

A Nonulcerated, Slowly Growing Mass of the Mandible

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The following Case Challenge is provided in conjunction with the American Academy of Oral and Maxillofacial Pathology.

Case Summary

A 38-year-old male presented with a slowly growing mass of the right mandible. The lesion displaced the mandibular canine and premolar teeth on the same side.

After you have finished reviewing the available diagnostic information, make the diagnosis.

Diagnostic Information

History of Present Illness

The patient reported that the lesion had been enlarging slowly for the past 21 years. The mass was not tender and the patient did not recall any history of trauma. No other information was available.

Medical History

The patient's medical history was unremarkable. He denied alcohol and tobacco use.

Clinical Findings

The clinical exam revealed a 3x2 cm "bony hard" swelling of the right mandible buccal to #27 and #28. The overlying mucosa was intact and no lingual extension was noted (Figure 1). The remainder of the oral cavity was within normal limits.

Radiographic Findings

The periapical radiograph demonstrated a well-defined mixed radiolucent/radiopaque lesion that caused divergence of the roots without resorption. An occlusal radiograph also revealed buccal expansion and scattered opacifications (Figures 2 and 3).

Biopsy Findings

On microscopic examination, the multiple sections examined revealed islands, strands, and sheets of polyhedral epithelial cells containing mildly pleomorphic nuclei set in a deeply eosinophilic cytoplasm (Figures 4-6).

Hyalinized, eosinophilic amorphous material was noted throughout the stroma. Basophilic calcifications composed of concentric rings also were identified (Liesegang rings). The lesion also demonstrated positive staining with Congo Red and apple/green birefringence under polarized light, a staining pattern consistent with amyloid (Figures 7 and 8).



Figure 1. Firm, nonulcerated mass of the right mandibular gingiva.



Figure 2. Periapical radiograph demonstrating a mixed radiolucent/radiopaque lesion between teeth numbers 28 and 29.



Figure 3. Mandibular occlusal radiograph demonstrating the mixed radiolucent/radiopaque nature of this lesion.

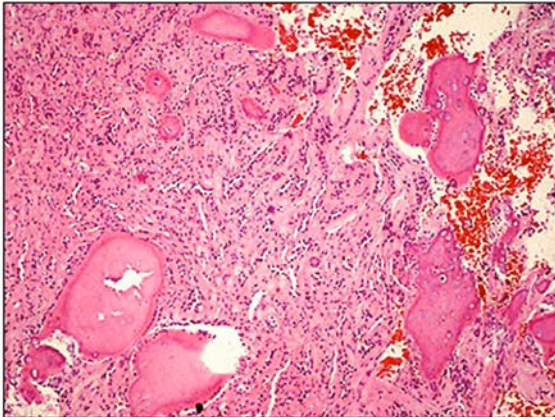


Figure 4. Low power photomicrograph of the incisional biopsy specimen.

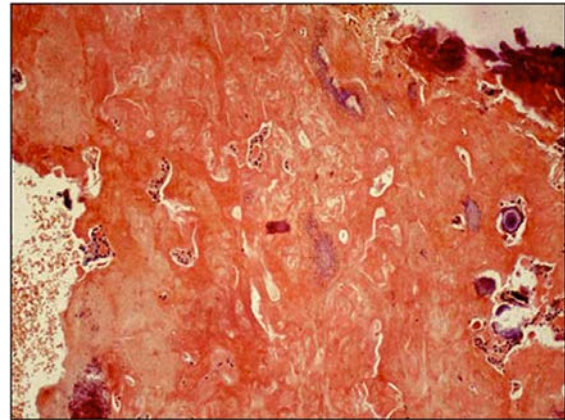


Figure 7. Low power photomicrograph demonstrating positive staining with Congo Red.

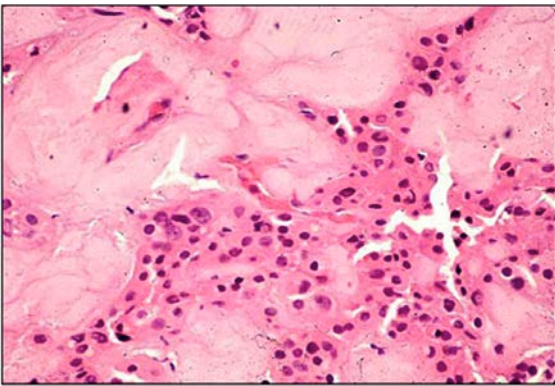


Figure 5. Medium power photomicrograph demonstrating areas of epithelial cells admixed with deposits of amorphous material.

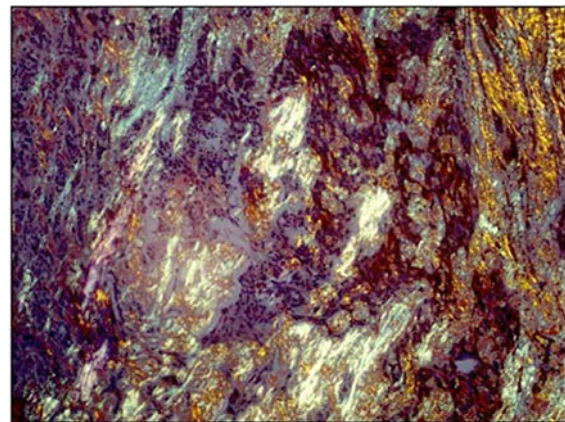


Figure 8. Congo Red stain with polarized light showing apple/green birefringence.

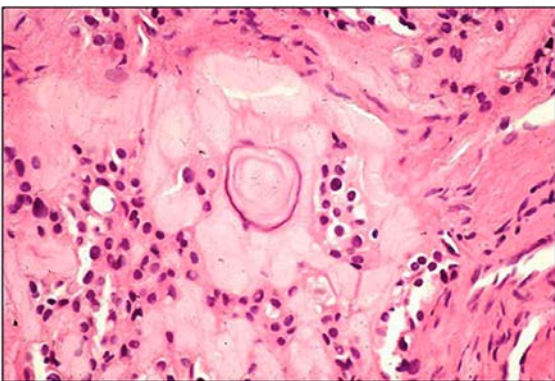


Figure 6. Medium power photomicrograph demonstrating a lamellar calcification.

Can you make the diagnosis?

A 38-year-old male presented with a slowly growing mass of the right mandible. The lesion displaced the mandibular canine and premolar teeth on the same side.



Select the Correct Diagnosis

- A. Calcifying Odontogenic Cyst (COC)
- B. Calcifying Epithelial Odontogenic Tumor (Pindborg tumor)
- C. Cemento-Ossifying Fibroma (COF)
- D. Osteosarcoma

Calcifying Odontogenic Cyst (COC)

Choice A. Sorry, this is not the correct diagnosis.

A calcifying odontogenic cyst (COC) may present with a clinical appearance that closely mirrors the lesion seen in this case. In addition, COCs affect the anterior regions of the jaws in a relatively high percentage of cases. The radiographic

presentation of COC is entirely consistent with the radiographic features seen here as well, and therefore it should definitely be included in the differential diagnosis. However, the histologic findings in this case do not demonstrate a cystic architecture with an ameloblastoma-like epithelial lining and the ghost cells characteristic of COCs.

Please re-evaluate the information about this case.

Calcifying Epithelial Odontogenic Tumor (Pindborg tumor)

Choice B. Congratulations! You are correct.

The calcifying epithelial odontogenic tumor (CEOT) was first described by Pindborg in 1955. It is a benign neoplasm commonly occurring intraosseously, but it also may occur in an extraosseous location. The tumor often presents as a painless mass with slow growth.¹ CEOT accounts for less than 1% of all odontogenic tumors. These tumors are seen more commonly in males than females and the average age is 36.9 years.¹ They are most often located in the premolar-molar region of the mandible, and 50% are associated with an unerupted tooth.²

Clinically, the intra-osseous CEOT appears as a painless, slow-growing mass. The peripheral (extra-osseous) variant appears as a painless, firm gingival mass.¹ The characteristic radiographic appearance is a uni- or multilocular radiolucency containing variably sized radiopacities. In early lesions, the calcified component may be undetectable on radiographs and the tumor will appear radiolucent. Some tumors remain radiolucent and do not develop opacities.³

Histologically, the tumor typically consists of sheets and strands of polyhedral and polymorphous squamous cells, often showing intercellular bridges.¹ Variation in nuclear size,

shape, and hyperchromaticity are often noted but do not indicate malignancy. Mitoses are very uncommon.³

A characteristic feature of the CEOT is the presence of an amorphous, eosinophilic extracellular material. The precise nature of this material is still uncertain but is thought to be a form of amyloid, which results from degradation of lamina densa material (basal lamina) secreted by the tumor epithelial cells.⁴ It generally stains as amyloid (i.e., positive staining with Congo Red) and will exhibit apple-green birefringence when viewed with polarized light.⁵ A pattern of calcification in a droplet-like fashion, often showing appositional basophilic concentric rings (Liseegang rings), is common but not always present.⁶

CEOT does not appear to be an aggressive lesion and conservative excision with a narrow rim of surrounding bone appears to be the treatment of choice.⁵ Lesions in the posterior maxilla should probably be treated more aggressively.⁵ Maxillary lesions are considered more aggressive and are more difficult to treat because the thick compact bone found in the mandible that confines tumor growth is not found in the maxilla. The combination of thin fragile bone with the proximity of the maxilla to the nasal cavity, paranasal sinuses, orbit, and vital structures at the base of the skull add a clinical dimension not present with mandibular tumors.⁷ Recurrence rate is reported to be about 15%.⁵ Malignant transformation is rare and the prognosis is good.

Cemento-Ossifying Fibroma (COF)

Choice C. Sorry, this is not the correct diagnosis.

Cemento-Ossifying Fibroma (COF) may present clinically with features that resemble the lesion seen in this case. It appears COFs occur across a wide age range, with the greatest number of cases encountered during the third and fourth decades of life. In addition, the mandibular premolar and molar area is the most common

site.⁵ Radiographically, presentation of COF is also consistent with the radiographic features seen here. However, histologically, COFs demonstrate replacement of normal bone by fibrous tissue and irregular woven bone trabeculae, lamellar bone, or cementum-like deposits. In the present case, the microscopic features did not demonstrate any new bone formation. In addition, epithelial islands and amyloid-like material should not be seen in COF.

Please re-evaluate the information about this case.

Osteosarcoma

Choice D. Sorry, this is not the correct diagnosis.

Osteosarcoma is an unlikely choice due to the long history of this lesion. The symmetrical growth pattern of the clinical lesion and the lack of surface ulceration and pain would be less consistent with a diagnosis of osteosarcoma. Radiographically, this lesion is well circumscribed

and does not demonstrate irregular root resorption or a moth-eaten appearance, additional features commonly associated with osteosarcoma in the jaws. Histologically, osteosarcoma demonstrates osteoid production by malignant mesenchymal cells, features that are absent in this case. Also, osteosarcomas are mesenchymal tumors, and this tumor shows a prominent epithelial component.

Please re-evaluate the information about this case.

References

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Note: Bio information was provided at the time the case challenge was developed.

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