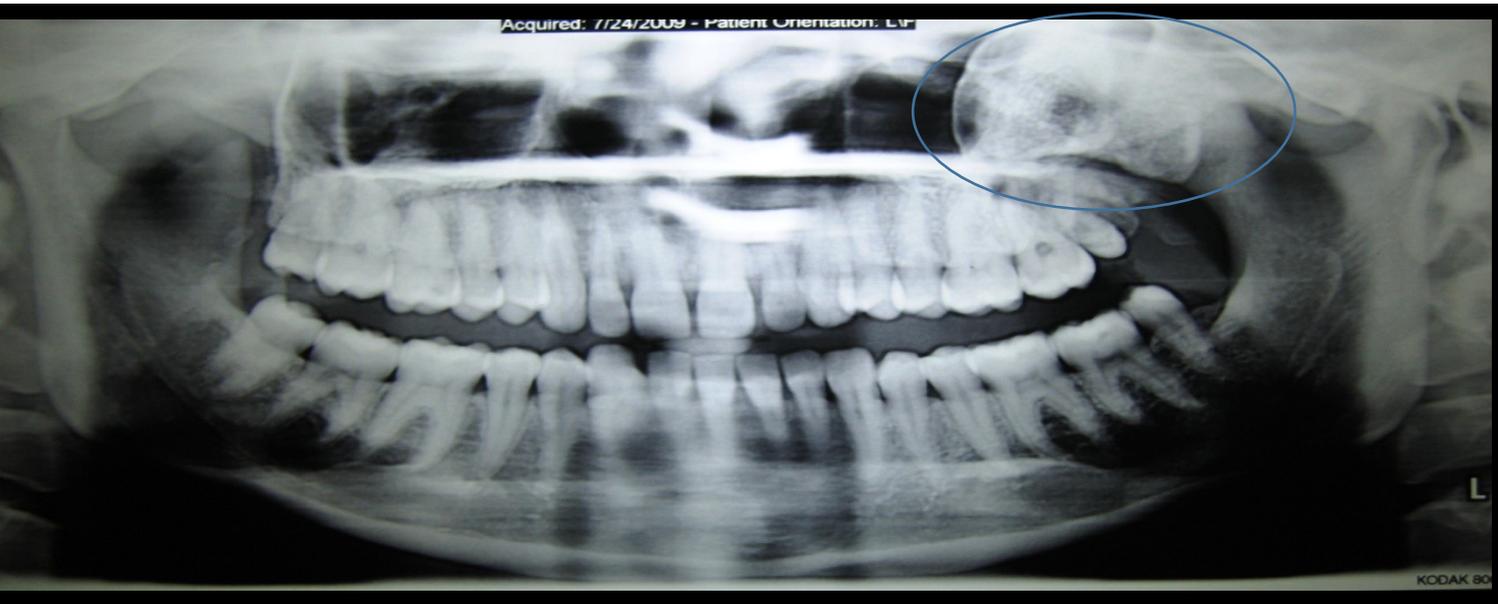


25 April 2020



40th AOMSI AMR

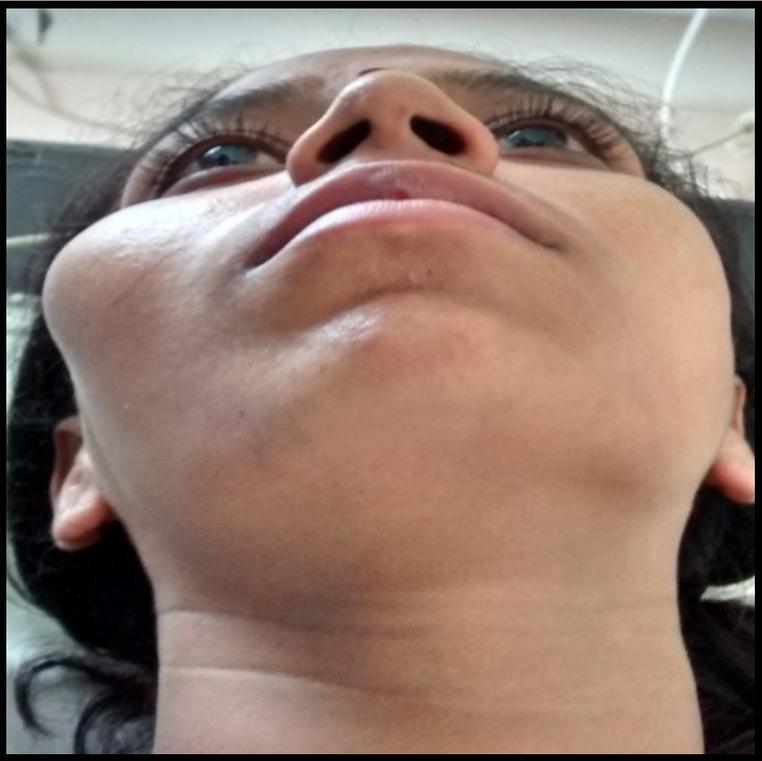
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# CASE REPORT-2 OSTEOCHONDROMA



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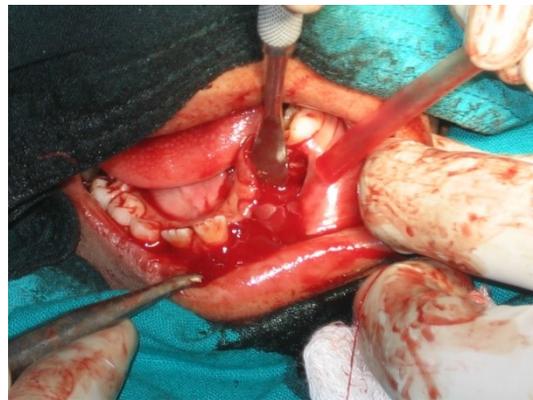
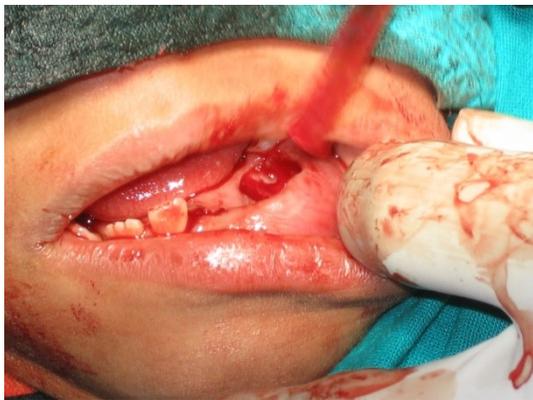




# ANEURYSMAL BONE CYST

- Aneurysmal bone cyst (ABC) is rare benign lesions of bone which are infrequent in craniofacial skeleton.
- ABC's are characterized by rapid growth pattern with resultant bony expansion and facial asymmetry.
- ABC is a benign cystic lesion of bone, composed of blood-filled spaces separated by connective tissue septa containing fibroblasts, osteoclast-type giant cells and reactive woven bone.
- Fifty percent of ABCs arise in the long bones and 20% in the vertebral column. It accounts for 1.5% of the nonodontogenic, nonepithelial cysts of the mandible.
- It is found more frequently in the mandible than the maxilla (3:1) with preponderance for the body, ramus and angle of the mandible.
- It affects young persons under 20 years of age with no gender predilection.
- ABC can be classified into three types. Conventional or vascular type (95%) manifests as a rapidly growing, expansive, destructive lesion causing cortical perforation and soft tissue invasion. The solid type (5%) may present as a small asymptomatic lesion first noticed as radiolucency on a routine radiograph or as a small swelling. A third form or mixed variant demonstrates features of both the vascular and solid types. It may be a transitory phase of the lesion because sudden activation or rapid enlargement of stable lesions has been reported.
- Treatment consisted of surgical curettage of the lesion. A one year follow-up showed restoration of facial symmetry and complete healing of the involved site

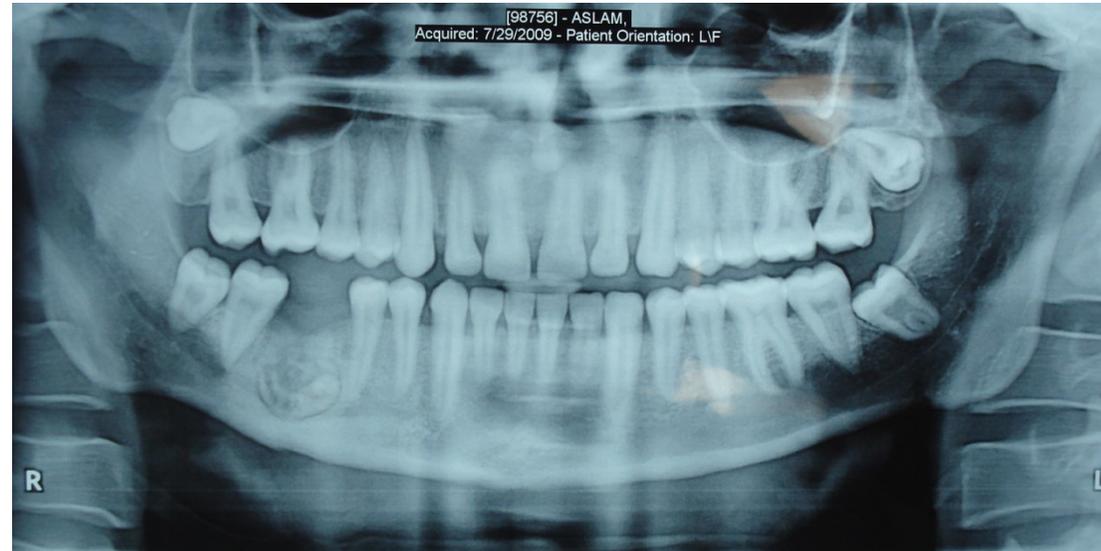
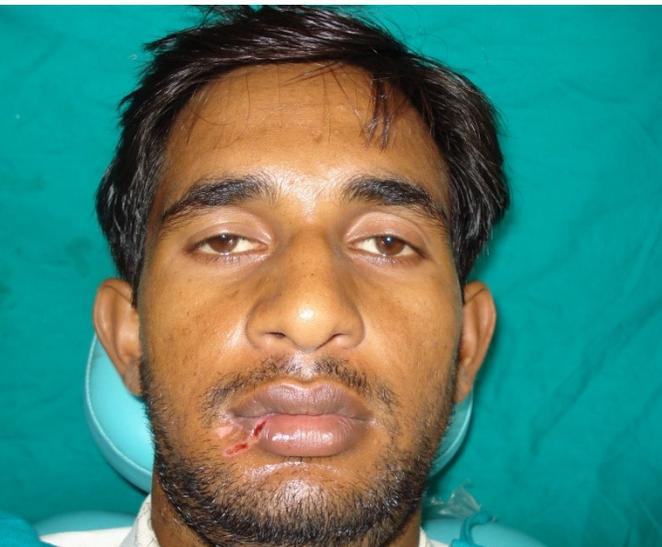


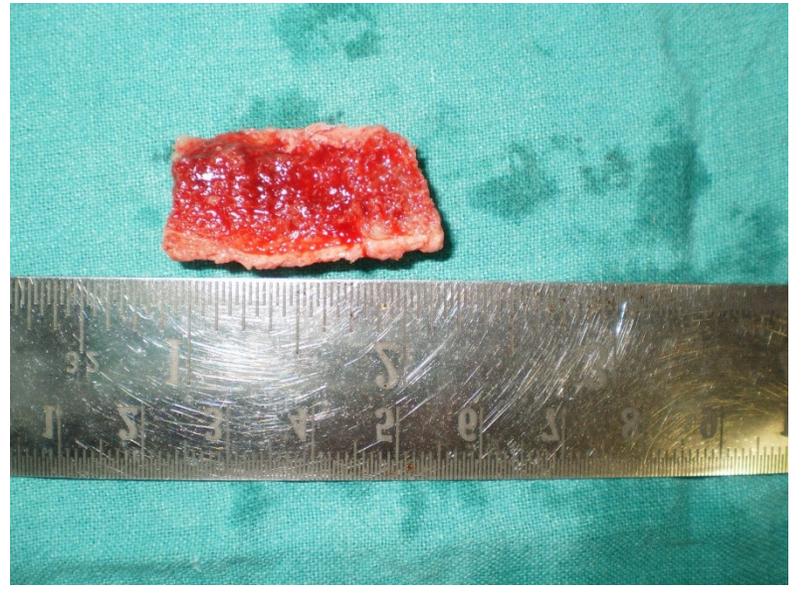
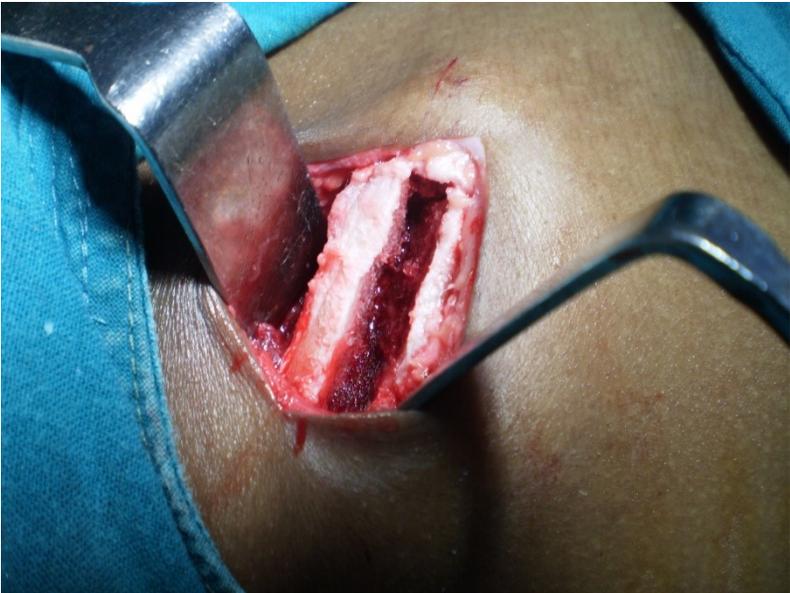




# FLORID CEMENTO-OSSEOUS DYSPLASIA

- is a type of fibro-osseous lesion
- FCOD is a very rare condition presenting in the jaws
- represents a reactive process in which normal bone is replaced by poorly cellularized cementum-like materials and cellular fibrous connective tissues.
- It is strictly localized to the tooth-bearing or edentulous areas, often occurring bilaterally with symmetric involvements.
- FCOD may be completely asymptomatic
- Found on routine radiographic examination, or may present with dull pain, alveolar sinus tract, and exposure of avascular bone into the oral cavity due to secondary infection.
- Rarely are these lesions expansile.





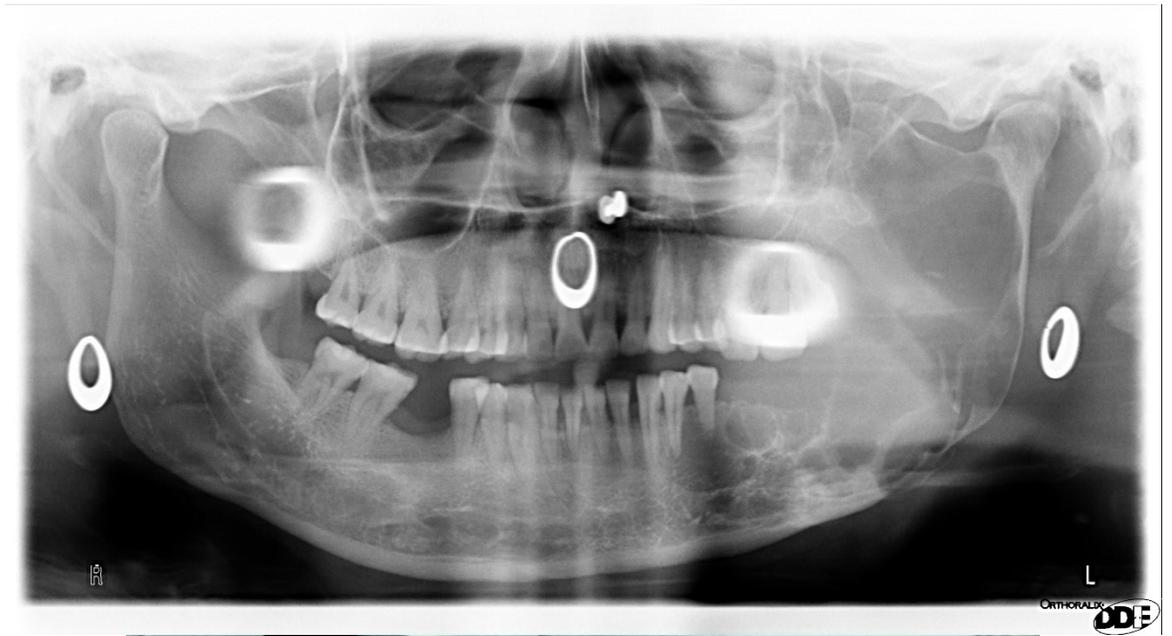


# THE CALCIFYING ODOTOGENIC CYST OR THE GORLIN CYST

- Is a benign odontogenic tumor of cystic type most likely to affect the anterior areas of the jaws.
- It is most common in people in their second to third decades but can be seen at almost any age.
- On radiographs, the calcifying odontogenic cyst appears as a unilocular radiolucency.
- In one-third of cases, an impacted tooth is involved.
- Microscopically, there are many cells that are described as "ghost cells", enlarged eosinophilic epithelial cells without nuclei.
- Praetorius (1981) proposed a classification for grouping CEOC as Type I (cystic type) and Type II (neoplastic type [dentinogenic ghost cell tumor])
- He further sub-divided the cystic variant (Type I) into three different types:
  - (a) Simple unicystic type,
  - (b) Odontome-producing type, and
  - (c) Ameloblastomatous proliferating type.

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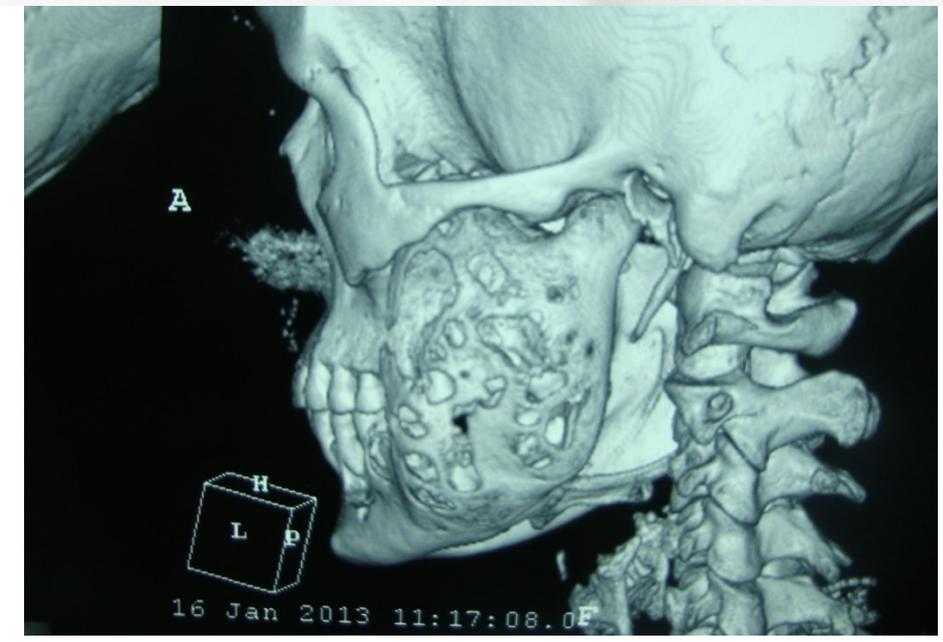
INSTITUTE OF DENTAL SCIENCE BAREILLY-243001 (U.P.) DEPARTMENT OF ORAL PATHOLOGY AND MICROBIOLOGY		TO BE FILLED BY THE RECEIVER	
HISTOPATHOLOGY REPORT		SIGNATURE	NAME
NAME: Mr./Miss./Ms. <i>Kapila</i>	AGE: <i>38y</i>	SEX: <input checked="" type="checkbox"/> MALE <input type="checkbox"/> FEMALE	OPD NO: <i>DS 1150</i>
BIOPSY No: <i>4-05/13</i>	PREVIOUS BIOPSY NO: <i>-</i>	PREVIOUS CYTOLOGY NO: <i>-</i>	DEPARTMENT: <i>DMFS</i>
REFERRED BY: <i>Dr. Bannant</i>			

Grossing:  
Received multiple pieces of tissue, soft in consistency white to brown in color.

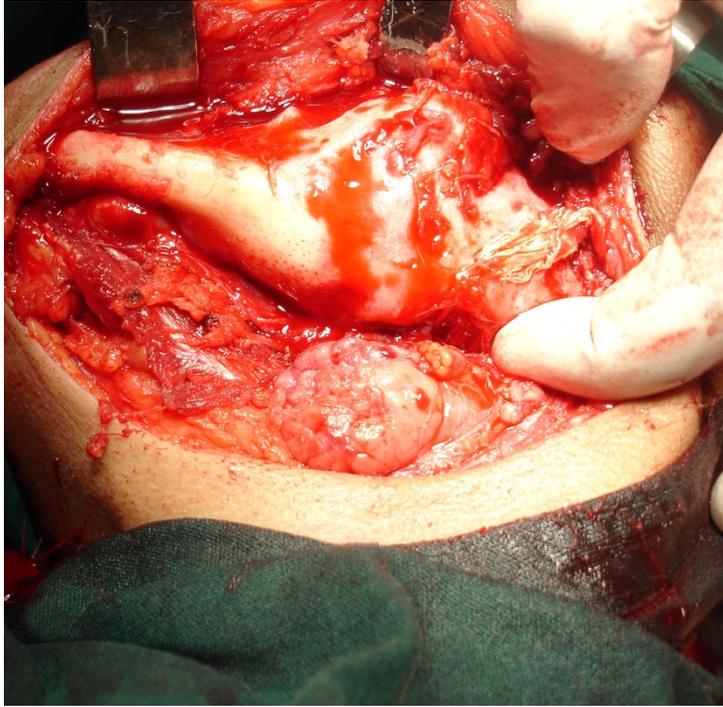
Histopathology:  
Microscopic features show cystic tissue, with epithelium overlying normal connective tissue. Epithelium consists of tall columnar cells with reverse polarity of the nucleus. Stellate reticulum cells are seen with areas of squamous metaplasia are present. Few gland cells are visible. Stroma show endothelial lined blood vessels and inflammatory component.

Features suggestive of  
'Calcifying Epithelial Cyst (Type I)'

Signature of HOD: *[Signature]*  
Signature of Oral Pathologist: *[Signature]*

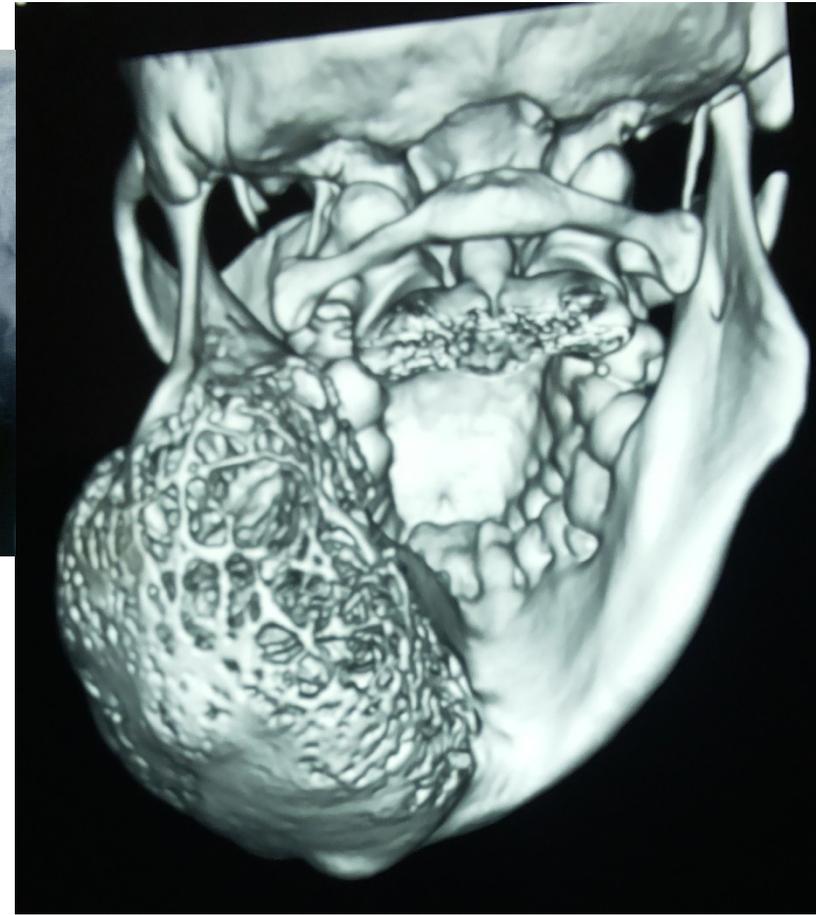


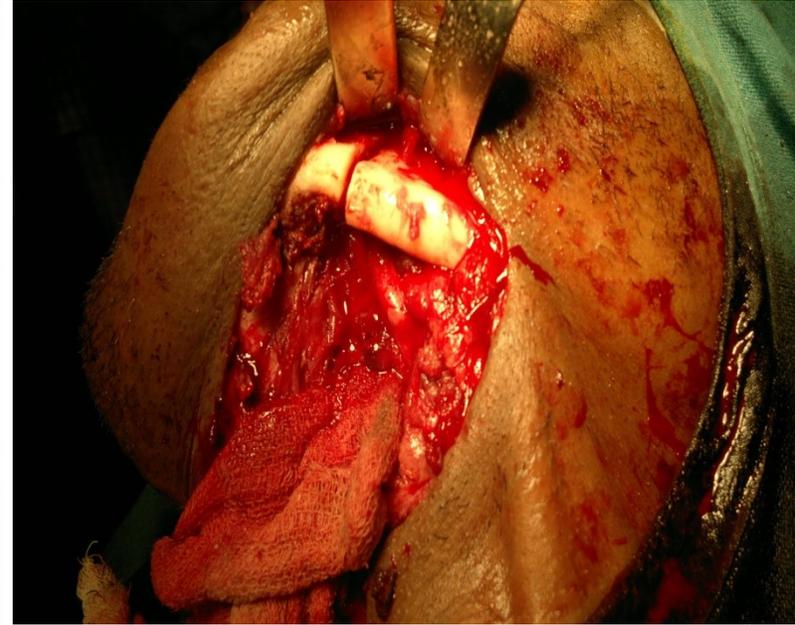
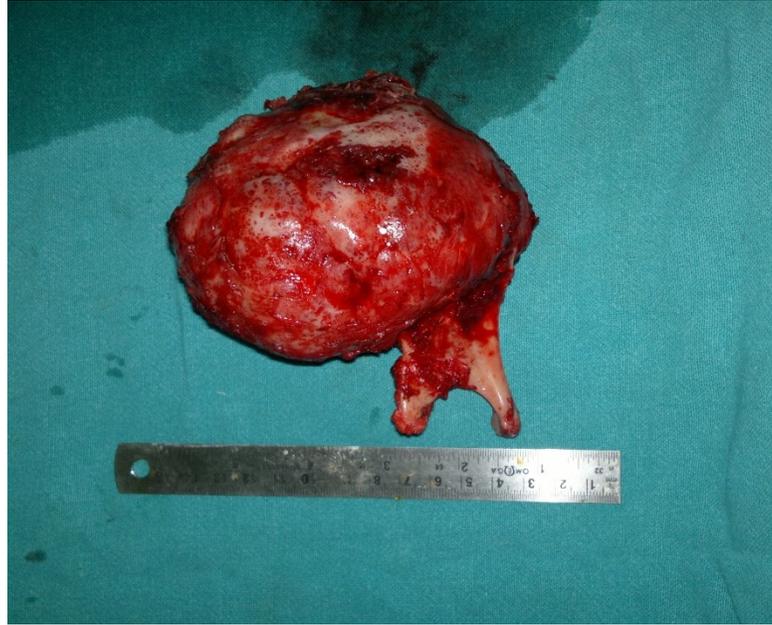
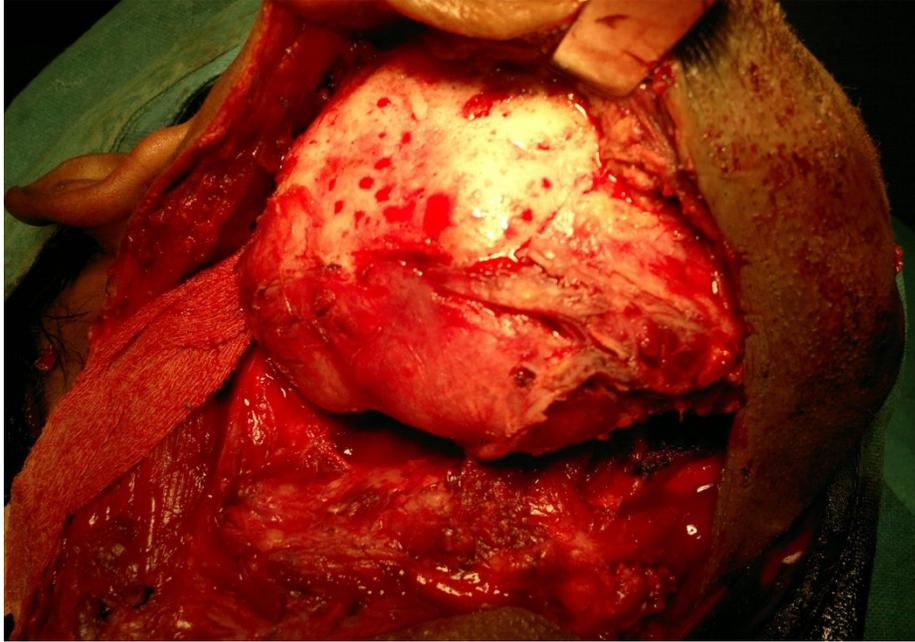
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# OSTEOSARCOMA

- Osteosarcoma (OS), a rare malignant bone tumour arising from primitive bone forming mesenchyme, most often arises in the metaphyses of long bones of the extremities.
- Bone or osteoid formation within the tumour is characteristic of an osteosarcoma.
- Craniofacial osteosarcoma (CFOS), most often located in the mandible or maxilla, accounts for only 5–13% of all osteosarcomas.
- In general, OS of the jaw is a high-grade lesion. Low-grade lesions are rare and represent less than 2% of all osteosarcomas.
- The primary presenting complaints are pain, swelling, paresthesia, and ulceration.
- Radiologically, the lesions may range from predominantly radiolucent to radio-opaque lesions, with any combinations thereof, depending on the degree of ossification.
- Similar to osteosarcoma of the extremities, adequate surgical resection is considered a mainstay of treatment.
- Because of easier resectability, and the ability to obtain negative surgical margins, mandibular osteosarcomas have a better prognosis than maxillary tumors.
- Head and neck OS are associated with a lower metastatic rate than are long bone OS and they have a better 5-year survival rate, ranging between 10% and 85%.



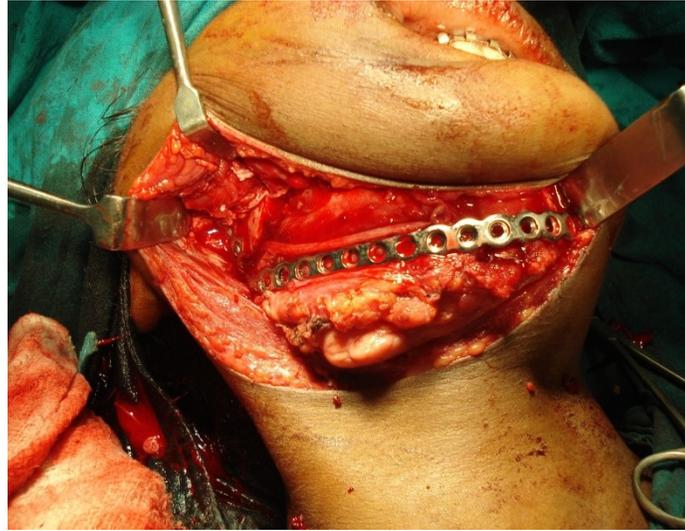
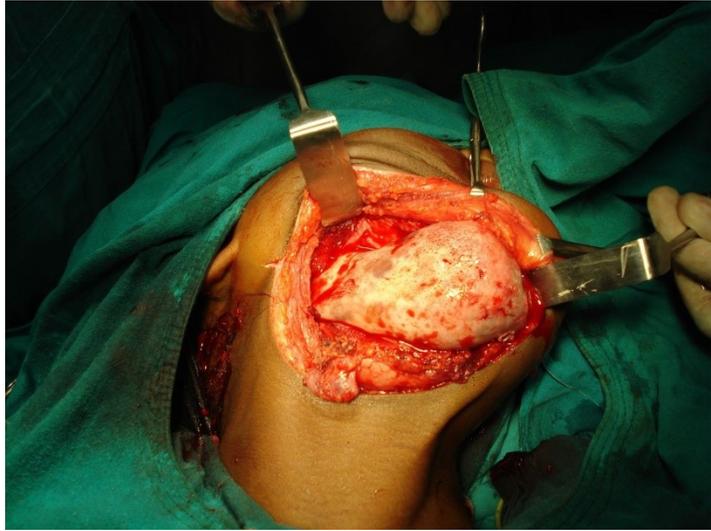




# JUVENILE OSSIFYING FIBROMA

- Juvenile ossifying fibroma (JOF) is a fibro-osseous neoplasm described as an actively growing lesion consisting of a cell-rich fibrous stroma, containing bands of cellular osteoid without osteoblastic lining, together with trabeculae of more typical woven bone.
- Small foci of giant cells may also be present. The lesion is nonencapsulated but well demarcated from surrounding bone.
- It is segregated in two distinct sub types. 1) Trabecular 2) psammomatoid variety.
- JOF commonly affects young children with slight male predilection with a mean age of 11 years
- Maxilla, paranasal sinus, orbit and fronto- ethmoid bone are the predilicted sites for the involvement of the JOF.
- Very few cases of mandibular JOF have been reported .
- Clinically in most instances JOF is slowly growing asymptomatic swelling causing facial asymmetry, but tumor can progress to considerable size and behaves, as aggressive lesion. Less commonly, pain and paresthesia is present.
- The presence of cortical thinning, perforation, tooth displacement and root resorption, are suggestive of feature of aggressive JOF,
- Non- aggressive JOF are treated by curettage and local excision. Aggressive JOF require the complete surgical excision, en-bloc or resection to prevent recurrences.

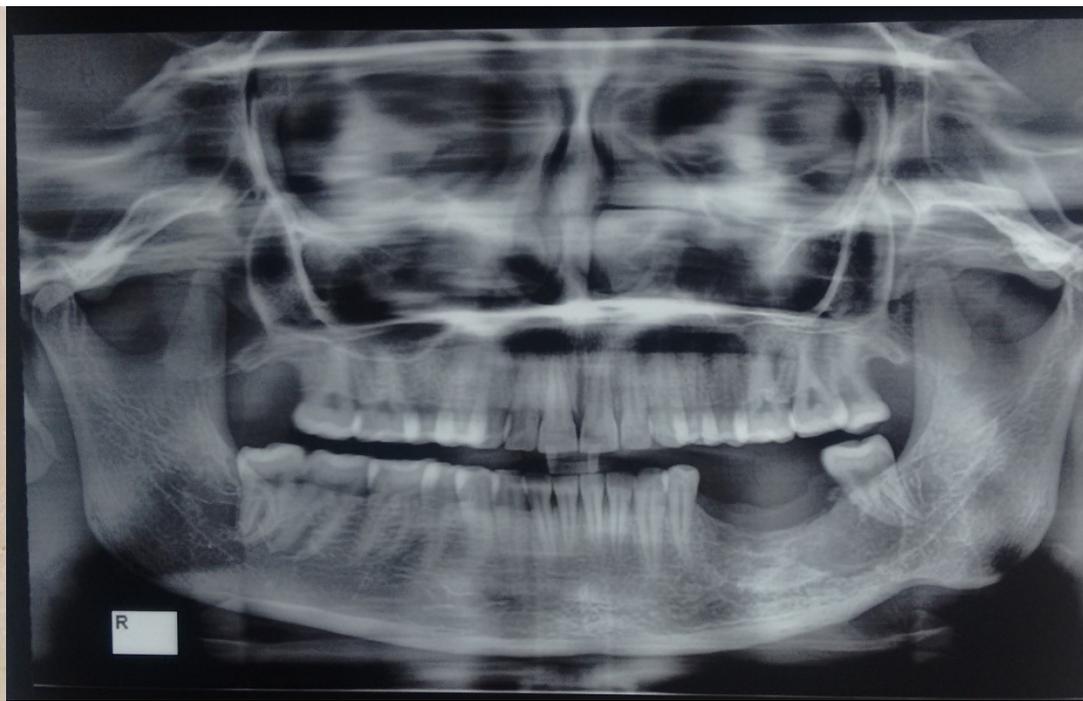


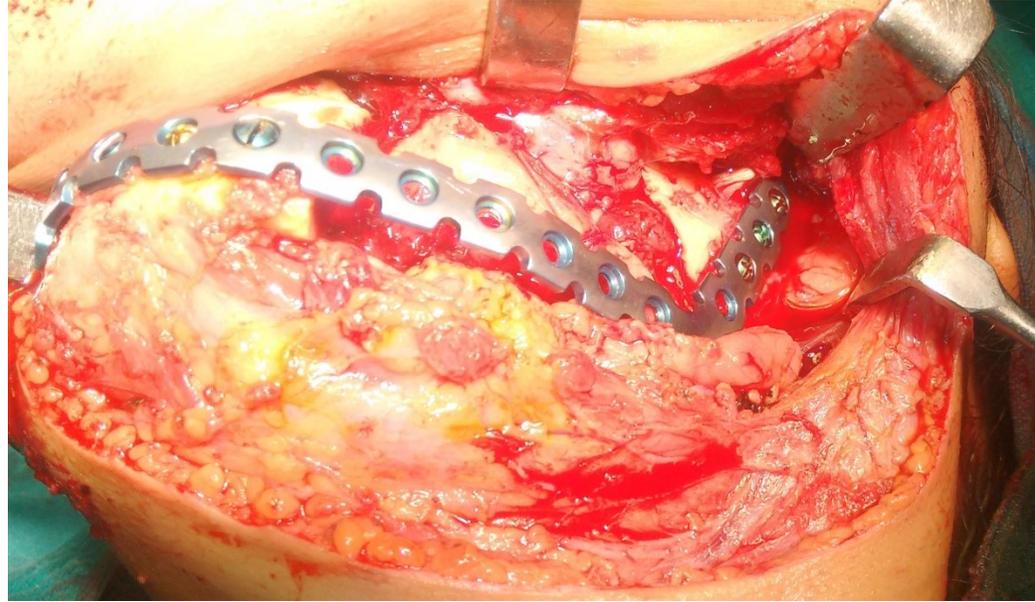
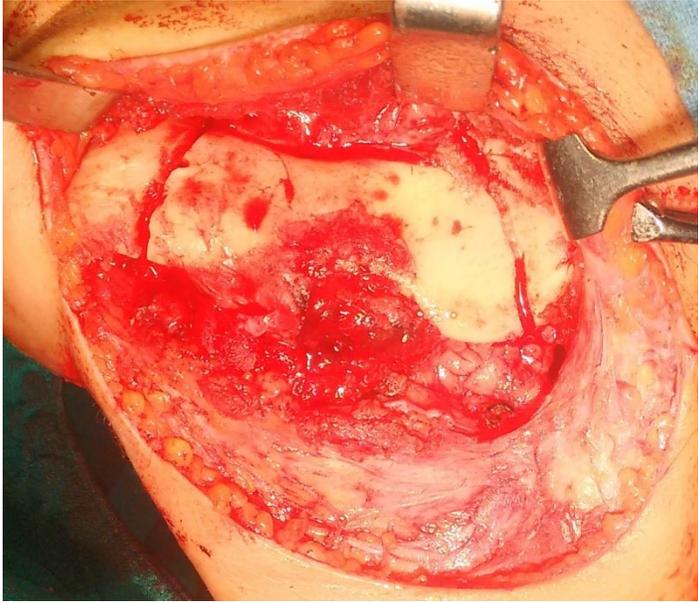




# CENTRAL GIANT CELL GRANULOMA (CGCG)

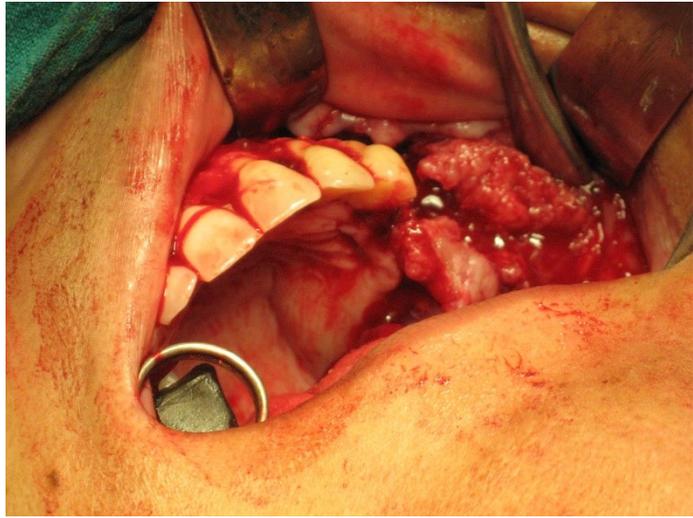
- The central giant cell granuloma (CGCG) is considered a benign, nonneoplastic lesion of bone found in a younger age group who are usually less than 30 years of age.
- CGCG accounts for fewer than 7% of all benign tumors of the jaws-with a prevalence for the mandible at a 65 to 75% rate and affecting females more often than males.
- The growth is an intraosseous lesion consisting of cellular fibrous tissue (fibroblasts) that contains multinucleated giant cells.
- CGCG is very slow growing. But, when seen in a more aggressive form, it exhibits rapid growth, swelling, loosening of the teeth, displacement of the teeth, and it penetrates the cortical bone.
- CGCG is seen as a multilocular lesion or, in rare cases, a unilocular lesion that has well-defined margins. The borders may have a scalloped appearance. The more aggressive form may depict not only root resorption but also perforation of cortical bone.
- The prognosis is good when complete removal is obtained,





Cental Giant cell Granuloma  
Case-2



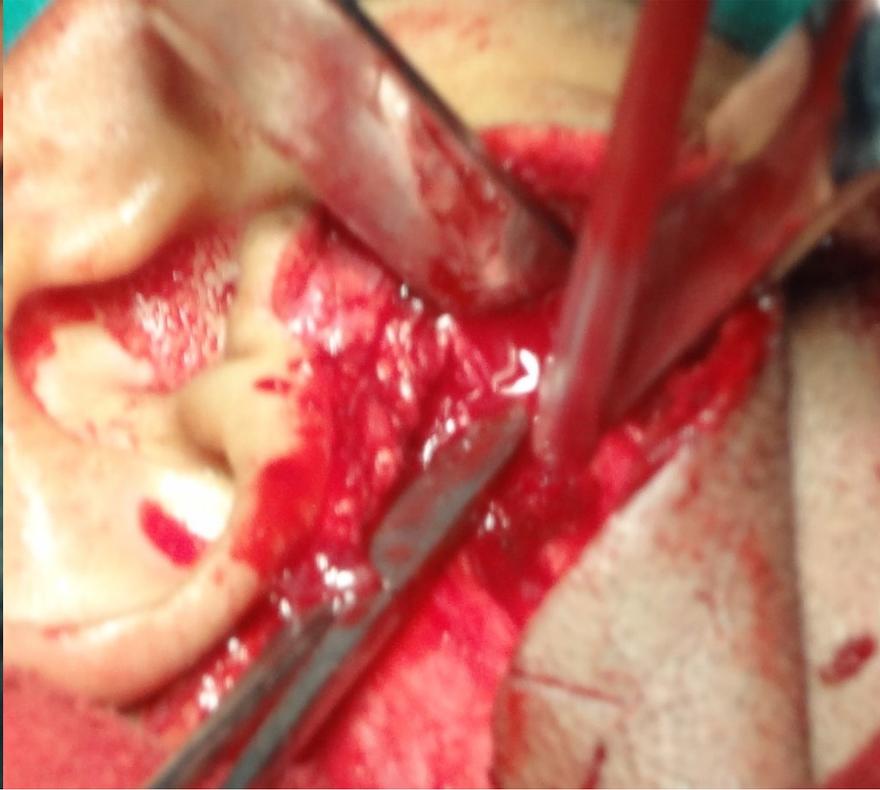




# TUBERCULAR OSTEOMYELITIS

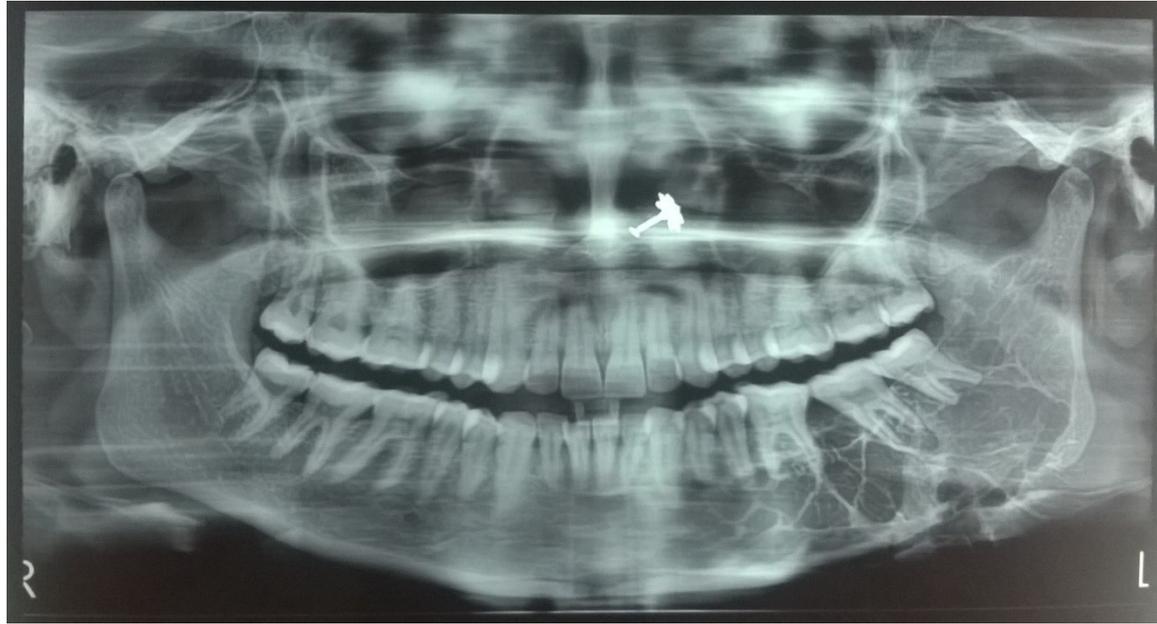
- Tuberculosis (TB), one of the oldest known microbial infectious diseases affecting humans has continued to burden our healthcare system over generations. Conventionally, primary TB usually manifests as a pulmonary infection.
- TB is characterized by a clinical spectrum of presentations such as persistent cough, evening rise in temperature, weight loss, and lassitude as well as atypical presentations that require a high index of suspicion.
- Extrapulmonary infections notably involve the musculoskeletal system more commonly than the respiratory system. A literature search revealed that most cases of TB of the jaw have in fact been primary lesions.
- Rarely, secondary oral manifestations associated with pulmonary infection are seen, which can appear as lesions on the gingiva, palate, lips, tongue, buccal mucosa, frenulum, and in the jaw bones.
- Anti-tubercular and condylectomy is the treatment of choice.

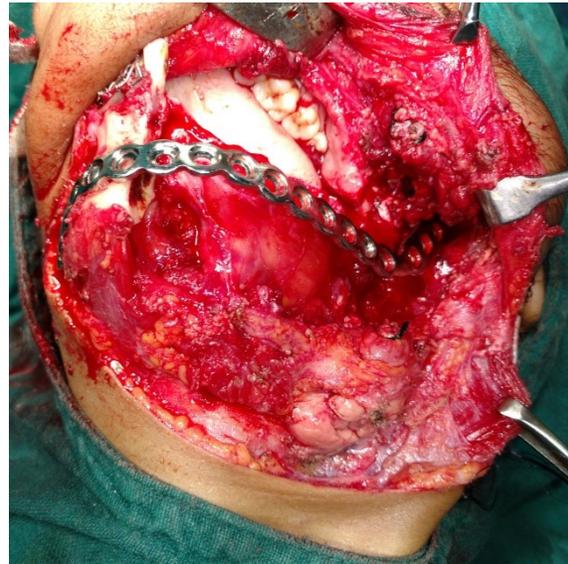
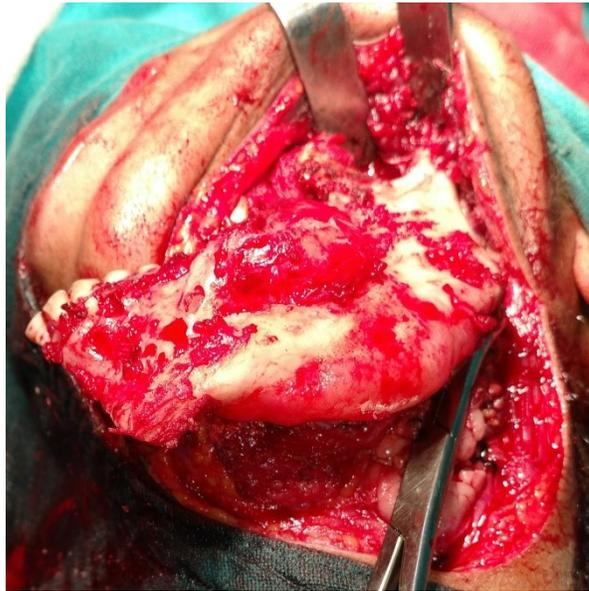


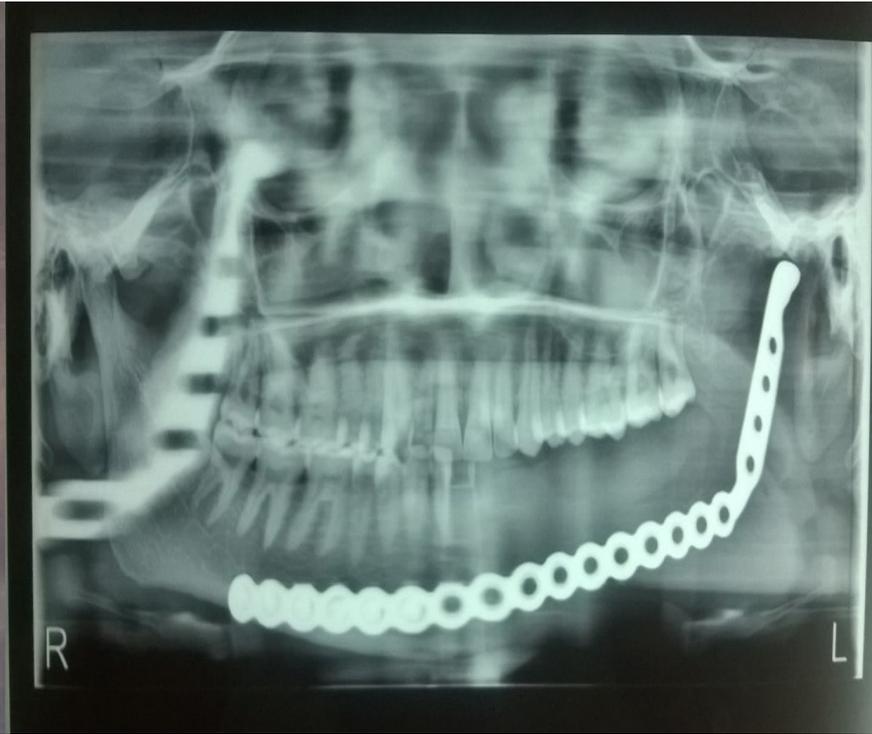


# ODONTOGENIC MYXOMA

- Odontogenic myxoma is a rare intraosseous neoplasm, which is benign but locally aggressive.
- It rarely appears in any bone other than the jaws, as reported in the clavicle or in ribs.
- It is considered to be derived from the mesenchymal portion of the tooth germ.
- Clinically, it is a slow-growing, expansile, painless, non-metastasizing, central tumor of jaws, chiefly the mandible.
- While of all the odontogenic tumors, odontogenic myxoma contributes only 3–11%.
- Odontogenic myxoma most frequently occurs in second or third decades of life, has a slight female predilection.
- it is a slow-growing, expansile, painless tumor, which may cause root resorption, tooth mobility, bone expansion, cortical destruction and facial distortion.
- Radiographically, the classic presentation is that of a multilocular radiolucency, with well-developed locules, consisting of fine trabeculae, arranged at right angles, known as the 'Tennis-racquet' or 'step-ladder' pattern. A 'sun-ray' or 'sun-burst' appearance has also been reported in the literature.
- For extensive lesions a more radical approach is more appropriate.







# ADENOMATOID ODONTOGENIC TUMOR

- Adenomatoid odontogenic tumor (AOT) is a rare odontogenic tumor
- It is predominantly found in young and female patients, located more often in the maxilla in most cases associated with an unerupted permanent tooth.
- Clinical features generally focus on complaints regarding a missing tooth.
- The lesion usually present as asymptomatic swelling which is slowly growing and often associated with an unerupted tooth.
- However, the rare peripheral variant occurs primarily in the gingival tissue of tooth-bearing areas
- Unerupted permanent canine are the the most often involved in AOTs.
- Whereas the follicular variant shows a well-circumscribed unilocular radiolucency associated with the crown and often part of the root of an unerupted tooth, the radiolucency of the extrafollicular type is located between, above or superimposed upon the roots of erupted permanent teeth.
- A review of the literature depicts a lesion in which conservative management produces a uniformly excellent outcome without recurrence



# CONDYLAR HYPERPLASIA

- Condylar hyperplasia (CH) can be defined as the excessive growth of one condyle over the contralateral side.
- Asymmetric facial deformities (AFD) and malocclusion are a clear consequence of CH.
- Generally, there is no pain associated with the affected joint, although joint noises linked to CH and deviation of the mouth opening towards the contralateral side have been described.
- ***Wolford has developed a simple classification that will allow the clinician to better understand the nature of the various CH pathologies:***

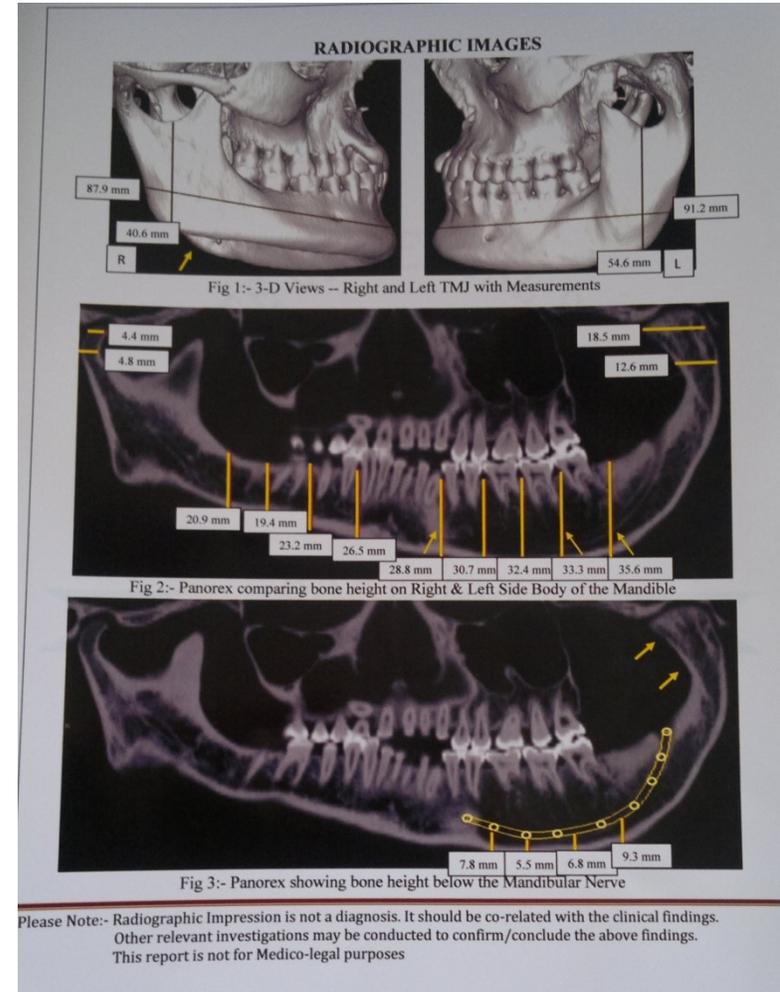
CH Type 1: This condition develops during puberty, is an accelerated and prolonged growth aberration of the normal condylar growth mechanism, is self-limiting but can grow into the 20's, and can occur bilaterally (CH Type 1A) or unilaterally (CH Type 1B).

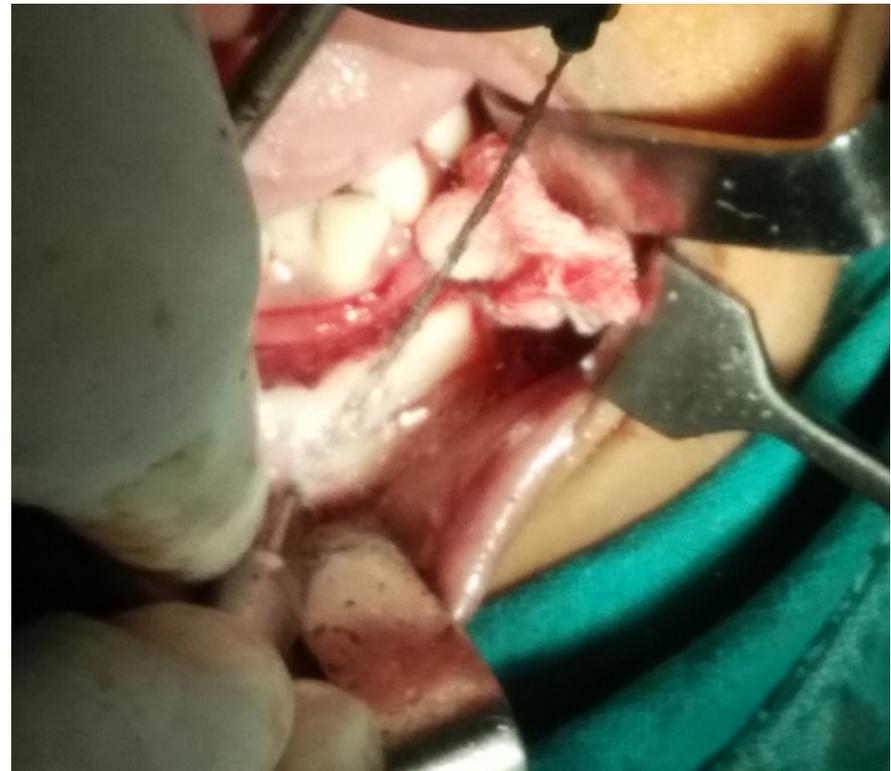
CH Type 2: These condylar pathologies can develop at any age (although 2/3s develop in the 2nd decade), are unilateral condylar vertical and/or horizontal over-growth deformities, and are the most common occurring mandibular condylar tumors; osteochondroma (CH Type 2A) and less common osteoma (CH Type 2B).

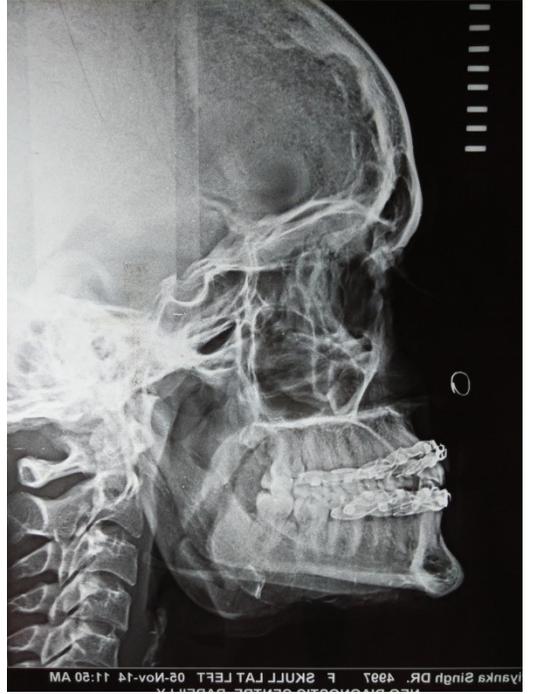
CH Type 3: These are other rare benign causing condylar enlargement.

CH Type 4: These are malignant conditions that can cause condylar enlargement.

- The surgical correction of asymmetry mainly depends on the degree of the asymmetry and the resulting malocclusion.
- Some authors have advocated condylectomy in actively growing condyles especially in immature patients.

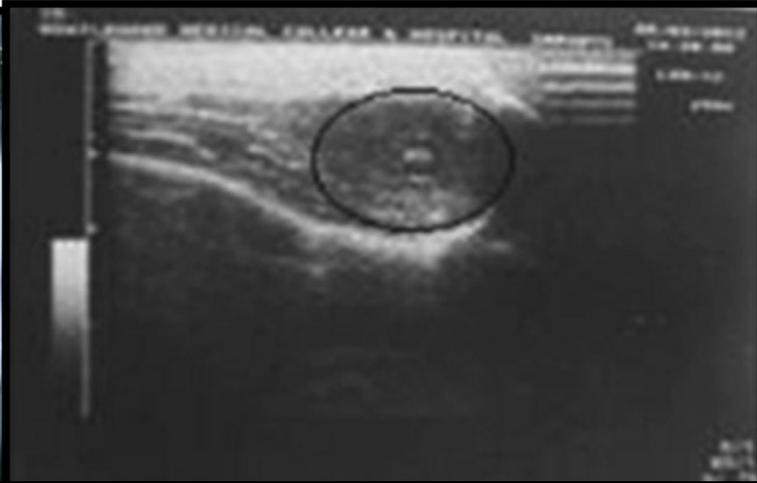
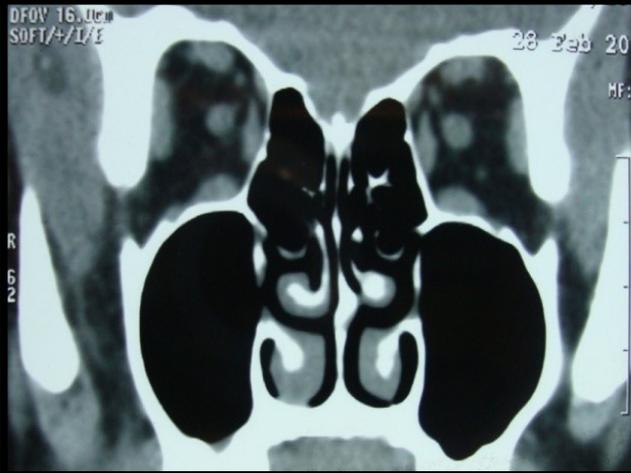






# CYSTICERCOSIS

- Cysticercosis is a zoonotic disease caused by the helminth *T. solium*. The disease is endemic in developing countries such as India in South-east Asia.
- Cysticercosis demonstrates a strong predilection for the central nervous system (CNS) and it is the most common cause of acquired epilepsy in India.
- Apart from the CNS, other organs such as skeletal muscle, the eyes, the heart and the lungs may be involved either on their own or as a part of disseminated disease.
- Solitary skeletal muscle or soft tissue cysticercosis has been reported and there is a report of isolated involvement of the temporalis muscle.
- The imaging characteristics on CT, USG and MRI usually suffice in establishing a diagnosis of cysticercosis.





**THANK YOU**