Odontogenic Tumors

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are unique to the jaws and originate from tissue associated with tooth development
The abnormal tissue in each of these tumors can be correlated with similar tissue in normal odontogenesis from inception to tooth eruption.

1. Remnants of dental lamina (rests of serres)
2. Remnants of root sheath of hertwig (rests of malasses)
3. Dental organ and its contents (tooth germ, dental sac, dental papilla)
4. Lining of odontogenic cysts (dentigorous)
5. Basal cell of oral mucosa.
Epithelial tumors

Odontogenic epithelium with mature, fibrous stroma without odontogenic ectomesenchyme:

Benign:

1. Ameloblastoma, solid / multicystic type
2. Ameloblastoma, extraosseous / peripheral type
3. Ameloblastoma, desmoplastic type
4. Ameloblastoma, unicystic type
5. Squamous odontogenic tumor
6. Calcifying epithelial odontogenic tumor
7. Adenomatoid odontogenic tumor
8. Keratocystic odontogenic tumor
Malignant:

1. Metastasizing (malignant) ameloblastoma
2. Ameloblastic Carcinoma
Ameloblastoma

Definition
The solid/multicystic ameloblastoma is a slowly growing, locally invasive, epithelial odontogenic tumor of the jaws with a high rate of recurrence if not removed adequately, but with virtually no tendency to metastasize.
Clinical picture

► The tumor occurs exclusively in the jaws, rarely in the sinonasal cavities.

► Approximately 80% occur in the mandible, with a marked predilection for the posterior region.

► Most maxillary examples occur in the posterior or anterior region.

► Small lesions are asymptomatic.
Radiographic picture

- Large lesions present as variably sized. Unilocular or multilocular radiolucencies resembling cysts and they may reveal scalloped borders.

- An unerupted tooth may be associated with resorption of the roots of adjacent teeth is common.

- Definitive diagnosis cannot be made radiologically.
Histopathology:

Types:
1. Follicular type
2. Plexiform type
3. Basaloid type
4. Desmoplastic type
a) The follicular pattern:

Consists of islands of odontogenic epithelium within a fibrous stroma.

1. The basal cells of these islands are columnar, hyperchromatic, and lined up in a palisaded fashion, their nuclei are displaced away from the basement membrane, and their cytoplasm is generally vacuolated.

2. The central cells may be loosely arranged, resemble stellate reticulum.

3. These areas often become cystic
Ameloblastoma - Histopathology

- Central mass of cells (stellate reticulum like)
- Peripheral cells *(tall columnar cells)*
- Nuclei hyperchromatic
- Vacuolisation
- Reversed polarity

Vicker & Gorlin's Criteria
Micro-cystis ameloblastoma
Macro-cystic ameloblastoma
Acanthomatous ameloblastoma show squamous metaplasia of the central cores of neoplastic epithelium sometimes with keratin formation
Granular ameloblastoma

They usually resemble the follicular type, but the epithelium, particularly in the central areas of the tumor islands, forms sheets of large eosinophilic granular cells. It is a slight more aggressive variant.
Spindle cell ameloblastoma

The central follicle is formed of spindle cells
Basal cell ameloblastoma

It consists of more darkly staining cells predominantly in a trabecular pattern with little evidence of palisading at the periphery. They have been mistaken for basal cell carcinomas.
b) The plexiform pattern

- Contains basal cells arranged in anastomosing strands
- Stellate reticulum like cells in the middle.
- The stroma is usually delicate, often with cyst-like degeneration.
Ameloblastoma - Histopathology

Plexiform pattern

anastomosing cords

cuboidal to columnar cells

stellate reticulum like cells

Does not strictly adhere to Vicker & Gorlin's criteria
Subtypes of the plexiform pattern

The plexiform ameloblastoma demonstrates subtypes that depend mainly on changes in connective tissue stroma.

1. Macrocystic ameloblastoma due to large cystic spaces within the stroma of the tumor

2. Hemangio-ameloblastoma due to the presence of large blood spaces within the connective tissue stroma
Prognosis and predictive factors

- Long-term follow up is essential, since recurrences have been noted more than ten years after the initial treatment. This is due to absence of capsule which allow direct invasion of the tumor within bone marrow spaces.
plexiform networks

cystic degeneration!
Ameloblastoma – desmoplastic type

Definition

Desmoplastic ameloblastoma is a variant of ameloblastoma with specific clinical, imaging and histological features.
Clinical pictures:

- The same age, sex and gender of multicyclic ameloblastoma
- The maxilla: mandible ratio is 1:1.
- Found predominantly in the anterior mandibular region.
- A painless swelling of the jaw bone represents the chief initial complaint.
- The size of the tumor varies between 1.0 and 8.5 cm in diameter.
Histopathology

- The epithelial tumor islands are very irregular or bizarre in shape.
- The epithelial cells at the periphery of the islands are cuboidal with occasional hyperchromatic nuclei.
- Myxoid changes of the juxta epithelial stroma are often found.
- Formation of metaplastic osteoid trabeculae (osteoplasia) may be present.
- A fibrous capsule is not present corresponding to the radiographically poorly defined tumor margin.
Desmoplastic Pattern

- densely fibrous stroma
- squeezes
- thin cords of epithelium
- islands less conspicuous
- central cells are spindle shaped to squamatoid
Radiographically,

- About 50% of A-Ds show a mottled, mixed radiolucency/radiopacity with diffuse margins and ill-defined borders suggesting a fibro-osseous lesion.

- Desmoplastic ameloblastoma. Notice irregularly shaped epithelial island surrounded by a narrow zone of loose-structured connective tissue embedded in desmoplastic stroma.

- Resorption of tooth roots and bone formation may occur.
Radiograph showing well-defined lesion with areas of calcification.
Ameloblastoma, extraosseous / peripheral type

Definition

The extraosseous / peripheral ameloblastoma is the extraosseous counterpart of the intraosseous solid / multicystic ameloblastoma
Clinical features / Imaging

- Extraosseous/peripheral ameloblastomas comprise 1.3-10% of all ameloblastomas.

- Age range: The mean age of patients (males: 53 years; females: 51 years) is significantly higher than for the intraosseous counterpart which has a mean age of 37 years.

- Sex: male > female, their ratio is 1.9 (M): 1 (F).

- Tooth-bearing areas, (gingiva) or alveolar mucosa in edentulous areas.

- Mandible > maxilla with ratio of 2.4:1.

- Painless, firm and exophytic growth with a smooth, pebbly or papillary surface.

- Superficial erosion or depression (saucerization or cupping) of the bone crest due to pressure resorption, there is rarely significant bone involvement.
Histopathology

- It consists of odontogenic epithelium with the same histomorphology cell types and patterns as seen in ameloblastoma.

- Some lesions are located entirely within the connective tissue of the gingiva, showing no continuity with the surface epithelium, whereas others seem to fuse with or originate from the mucosal epithelium.
Prognosis

- Peripheral ameloblastoma don’t show invasive behavior and conservative excision is the treatment of choice.
- The recurrence rate is low and Long-term follow up is recommended.
Ameloblastoma, unicystic type

Definition

The unicystic ameloblastoma (UA) represents an ameloblastoma variant, presenting as a cyst.
Clinical features.

- Cases associated with an unerupted tooth
- show a mean age of 16 years as opposed to 35
- The mean age is significantly lower than that for ameloblastoma.
- There is no gender predilection
- 5 to 15% of all ameloblastomas are of the unicystic type.
- More than 90% of cases involve the posterior mandible
- Asymptomatic, sometimes presenting as a swelling of the posterior mandible.
- Up to 80% are associated with an unerupted mandibular third molar.
Clinical features

- young patients
- mean age = 23 yrs
- Predilection
  - Male = Female
- may cause swelling
  - pain rarely
  - lip numbness
- usually asymptomatic
- mandibular posterior region!
Radiographically

- unilocular radiolucency
- Root resorption may occur
Radiographic features

- Unilocular lesion surrounding unerupted tooth mimicking dentigerous cyst
- Unilocular lesion in a "non-dentigerous" relation!
- Root resorption
Three histopathologic variants exist.

1. The luminal variant is a cystic lesion lined by ameloblastomatous epithelium (tumor is confined to the lining epithelium).

2. Intraluminal extensions may occur. These extensions usually exhibit a plexiform epithelial pattern (tumor extends within cystic cavity). There is no tumor infiltration into the fibrous wall for both variants.

3. The mural variant, the cyst wall is infiltrated by ameloblastomas epithelium that exhibits either a follicular or plexiform pattern. Sometimes all variants may occur in the same lesion.
cystic tumor

- morphology
- behaviour

lumen
- ameloblastic epithelial lining
- capsule/wall
Pathogenesis:

1. Dentigerous cyst

Epithelium - ameloblastic transformation
2- Reduced Enamel Epithelium

ameloblastic transformation of lining
3- Microcystic fusion
Histopathology

1. Luminal type

Vickers and Gorlin criteria!!!
2. Intraluminal type

Nodule of epithelium in lumen

PLEXIFORM UNICYSTIC AMELOBLASTOMA!
3. Mural type

follicular islands
unicystic ameloblastoma
requires microscopy
for final diagnosis
Treatment

unicystic ameloblastoma → can be misdiagnosed as odontogenic cyst
  
  compared to ameloblastoma
  
  less
  
  aggressive approach
  
  conservative approach
  
  enucleation & radiographic observation
  
  lower recurrence rate
  
  aggressive approach
  
  aggressive approach??
  
  local resection
  
  follicular islands may invade bone
  
  confirmed microscopically

MURAL TYPE
Calcifying epithelial odontogenic tumor, C.E.O.T (Pindborg tumor).

- **Definition**

  CEOT is a locally invasive epithelial odontogenic neoplasm, characterized by the presence of amyloid material that may become calcified.
Clinical picture

- CEOT accounts for approximately 1% of all odontogenic tumors
- Age between 20 and 60 years with a mean around 40 years
- There is no gender predilection.
- Most cases are intraosseous, approximately 6% arise in extraosseous locations.
- Intraosseous tumors affect the mandible more often than the maxilla with a ratio of 2:1.
- Peripheral lesions usually occur in the anterior gingiva
- Asymptomatic slow-growing expansile mass of the jaw
- Peripheral gingival lesions are firm painless masses.
Radiographically,

- Most CEOTs present as mixed radiolucent-radiopaque lesions
- Unilocular or multilocular.
- Unerupted tooth, most often a mandibular third molar, is associated with the lesion.
Histopathology

- The tumor consists of a fibrous stroma with islands and sheets of polyhedral epithelial cells.

- These cells show:
  1. Abundant eosinophilic cytoplasm
  2. Sharply defined cell borders
  3. Well-developed intercellular bridges.
  4. Their nuclei are frequently pleomorphic, with giant nuclei being common.
  5. Within or around the sheets of tumor cells are eosinophilic, homogeneous.

- Hyaline material
  1. This material is often calcified in the form of concentric rings is present.
  2. This material proved to be amyloid in nature.
Higher magnification of epithelial sheet. **Note the nuclear polymorphism** and intercellular bridges (H&E, x 150).
Figure 4b: Histopathological image
Adenomatoid odontogenic tumor

Definition

- Adenomatoid odontogenic tumor (AOT) is composed of odontogenic epithelium in a variety of histoarchitectural patterns, embedded in a mature connective tissue stroma and characterized by slow but progressive growth.
Clinically

- 2-7% of all odontogenic tumors.
- Age: second decade of life and 90% are found before the age of 30.
- The male: female ratio is 1:1.9.
- The AOT almost exclusively occurs intraosseous with a preference for the maxilla over the mandible with a ratio of 2.1:1.
- The rare peripheral type occurs almost exclusively in the anterior maxillary gingiva.
Intra osseos AOT occurs mostly in maxillary canines region, most AOTs are asymptomatic.

When growth it causes cortical expansion, it may present as a palpable bony-hard swelling

AOTs may cause displacement of neighboring teeth.

A peripheral variant is present and demonstrated as a fibroma or an epulis-like lesion of the gingiva at the maxillary canine region.
Radiographical picture

- The intraosseous, AOT, shows a well-defined, unilocular radiolucency resemble a dentigerous cyst when associated with impacted teeth.
- If not associated with an unerupted tooth, AOT presents as a unilocular radiolucent lesion.
- In two thirds of the intraosseous variant, the radiolucency shows discrete radiopaque foci.
- The peripheral variant may disclose erosion (saucerization) of the alveolar bone crest.
**Histopathology**

- Variably sized solid nodules of cuboidal or spindle cells of odontogenic epithelium forming nests with minimal stromal connective tissue.

- Between the epithelial cells eosinophilic amorphous material is present formed of amorphous amyloid-like material and globular masses of calcified substances.

- Tubular or duct-like appearances are present between the cells lined by a single row of columnar epithelial cells, with the nuclei polarized away from the luminal surface.
The duct-like spaces represent pseudo Lumina formed by secretion of the columnar epithelial cells.

The lumen may be empty or contain eosinophilic material or cellular debris.

The duct-like structures may not be present in all AOTs.

Occurrence of a hyaline, dysplastic material or calcified osteodentin.
Conductive tissue

- Tubular arrangement of tumor epithelial cells
- Solid areas of tumor epithelial cells
- Sheets of adenocarcinomatous epithelial cells
- Convulated band arrangement of tumor epithelial cells
Keratocystic odontogenic tumor

Definition

- A benign uni- or multicystic, intraosseous tumor of odontogenic origin, with a characteristic lining of parakeratinized stratified squamous epithelium and potential aggressive, infiltrative behaviour.

- It may be solitary or multiple. The latter is usually one of the stigmata of the inherited naevoid basal cell carcinoma syndrome (NBCCS).
Clinically

- The traditional name is odontogenic keratocyst (OKC), which stresses the benign behavior of this lesion.
- The term keratocystic odontogenic tumor (KCOT) as it better reflects its neoplastic nature.
- Other names include odontogenic keratocystoma and primordial cyst.
- Age: second and third decades.
- The mean age of patients with multiple KCOTs, with or without the NBCCS, is lower than those with single non-recurrent KCOTs.
- Males are more involved than females.
The mandible is involved more frequently than the maxilla.

About one-half originate at the angle of the mandible, extending anteriorly and superiorly.

The most important clinical feature of the KCOT is its potential for locally destructive behavior, its recurrence rate, and its tendency to multiplicity, particularly when associated with the Nevoid Basal Cell Carcinoma Syndrome.

Patients may complain of pain, swelling or discharge.

These tumors may reach a large size prior to discovery.

KCOT may penetrate cortical bone and involve adjacent structures.
Radiographically

- Multilocular radiolucency in mandibular lesions
- KCOTs may appear as small, round or ovoid unilocular radiolucencies or may be larger with scalloped margins
- A mandibular radiolucency may involve body, angle and ascending ramus.
- The radiolucencies tend to be well-demarcated with distinct sclerotic margins.
- Adjacent teeth may be displaced but root resorption occurs rarely.
- CT scans and cone beam radiographs may be helpful in detecting cortical perforation and assessment of soft tissue involvement
Histopathology

- The KCOTs are lined by a regular parakeratinized stratified squamous epithelium,
- Usually about 5-8 cell layers thick and without rete ridges.
- There is a well-defined, often palisaded, basal layer of columnar or cuboidal cells.
- The nuclei of the columnar basal cells tend to be oriented away from the basement membrane and are often intensely basophilic.
- This is an important feature in distinguishing KCOT from jaw cysts with keratinization. The parakeratotic layers often have a corrugated surface.
Desquamated keratin is present in many of the cavities.
Mitotic figures are found frequently in the suprabasal layers.
Some linings may show features of epithelial dysplasia but malignant transformation to squamous cell carcinoma is rare.
In the presence of an intense inflammatory process, the epithelial lining loses its characteristic cellular and architectural features.
Cystic jaw lesions that are lined by orthokeratinizing epithelium do not form part of the spectrum of a keratocystic odontogenic tumor (KCOT).
Causes of high recurrence rate

1. Mitotic activity of the lining which causes the presence of daughter cysts in the connective tissue wall
2. Friable cyst lining easily detached and implanted during surgical removal