ODONTOGENIC TUMORS

DR. VISHAL BANSAL
Introduction

Neoplasia literally means "new growth."
A neoplasm, as defined by Willis, is "an abnormal mass of tissue the growth of which exceeds and is uncoordinated with that of the normal tissues and persists in the same excessive manner after the cessation of the stimuli which evoked the change."
• Malignant tumors are collectively referred to as cancers, derived from the Latin word for crab—that is, they adhere to any part that they seize in an obstinate manner, similar to a crab's behavior.

• Malignant, as applied to a neoplasm, implies that the lesion can invade and destroy adjacent structures and spread to distant sites (metastasize) to cause death.
ODONTOGENIC TUMORS

• Odontogenic tumors are lesions derived from the epithelial and/or mesenchymal remnants of the tooth forming apparatus.

• They are therefore found exclusively in the mandible and maxilla (and occasionally gingiva).
• The etiology and pathogenesis of this group of lesions are unknown. Clinically, odontogenic tumors are typically asymptomatic, although they may cause jaw expansion, movement of teeth, root resorption and bone loss.
Box 15-3 Classification of Odontogenic Tumors

A. Tumors of odontogenic epithelium
   1. Ameloblastoma
      a. Malignant ameloblastoma
      b. Ameloblastic carcinoma
   2. Clear cell odontogenic carcinoma
   3. Adenomatoid odontogenic tumor*
   4. Calcifying epithelial odontogenic tumor
   5. Squamous odontogenic tumor

B. Mixed odontogenic tumors
   1. Ameloblastic fibroma
   2. Ameloblastic fibro-odontoma
   3. Ameloblastic fibrosarcoma
   4. Odontoameloblastoma
   5. Compound odontoma
   6. Complex odontoma

C. Tumors of odontogenic ectomesenchyme
   1. Odontogenic fibroma
   2. Granular cell odontogenic tumor
   3. Odontogenic myxoma
   4. Cementoblastoma
Classification of odontogenic tumors of the jaws

A. Epithelial odontogenic tumors

1. Tumors producing minimal inductive changes in connective tissue
   - Ameloblastoma (adamantinoma)
   - CEOT (Pindborg tumor)
   - AOT (adenoameloblastoma)

2. Tumors producing extensive inductive changes in C.T.
   - Ameloblastic fibroma
   - Ameloblastoma fibro-odontoma
   - Odontoameloblastoma (ameloblastic odontoma)
   - Odontoma (compound-composite, complex-composite)
Mesodermal odontogenic tumor

1. Central odontogenic fibroma
2. Odontogenic myxoma (myxofibroma)
3. Cementoma:
   a. Periapical cemental dysplasia
   b. Cementifying fibroma
   c. Benign cementoblastoma
4. Dentinoma
C. Tumors of unknown origin
1. Melanotic neuroectodermal tumor of infancy (melanotic progonoma, retinal anlage tumor)

D. Malignant odontogenic tumors
1. Odontogenic carcinoma
   a. Primary intraosseous carcinoma
   b. Malignant ameloblastoma

2. Odontogenic sarcoma
   a. Ameloblastic fibrosarcoma (ameloblastic sarcoma)
   b. Ameloblastic odontosarcoma
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RADIOGRAPHIC PATTERN

SOAP BUBBLE - Large loculations
HONEY COMB - Small loculation

These are corticated cyst like spaces which are visible because of the alignment of the vessels in the directions of X-ray beam.

TENNIS RACKET APPEARANCE - Vertical septa instead of loculations present

SUNBURST APPEARANCE - Trabeculae radiating from the centre of the lesion resembling the spoke of the wheel

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Honey comb appearance

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EPITHELIAL TUMOURS

AMELOBLASTOMA

• It is usually unicentric, non functional intermittent in growth, anatomically benign and clinically persistent – Robinson.

• Adamantinoma- Malassez 1885
• Ameloblastoma – Churchill 1934
Pathogenesis

• This neoplasm originates within the mandible or maxilla from epithelium that is involved in the formation of teeth.
• epithelial sources include:
  • 1) The enamel organ,
  • 2) Odontogenic rests (rests of Malassez, rests of Serre),
  • 3) Reduced enamel epithelium,
  • 4) The epithelial lining of odontogenic cysts, especially dentigerous cysts.
  • 5) Disturbances of developing enamel organ
  • 6) Basal cells of surface epithelium of the jaws.
  • 7) Heterotrophic epithelium
ODONTOGENIC TUMORS AND THEIR SUSPECTED ORIGINS

AMELOBLASTOMA

BENIGN

• Solid/multicystic ameloblastoma → Remnants of dental lamina.

• Peripheral ameloblastoma → Remnants of dental lamina.

• Desmoplastic ameloblastoma → Epithelial rests of malassez.

• Unicystic Ameloblastoma → preexisting odontogenic cyst.
  ▪ Squamous odontogenic tumor → Epithelial rests of malassez located in PDL
  ▪ Calcifying epithelial OT → Rests of dental lamina

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- Adenomatoid OT → Dental lamina or its remnants

- Ameloblastic fibroma

- Ameloblastic fibro-odontoma

- Ameloblastic fibrodentinoma → intermediate stage of AF & AFO

- Complex odontoma → Unknown

- Compound odontoma → AF if they left untreated

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- Odontogenic fibroma → elusive and controversial origin

- Odontogenic myxoma → DP, DF & periodontal tissues possible germ center

- Cementoblastoma → Cellular cementum
MALIGNANT: ODONTOGENIC CARCINOMAS

- Metastasizing, Malignant Ameloblastoma → same as nonmetastasizing, usually preceded by local recurrences of tumor.

- Ameloblastic Carcinoma → Not clear, may originate from ex ameloblastoma

- Primary intraosseous squamous cell carcinoma(solid) → unknown

- PIOSCC from OC → unknown
UNICYSTIC AMELOBLASTOMA

- The unicystic ameloblastoma represents an ameloblastoma variant, presenting as a cyst.
- UA arises from pre-existing odontogenic cyst in particular a dentigerous cyst.

CLINICAL PRESENTATION

- Age: 1\textsuperscript{st} and 2\textsuperscript{nd} decayed
- Gender: male : female, 1.6 : 1 however, when tumor is not associated with an unerupted tooth gender ratio is reversed to male : female ratio 1:1.8.
- Location: jawbone marked predominance for mandible – posterior mandible, ascending ramus most often affected.
- Local swelling, occasional pain
- Discharge or drainage in case of secondary infection.

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To standardized the terminology of ameloblastomas associated with cyst and to recommend curative surgical approaches based on evidence of invasion:

- Ameloblastoma in situ

- Mural ameloblastoma in situ

**HISTOLOGIC SUBDIVISION OF UNICYSTIC AMELOBLASTOMA**

(Ackermann et al)

- 1 – Luminal UA
- 1.2 - Luminal and intraluminal UA
- 1.2.3 - Luminal, Intraluminal, and Intramural UA
- 1.3 – Luminal & Intramural UA

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Microinvasive ameloblastoma:

Intramural microinvasive

Transmural microinvasive

Invasive ameloblastoma:
arising from lining of a cyst

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RADIOGRAPHIC PRESENTATION

Unilocular – predominates
Multilocular

TREATMENT & RECURRENCE RATE

1 and 1.2 may be treated conservatively 
(by Ackerman et al)

1.2.3 & 1.3 – should be treated aggressively same as SMA

RR:
1- 25%
1.2- 10.7 %
1.2.3 & 1.3 No value ,poor follow up

Recurrence- many year after surgery

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- PIOSCC from KOC → unknown

- Clear cell odontogenic carcinoma → probably arises from residues of DL or rests of malassez

- Ghost cell odontogenic carcinoma → unknown
  - Ameloblastic fibrosarcoma → not be fully established

- Ameloblastic fibrodentinosarcoma and Fibro-odontosarcoma → unknown

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• For many years it was regarded as a single, distinct clinical entity with a broad spectrum of histological features. The tumour is slow growing, locally aggressive, and capable of causing large facial deformities. It has high recurrence rate if they are not widely and carefully excised. Metastasis is rare.

• In recent years, it is shown that not all lesions with histologic features ameloblastoma have the same potential for destruction, recurrence, and even metastasis.
• Three clinical subtypes of ameloblastoma are generally recognized for treatment purposes
  • (1) Common polycystic ameloblastoma
  • (2) Unicyctic ameloblastoma
  • (3) Peripheral (extraosseous) ameloblastoma.
CONVENTIONAL SOLID OR MULTICYSTIC INTRAOSSEOUS AMELOBLASTOMA

Clinical and radiographic features

- Third to seventh decades of life.
- No significant gender predilection.
- About 85 per cent of tumours occur in the mandible, of which some 70 per cent arise in the molar region and ascending ramus, 20 per cent in the premolar region, and 10 per cent in the incisor region.
- About 15 per cent of tumours occur in the maxilla.
- In the maxilla, most also occur in the molar region but about 15 per cent involve the antrum.
- The tumour is often asymptomatic and diagnosed during routine radiographic examination.
- Presents as painless swelling or expansion of the jaw. When untreated the lesion may grow slowly to massive proportions.
- Pain parasthesia are uncommon even with large tumours.
• **RADIOGRAPHIC FEATURE**

• Multilocular radiolucent lesion.
• “soap bubble” appearance when the radiolucent loculations are large
• “honeycombed” when the loculations are small.
• Buccal and lingual cortical expansion is frequently present.
• Resorption of the roots of teeth adjacent to the tumour is common. May be associated with an unerupted tooth often a mandibular third molar.
• Solid ameloblastomas may radiographically appear as unilocular radiolucent defects, which may resemble almost any type of cystic lesion, but they often show irregular scalloping.
Management of ameloblastoma

- MARGINAL RESECTION (1 ..1.5 CM )

With CONTINUITY DEFECT

without continuity defect

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DESMOPLASTIC AMELOBLASTOMA

- Is a benign locally infiltrative epithelial neoplasm believed to be a variant or subtype of the SMA

CLINICAL PRESENTATION

- Age—17-72 yrs
- More in male than female
- Maxilla – mandibular ratio 1:1 ratio, predominantly in mandibular anterior region
- The size of tumor varies between 1.0 and 8.5 cm in diameter
- Painless swelling in most of the cases initially

PATHOGENESIS

Oxytalan fibers identified in stromal tissue of 1 case reported by Kawai et al. This finding was interpreted by authors as indicating a tumor derivation from epithelial rests of malassez in peridontal membrane of a related tooth

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• The desmoplastic ameloblastoma has a marked predilection to occur in the anterior regions of the jaws, particularly the maxilla.

• Radiographically, this type seldom suggests the diagnosis of ameloblastoma and usually resembles a fibro-osseous lesion because of its mixed radiolucent and radiopaque appearance, which is due to osseous metaplasia within the dense fibrous septa that characterize the lesion, and not because the tumour itself is producing a mineralized product.
• HISTOPATHOLOGICAL FEATURES

• Conventional solid or multicystic intraosseous ameloblastomas show a remarkable tendency to undergo cystic change. Several microscopic subtypes of conventional ameloblastoma are recognized, but these patterns generally have little bearing on the behaviour of the tumor.
Radiographic Feature

HISTOPATHOLOGY

Animal like configuration or outline of the tumor is pathognomonic

Epithelial cells at the periphery are cuboidal with hyperchromatic nuclei

Central part of the tumor appear hypercellular

TREATMENT & RECURRENCE RATE

• Treatment : Resection

• RR – Acc to WHO 1992 classification, “DP Ameloblastoma has lower recurrence rate than other ameloblastoma”

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• FOLLICULAR AMELOBLASTOMA

• Also known as simple ameloblastoma,
• most common pattern.

• Islands of epithelium resemble enamel organ epithelium in a mature fibrous connective tissue stroma.

• The epithelial nests consist of a core of loosely arranged angular cells resembling the stellate reticulum of an enamel organ.

• A single layer of tall columnar cells (with nuclei at the opposite pole to the basement membrane i.e. reversed polarity) ameloblastoma-like cells surrounds this central core. Cyst formation is common and may vary from microcysts which form within the epithelial islands, to large macroscopic cysts several centimetres in diameter.

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• PLEXIFORM PATTERN

• Consists of long, anastomosing cords or larger sheets of odontogenic epithelium presenting as a network of interconnecting strands of cells.

• Each of these strands are bounded by columnar or cuboidal ameloblast-like cells surrounding more loosely arranged epithelial cells.

• The supporting stroma tends to be loosely arranged and vascular.

• Cyst formation is relatively uncommon in this variety, when it occurs, it is more often associated with stromal degeneration rather than cystic change within the epithelium.

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• **ACANTHOMATOUS AMELOBLASTOMA**

• When extensive squamous metaplasia, often associated with keratin formation, occurs in the central portions of the epithelial islands of a follicular ameloblastoma, the term acanthomatous is applied. Histopathologically it resembles squamous cell carcinoma or squamous odontogenic tumor.
• GRANULAR CELL AMELOBLASTOMA

• Ameloblastomas may show transformation of groups of lesional epithelial cells to granular cells having abundant cytoplasm filled with eosinophilic granules that resemble lysosomes ultrastructurally and histochemically.

• This variant has been seen in young patients and in clinically aggressive tumors
• DESMOPLASTIC AMELOBLASTOMA

• This type of ameloblastoma contains small islands and cords of odontogenic epithelium in a densely collagenised stroma. Immunohistochemical studies have shown increased production of the cytokine known as transforming growth factor β in association with this lesion, suggesting that this may be responsible for the desmoplasia.

• BASAL CELL AMELOBLASTOMA

• These lesions are composed of nests of uniform basaloid cells, and they may histologically look similar to basal cell carcinoma. No stellate reticulum is present in the central portions of the nests. Peripheral cells tend to be cuboidal rather than columnar. Less common variant.
• Salient features in treatment and prognosis

• Treatment options range from simple enucleation and curettage to en bloc resection. Recurrence rates of 50% to 90% have been reported after curettage and even 5-year disease free periods do not indicate a cure.

• Marginal resection is the most widely used treatment, but recurrence rates of up to 15% have been reported after marginal or block resection. Many surgeons advocate that the margin of the resection should be at least 1.0cm past the radiographic limits of the tumor.

• Ameloblastomas of posterior maxilla are particularly dangerous because of the difficulty of obtaining an adequate surgical margin around the tumor.
Some studies show that ameloblastomas may be radiosensitive, radiotherapy has seldom been used as a treatment modality because of the intraosseous location of the tumor and the potential for secondary radiation – induced malignancy developing in a relatively young patient population.

The conventional tumor is a persistent, infiltrative neoplasm that may kill the patient by progressive spread to involve vital structures. Most of these tumors, however, are not life-threatening lesions.
UNICYSTIC AMELOBLASTOMA

Unicystic ameloblastoma account for 10% to 15% of all intraosseous ameloblastomas. Whether it originates as a neoplasm or it is the result of neoplastic transformation of a non-neoplastic cyst epithelium is debatable. Both mechanisms probably occur, but proof of which is involved in an individual patient is virtually impossible to obtain.

Clinical and radiographic features

They are most often seen in younger patients, with about 50% diagnosed during second decade of life, average age 23 years. More than 90% of these tumors are seen in mandible, usually in the posterior region and are often asymptomatic, although large lesions may cause a painless swelling of the jaws.

This lesion typically appears as a circumscribed radiolucency that surrounds the crown of an unerupted mandibular third molar, clinically resembling a dentigerous cyst. The diagnosis is complete only after microscopic study of specimen.
• HISTOPATHOLOGICAL FEATURES

• Three variants have been described.

• 1) Luminal unicystic ameloblastoma- tumor confined to the luminal surface of the cyst. The lesion consists of a fibrous cyst wall with a lining that consists totally or partially of ameloblastic epithelium. A single layer of tall columnar or cuboidal cells (with nuclei at the opposite pole to the basement membrane i.e. reversed polarity and basilar cytoplasmic vacuolization. The overlying epithelium shows loosely arranged angular cells resembling the stellate reticulum of an enamel organ.

• 2) Intraluminal unicystic ameloblastoma

• One or more nodules of ameloblastoma project from the cystic lining into the lumen of the cyst. These nodules may be relatively small or largely fill the cystic lumen. In some cases, the nodule of tumor that projects into the lumen demonstrates an oedematous, plexiform pattern that resembles the plexiform pattern seen in conventional ameloblastomas-referred as plexiform unicystic ameloblastomas.

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3) Mural unicystic ameloblastoma

In this variant the fibrous wall of the cyst is infiltrated by typical follicular or plexiform ameloblastoma. The extent and depth of the ameloblastic infiltration may vary considerably. With any presumed unicystic ameloblastoma, multiple sections through many levels of the specimen are necessary to rule out the possibility of mural invasion of tumor cells.
• Salient features in treatment and prognosis
• These tumors are usually treated as cysts and enucleation is done. However patients are kept under long term followup. Recurrence rates of 10% to 20% have been reported after enucleation and curettage of conventional solid and multicystic intraosseous ameloblastomas
• **PERIPHERAL (EXTRAOSSEOUS) AMELOBLASTOMA**

• It is uncommon and accounts for about 1% of all ameloblastomas. It probably arises from rests of dental lamina beneath the oral mucosa or from the basal epithelial cells of the surface epithelium.

• **Clinical and radiographic features**

  • It is usually a painless, nonulcerated sessile or pedunculated gingival or alveolar mucosal lesion. The clinical features are nonspecific, and most lesions are clinically considered to represent a fibroma or pyogenic granuloma. Most examples are smaller than 1.5cm, but larger lesions have been reported. mostly seen in middle aged persons, with an average age of 52 years.

  • They are most commonly found on posterior gingival and alveolar mucosa, more common in mandibular than in maxillary areas. In some cases, the superficial alveolar bone becomes slightly eroded, but significant bone involvement does not occur.
PERIPHERAL AMELOBLASTOMA

- Also known as extra-osseous ameloblastoma, soft tissue ameloblastoma, ameloblastoma of mucosal origin or ameloblastoma of gingiva.

- Remnants of dental lamina & Surface epithelium

- It occurs in soft tissue overlying tooth bearing areas of maxilla & mandible

- Do not invade underlying bone.

**CLINICAL PRESENTATION**

- Age – men: 5th & 6th, women: 4th decayed, Gender – M > F

- Painless, sessile, firm and exophytic growth

- Surface relatively smooth but in several cases granular or pebbly or papillary or warty appearance.

- Size ranges from: 0.3 to 4.5 cm
Radiographic Feature

- No bony involvement
- Superficial bony depression-cupping or saucerization (caused by pressure resorption)

Histopathology

Epithelium composed of columnar basal cells & stellate reticulum are rarely visible

In number of cases of PA: clear cells or vacuolated cells occur as district clusters or in direct transition from ameloblastic tumor cells

Treatment & Recurrent Rate

- 1<sup>st</sup> treatment of choice is conservative supraperiosteal surgical excision with adequate free margins
- RR – 16 to 19%

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• **MALIGNANT AMELOBLASTOMA AND AMELOBLASTIC CARCINOMA**

  The frequency of malignant behaviour in ameloblastomas is difficult to determine but probably occurs in far less than 1% of all ameloblastomas.

• **MALIGNANT AMELOBLASTOMAS**

  Tumor that shows the histopathologic features of ameloblastoma, both in the primary tumor and in the metastatic deposits.

• **AMELOBLASTIC CARCINOMA**

  Tumor that has cytologic features of malignancy in the primary tumor, in a recurrence, or in any metastatic deposit. These lesions may follow a markedly aggressive local course, but metastases do not necessarily occur.
Clinical and radiographic features

Malignant Ameloblastomas – observed in patients who range in age from 4 to 75 years (mean age 30 years). For patients with documented metastases, the interval between the initial treatment of the ameloblastoma and first evidence of metastasis varies from 1 to 30 years.

Metastasis from ameloblastomas is most often found in the lungs which is sometimes regarded as aspiration or implant metastases. However, the peripheral location of these lung metastases suggests that they must have occurred by blood or lymphatic routes rather than aspiration.

Cervical lymph nodes are the most common site for metastasis of an ameloblastoma. Spread to vertebrae, other bones, and viscera has also been confirmed.

The radiographic findings of malignant ameloblastomas may be essentially the same as those in typical nonmetastasizing ameloblastomas. Ameloblastic carcinomas are often more aggressive lesions with ill-defined margins and cortical destruction.

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HISTOPATHOLOGICAL FEATURES

• Malignant ameloblastomas, the primary jaw tumor and the metastatic deposits show no microscopic features that differ from those of ameloblastomas with a completely benign local course.

• In ameloblastic carcinomas, the metastatic deposits or primary tumor shows the microscopic pattern of ameloblastoma in addition to cytologic features of malignancy. These include an increased nuclear to cytoplasmic ratio, nuclear hyperchromatism, and the presence of mitoses. Necrosis in tumor islands and areas of dystrophic calcification may also be present.

Salient features in treatment and prognosis

• The prognosis of patients with malignant ameloblastomas appears to be poor. Lesions designated as ameloblastic carcinoma have demonstrated a uniformly aggressive clinical course with perforation of the cortical plates of the jaw and extension of the tumor into adjacent soft tissues.
CALCIFYING EPITHELIAL ODONTOGENIC TUMOR (PINDBORG’S TUMOR)

Calcifying epithelial odontogenic tumor, widely known as the Pindborg’s tumor is an uncommon lesion that accounts for less than 1% of all odontogenic tumors. Although the tumor is clearly of odontogenic origin, its histogenesis is uncertain. The tumor cells bear close morphological resemblance to the cells of the stratum intermedium of the enamel organ. Some investigators have suggested that the tumor arises from dental lamina remnants based on its anatomic distribution in the jaws.

Clinical and radiographic features

It is most often encountered in patients between 30 and 50 years of age, no sex predilection. About 2/3rd of all cases have been found in the mandible, most often in the posterior areas. A painless, slow-growth swelling is the most common presenting sign.

Radiographically, the tumor shows a unilocular or, more often, a multilocular radiolucent defect. The margins of the lytic defect are often scalloped. The lesion may be entirely radiolucent, but the defect may contain calcified structures of varying size and density. The tumor is frequently associated with an impacted tooth, most often a mandibular third molar, with calcification around the crown of the impacted tooth.
RADIOGRAPHIC FEATURE

DIFFERENTIAL DIAGNOSIS

Common radiolucent-radiopaque lesion – calcifying odontogenic cyst

If present young patients – AFO & desmoplastic Ameloblastoma

Treatment & Recurrence
- Small tumor: conservative treatment
  - Large tumor: local resection

- Recurrence rate: Pindborg reported 14% RR – in case of inadequate t/t
• HISTOPATHOLOGICAL FEATURES

• Shows discrete islands, strands, or sheets of polyhedral epithelial cells in a fibrous stroma. The cellular outlines are distinct and intercellular bridges may be noted. The nuclei show considerable variation, giant nuclei and nuclear pleomorphism may be seen which is not indicative of malignancy. Large areas of amorphous, eosinophilic, hyalinised (amyloid-like) extracellular material is often seen. Tumor islands frequently enclose masses of this hyaline material; this results in cribriform appearance. Calcifications, which are a distinctive feature of the tumor, develop within the amyloid-like material and form concentric rings (Liesegang ring calcifications) which tend to fuse to form large, complex masses.

• Variants –

• Some tumors consist of large sheets of epithelial cells with minimal production of amyloid-like material and calcifications.

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• Others show large diffuse masses of amyloid like material that contain only small nests or islands of epithelium.

• A clear cell variant is described, in which clear cells constitute a significant portion of the epithelial component.

• Amyloid like material in this tumor has been extensively investigated by histochemical, immunohistochemical, and biochemical methods and electron microscopy. But its exact nature is still not known. Conflicting reports say it is either type iv collagen or keratin. The material as amyloid i.e. positive staining results with congo red staining it exhibits apple-green birefringence when viewed under polarized light.

• Salient features in treatment and prognosis

• It tends to be less aggressive compared to ameloblastoma. Conservative local resection to include a marrow rim of surrounding bone appears to be the treatment of choice. Lesions in the posterior wall of maxilla should be treated more aggressively. Recurrence rate of about 15% is seen, and overall prognosis is good.

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ADENOMATOID ODONTOGENIC TUMOR

• The adenomatoid odontogenic tumor represents 3% to 7% of all odontogenic tumors. It was formerly considered to be a variant of the ameloblastoma and was designated as “adenoameloblastoma”. Because of its slow growth and circumscription of the lesion it is sometimes classified as a hamartoma rather than a true neoplasm. Although there is evidence that the tumor cells are derived from enamel organ epithelium, investigators have also suggested that the lesion arises from remnants of dental lamina.
• Clinical and radiographic features

• These tumors are largely limited to younger patients and two thirds are diagnosed when patients are 10 to 19 years of age, uncommon over age of 30 years. It more commonly occurs in the anterior portions of the jaws and is found twice as often in the maxilla as in the mandible. Females affected twice as often as males.

• Most adenomatoid tumors are small and seldom exceed 3.0cm in greatest diameter. Peripheral extraosseous forms are seen but are rare. These usually appear as small sessile masses on the facial gingiva of the maxilla. Clinically it is difficult to distinguish these lesions from common gingival fibrous lesions.

• Frequently asymptomatic and are discovered during the course of a routine radiograph. Larger lesions cause a painless expansion of the bone. In about 75% of cases the tumor appears as a circumscribed, unilocular radiolucency that involves the crown of an unerupted tooth, most often a canine. This variety is known as follicular adenomatoid odontogenic tumor.

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• Less often it may be present as a well-delineated unilocular radiolucency that is not related to an unerupted tooth, but rather is located between the roots of erupted teeth (extrafollicular type).

• The lesion may appear completely radiolucent; often, however it contains fine (snowflake) calcifications. This feature may be helpful in differentiating the adenomatoid odontogenic tumor from a dentigerous cyst.
• HISTOPATHOLOGICAL FEATURES

• It is a well defined lesion that is usually surrounded by a thick, fibrous capsule. When the lesion is bisected, the central portion of the tumor may be essential solid or may show varying degrees of cystic change. Microscopically, the tumor is composed of spindle-shaped epithelial cells that form sheets, strands or whorled masses of cells in a scant fibrous stroma. The epithelial cells may form rosettelike structures about a central space, which may be empty or contain small amounts of eosinophilic material which may stain for amyloid.

• The tubular or duct like structures, which are the characteristic of adenomatoid tumor may be prominent, scanty, or even absent in a given lesion. These consist of a central space surrounded by a layer of columnar or cuboidal epithelial cells. The nuclei of these cells tend to be polarized away from the central space. The mechanism of formation of these tubular structures is not entirely clear but is likely because of secretory activity of the tumor cells, which appear to be preameloblast. These structures are not true ducts, and no glandular elements are present in the tumor.

• Small foci of calcification may also be scattered throughout the tumor, which may be interpreted as abortive enamel formation.

• Some adenomatoid tumors contain larger areas of matrix material or calcification interpreted as dentinoid or cementum.
• DIFFERENTIAL DIAGNOSIS
• Calcifying epithelial odontogenic tumor
• Odontoma
• Calcifying odontogenic cyst

• Salient features in treatment and prognosis
• This tumor is completely benign; because of its capsule, it enucleates easily from the bone. Aggressive behaviour has not been documented and recurrence after enucleation is seldom
EPITHELIAL ODONTOGENIC TUMOR

TUMORS PRODUCING EXTENSIVE INDUCTIVE CHANGES IN CONNECTIVE TISSUE

1. AMELOBLASTIC FIBROMA
2. AMELOBLASTIC FIBRO-ODONTOMA
3. ODONTO AMELOBLASTOMA
4. ODONTOMA
   • COMPOUND -COMPOSITE
   • COMPLEX- COMPOSITE
Ameloblastic fibroma

• Uncommon odontogenic neoplasm that produce inductive changes in connective tissue.
• Histomorphologic appearance to ameloblastoma
• Reported most frequently under the term “soft-mixed odontoma”
Pathogenesis

- Ameloblastic fibroma is epithelial origin, inductive effect stimulates tumor like proliferation of connective tissue, resulting in characteristics of secondary mixed tumor
• Clinical features:
  • 1.5 to 41 yrs (mean age 25 years)
  • Vast majority in mandibular posterior region

• Radiographic appearance:
  • Small lesion unilocular
  • Large lesion – multilocular

• Treatment considerations
  • Tumor expands by expansion and not by infiltration
  • Enucleation & curettage – 17% recurrence
  • Recurrence - resection

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AMELOBLASTIC FIBRO- ODONTOMA

• WHO defined as a neoplasm having general features of an ameloblastic fibroma but that also contains enamel and dentin.
• Odonto-ameloblastoma----a very rare neoplasm characterized by presence of enamel, dentin and odontogenic epithilium resembling that of an ameloblastoma, both in structure and behaviour.
• Ameloblastic fibro-odontoma-----much more common and less aggressive than odonto-ameloblastoma.

• Pathogenesis :-
• **Ameloblastic fibro-odontoma** appears to arise from an abnormal proliferation of odontogenic epithilium from permanent tooth germ. Abnormal proliferating epithilium exerts an organizing effect on mesodermal element with formation.
Clinical and radiological features (AFO)

- Usually in children with an average age of 10 years,
- rarely seen in adults.
- Ameloblastic fibro-odontomas occur more frequently in the posterior regions of the jaws, (maxilla=mandible)
- no gender predilection.
- Expansile lesion that exhibits little tendency to infiltrate bone
- Grows slowly, but capable to attain large size
- Swelling, mild pain, delayed eruption of teeth

Radiologically,
- Tumor shows a well-circumscribed unilocular or, rarely, multilocular radiolucent defect that contains a variable amount of calcified material with the radiodensity of tooth structure. The calcified material within the lesion may appear as multiple small radiopacities or as a solid conglomerate mass. An unerupted tooth is present at the margin of the lesion, or the crown of the unerupted tooth may be included within the defect.
HISTOPATHOLOGICAL FEATURES

• Soft tissue components are similar to that of ameloblastic fibroma. The calcifying elements consists of foci of enamel and dentin matrix formation in close relation to the epithelial structures.

• The more calcified lesions show mature dental structures in the form of rudimentary small teeth or conglomerate masses of enamel and dentin. A similar tumor in which the calcifying component consists of only dentin matrix and dentinoid material has been designated by some as ameloblastic fibro-dentinoma.
Treatment

• Careful curettage of all tumor and capsule fragment because of peripheral tissue is often the site of more active, proliferating ameloblastic component.

• The lesion usually separates easily from its bony bed.

• The tumor is well circumscribed and does not invade the surrounding bone.

• Prognosis is excellent, and recurrence after conservative removal is unusual.
ODONTOMA

• Tumor of odontogenic origin
• Growth in which both epithelial & ectomesenchymal cells exhibit complete or incomplete differentiation of tooth formation.
• Composite word to designate which contains four dental tissues (enamel, dentin, cementum, pulp).
• Pathogenesis - unknown. Most acceptable theory is that odontomas develop from enamel organ or from dental lamina in place of normal tooth or possibly from supernumerary lamina in association with follicle of unerupted tooth.

• Classification:

  Compound – composite odontoma: it consists of formed calcified tooth like structures or miniature dwarfed teeth.
complex:

It is a malformation in which all dental tissues are represented with the individual tissues being well formed but occurring in a disorderly pattern.

Irregular mass bearing no morphologically similarity to the rudimentary teeth.

Haphazard arrangement of calcified dental structure.
• Age: 1-2 decades (10-70yrs)
• Sex: M=F
• Site: Max = Mand
• Complex: mandible 67% (Posterior)
• Compound: maxilla (Anterior)

Generally asymptomatic, slow growing, noninfiltrating malformation, once enlarges than notice. Interference of odontoma—cause failure of eruption of permanent teeth.

RADIOGRAPHIC FEATURES:
COMPOUND ODONTOMA:
Radiopaque Mass with anatomic similarity to normal tooth. lesion is well demarcated from the surrounding bone by a thin radiolucent line representing the follicular capsule.
• **Complex odontoma**: small, large, huge, irregular or ovoid smooth densely radiopaque mass surrounded by a radiolucent zone.

• **Mixed radiolucent /radiopaque lesion.**
• Asymptomatic
• No bony expansion
• Commonly detected on radiographs
• Associated unerupted or impacted teeth.

**MANAGEMENT:** Surgical Removal

**RECCURRENCE:** Rare
Mesodermal odontogenic tumors

1. Central odontogenic fibroma
2. Odontogenic myxoma (myxofibroma)
3. Cementoma
   • Peripheral cemental dysplasia
   • Cementifying fibroma
   • Benign cementoma
4. Dentinoma
Central odontogenic fibroma

• Extremely rare benign neoplasm

• Central odontogenic fibroma and odontogenic myxoma are essentially similar, varying only in maturity

• Pathogenesis:-

• derived from one of the mesenchymal components of tooth germ, either the dental follicle, dental papilla or PDL
CLINICAL FEATURES

- Age----11-67 yrs
- Male=female
- Progressive enlargement of jaw---with accompanying facial deformity, resulting displacement of adjacent tooth
- More predilection on mandible
Radiographic features

• Multilocular radiolucent lesion that involve relatively large portion of jaws, associated with unerupted or displaced teeth.

• **Histology:-**

  • Similar to myxoma

  • More abundance of collagen and great cellularity of odontogenic fibroma
Treatment

• Enucleation and aggressive curettage appear to be treatment of choice (tumor in most instances is not encapsulated, the fibrous mass appears to separate easily from surrounding osseous tissue)
Odontogenic myxoma (myxofibroma)

• Mesenchymal neoplasm thought to be arise from mesodermal portion of tooth germ

• **Pathogenesis:** classified in two different varieties

  • (a) odontogenic:- arise from mesenchymal papilla of developing tooth, either before or after calcification has begin.

  • (b) osteogenic type:- origin from disturbed mesenchymal focus in the bone, arise away from tooth bearing area of jaw, behaved more aggressively.

• Myxomas of the jaws to be odontogenic fibroma that have undergone myxomatous degeneration
Clinical features

- Common in 2nd and 3rd decades
- Male=female
- Maxilla is more common than mandible
- Posterior region affected more than anterior region
- Asymptomatic swelling of the jaws, in which overlying mucosa is uninvolved.

- MACROSCOPICALLY:
  - Gelatinous texture, grey to white Shiny material

- HISTOLOGICALLY:
  - Stellate shaped cells with long anastomosing cytoplasmic processes in a granular, basophilic ground substance, rich in hyaluronic acid, Odontogenic myxoma are active cells synthesizing myxoid material and small number of collagen fibers.
RADIOGRAPHIC FEATURES:
• Honey comb appearance
• Tennis racket appearance---because of fine trabeculae of bone transverse the lesion giving it a compartmentalized appearance
• Soap bubble appearance

DIFFERENTIAL DIAGNOSIS:
• Ameloblastoma
• Haemangioma

TREATMENT: nonencapsulated, infiltrating and jelly like consistency, difficult to eradicate by local management, Wide Surgical excision of the tumor, resection with or without continuity defect.

RECURRENCE:33% (Long term follow up)

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CEMENTOMA

• Cementomas are diversified group of nonrelated lesion producing cementum like material
• Cementum is a complex calcified tissue similar to bone both in morphology and in composition
Periapical cemental dysplasia

- Most common cementum producing lesion
- Mostly seen in blacks
- Oftenly---multiple lesions----
- Approx. 77% in mandibular anterior region Or cementoma, periapical osteofibrosis, periapical dysplasias
Radiographic features

• Lesion has 3-distinct stages

• 1\textsuperscript{st} ---”Osteolytic” stage------radiolucent like lesion, surrounding apex of teeth

• D/D-----periapical granuloma

• 2\textsuperscript{nd} ---”Intermediate” stage-----referred as cementoblastic, in which lesion is partially calcified and shows central area of radiopacity
• IIIrd ----”Mature” stage----completely radiopaque lesion

• Surrounded by thin radiolucent line

• Periapical cemental dysplasia has limited growth potential and rarely attains a size larger than 5mm
Treatment

• Observation only------because of limited growth capacity
• Surgical removal renders the adjacent teeth non-vital
Cementifying fibroma

• Previous thoughts:- it is a bone reactive lesion such as fibrous dysplasia
• Current thoughts:- odontogenic neoplasm

Pathogenesis:-
Arise from undifferentiated mesenchymal cells found in PDL
CLINICAL FEATURES

• Centrally located lesions
• Slow growing, expansile lesion
• Most commonly found in premolars or molar region of middle aged persons
• 3/4th occur in mandible
• Lesion is asymptomatic until growth produces a noticeable swelling and mild deformity
• Displacement of teeth in the area of tumor is a common sign
Radiographic features

• Early stage:- unilocular (commonly), multilocular, well demarcated and circumscribed from surrounding bone

• As lesion matures, there is increasing calcification with varying amount of radiopaque material becoming dispersed throughout radiolucent area.
Pathologic features

• Mixture of cellular connective tissue and scattered rounded masses or droplets of cementum

• In mature cementifying fibroma, cementoblasts may be prominent
Surgical considerations

• Expansile growth that shows no tendency toward infiltrating growth pattern

• Small lesion------conservative therapy (enucleation and curetage)

• Unlimited growth capacity-----when attain large size, chances of pathological fractures and resection may be the treatment.
Benign cementoblastoma
(true cementoma)---true neoplasm of tooth cementum

• Pathogenesis:-
Mesenchymal odontogenic tumor originates from connective tissue of PDL and/or apical portion of dental follicle

• Clinical features:-
Painless, bony expansion, common in mandible
Lesion may cause pain due to pressure on contents of inferior alveolar canal
Radiographic features

• Solitary radiopacity, confluent with tooth root
• Evidence of root resorption is a common finding
• Immature——radiolucent
• Mature——central area may have “sun burst” appearance with uniform peripheral radiolucent zone.
Treatment

- Small lesion: generally delivered by extraction of offending tooth
- Large lesion: enucleation and curettage
DENTINOMA

• Rare odontogenic tumor, predominantly of dentin with some soft tissue and cementum. Contains no enamel

• 2 types- 1. immature dentinoma (fibroameloblastic type) primarily soft tissue components.
  2. mature type(mature dentinoma) primarily of calcified tissue.
• PATHOGENESIS- Lack of odontogenic epithelium. Inductive theory suggests odontogenic epithelium is not present it must have existed at early stage of tumor development, exerting an organizing influence on mesenchymal cells and inducing them to lay down dentin matrix.

• Clinical and radiographic features-
  age  4-55 yrs.
Mandibular molar and angle region has more predeliction.
Well defined radiolucency with varying degree of radiopacity

**Treatment:**- complete removal and aggressive curettage
(although rare tumor ---enough literature isn’t available in terms of treatment and recurrence)

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Tumors of unknown origin

Melanotic neuroectodermal tumour of infancy (melanotic progonoma, retinal anlage tumor)

• Clinically distinctive benign neoplasm of neuroectodermal in origin
• Never congenital, develop in infancy.
• Originates – neural crest cells overgrowth
Malignant odontogenic tumors

• 1. odontogenic carcinoma
  a. Primary intraosseous carcinoma
  b. Malignant ameloblastoma

• 2. odontogenic sarcoma
  a. Ameloblastic fibrosarcoma
  b. Ameloblastic odontosarcoma
Extreme rare malignancy
Develops from bone from odontogenic epithilium or enclaved epithilium at site of fusion of embryonic facial process

**CLINICAL FEATURES:-**
- Mandible is more common than maxilla (75-80%)
- 6\textsuperscript{th}-7\textsuperscript{th} decades of life
- M:F = 2:1
- Metastasis in local lymph nodes or sometimes distant metastasis
Pathologic features

• Absence of prickle cell layer suggests origin from odontogenic epithilium

• Surgical consideration:-
  Treatment just like sq. cell carcinoma
  1. Wide excision+ neck dissection
  2. Radiation therapy------better response
Primary intraosseous carcinoma (a/c to ELZAY)

• 3-types:-
  1. Those arising from odontogenic cyst
  2. Those arising from ameloblastoma
     a. Well differentiated malignant ameloblastoma
     b. Poorly differentiated (ameloblastic carcinoma)
  3. Arising de novo from odontogenic epithilium residue
• Malignant ameloblastoma: -
  Ameloblastoma which has given evidence of truly malignant behaviour, judged chiefly by occurrence of metastasis, but in which metastatic lesions have shown no histologic differences from primary tumor.

• Ameloblastic carcinoma: -
  Ameloblastoma in which there has been obvious histologically malignant transformation of epithelial component and in which tumor behaved in malignant fashion so that metastatic lesions don’t bear resemblance to primary odontogenic tumor.
Clinical features and pathogenesis

1. Cytologically benign (basic pattern of ameloblastoma + hypercellularity + cellular atypia)
2. Locally aggressive
3. Undergo regional lymph nodes metastasis, but it is not required to show metastasis to confirm the diagnosis.
4. Large tissue masses with ulceration, significant bone resorption, tooth mobility
Diagnostic workup

1. Neck examination
2. CT
3. Chest radiography

Generally intraosseous tumors-----T4 by location

Treatment:- 2-3 cm wide excision either prophylactic or therapeutic neck dissection
Malignant ameloblastoma

- Two main routes of metastasis: lymphatics and blood vessels
- Regional L.N’s are most frequent site of metastasis

- **Surgical treatment:**
  wide excision and neck dissection
Odontogenic sarcoma

- Ameloblastic fibrosarcoma:

Defined as a neoplasm with a structure similar to the ameloblastic fibroma—but in which mesodermal component shows features of malignancy
Clinical features

• 2\textsuperscript{nd} and 3\textsuperscript{rd} decade
• More common in mandible
• Pain, swelling, ulceration and paresthesia
• Moderately rapid growth pattern

Radiographic features:
Extensive area of destruction with irregular and indistinct margins
Sometimes --- multilocular radiolucencies
Surgical consideration

• Wide surgical excision with soft tissue + chemotherapy (Actinimycin D, Vincristine, Cyclophosphamide)
Ameloblastic odontosarcoma

• Similar to ameloblastic sarcoma, but in which limited amount of dentin and enamel

• Surgical management:-
  clinical behaviour and surgical management similar to ameloblastic fibrosarcoma
THANKYOU