

## **ODONTOGENIC TUMORS**

Odontogenic tumors are derived from the epithelial and/or mesenchymal remnants of the tooth-forming apparatus. Therefore, they are found exclusively in the mandible or maxilla (and occasionally gingiva) and must be considered in the differential diagnosis of lesions involving these sites. The important & common types of odontogenic tumors:

### **I- Benign Epithelial Odontogenic Tumors:**

- 1-Ameloblastoma
- 2-Calcifying Epithelial Odontogenic Tumor (CEOT).
- 3-Adenomatoid Odontogenic Tumor (AOT).
- 4-Calcifying Odontogenic Cyst (COC).

### **II-Benign Mesenchymal Odontogenic Tumors:**

- 1- Odontogenic Fibroma.
- 2- Odontogenic Myxoma.
- 3- Cementoblastoma.

### **III-Benign Mixed Odontogenic Tumors (Epithelial & Mesenchymal)**

- 1- Ameloblastic Fibroma.
- 2- Odontoameloblastoma.
- 3- Ameloblastic Fibroodontoma.
- 4- Odontoma.

### **IV-Malignant Epithelial Odontogenic Tumors:**

- 1- Odontogenic Carcinoma.

### **Benign Epithelial Odontogenic Tumors:**

#### **Ameloblastoma:**

It is the most common clinically significant odontogenic tumor. Ameloblastoma originates from odontogenic epithelial origin that may remain within the alveolar soft tissue & bone. It may arise from rests of

dental lamina, enamel organ, epithelial lining of an odontogenic cyst or from the basal cells of the oral mucosa.

Ameloblastomas are slowly growing, locally invasive benign tumors. They occur in three different clinic-radiographical types:

1-Conventional or common (multicystic) intraosseous ameloblastoma.

2-Unicystic ameloblastoma.

3-Peripheral (extraosseous) ameloblastoma.

### **Conventional, Common, Solid (Multicystic) Ameloblastoma**

- A benign, aggressive tumor that is invasive persistent.

-Encountered in patients over a wide age range, mean age is 40 years

-Females & males are equally affected

-Mandibular molar-ramus area most commonly affected site, less commonly in the maxilla.

**Clinically**, presented as painless, swelling or expansion. If untreated it may grow slowly to a massive or grotesque deformity.

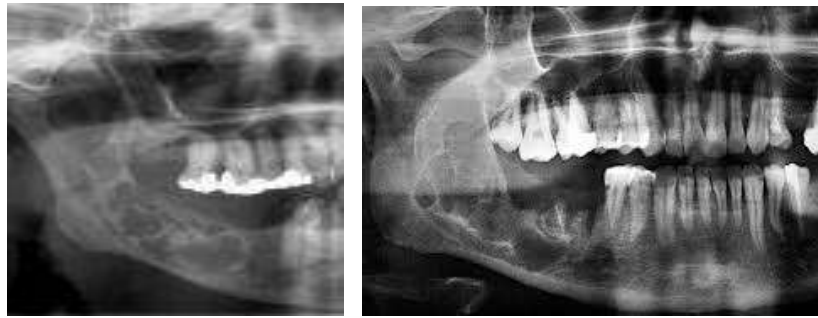
-A characteristic feature of this type of ameloblastoma is the tendency to expand the bony cortices, due to its slow growth pattern which allows the time for the periosteum to produce a thin shell of bone ahead of expanding lesion. This thinned outer shell of bone cracks easily during palpation this is called (Egg Shell Cracking) which is a diagnostic sign of ameloblastoma.



### **Radiographically:**

Multilocular radiolucency (RL), described as having a (soap bubble) appearance when the radiolucent loculations are large and being as

(honey comb) appearance when the loculations are small. Root resorption of adjacent teeth is common. In many cases unerupted tooth often mandibular third molar is associated with the radiolucent lesion.



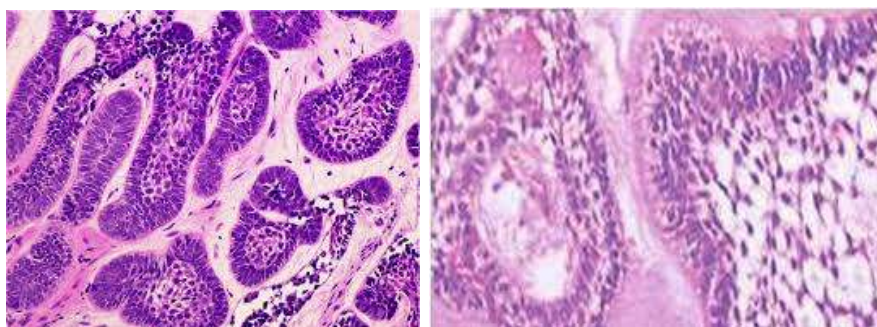
### **Histopathology:**

-Numerous histopathological patterns may be seen in this type of ameloblastoma. Some exhibit a single histopathological subtype, others may display several histopathological patterns within the same lesion. These patterns not affect tumor behavior.

- Common to all subtypes is the presence of palisading columnar cells around epithelial nests in a pattern similar to ameloblasts of the enamel organ. The ameloblast like cells have nuclei at the opposite pole of the basement membrane called the (Reversed Polarization). Central cells of these nests are loosely arranged polyhedral or angular cells that mimic the stellate reticulum of the enamel organ.

- The most common histopathological patterns are:

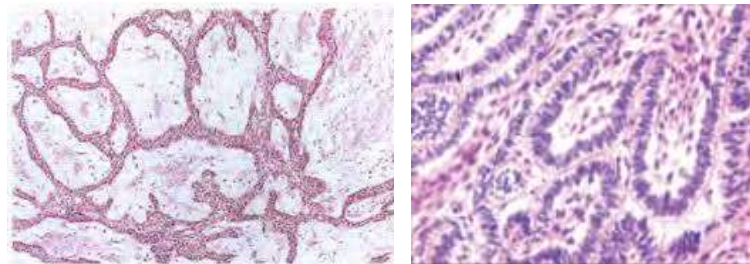
1- **Follicular Pattern:** The most common & recognizable. Islands or strands of epithelium resemble early stages of tooth development, in a mature fibrous C.T stroma.



The epithelial arrangement have an outer border composed of the palisaded ameloblast-like cells with reversed polarity, and centrally a stellate reticulum-like cells are seen which sometimes undergo areas of

degeneration forming central microcysts. This cystic degeneration may be due to ischemia within the large islands of epithelial proliferation.

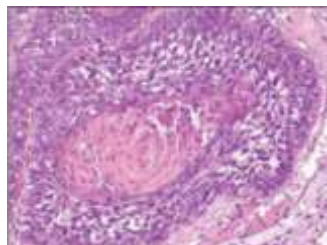
**2-Plexiform Pattern:** Consist of long anastomosing cords in a fish-net or mesh arrangement. The cords of epithelium are bounded by columnar or cuboidal ameloblast-like cells, these basal cells do not resemble ameloblasts, because they lack the distinctive reversed polarization of the nucleus.



Large & small cyst-like areas are present that are not necessarily caused by degeneration of epithelium, but as a result of strangulation & degeneration of CT stroma by the proliferating epithelia.

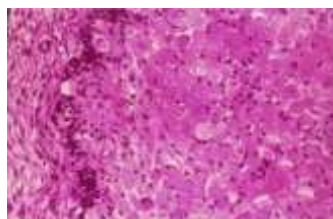
### **3-Acanthomatous Pattern:**

The follicular tumor islands shows extensive squamous Metaplasia, often associated with keratin formation in the shape of keratin pearls, in the central portions.



### **4-Granular Cell Pattern:**

Ameloblastomas may sometimes show transformation of groups of tumor epithelial cells to granular cells. These cells have abundant cytoplasm filled with eosinophilic granules.



**5-Desmoplastic Pattern:** This pattern contains small islands & cords of cuboidal & darkly stained odontogenic epithelium in a dense scar-like collagenized stroma.

-This variant has a mixed radiolucent / radioopaque appearance that resembles fibro-osseous lesions.

### **Unicystic Ameloblastoma: (UA)**

-It is different from common solid ameloblastoma in the clinical, radiographical, histopathologic features & its treatment.

- Whether it originates de novo from preexisting odontogenic epithelial remnants or transformed from normal cyst lining, cannot be determined.

-It is mostly affect younger patients in the second decade of life.

-The mandibular posterior region most commonly involved.

-Most of these lesions are found during biopsy examination of a large unilocular cyst commonly associated with the crown of an impacted tooth in a dentigerous cyst relationship & usually associated with a severely displaced mandibular third molar.

### **Radiographically:**

Well defined unilocular radiolucency that surround the crown of unerupted tooth.

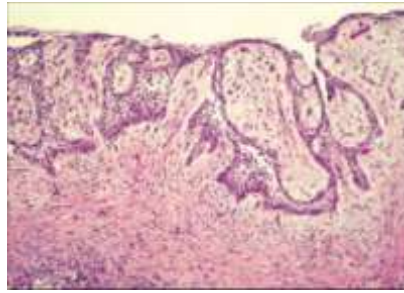


### **Histopathology:**

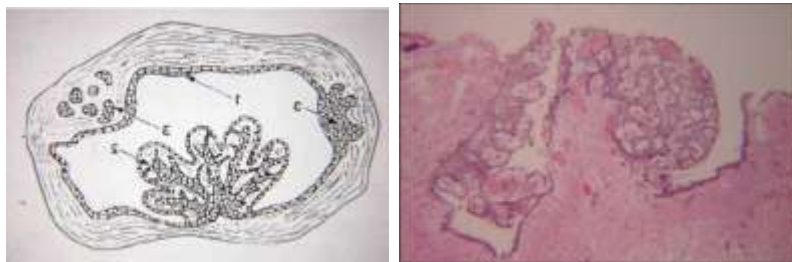
- Three histopathologic variants of unicystic ameloblastoma

First: The **Luminal** UA, the tumor is confined to the luminal surface of the cyst. The lesion is composed of fibrous cyst wall surrounding a solitary large fluid-filled lumen with a lining that consist of ameloblastic

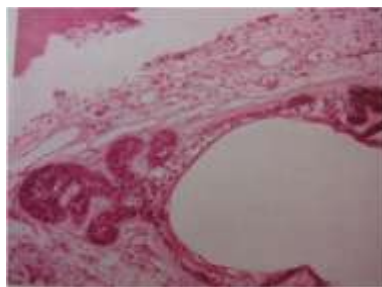
epithelium, which is hyperchromatic layer of palisaded basal cell layers resemble stellate reticulum.



Second: **Intraluminal** UA or Plexiform UA in this variant one or more ameloblastoma nodules project from the cystic lining into the lumen of the cyst. Sometimes these nodules demonstrates plexiform pattern similar to that presented in the plexiform pattern of conventional ameloblastoma.



Third: **Mural** UA in this subtype, the fibrous wall of the cyst is infiltrated by typical follicular or plexiform ameloblastoma. The extent and depth of infiltration considerably variable from UA to another.



### **Peripheral (Extraosseous) Ameloblastoma :**

- Uncommon, arises from the dental lamina beneath the oral mucosa or from basal epithelial cells of the surface epithelium.
- Histologically, resemble the intraosseous conventional ameloblastoma.

**Clinically:** -appears as firm, sessile nodule of the gingiva with smooth surface. Most commonly found on the posterior gingival & alveolar mucosa, and they more common on the mandibular than maxillary area.

**Radiographically:** Soft tissue lesion does not show radiographical changes only superficial saucerization of the cortical plate.

**Histologically:** Islands of ameloblastic epithelium that occupy the lamina propria underneath the surface epithelium. The tumor islands most commonly show the follicular & plexiform pattern of intraosseous ameloblastoma.

Treatment of Ameloblastoma: depend on the clinical type ranges from local excision to block resection.

### **Calcifying Epithelial Odontogenic Tumor (Pindborg Tumor) (CEOT):**

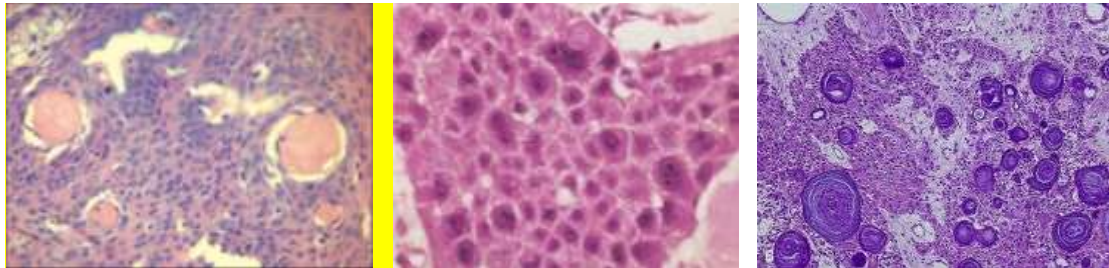
- It is locally aggressive tumor arise from rests of dental lamina. The tumor cells resemble cells of stratum intermedium of enamel organ.
- It affects people from 30-50 years of age.
- CEOT occurs either a central (intraosseous), or peripheral (extraosseous)
  - Intraosseous CEOT, most of the cases involve the mandible, especially the posterior parts, Presented as a painless, slowly growing swelling.
- Extraosseous CEOT, most commonly occurs in the anterior part of the mouth as a superficial soft tissue swelling of the gingiva in the tooth-bearing area & edentulous areas of the jaws.

**Radiographically:** Unilocular or multilocular radiolucent defect, containing radioopaque calcified structures of variable sizes. The tumor is frequently associated with crown of unerupted tooth, most often mandibular third molar. Calcifications often more prominent around the crown of the impacted tooth.



### **Histopathologically:**

- Discrete islands, strands, or sheets of polyhedral epithelial cells in a fibrous stroma. - The cellular outlines of epithelial tumor cells are distinct, and intercellular bridges are prominent.
- The nuclei exhibit pleomorphism, multinucleation, prominent nucleoli & hyperchromatism, mitotic figures are rare.
- Presence of large areas of amorphous, eosinophilic (amyloid — like) extracellular material found between the epithelial sheets..
- Calcifications are distinctive feature of the tumor scattered throughout the epithelia & C.T.
- The amyloid-like material stains positively as amyloid with amyloid stains like: Congo-red and *Thioflavin T* stains.



Treatment: The biological behavior of CEOT is similar to ameloblastoma.

Conservative local resection including narrow rim of the surrounding normal bone is the treatment of choice.

### **Adenomatoid Odontogenic Tumor (AOT):**

- It is an epithelial odontogenic tumor, with an inductive effect on the odontogenic mesenchyme.
- Odontogenic lesion containing duct like or gland like structures
- It is a benign tumor, but may be considered as a hamartoma.
- It arise from reduced enamel epithelia of post secretory phase of enamel organ development.

### **Clinically:**

- AOT largely confined to the young patients in the 2nd decade of life, with female predilection.



- It is usually associated with an impacted tooth & often causes failure of tooth eruption.
- It asymptomatic, it has a striking tendency to involve the anterior portion of the jaws, & found twice as often in the maxilla as in the mandible.

**Radiographically:** well-circumscribed unilocular radiolucency that involve the crown of an unerupted tooth, most often a canine. This appearance difficult to be differentiated from dentigerous cyst.



The differences between them are:

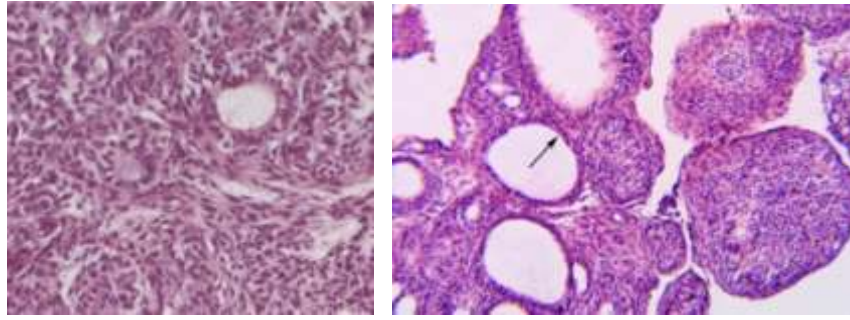
- 1 The radiolucency extends apically beyond the cemento-enamel junction.
- 2 The presence of radioopaque flacks sometimes.

### **Histopathologic Features:**

A well-defined lesion surrounded by thick fibrous capsule. The tumor composed of spindle-shape epithelial cells that forms sheets, strands or whorled pattern of cells in a scanty fibrous stroma.

Rosettes, tubular or duct-like structures of columnar epithelial cells give the lesion the glandular pattern& So the tumor is called (adenomatoid), but in fact they are not true ductal structures & no glandular tissues are present in this tumor.

These duct-like structures may be empty, or contain Small amounts of eosinophilic material stain positive with amyloid. Small foci of calcifications may be scattered among the tumor epithelial sheets



Treatment:

The tumor is completely benign, well encapsulated, it easily enucleated from bone with no tendency for recurrence.

### **Calcifying Odontogenic Cyst (COC): (Ghost Cell Cyst)**

It is an uncommon lesion , although it is widely considered as a cyst, many investigators prefer to classify it as a neoplasm. The solid variant is known as (Odontogenic Ghost Cell Tumor). It is believed to develop from odontogenic epithelial remnants within gingiva, mandible or maxilla.

#### **Clinical Features:**

- It affect young patients in the second decade of life, predilection for females.
- Most of the cases are seen in the maxilla.
- The central (intraosseous) lesions cause painless expansion of the buccal& lingual cortices.
- Rarely, peripheral (extraosseous) masses involving the gingiva of older than 50 years patients found anterior to the first molar region.

#### **Radiographical Features:**

COC presented as unilocular or multilocular radiolucency with well demarcated Margins. Within the radiolucency there may be scattered of irregular sized calcifications. In many cases the radiolucent lesion is associated with crown of unerupted tooth.

#### **Histopathological Features:**

- Many COCs presented as well-defined cystic cavity with a fibrous tissue wall & a lining of odontogenic epithelium, other lesions intraluminal epithelial proliferation obscure the cyst lumen and produce a solid tumor.

- The epithelial lining similar to that of ameloblastoma Composed of outer layer of palisaded columnar basal Cells & inner layer of stellate reticulum.

-Enlarged eosinophilic epithelial cells without visible Nuclei known as Ghost Cells are present within the Stellate reticulum-like area.

- Multiple spherical & diffuse calcifications within the Epithelia & C.T are present also.

Treatment: Cystic lesion is treated by enucleation as a non-neoplastic cyst, whereas Solid lesions may be treated more aggressively.

## **Benign Mesenchymal Odontogenic Tumors:**

### **Odontogenic Fibroma:**

#### **1-Central Odontogenic Fibroma:**

- Uncommon, it is the central intraosseous counterpart of peripheral odontogenic fibroma.

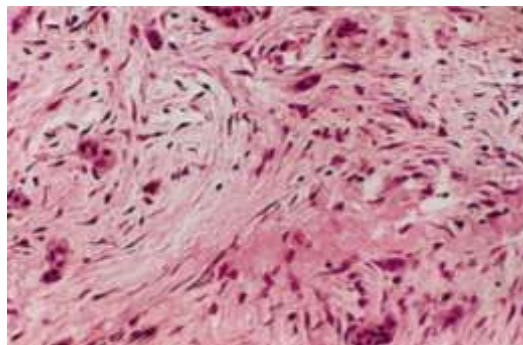
- It may originates from periodontal ligament or dental pulp.

- It affect any age, in the maxilla & mandible in the posterior portions.

- Associated with crown of unerupted tooth, small lesion is asymptomatic, but larger lesions produce localized boney expansion and loosening of teeth.

- **Radiographically:** Similar to conventional or common ameloblastoma.

- **Histopathology:** A cellular, hyalinized C.T in a scar-like pattern.



- Treatment: Enucleation with high recurrence rate.

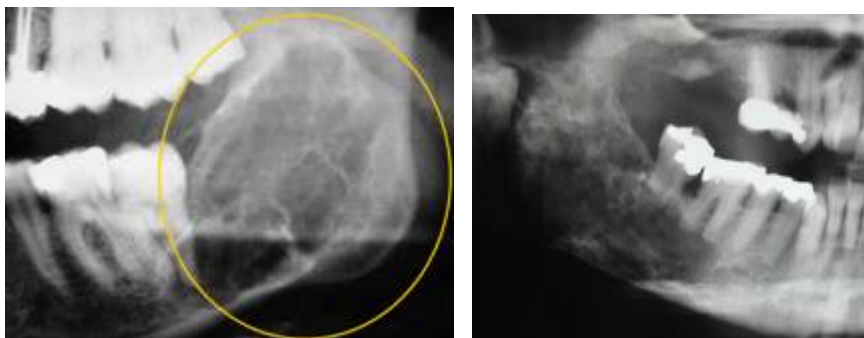
## 2-Peripheral Odontogenic Fibroma:

- Uncommon, it is the extraosseous counterpart of central odontogenic fibroma.
- Firm, slowly growing, sessile gingival mass covered by normal-appearing mucosa.
- It affect people at any age, often presented in the anterior gingiva of the mandible.
- **Histopathologically:** The lesion is composed of mixture of dense C.T that separates localized areas of loose C.T. Small epithelial islands can be seen near the Surface adjacent to thin elongated rete pegs. - Treatment: Local excision.

## Odontogenic Myxoma:

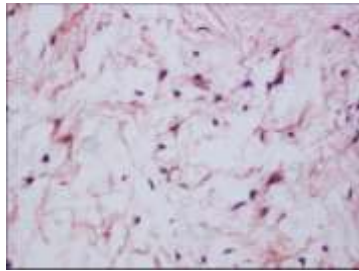
- It is a Benign mesenchymal tumor that mimics microscopically the dental pulp. - Locally aggressive intraosseous tumor occurs in tooth-bearing area of either jaw.
- Affect people in a mean age of 30 years.
- Most lesions are slowly growing, painless swelling that sometimes displace teeth. - Mandible most often involved than the maxilla, in the premolar-molar area extending to the ramus.

**Radiographically:** Myxoma appears as multilocular radiolucency with a honey comb or soap-bubble pattern. It displaces or cause resorption of the roots of teeth in the tumor area.



**Histopathology:** Gross examination of myxoma revealed gelatinous or loose structure. Microscopically, the tumor composed of haphazardly arranged stellate, spindle shaped & round cells in an abundant, loose myxoid stroma that contain only a few collagen fibrils.

In the periphery, the myxomatous tissue penetrates the trabecular spaces producing islands of residual bone, this leads to difficulty in removing the lesion.



Treatment: Surgical excision is the treatment of choice. Because of the loose, gelatinous consistency, curettage may result in incomplete removal of the neoplasm. High recurrence rate is due to absence of capsule. The removal of intact specimen of this gelatinous nature neoplasm is important to reduce the chance of recurrence.

### **Cementoblastoma: (True Cementoma):**

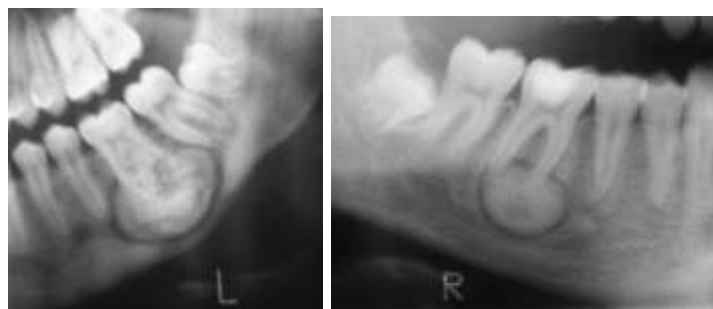
An odontogenic tumor of cementoblasts, in a histopathological features identical to bone tumors.

-Mostly occurs in the mandible, in the molar & premolar region mainly involving the first permanent molar.

-No significant sex predilection, predominantly in children & young adults.

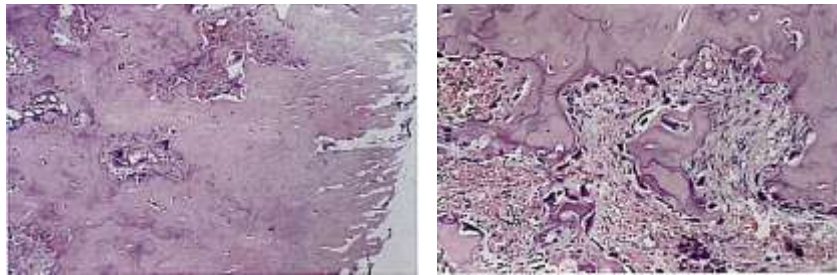
- Pain and swelling may be present, signs of local aggressiveness behavior may be observed, including bony expansion, cortical erosion or displacement of adjacent teeth.

**Radiographically:** the tumor appears as a radioopaque mass that is fused to one or more tooth roots & surrounded by a thin radiolucent rim.



**Histopathological features:** it resembles closely osteoblastoma with primary distinguishing feature being tumor fusion with the involved tooth.

It consists of sheets & thick trabeculae of mineralized material with irregularly placed lacunae & prominent reversal. Cellular fibrovascular tissue is present between mineralized trabeculae. The periphery of the lesion, corresponding to the radiolucent zone on the radiograph, is composed of uncalcified matrix, which often is arranged in radiating columns.



Treatment: surgical extraction of the tooth together with the attached calcified mass.

### **Benign Mixed Odontogenic Tumors (Epithelial & Mesenchymal)**

#### **Ameloblastic Fibroma & Ameloblastic Fibro-odontoma:**

- Ameloblastic fibroma & ameloblastic fibro-odontoma are considered together because they appear to be slight variation of the same process.
- Except for the presence of an odontoma people affected with either of these two lesions share similar features of age, gender, & location.
- A benign mixed odontogenic tumor in which the epithelium and mesenchymal tissues are both neoplastic.
- It is predominantly occur in children & young adults, the mandibular molar-ramus area is the favored location.
- It is presented as slowly growing, asymptomatic jaw expansion.

#### **Radiographic Features:**

Unilocular or multilocular radiolucency, usually surrounded by sclerotic margin may be associated with crown of impacted tooth.

#### **Histopathological Features:**

- The tumor is composed of cell-rich mesenchymal tissue resembling primitive dental papilla mixed with proliferating odontogenic epithelium.
- The odontogenic epithelium presented as narrow cords or form small islands that resemble cap & bell stage of the developing enamel organ
- Zones of hyalinization & focal areas of calcifications are often seen surrounding the epithelial components of the lesion.



-In ameloblastic fibro-odontoma, one or more foci contain enamel & dentine. This may be in form of a compound or complex odontoma.



#### Differential Diagnosis:

- Calcifying odontogenic cyst.
- Calcifying epithelial odontogenic tumor.
- Developing odontoma.
- Possibly adenomatoid odontogenic tumor.

#### Treatment:

- The lesion mostly is well-encapsulated & easily separated from the surrounding bone by enucleation.

Recurrence is high due to inadequate removal of the multilocular lesion.

#### **Odontoma:**

-Mixed odontogenic tumor, it is composed of both epithelial & mesenchymal dental hard tissues. These fully differentiated tissues are enamel, dentine, cementum & pulp tissues. Biologically, odontomas can be regarded as hamartomas rather than neoplasms.

-The calcified tissues of odontoma presented either as numerous miniature or rudimentary teeth known as Compound Odontoma, or they may appear as amorphous conglomeration of hard tissues, known as Complex Odontoma



- Odontoma represents 70% of all odontogenic tumors.

-Most commonly found in the maxilla than in the mandible as asymptomatic swelling.

- Compound odontoma: occurs in the anterior part of the maxilla, either over a crown of unerupted tooth or between roots of erupted teeth.

**Radiographically**, appears as multiple radioopaque structures resemble tiny teeth in a single focus. It may contain as few as 2-3 miniature tooth-like structures or as many as 20-30.



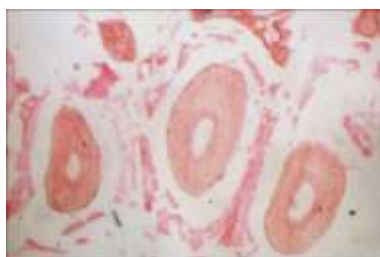
- Complex Odontoma: Mostly found in the posterior part of mandible over impacted tooth & can reach a size up to several centimeters. Radiographically, appears as a solid radiopaque mass, individual tooth-like structures are absent.





### **Histopathology:**

- Compound odontoma: Enamel, dentin & pulp tissue of the tooth-like structures are arranged in an orderly pattern with the surrounding capsule.



- Complex odontoma: composed of a single, disorganized mass of enamel, dentin & pulp with no recognizable tooth shape.



Treatment: Enucleation from the surrounding bone.

### **Malignant Odontogenic Tumors:**

#### **1- Malignant ameloblastoma:**

- Some ameloblastomas that appear cytologically benign show a metastasis to regional lymph nodes or to other sites such as lung.
- In many cases these lesions recurred repeatedly, requiring multiple surgical procedures.

#### **2-Ameloblastic carcinoma**

Differs from malignant ameloblastoma in that its epith, are composed of cytologically malignant cells.

**3-Odontogenic Carcinoma:** an aggressive, destructive & highly infiltrative intraosseous lesion of the jaws, characterized by high recurrence rate. It is composed of poorly differentiated epithelial cells & clear cells in a pattern resemble that of early odontogenesis.

Radiographically: Diffuse honey-comb radiolucency.

Histopathologically: Epithelial structures with malignant features surrounded by myxomatous C. T.