Odontogenic tumors

Markéta Hermanová

Odontogenic tumors

- Tumors derived from the dental formative tissues
- Uncommon, some are exceedingly rare, represent 1 % of all tumors of oral cavity.
- Derived from epithelial, ectomesenchymal and mesenchymal tissues.

Classification of odontogenic tumors

- According biological behavior
- Benign
- Malignant

- According localisation
- Intraosseous
- Extraosseous

Classification of odontogenic tumors

- **Histogenetic/histomorphological classification**
- **Epithelial**
  - Without odontogenic mesenchyme
  - With odontogenic mesenchyme
- **Mesenchymal**
- **Tumors of debatable origin**
  - Melanotic neuroectodermal tumor of infancy
  - Congenital gingival granular cell tumor (epulis congenita)

Benign epithelial odontogenic tumor

- **Without odontogenic mesenchyme**

Ameloblastoma

Squamous odontogenic tumor

Calcifying epithelial odontogenic tumor

Adenomatoid odontogenic tumor

Keratinising cystic odontogenic tumor

- **With odontogenic mesenchyme**

Ameloblastic fibroma and fibrodentinoma

Ameloblastic fibro-odontoma

Odontoameloblastoma

Calcifying odontogenic cyst and dentinogenic ghost cell tumor

Complex odontoma

Compound odontoma

Benign mesenchymal odontogenic tumours

- **Odontogenic fibroma**
  - benign, rare, mandible, maxilla

- **Myxoma**
  - benign, relapsing, mandible, maxilla

- **Cementoblastoma**
  - benign, relapsing, associated with the root of a tooth
  - Calcified cementum-like tissue containingscattered cells lying in lacunae

Malignant odontogenic tumors

- **Odontogenic carcinomas**

Malignant ameloblastoma/ameloblastic carcinoma

Primary intraosseous carcinoma (without accociation with oral mucosa)

Clear cell odonogenic carcinoma

Malignant variants of other epithelial tumors

Malignant change in odontogenic cysts

(metastatic spreading into lymph nodes, lungs, bones,…)

- **Odontogenic sarcomas**

Ameloblastic fibrosarcoma

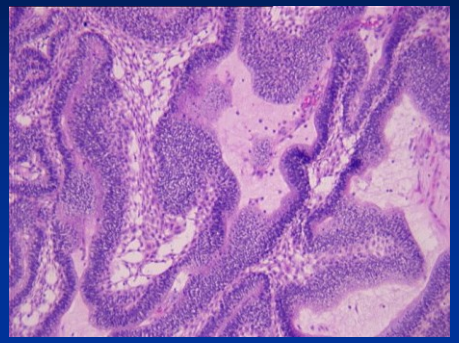
Ameloblastic fibro-odontosarcoma

Ameloblastoma

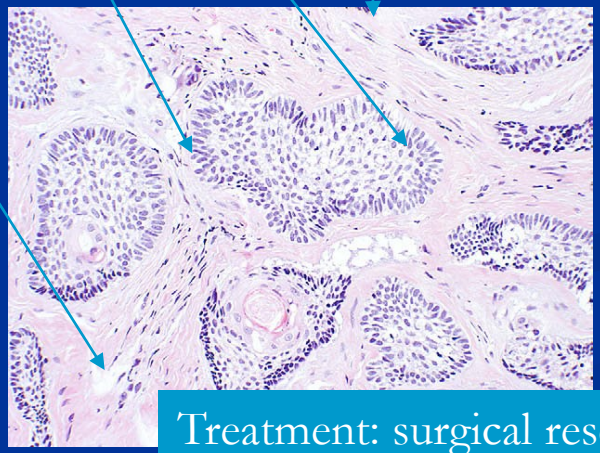
- Benign, local invasive, rellapse after years

- Subtypes:
  - Solid/multicystic
  - Extraosseous/peripheral (soft tissues above mandibula, older patients)
  - Desmoplastic (anterior maxilla and mandible)
  - Unicystic

Solid multicystic ameloblastoma



- 3.-5. decade, molar region/ascending ramus of the mandible (70 %) + posterior maxilla
- Two main histological patterns: follicular and plexiform
- *Variants:* basaloid, granular, acantomathous, keratoacantomathous
- Columnar/cuboidal peripheral cells considered to be preameloblasts)
- Mature fibrous stroma, does not contain enamel or dentin

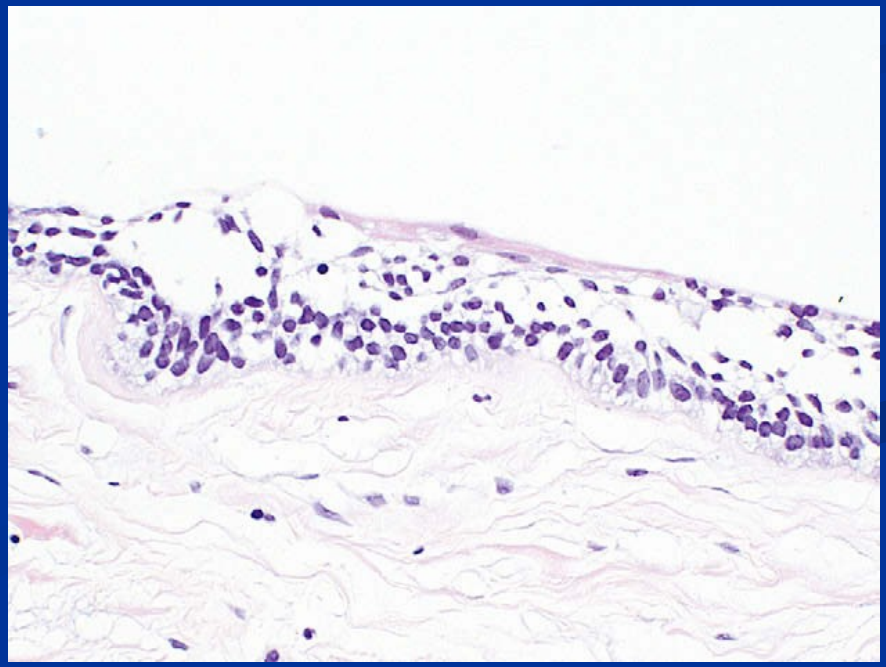


Treatment: surgical resection with a margin of normal bone

Unicystic ameloblastoma

- Younger patients

- Often associated with an unerupted tooth, macroscopically looks like dentigerous cyst
- Cystic lesion lined by ameloblastomatous epithelium, with palisading of basal layer of the cells
- Luminal amd mural variants
- Luminal variants treated by enucleation and curettage;
  mural variants treated by resection



Other benign epithelial odontogenic tumors

- **Squamous odontogenic tumor**
- benign, locally aggressive, rare, in mandible, M>F, 4th decade
- nests of squamous epithelium in fibrous stroma

- **Calcifying epithelial odontogenic tumor (Pindborg´s tumor)**
- benign, locally aggressive, rare, in mandible, M>F, 4th decade (molar and premolar region)
- sheets of pleomorphic epithelial cells, amyloid-like material, calcification
- assoc. with Gorlin-Goltz syndroma

Other benign epithelial odontogenic tumors

- **Adenomatoid odontogenic tumor**
  - benign, hamartoma?, 2nd, 3rd decade, anterior part of the maxila, often assoc. with unerupted tooth
  - solid nodules and tubular structures + eosinophilic material, calcification; treated by enucleation

- **Keratinising cystic odontogenic tumor = odontogenic keratocyst (WHO 2005 true tumor)**
  - benign, locally aggressive
  - Mandibular angle (50 %), 60 % relapsing; treated by enucleation?

Odontogenic keratocysts

- Bimodal age distribution – 2nd-3rd decades and 5th decade
- Few symptoms; cause little expansion; may reach large size
- Unilocular/multilocular radiolucency; may mimic dentigerous cyst
- More common in mandibula than in maxila
- Tendency to recur
- May be multiple; associated with nevoid basal cell carcinoma syndrome

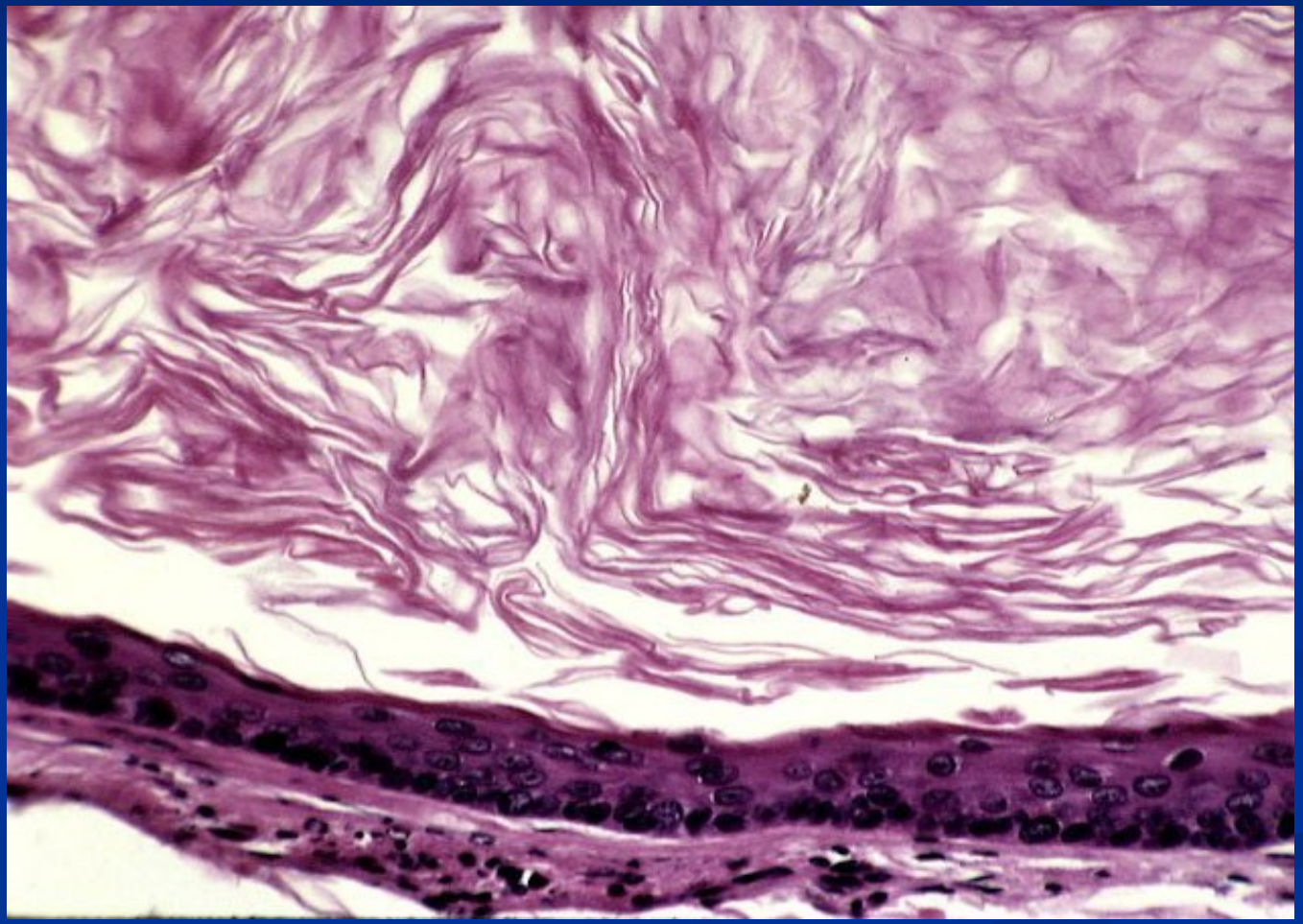
Naevoid basal cell carcinoma syndrome (Gorlin-Goltz syndrome)

- AD
- **Multiple naevoid BCC + multiple odontogenic keratocysts + skeletal abnormalities (rib abnormalitites, vertebral deformities, polydactyly, cleft lip/palate) + calcified falx cerebri + brain tumours**
- Mutation in tumor supperssor gene PTCH (9q)
- Mutations of PTCH affect the normal function of *Hedgehog* signalling pathway
- *Hedgehog* signalling pathway controls transcription of the genes involved in the developlment, patterning, and growth of numerous tissues and organs

Odontogenic keratocysts

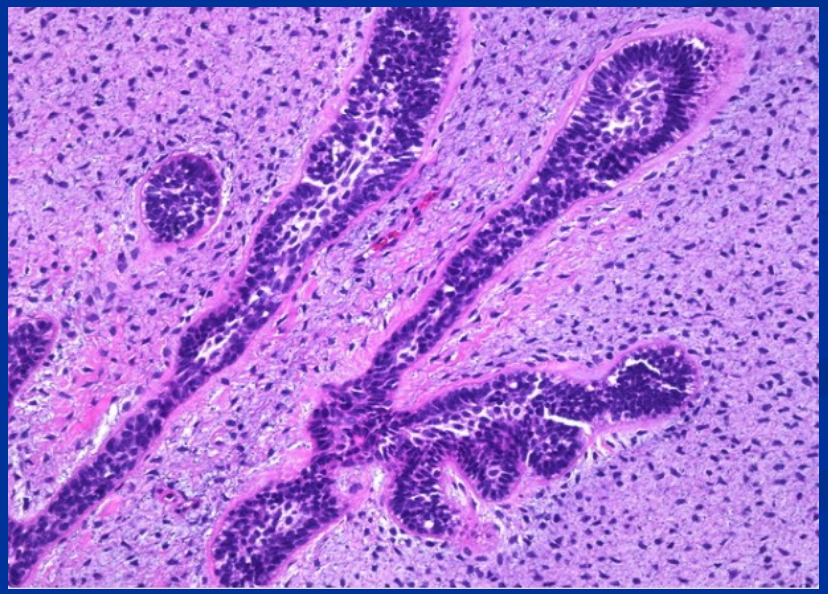
- **Thin, easily torn wall**
- **Lined by an even layer of parakeratinized squamous epithelium**
- **Palisaded basal cell layer**
- **Satellite cysts in capsule**
- **Tendency to recur due to difficulty of surgical removal**
  - thin, easily ruptured wall
  - Projection into cancellous spaces easily torn
  - Satellite cysts in capsule
- **Cyst enlargement involves**
  - Focal areas of active growth of the cyst wall
  - Extension of proliferating areas along cancellous spaces
  - Production of bone resorbing factors

Odontogenic keratocyst



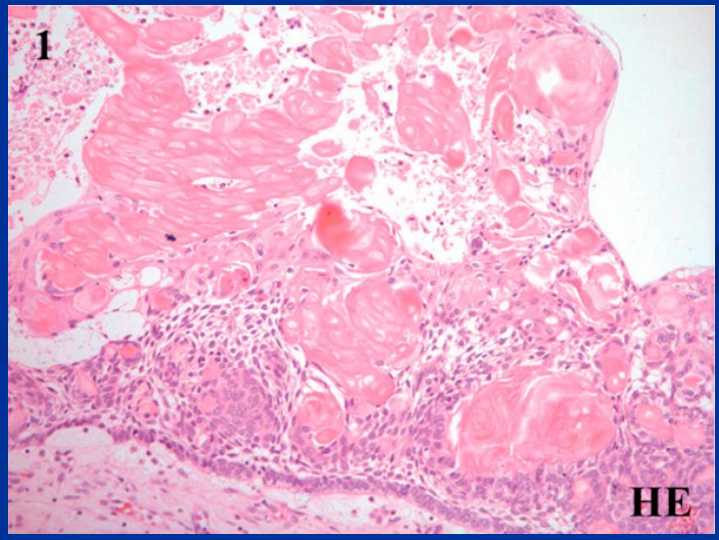
Ameloblastic fibroma, fibrodentinoma, fibro-odontoma

- Benign tumors, fibro-odontoma v.s. hamartoma
- Well circumscribed, mainly in 1st and 2nd decade, posterior mandible
- Odontogenic epithelium + cellular mesenchymal tissue
- Fibrodentinoma contains dentin, fibro-odontoma enamel and dentin
- Must be distinguished from ameloblastoma, different treatment (curretage)!



Calcifying cystic odontogenic tumor = calcifying odontogenic cyst (Gorlin´s cyst)

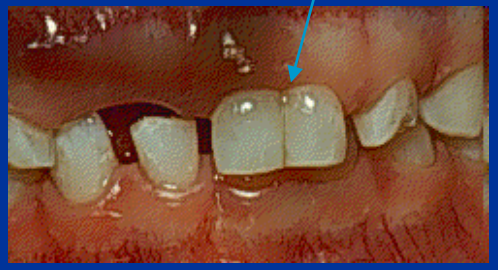
- WHO 2005 true tumor
- Wide age range
- Anterior mandible, maxilla, gingiva
- Cyst lined by ameloblastic epithelium, „ghost cells“→calcification, „dentinoid“ at basal layer of epithelium in supporting fibrous tissue
- Tend to recur, sometimes associated with ameloblastoma
- Treatment: enucleation

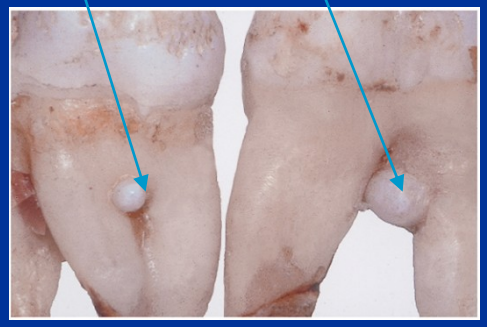


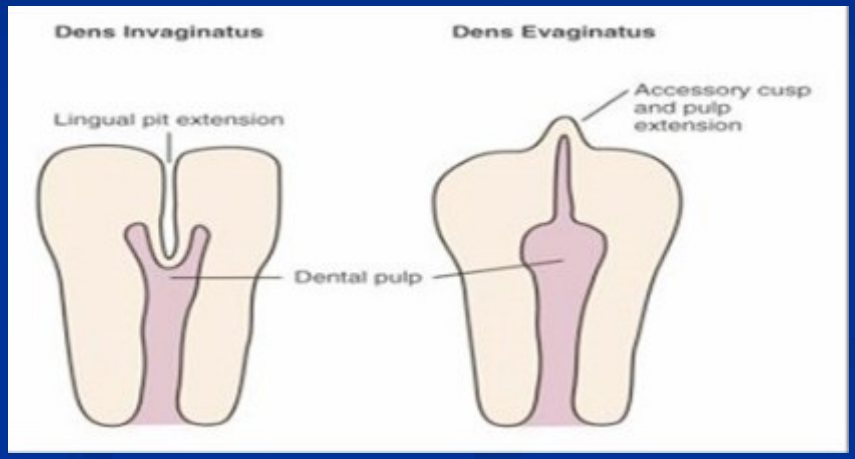
Odontomas

(true tumors?, hamartomas, congenital anomalies?)

- Invaginated (dens-in-dente, dens invaginatus)
- Evaginated (dens evaginatus)
- Enamel pearls/enamelomas
- „Double tooth“/geminated odontoma





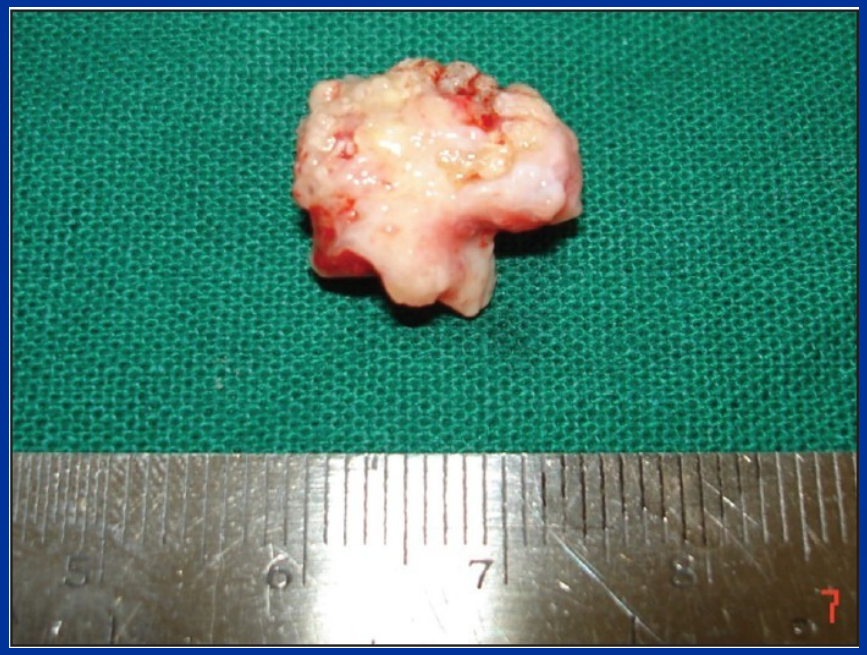


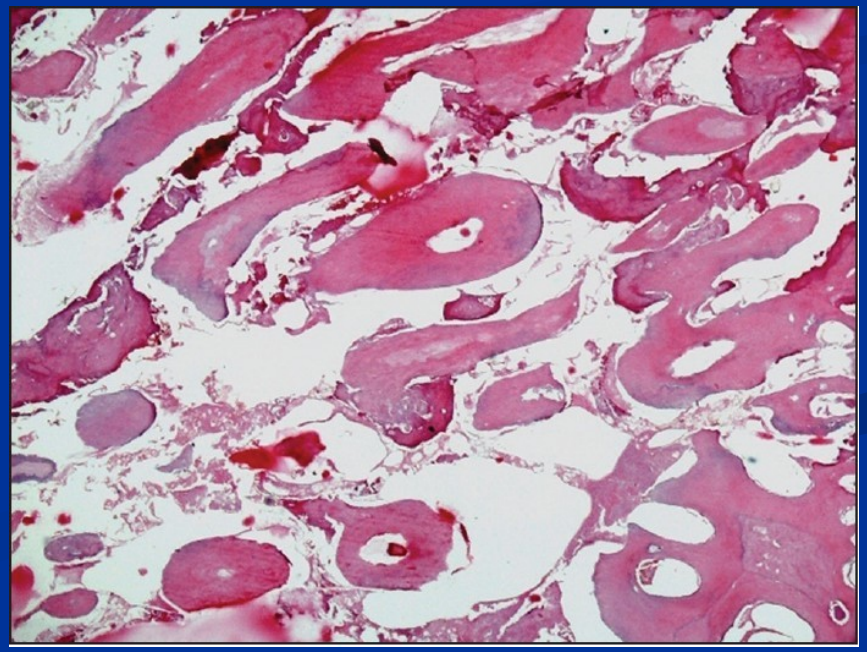
- **Complex odontoma**
- **Compound odontoma**

Complex odontoma

- Develompmental lesion resulting in disorganized mass of dental tissue

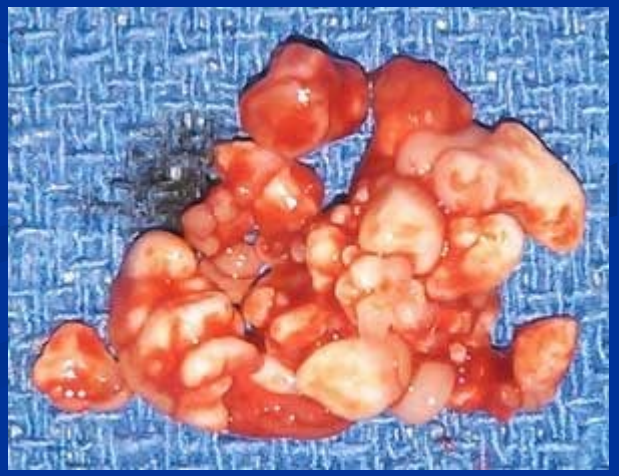
- 2nd/3rd decade, predominantly molar region mandible, omay overlie/replace a tooth





Compound odontoma

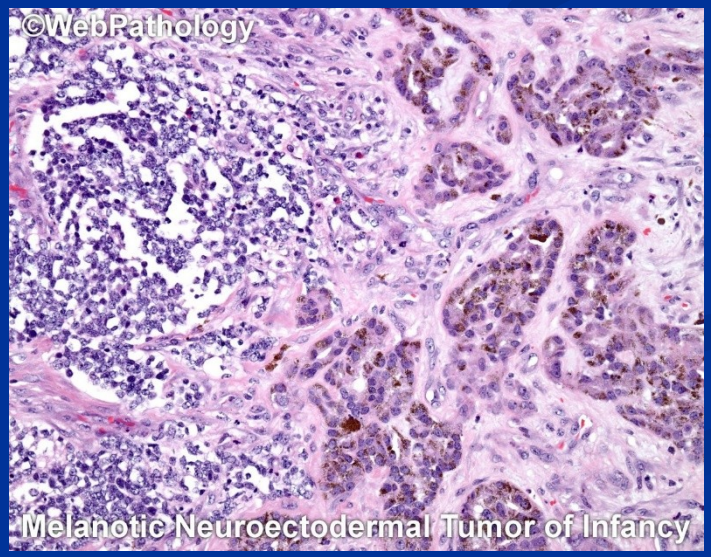
- 1st/2nd decade; predominantly anterior maxilla
- Developmental lesion resulting in the formation of a bag of discrete denticles
- Denticles comprise enamel, dentine, cementum, and pulp in their normal anatomical relationship
- Separate dinticles identifiable on radiograph



Melanotic neuroectodermal tumor of infancy

Synonyms: melanotic progonoma, retinal anlage tumor,…

- Very rare
- Tumor of infancy, 80 %<6 month, 95 % <1 year of age
- F:M: 2:1
- maxilla>mandible>skull
- Rapidly growing pigmented mass
- Local recurrences, metastatic (7 %): lymph nodes, liver, bones

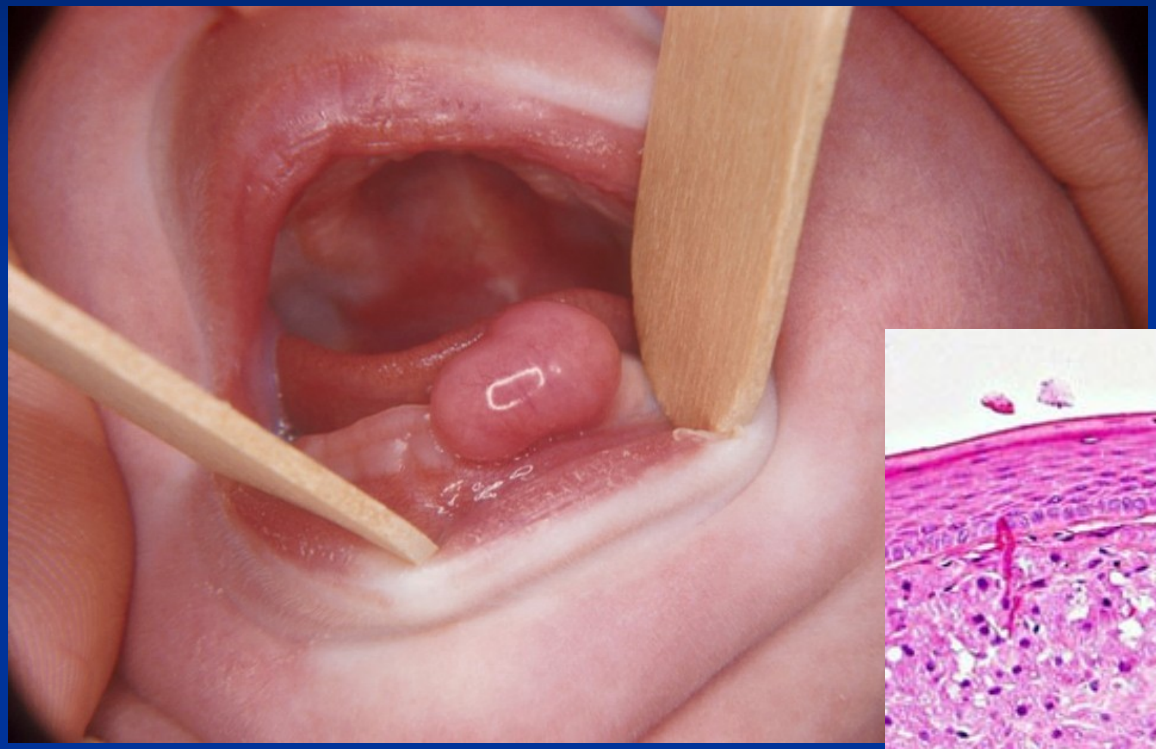


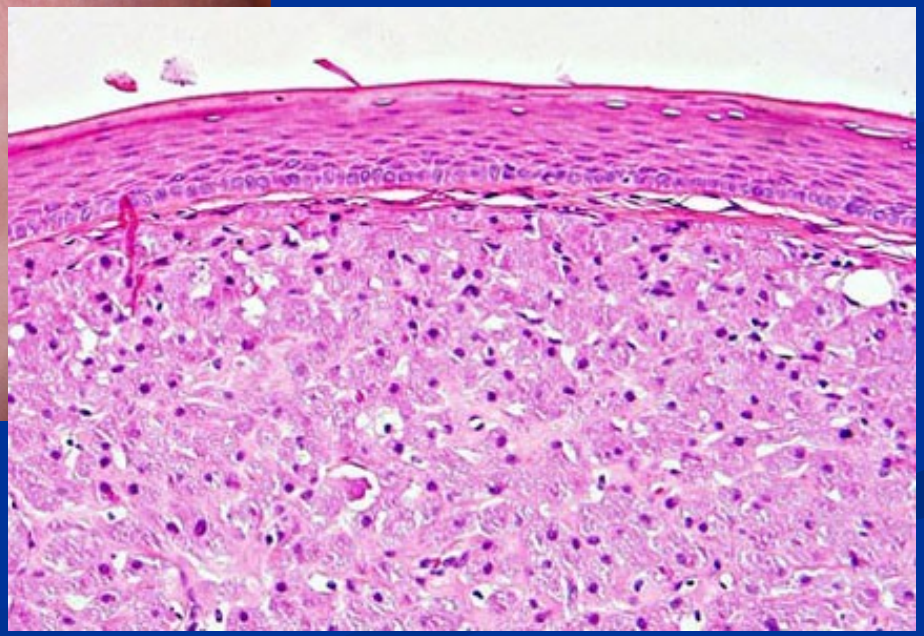
Congenital epulis of the newborns – congenital gingival granular cell tumor:

- Incisor region of the maxilla, F>M
- Closely packed granular cells covered by flattened squamous epithelium
- Benign lesion, unknown etiology

(reactive?,….neoplastic???, but unrelated to granular cell tumor of the tongue)

Congenital epulis of the newborns – congenital gingival granular cell tumor





Take home message

- Odontogenic tumors are rare, but occur
- NOT ONLY ameloblastoma
- Often benign x locally aggressive
- Not to rely only on radiography, histopathological examination of diagnosis is necessary!
- Secondary inflammation can cover some signs

Thank you for your attention……..