

A Unilocular Radiolucency of the Anterior Maxilla

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

Case Summary

An 18-year-old white male presented to his dentist for evaluation of a fracture to the crown of his maxillary right central incisor. During radiographic evaluation of the region, a 1x1 cm asymptomatic radiolucency was discovered between the roots of a maxillary right canine and lateral incisor.

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

Diagnostic Information

Oral Findings

On clinical examination, slight labial cortical bone expansion was palpable overlying the lesion. Pulp testing indicated all teeth in the area were vital.

Radiographic Findings

A periapical radiograph revealed a well-circumscribed, round, 1 x 1 cm radiolucency between the roots of the maxillary right canine and lateral incisor (Figure 1). The lesion exhibited a sclerotic border, and no obvious central calcifications could be identified.

Excisional Biopsy and Photomicrographs

A biopsy of the mass was performed under local anesthesia. After reflection of a buccal flap, the cortical plate appeared slightly expanded and thinned. The cortical bone was removed, and a solid soft tissue mass was easily curetted from the bony cavity. Multiple irregular fragments of soft tissue were submitted for microscopic examination.

The tissue sections showed an epithelial proliferation exhibiting a variable microscopic pattern. At low power, the lesion showed solid, nodular masses of basophilic cells intermixed with less cellular areas (Figures 2 and 3).

The epithelium in the solid zones consisted of small, tightly packed polygonal to spindle-shaped cells. The less cellular areas were characterized by ribbon-like cords of basophilic cells with little background stroma.

A characteristic finding in the solid zones was the presence of occasional duct-like structures lined by a row of cuboidal to columnar epithelial cells (Figures 4 and 5).

In addition, focal dystrophic calcifications were present, sometimes being found in association with slightly larger, eosinophilic cells exhibiting a squamoid appearance (Figure 6).

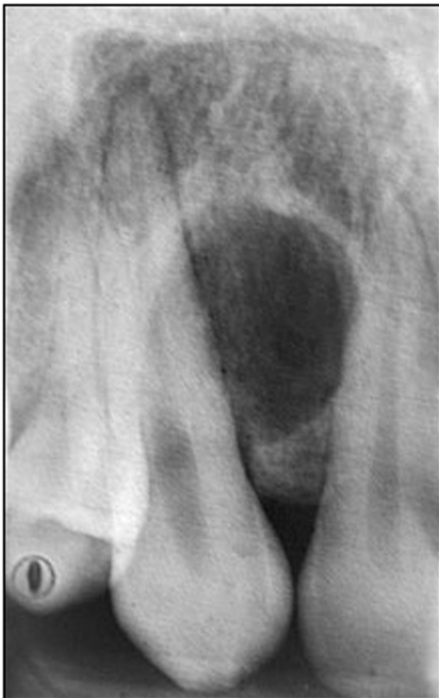


Figure 1. A well-circumscribed unilocular radiolucency between the roots of the maxillary right canine and lateral incisor.

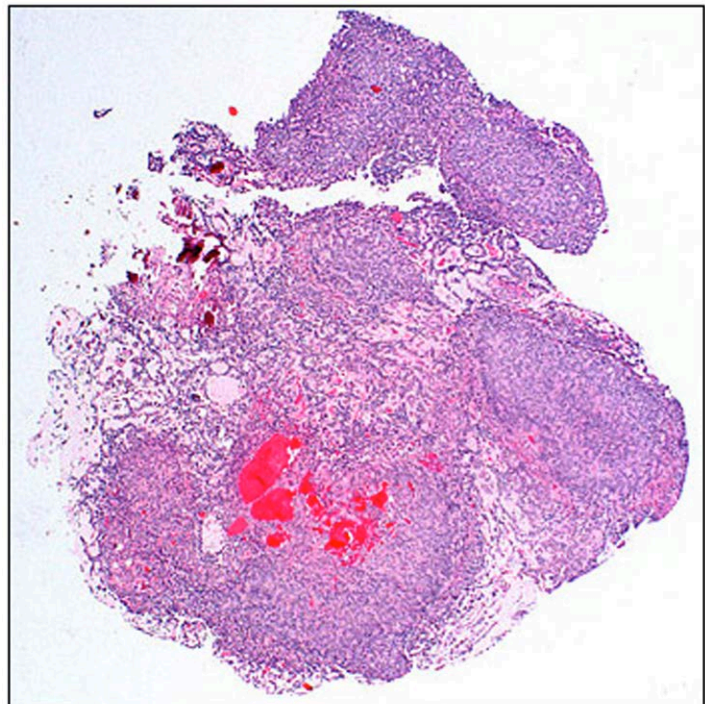


Figure 2. Low-power scanning photomicrograph of one portion of the lesion showing nodular groups of basophilic cells (Hematoxylin and eosin stain).

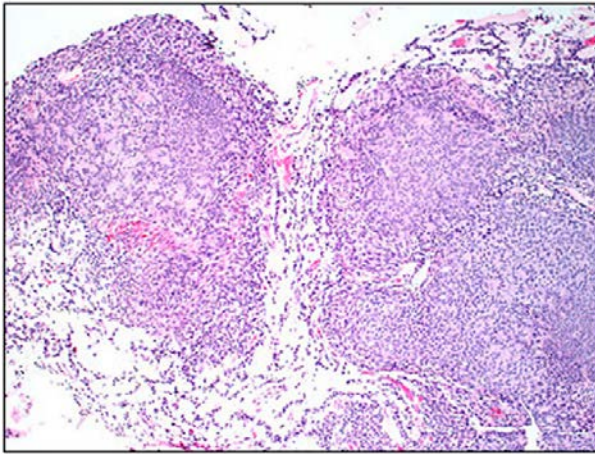


Figure 3. Medium-power photomicrograph showing nodular groups of epithelial cells intermixed with looser ribbon-like strands (Hematoxylin and eosin stain).

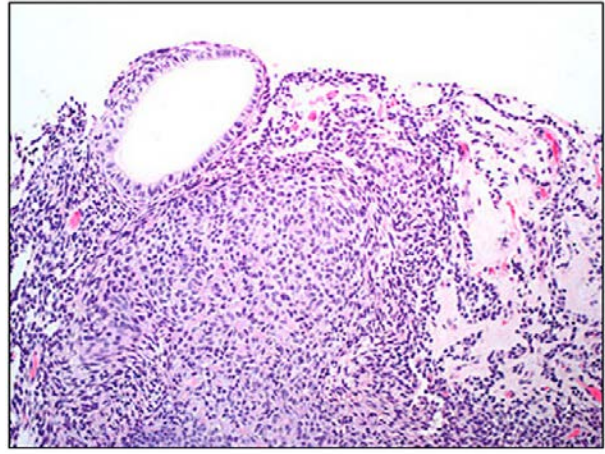


Figure 4. Medium-power photomicrograph showing a duct-like structure in the upper left corner of the image (Hematoxylin and eosin stain).

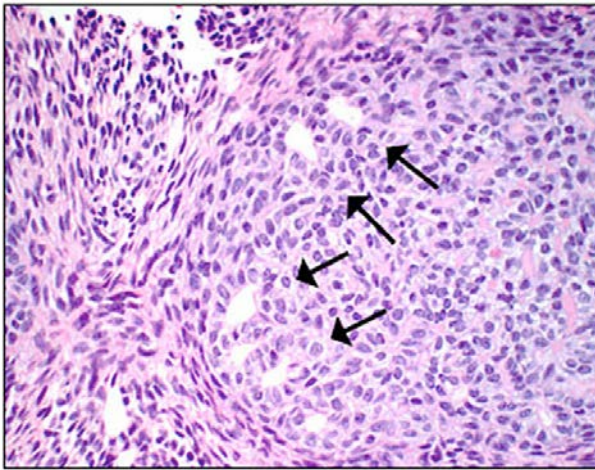


Figure 5. High-power photomicrograph showing multiple small duct-like structures surrounded by cuboidal epithelial cells (arrows) (Hematoxylin and eosin stain).

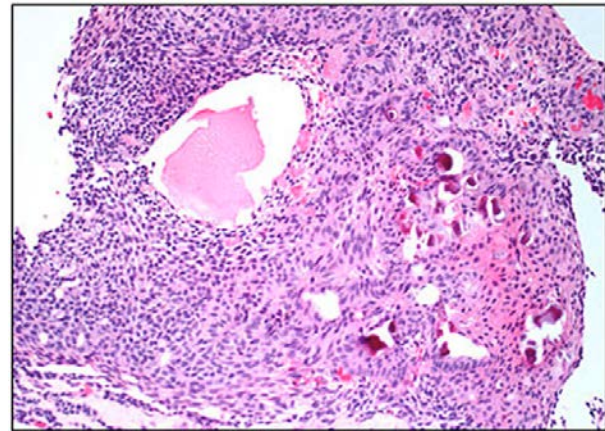


Figure 6. Medium-power photomicrograph showing small dystrophic calcifications in association with squamoid cells (Hematoxylin and eosin stain).

Can you make the diagnosis?

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Select the Correct Diagnosis

- A. Adenomatoid Odontogenic Tumor
- B. Globulomaxillary Cyst
- C. Odontogenic Keratocyst
- D. Ameloblastoma

Adenomatoid Odontogenic Tumor

Choice A. Congratulations! You are correct.

The adenomatoid odontogenic tumor (AOT) is an uncommon neoplasm representing from 3% to 7% of all odontogenic tumors.¹⁻³ The term “adenomatoid” refers to the characteristic gland-like tubular or ductlike structures found microscopically. When this tumor was first described, it was known by the name “adenoameloblastoma” under the incorrect assumption it represented a variant of ameloblastoma. However, because of its different clinical features and biologic behavior, it is widely accepted today this lesion represents a separate, distinct tumor.

Many of the basic clinical features of the adenomatoid odontogenic tumor can be enumerated by recalling the fraction “2/3:”

- Two-thirds of AOTs are diagnosed in patients in the second decade of life.
- Two-thirds of AOTs occur in females.
- Two-thirds of AOTs occur in the maxilla.

The AOT also demonstrates a striking predilection for the anterior portions of the jaws, with 80% occurring anterior to the first bicuspid. Rarely does this tumor ever develop in the molar/ramus region. These various clinical characteristics are in sharp contrast to the ameloblastoma, which occurs most often in the posterior mandible.

Most AOTs are relatively small tumors, seldom surpassing 3 cm in diameter. In 75% of cases, the tumor is associated with the crown of an unerupted tooth, most frequently a canine. The lesion is usually asymptomatic, often being discovered as an incidental finding or when radiographs are obtained to determine why a tooth has failed to

erupt. Rare extraosseous examples have been reported in the gingival soft tissues.⁴

Radiographically, the AOT presents as a well-circumscribed unilocular radiolucency. When associated with an impacted tooth, this lucency typically surrounds the crown and resembles a dentigerous cyst. In some cases, the radiolucency will extend down the lateral root surface beyond the cemento-enamel junction, which may help to distinguish the lesion from a dentigerous cyst. Some AOTs will exhibit fine snowflake calcifications within the lucency.

Microscopically, the AOT is usually well-circumscribed and surrounded by a thick fibrous capsule. The tumor is composed of spindle-shaped and polygonal epithelial cells arranged in sheets and whorled aggregates (“Zellballen”). Between the nodular cell groups, the tumor is often less cellular and characterized by narrow cords with a plexiform pattern. The most characteristic feature is the presence of scattered rosettes and ductlike structures surrounded by palisaded cuboidal to columnar cells. Scattered dystrophic calcifications are also a common finding.

Some AOTs show areas of differentiation with amyloid-like deposits consistent with the calcifying epithelial odontogenic tumor (CEOT), although such tumors still behave in the same fashion as ordinary AOTs.⁵⁻⁷ The focal squamoid zones with calcifications in the present case were felt to be suggestive of CEOT differentiation, although special stains for amyloid were negative.

The prognosis for the AOT is excellent. Because the tumor is typically well-encapsulated, it usually enucleates easily from the bone and rarely, if ever, recurs.

Globulomaxillary Cyst

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

The so-called “globulomaxillary cyst” was once theorized to represent a fissural cyst developing from epithelium entrapped during fusion of the globular portion of the medial nasal process with the maxillary process.⁸ Purported examples of this cyst occurred between the maxillary lateral incisor and canine teeth. The classic radiographic appearance was described as an inverted pear-shaped radiolucency, which often was associated with lateral tipping of the adjacent tooth roots.

However, most experts today believe no epithelial entrapment should occur in this region during embryonic development.^{9,10} Instead, almost all cysts in the globulomaxillary region can be explained on an odontogenic basis. Most examples show significant inflammation of the cyst lining and are consistent with periapical cysts arising from adjacent nonvital teeth (usually the lateral incisor). Developmental odontogenic cysts, such as the odontogenic keratocyst and lateral periodontal cyst, also can occur in this area. Therefore, the concept of a fissural “globulomaxillary cyst” should be discarded in favor of an odontogenic origin for most cysts in this region.

Please re-evaluate the information about this case.

Odontogenic Keratocyst

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

The radiographic finding of a well-defined radiolucency between the roots of teeth would be consistent with this diagnosis; however, microscopic examination did not show features of an odontogenic keratocyst.¹¹ Histopathologic features characteristic of an odontogenic

keratocyst include a cyst with an epithelial lining exhibiting a corrugated superficial parakeratin layer, a palisaded and hyperchromatic basal cell layer, and a uniform thickness (approximately 6 to 8 layers thick).¹² In addition, although it is possible for an odontogenic keratocyst to occur in the anterior maxilla, this lesion most often arises in the posterior mandible.¹³

Please re-evaluate the information about this case.

Ameloblastoma

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

The ameloblastoma classically presents as an expansile, multilocular radiolucency involving the posterior mandible.^{14,15} However, occasionally ameloblastomas may present as smaller unilocular radiolucencies involving the posterior mandible or other regions of the jaws, thereby mimicking other odontogenic tumors or odontogenic cysts. Nevertheless, the biopsy specimen of the current case did not show

microscopic features of an ameloblastoma. Conventional ameloblastomas typically exhibit a proliferation of odontogenic epithelium forming islands, anastomosing cords, or sheets with a peripheral layer of tall, columnar cells resembling ameloblasts and a central core of loosely arranged, angular cells resembling stellate reticulum. The nuclei of the columnar cells at the periphery tend to exhibit reverse polarization (i.e., the nuclei are oriented away from the basement membrane). Additional less common histopathologic variants of ameloblastoma also exist.

Please re-evaluate the information about this case.

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

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