NEXT GENERATION LEARNING

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Creating a Better Pathologist
How to Diagnose the Most Common Odontogenic Lesions

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No Conflicts of Interest to Report
Objectives

✓ Learn to utilize the radiograph and clinical information in odontogenic lesions to help narrow the differential diagnosis

✓ Understand the associated clinical differential diagnosis and the associated treatments of each

✓ Consider the most common entities in the differential diagnosis
Odontogenic Cystic Lesions

1) Keratocystic odontogenic tumor
2) Dentigerous cyst
3) Ameloblastomas with cystic changes
4) Radicular (periapical) cyst
5) Hyperplastic dental follicle

Fibro-Osseous Lesions

1) Fibrous dysplasia
2) Cemento-ossifying fibroma
Odontogenic Cystic Lesions
Clinical history from a pathology requisition

A. Cyst from mandibular area #32

1) What is the oral surgeon actually asking?

2) What do they really want to know?
I've already enucleated this cyst that I have sent to the laboratory. Is there anything else I need to do or worry about?

What the oral surgeon really means:

A. Cyst from mandibular area #32
A. Cyst from mandibular area #32

Impacted molar with a cyst

Let’s consider this patient from the oral surgeon’s perspective
Radiographic differential:

- Keratocystic odontogenic tumor
- Dentigerous cyst
- Unicystic Ameloblastoma
- Ameloblastoma with cystic change

All of these radiographic diagnoses are correct. Histology is the key.
Keratocystic odontogenic tumor

Dentigerous cyst

Unicystic Ameloblastoma

Ameloblastoma with cystic change

Can all of these lesions be treated with simple enucleation? No! All require different therapies.
Cyst associated with an impacted molar

What if this was a keratocystic odontogenic tumor?

What’s the worst that could happen if nothing is done or simple enucleation performed?
Keratocystic odontogenic tumor

This radiograph represents what the oral surgeon worries is going to happen if they let a keratocystic odontogenic tumor ‘get away’.
Formerly named parakeratinizing odontogenic keratocyst, now classified as a neoplasm.

Genetic mutations and deletions in \textit{PTCH} gene (9q22.3)

Recurrences of 30-60\% with simple enucleation alone
- thin squamous keratinized lining
- usually parakeratinized
- palisaded basal layer (key finding)
- often a corrugated surface

Keratocystic odontogenic tumor
Keratocystic odontogenic tumor
Keratocystic odontogenic tumor

Prominent basal layer
Keratocystic odontogenic tumor

When a patient has more than one keratocystic odontogenic tumor, this very likely represents **Basal Cell Nevus** syndrome

*(Basal Cell Carcinoma Nevus Syndrome, commonly known as Gorlin or Gorlin-Goltz Syndrome)*

Often presents in the first and second decade of life
Treatment: Keratocystic Odontogenic Tumor

Requires additional surgical procedure with complete curettage and chemical cautery of the area of the cyst with Carnoy’s solution and in some cases, removal of some bone (peripheral ostectomy).

Long term (10 yr) followup with radiograph every year for 5 years and then every other year thereafter.
Dentigerous cyst

This is also a squamous lined cyst on histology. But, we are looking for what is **NOT** present:

- no parakeratosis
- no palisaded basal layer
- no surface corrugation

We need to make sure this is not a keratocystic odontogenic tumor.
We have just seen this radiograph of a keratocystic odontogenic tumor.

From this radiograph, could this be a dentigerous cyst?

Yes!

Histology is key.
25% of all jaw cysts

Most commonly involves third molars

Any unerupted tooth maybe affected

Often associated with impacted teeth
- nonkeratinized stratified squamous epithelial lining
- fibrous connective tissue wall
- inflammation +/-
- no palisaded basal layer
- no parakeratosis

Dentigerous cyst
Dentigerous cysts
Keratocystic Odontogenic Tumor

Dentigerous Cyst
Treatment:
Dentigerous cyst

Enucleation of the cyst with the tooth.

No further treatment required.
Ameloblastoma

Histologically benign epithelial odontogenic tumor that has an aggressive behavior
Ameloblastoma

Reverse nuclear polarity (nuclei pulled away from basement membrane)

Stellate reticulum
Most ameloblastomas are solid, but ameloblastomas usually contain variably sized cysts.

In some tumors, the entire lesion is cystic.
UNICYSTIC AMELOBLASTOMA

Definition:
Ameloblastoma epithelium lines the cyst

AND

Must have no epithelium invading the cyst wall
Unicystic ameloblastoma: Treatment

Complete curettage, chemical cautery

Followup: Long term with radiographs
If the lesion looks like unicystic ameloblastoma but the epithelium invades the wall, the terminology is ‘cystic ameloblastoma’

The treatment is same as for solid ameloblastoma:

Surgical resection with 1 cm surgical margins beyond the radiographic border of the lesion
In the cases presented so far, we have noted that the radiograph has limited value as to establishing a specific diagnosis.

But that is not the case for all cystic lesions.

Radicular (periapical) cysts cannot be diagnosed without the radiograph.
Periapical (Radicular) Cyst
Periapical (radicular) cysts

Arise when epithelium around root is stimulated by inflammation

Histopathologic features may be identical to inflamed dentigerous cysts, therefore you need radiographic and/or clinical description for separation

Periapical cysts are treated with simple enucleation with removal of the tooth
Inflammation

No parakeratosis

No palisaded basal layer
An inflamed dentigerous cyst can mimic a radicular cyst exactly.

The radiograph is required.
Periapical (radicular) cyst

Based only the histology, could this be an inflamed dentigerous cyst?

Yes. The radiograph and clinical findings would be required to separate the two.
Is it clinically imperative to separate dentigerous cyst from periapical cyst?

No

One would always like to be correct with matching clinical and histologic findings but essentially the treatment has been performed and no other therapy is required.
Hyperplastic dental follicle

Hyperplastic dental follicle represents an enlargement of the follicle surrounding an unerupted tooth.

The stroma is often edematous/myxoid
Hyperplastic dental follicle

The specimen will be sent to the lab as ‘odontogenic cyst’ or ‘dental follicle’

‘Radiographic distinction between a small dentigerous cyst and an enlarged dental follicle is difficult and may be an academic exercise’

General rule:
If the cyst is > 1.0 cm, designated dentigerous cyst
Hyperplastic dental follicle

Reduced enamel epithelium
Hyperplastic dental follicle

Odontogenic rests are common in hyperplastic dental follicle.

This is not ameloblastic fibroma.
Mental checklist when approaching *cysts* in the jaw:

Associated with unerupted tooth and:
1) no basal layer palisading
2) no corrugated surface
3) no parakeratosis

→ *Dentigerous cyst*

Does it have a palisaded basal layer, a corrugated surface and parakeratosis?

→ *Keratocystic odontogenic tumor*

Are there ameloblastomatous changes?

→ *Ameloblastoma, unicystic vs cystic with wall invasion*
What do you do if you have no history or radiographic information yet see what appears to be a dentigerous cyst?

The diagnostic line should be:

“Squamous lined cyst”

with a comment that there are no features of keratocystic odontogenic tumor or ameloblastoma.
In every pathology report, even where I have the history or radiograph and the diagnosis is one of these three:

- dentigerous cyst
- hyperplastic dental follicle
- radicular cyst

I still always note somewhere in the report that there are no features of keratocystic odontogenic tumor or ameloblastoma.
Dentigerous cyst

Periapical (radicular) cyst
Keratocystic Odontogenic Tumor
Unicystic Ameloblastoma

Ameloblastoma with cystic change
Fibro-osseous Lesions of the Jaws

Fibrous dysplasia

Cemento-ossifying fibroma

Fibro-osseous dysplasia
Fibro-osseous Lesions of the Jaws

Fibrous dysplasia
Cemento-ossifying fibroma

Fibro-osseous lesions of the jaws can appear very similar histologically. Although there can be some histologic hints, true separation is most safely carried out with the addition of clinical and radiographic findings.
Fibrous Dysplasia

- Developmental condition characterized by replacement of normal bone by a cellular fibrous connective tissue and irregular islands and trabeculae
- Caused by a post-zygotic mutation in GNAS1 gene
Fibrous Dysplasia

**MONOSTOTIC**
- Single bone
- 80% of jaw cases

**CRANIOFACIAL**
- Multiple adjacent bones (maxilla, temporal bone or others)

**POLYOSTOTIC**
- Multiple bones
- Jaffe-Lichtenstein syndrome
  - *Café-au-lait* pigmentation
- McCune-Albright syndrome
  - *Café-au-lait* pigmentation and endocrinopathies (hyperthyroidism, pituitary adenoma, others)
Fibrous Dysplasia

Diffuse nature of bone expansion with no discrete lesion borders
Fibrous Dysplasia

Irregular trabeculae of metaplastic woven bone

Fibrous connective tissue with varying cellularity

Blends with normal bone at lesion periphery (no capsule)

Bone spherules

Few osteoblasts
Fibrous Dysplasia

Relatively uniform bone trabeculae & connective tissue
Fibrous Dysplasia

No capsule, blends with surrounding bone
Cemento-ossifying fibroma: Gross findings

Note circumscription due to presence of a capsule
Cemento-ossifying fibroma

Bone trabeculae & basophilic cellular spherules which resemble cementum

Spherules of cementum like material often demonstrate peripheral brush borders with blending into the adjacent connective tissue

Trabeculae are variable in size and show a mixture of woven and lamellar patterns

Peripheral osteoblastic rimming is usually present
Cemento-ossifying fibroma

Irregular trabeculae of osteoid & bone in varying stages of mineralization

Fibrous connective tissue
Cemento-ossifying fibroma

Acellular cementum spherules
Cemento-ossifying fibroma

Note spherules with a ‘brush border’ blending into surrounding stroma
Cemento-ossifying fibroma

Osteoblastic rimming
Fibrous dysplasia has a more uniform pattern of osseous formation than cemento-ossifying fibroma

Fibrous Dysplasia  Cemento-ossifying fibroma
Fibrous dysplasia or Cemento-ossifying fibroma?

A mix of mineralization patterns resembling bone or cementum along with abundant fibrous tissue is much more suggestive of cemento-ossifying fibroma over fibrous dysplasia.

Differentiating these lesions still usually requires clinical and radiographic correlation. No distinct cortical border is seen in dysplasias.
Treatment of Fibro-osseous Lesions

*Fibrous Dysplasia:*  
Various modalities; therapy not always required  
Recontouring can be performed for asymmetry

*Cemento-ossifying fibroma:*  
Conservative excision