LECTURE # : 12

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Above is a photograph showing an alveolar bone swelling. It is not an inflammatory swelling because it is not red, it’s painless, wasn’t rapid to develop, and it caused mobility of teeth.

**DDx:**
- Giant cell lesion.
- Central giant cell granuloma.
- Bone tumors (because the swelling was from a bony origin, gingiva and CT tumors were excluded).
- Odontogenic tumors.
- Cystic lesions (some cause bone expansion).
- Fibro-osseous lesions (can cause progressive enlargement of bone).

**DX:**
*Odontogenic tumor*, in the lower right region, with swelling buccally and lingually and bone expansion without causing ulceration. In such cases, more investigations such as radiographs are required, and the area should be explored with biopsies.

Some appear extra-oraly. After excluding infections, you are left with the diagnosis of bone swellings, which call for more investigations.

**Odontomes**

- Odontomes by definition are hamartomas/developmental anomalies of teeth.
- Hamartomas: collections of dental tissues that do not look like teeth.
- *Few are classified as Odontogenic tumors (neoplasms).*

- Odontomes vs odontogenic tumors:
  Odontomes are hamartomas, not neoplasms.
Types of odontomes:

(1) Invaginated Odontomes (aka Dens Invaginatus), divided into coronal and radicular types.
(2) Evaginated Odontomes (aka Dens Evaginatus)
(3) Enamel pearl (aka enameloma)
(4) Complex and Compound Odontomes.

<table>
<thead>
<tr>
<th>Clinical Presentation</th>
<th>(1) Invaginated Odontomes/Dens Invaginatus (Coronal IO)</th>
<th>(2) Evaginated Odontomes/Dens Evaginatus</th>
</tr>
</thead>
<tbody>
<tr>
<td>- Deepening of the cingulum/lingual pit, giving the appearance of a tooth developing inside the tooth (dense in dente).</td>
<td></td>
<td>- Presents as an extra/accessory cusp, either in the cingulum area, or between cusps.</td>
</tr>
<tr>
<td>- Mainly involves the palatal surfaces of upper lateral incisors, often bilateral.</td>
<td></td>
<td>- This is sometimes associated with pulp extension; pulpitis.</td>
</tr>
<tr>
<td>- Asymptomatic. Pt. presents with cingulum caries, since these areas retain plaque and are difficult to clean. When felt with a probe it shows a very deep cingulum pit.</td>
<td></td>
<td>- Mainly affects central incisors and premolars. When seen in upper anterior teeth, they’re referred to as Talon cusps; a cusp on the cingulum area.</td>
</tr>
<tr>
<td>- Variable in degree; most cases are minor where clinically the shape of the tooth will not differ and we only observe caries.</td>
<td></td>
<td>- There are some racial differences (monogoloid) such as: In lower premolars, an extra cusp is commonly present in the groove between the buccal and lingual cusps in some areas of Asia. This cusp is more prominent than the B &amp; L cusps; more prone to attrition and fracture, and because it has pulpal extention, pulpitis is expected even before closure of the root (early stages).</td>
</tr>
<tr>
<td>- In severe cases, the lateral incisor looks dilated; change in tooth shape.</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
| Diagnosis          | a) Caries at the cingulum area.  
|                   | b) Pulpitis or a periapical lesion, if caries were left for long.  
|                   | c) Accidental radiographic finding.  
|                   | d) Chronic alveolar abscess, which is a presentation of IO, where we can see a sinus, parulis (associated with chronic alveolar abscess).  
|                   | e) The patient may present with dilated teeth.  
|                   | f) Invaginated odontomes.  

Consequences of the extra cusp with extending pulp in evaginated odontomes:

1. Easy attrition.
2. Prone to early pulp exposure and pulpitis i.e before root completion (open apex).

| Radiographic Examination | Shows:  
|--------------------------|---------------------------------------------------------------  
|                          | - An invagination that is lined with enamel and is continuous with the surface.  
|                          | - Variable in degree of severity.  
|                          | - Appearance of a tooth within a tooth (dens in dente), causing dilatation of tooth; distant or dilated odontome.  

| Histologically | - The invagination is lined by hypomineralized enamel and defective dentine, especially at the base, which cause cracks facilitating easy communication with the pulp; faster formation of pulpitis.  
|               | - Enamel could be absent/deficient in some areas.  

Before tooth eruption, the follicle around the tooth is filled with connective tissue extending to the inside, however after eruption, the dissolution of that connective tissue leads to plaque retention and eventually pulpitis.

<table>
<thead>
<tr>
<th>Pathogenesis</th>
</tr>
</thead>
<tbody>
<tr>
<td>- Unknown.</td>
</tr>
<tr>
<td>- The cingulum pit appears during the development of the dental papillae and enamel organ, so it is thought to be due to active proliferation of the enamel organ deeper than necessary into the dental papillae representing the early pulp.</td>
</tr>
</tbody>
</table>

A case of an invaginated odontome:
The figure shows a deep lingual pit on the cingulum that is more extended than normal.

So Coronal Invaginated odontomes have different types/degrees:

<table>
<thead>
<tr>
<th>Type 1:</th>
<th>Type 2:</th>
<th>Type 3:</th>
</tr>
</thead>
<tbody>
<tr>
<td>Observed as a deep invagination within the crown, above the cementoenamel junction.</td>
<td>Deeper/more severe, and is below the cementoenamel junction; reaching the root.</td>
<td>Extends towards the apex of the tooth reaching the periodontal ligament space. Seen as a tooth is developing inside a tooth.</td>
</tr>
</tbody>
</table>

Conclusion:
- The severity is variable, and as the severity increases, the shape of the tooth becomes more different and more dilated; dilated odontome- The tooth becomes conical in shape.
Invaginated odontomes are very common, and you might come across them daily in your clinic.

Examples on Type 2 Coronal Invaginated Odontomes:

1) Notice how the extension is lined with enamel (opaque line), and is adjacent to the pulp therefore plaque accumulation of plaque and caries can reach the pulp quite easily.

2) Notice how the root appears conical in shape.

➢ Radicular Invaginated Odontome are rare and further divided into:

1. Axial infolding:
   - Appears as an axial groove
   - e.g. the tooth has one root, and appears to be dividing into two roots.

2. Saccular invagination:
   - Appears as a hole in the root with the invagination inside of it.
   - The sac extends to the root area, and is lined by enamel even in the root.

Q: What is the source of this enamel that appears in the root area?

A: Hertwig epithelial root sheath.
(3) Enamel pearl (aka enameloma, a misnomer)
- Small droplet of enamel, most commonly seen at the area bifurcation of maxillary molars.

**Clinically:**
- Situated below the gingiva; cannot be seen, and therefore asymptomatic.
- Usually noticed after extraction of the affected tooth.

**Radiographically:**
- May be seen in radiographic images as an area of radiopacity.

**Histologically:**
E, E & D, E & D & P
May be completely made of enamel, or both enamel and dentine, and may also have a pulpal extension inside.

**Pathogenesis:**
Budding of ERSH

- In cases of periodontitis, the progression of the disease in areas with enamel pearls is faster, and a deeper pocket is observed because this area will be more difficult to clean; the nodule aids in plaque accumulation.

(4) Compound and Complex Odontomes

- Classified under odontogenic tumors
- As mentioned before, they are hamartomas and not tumors.

- Hamartomas reach a fixed size, and therefore do not resemble tumors in their pattern of growth. Hamartomas are benign

- Growth of these odontomes usually takes place in the first and second decades of life (before 20 years of age).

- Compound and Complex Odontomes are usually associated with permanent teeth, with a mean age of onset/presentation of 14-15 years.
**Clinically:**

<table>
<thead>
<tr>
<th>Compound odontomes</th>
<th>Complex odontomes</th>
</tr>
</thead>
<tbody>
<tr>
<td><img src="image1.png" alt="Image" /></td>
<td><img src="image2.png" alt="Image" /></td>
</tr>
<tr>
<td>- Look like a sac of several tooth-like structures/denticles.</td>
<td>- Mass of haphazardly arranged enamel, dentine, cementum and pulp.</td>
</tr>
<tr>
<td>- These structures are made of enamel, dentine and pulp but are smaller and of a different shape than normal teeth.</td>
<td>- Do not resemble teeth</td>
</tr>
<tr>
<td>- surrounded by a radiolucent well defined margin, which and this helps in clinical management and removal.</td>
<td>- surrounded by a radiolucent margin</td>
</tr>
<tr>
<td>- They are not expected to recur after removal.</td>
<td>- hard mass surrounded by a capsule so is easily removed by surgery</td>
</tr>
<tr>
<td>- Most common site of their formation is in the inter-canine region of the maxilla/anterior maxilla, but they could affect other areas.</td>
<td>- Cause complications to adjacent teeth, such as dilacerations, ectopic eruption or prevention of eruption.</td>
</tr>
<tr>
<td></td>
<td>Most commonly occur in the premolar and molar region in the mandible, but can arise in other areas as well.</td>
</tr>
</tbody>
</table>

**Diagnosis:**
- It is occasionally a **chance radiographic finding**, before any symptoms arise.

- May cause **bone expansion**:
  The patient presents with bone expansion in that region, or complaining of a missing tooth (odontomes may replace missing teeth), impaction, or delay of eruption.

- Rarely, a patient might present with an **erupted odontome** (looks like exposed bone).
Radiographic imaging:
- Initially, they show no calcification (just like teeth).
- In very early stages, it presents as a radiolucent lesion with some radiopacity.
- As time passes it becomes more calcified.

At the end stage:

<table>
<thead>
<tr>
<th>Compound odontomes</th>
<th>Complex odontomes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Appear as unilocular radiolucencies</td>
<td>Appear as a complex, solid, radiopaque</td>
</tr>
<tr>
<td>containing multiple small denticles,</td>
<td>mass with a radiolucent zone.</td>
</tr>
<tr>
<td>variable in number.</td>
<td></td>
</tr>
</tbody>
</table>

Histologically:

<table>
<thead>
<tr>
<th>Compound odontome</th>
<th>Complex odontome</th>
</tr>
</thead>
<tbody>
<tr>
<td>- Denticles are separated by fibrous</td>
<td>- They appear cellular in very early stages.</td>
</tr>
<tr>
<td>tissue.</td>
<td>- Disorganized/haphazardly arranged well-formed</td>
</tr>
<tr>
<td>- Enamel dentine, cementum, pulp are</td>
<td>masses of enamel, dentine, cementum and pulp.</td>
</tr>
<tr>
<td>normally arranged and surrounded by</td>
<td></td>
</tr>
<tr>
<td>follicles.</td>
<td></td>
</tr>
</tbody>
</table>

- All features resemble teeth, except in that they are multiple with different shapes and sizes.
Odontogenic tumors

- By definition: tumors that develop from dental tissues; enamel organ, dental papilla, or any part of the tooth can develop a tumor.
- Odontogenic tumors are classified into either benign or malignant.

Benign tumors: most common

- Benign tumors are further divided according to the origin of the tissue causing the tumor into: epithelial, mesenchymal, or mixed lesions.

Keratocysts were newly classified as odontogenic tumors, and are now called Keratinizing Cystic Odontogenic tumors. The last classification with keratocysts as tumors was released in 2005. It’s been debated that it should be classified back into a cyst, but no official update for the classification was made up to this year. Recall that keratocysts are the most important cysts in regards to prognosis, with a high recurrence rate.

Most common/important odontogenic tumor: Ameloblastoma

> **Most common odontogenic tumor debate:**
> If you believe in the classification of keratocysts as tumors, then Keratinizing Cystic Odontogenic tumors are the most common odontogenic tumors, otherwise, it’s ameloblastoma.

We are going to discuss many tumors, some are rare with a percentage of less than 1% of all odontogenic tumors, so you need to focus more on ameloblastomas and odontomes (classified as tumors) because they are the most common. Adenomatoid odontogenic tumors and calcifying odontogenic epithelial tumors are also common. Other tumors are very rare and you’re likely to never face them.
Epithelial Benign lesions:

1. Without odontogenic mesenchyme.
   - Mainly originate from the odontogenic epithelium.
   - Most important and most common type is **Ameloblastoma** (as important as pleomorphic adenomas in the salivary glands)
   - Other types that we might encounter are listed in the figure above.

2. With odontogenic mesenchyme (aka mixed tumors)
   Where the tumor originates from both odontogenic epithelium and mesenchyme of teeth.

   - Some tumors are purely mesenchymal in origin, such as:
     - Odontogenic fibroma
     - Odontogenic myxoma
     - Cementoblastoma, an interesting tumor, and the only tumor of the cementum.

Malignant tumors: more rare
- All tumors mentioned previously can have a malignant counterpart

A. Odontogenic carcinomas
   1- Malignant ameloblastoma
   2- Ameloblastic carcinoma.
   3- **Primary intraosseous squamous cell carcinoma.**
      This is a lesion inside the bone showing features of squamous cell carcinoma.
   4- Malignant variant of other epithelial tumors.
   5- Clear-cell odontogenic carcinoma.
   6- Malignant change in odontogenic cysts.

B. Odontogenic sarcomas
   - Ameloblastic Fibrosarcoma
   - Ameloblastic fibro-odontosarcoma

Tumors of debatable origin: some say odontogenic, and

1- Melanotic neuroectodermal tumor of infancy
2- Congenital gingival granular cell tumor (congenital epulis)

Recall:
There is a granular cell tumor that affects the tongue.

Types of Epulis:
- Fibrous
- Vascular/pyogenic/pregnancy
- Giant cell
This table shows a study of the prevalence of odontogenic tumors.

- Ameloblastomas: 50%
- Odontomes: 13%
- Adenomatoid odontogenic tumors: 4%
- Other tumors are rare to develop (less than 1%).

Keratocysts/KCOTs are actually more prevalent than ameloblastomas, but because these studies are hospital based, cystic lesions are not sent to the lab.

### Ameloblastoma
#### solid/multicystic

Management: resection of tumor with a border of normal tissue.

- **Multicystic:** multiple cysts inside tumor.
- The term Ameloblastoma refers to the solid, multicystic type and not the unicystic type.

- Ameloblastomas are the most important, and common odontogenic tumors.
- A portion of patients with bone swellings is expected to have ameloblastoma.

- Ameloblastomas are **benign but locally invasive** tumors.
- Local invasion indicates a **high recurrence rate** and more difficult surgery where safety margins need to be considered to reduce the LRR.

### Clinically:

- **Age of onset:**
  Most commonly occurs between the ages 30-60 (middle-age), but can occur at any other age. However it is not commonly seen below 20 years of age.

- **Gender:**
  No gender variations.

Management: resection and bone grafting. Since it is invasive, safety margins should be taken and therefore a part of the mandible is also removed.
Site:
Most common site is the **posterior mandible/body of angle of mandible** (same as keratocysts)

Pattern of growth:
Slowly and gradually grows over time, and can reach huge sizes, especially in areas of poor education and no early management.

Ameloblasts are **massive tumors**.

If not dealt with, it might continue to perforate the bone and extend into soft tissues, making the management more difficult in locating the margins.

Teeth:
It’s an asymptomatic tumor at the beginning, but can later cause **dental symptoms** associated with the area of expansion such as displacement, resorption, tilting, drifting, mobility and impaction of nearby teeth.

Might affect occlusion.

Behaviour:
Even in very severe cases, **ulceration of the skin doesn’t take place** since it’s not malignant. **May perforate the bone.**

Ameloblasts don’t perforate bone because they are slow-growing tumors. Usually, malignant/fast growing tumors cause perforations because the bone isn’t given a chance to compensate with bone formation from the outside. Slow-growing tumors end up with huge sizes without perforation of the cortex; no perforations in ameloblastomas.

Perforations can be seen in some types of radiographs, and they are indicative of **poor prognosis**, because they mean the tumor is progressing into the soft tissues, and margins of the tumor aren’t clear within soft tissues, making for a higher LRR after surgery.

(Perforations → higher LRR)

**Radiographic imaging:**
- Shows:
  1) multilocular radiolucency, with a soap bubble appearance.
  2) Root resorption.
  3) May be associated with impacted teeth.

- In some cases it might appear **unilocular**, however they’re usually multilocular. In these cases they might be confused with **cystic lesions**, but **biopsies verify ameloblastoma**.

DDx: keratocysts and other odontogenic tumors.

Resorption is more common with keratocysts, whereas bone expansion is more common with ameloblastomas. Keratocysts only cause expansion in late stages.