Odontogenic tumors

lesions in this group including hamartomas (tumor like anomalies or malformations) benign and malignant neoplasms with metastatic capabilities. They may be found within the maxillofacial skeleton (central) or may be located in the soft tissue overlying the tooth-bearing regions and in the alveolar mucosa of edentulous segments of the jaws (peripheral). Several classification systems based on histologic patterns have been developed for this complex group of lesions. Common to all is the division of tumors into those composed of odontogenic epithelial elements, those composed of odontogenic ectomesenchyme, and those that are proliferations of both epithelium and ectomesenchyme. On the basis of biological behavior, odontogenic tumors range from clinically trivial (i.e., benign, no recurrence potential) to malignant tumors.

Epithelial Tumors

Ameloblastoma

Slowly growing locally aggressive tumor that run a benign course. Ameloblastomas occur in three different clinic-radiographic situations

1. Conventional (solid or multicystic)
2. Unicystic
3. Peripheral (extraosseous)

Pathogenesis

The potential epithelial sources for ameloblastoma include

1. Remnants of dental lamina (rests of Serres)
2. Epithelial rests of Malassez
3. Reduced enamel epithelium

4. the epithelial lining of odontogenic cysts (especially dentigerous cysts)
5. Basal cell layer of surface epithelium

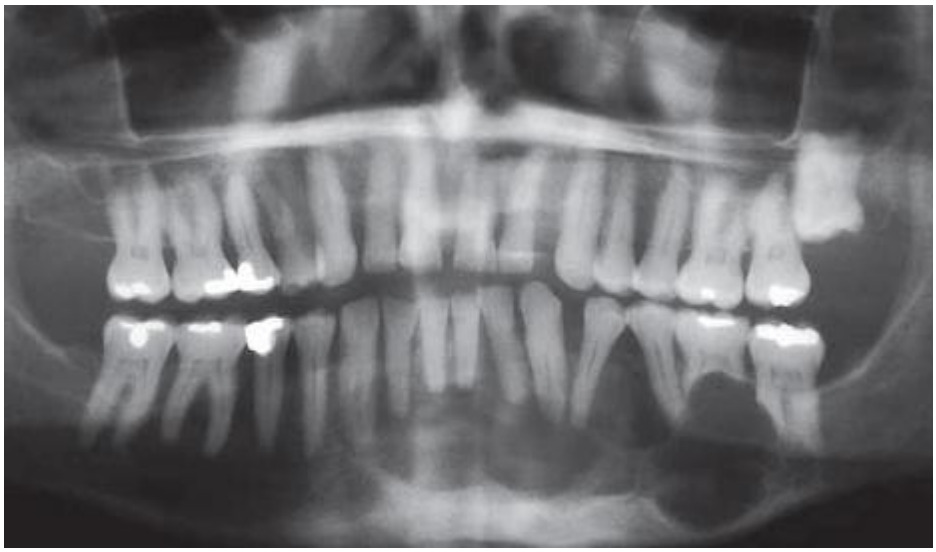
The trigger or stimulus for neoplastic transformation of these epithelial residues is unknown.

Clinical Features

Ameloblastoma occurs predominantly in the third to fifth decades of life. (The rare lesions occurring in children are usually cystic and appear clinically as odontogenic cysts). There appears to be no gender predilection for this tumor. Ameloblastomas occur predominantly in the mandible, particularly in the molar-ramus area. In the maxilla, the molar area is more commonly affected than the premolar and anterior regions. Lesions usually are asymptomatic and are discovered during routine radiographic examination or because of painless jaw expansion. tooth movement or malocclusion may be the initial presenting sign. Unicystic type usually occur in younger age group and its usually asymptomatic.

Radiographical features:

Ameloblastomas are osteolytic, typically found in the tooth-bearing areas of the jaws. The most typical radiographic feature is that of a multilocular radiolucent lesion. The lesion is often described as having a “soap bubble” appearance (when the radiolucent loculations are large) or as being “honeycombed” (when the loculations are small). Buccal and lingual cortical expansion is frequently present. Resorption of the roots of teeth adjacent to the tumor is common. In many cases an unerupted tooth, most often a mandibular third molar, is associated with the radiolucent defect. Unicystic Ameloblastomas radiographically appear as unilocular radiolucent defects, which may resemble almost any type of cystic lesion. The margins of these radiolucent lesions, however, often show irregular scalloping.



Cystic ameloblastoma occupying the body of the mandible.

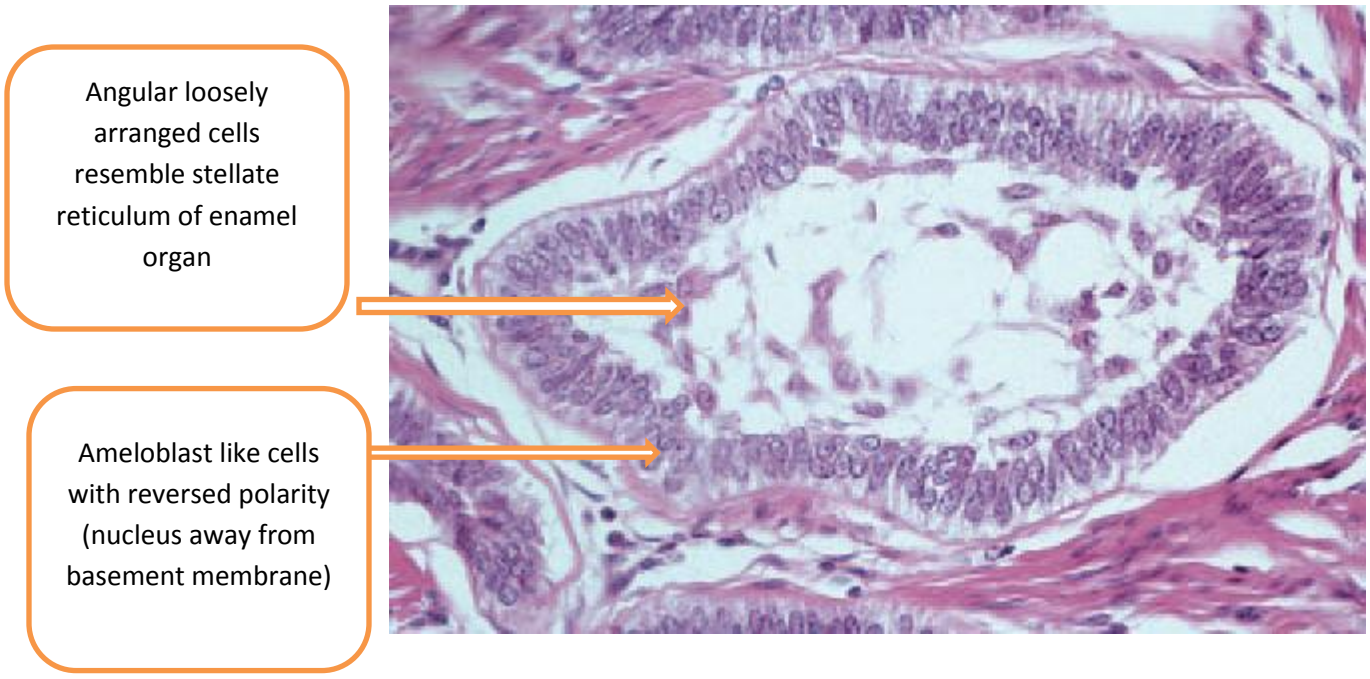
Histopathological features

Conventional solid or multicystic intraosseous ameloblastomas show a remarkable tendency to undergo cystic change; grossly, most tumors have varying combinations of cystic and solid features. The cysts may be seen only at the microscopic level or may be present as multiple large cysts that include most of the tumor. Several microscopic subtypes of conventional ameloblastoma are recognized, but these microscopic patterns generally have little bearing on the behavior of the tumor. Large tumors often show a combination of microscopic patterns.

The **follicular** and **plexiform** patterns are the most common. Less common histopathologic patterns include the **acanthomatous, granular cell, desmoplastic,** and **basal cell** types.

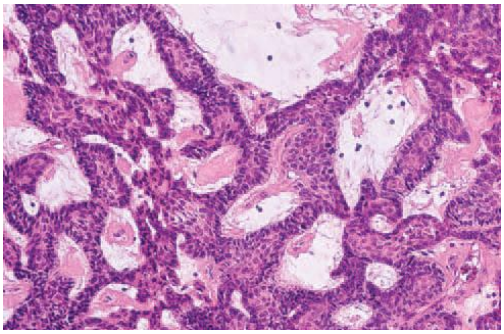
Follicular pattern:

The follicular histopathologic pattern is the most common and recognizable. 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 ameloblast-like cells surrounds this central core. The nuclei of these cells are located at the opposite pole to the basement membrane **(reversed polarity)**. In other areas, the peripheral cells may be more cuboidal and resemble basal cells. Cyst formation is common and may vary from microcysts, which form within the epithelial islands, to large macroscopic cysts, which may be several centimeters in diameter



Plexiform pattern:

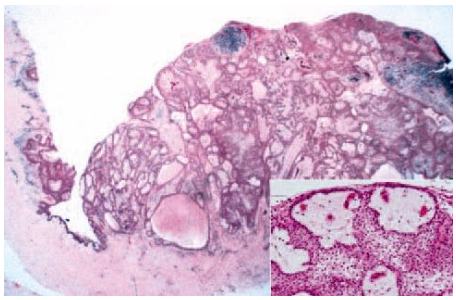
The plexiform type of ameloblastoma consists of long, anastomosing cords or larger sheets of odontogenic epithelium. The cords or sheets of epithelium are bounded by columnar or cuboidal ameloblast-like cells surrounding more loosely arranged epithelial cells. The supporting stroma tends to be loosely 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.



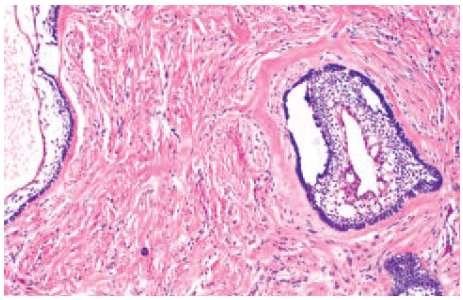
Ameloblastoma plexiform pattern

Three histopathologic variants of unicystic ameloblastoma have been described. In the first type **(luminal unicystic ameloblastoma)**, the tumor is 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. This demonstrates a basal layer of columnar or cuboidal cells with hyperchromatic nuclei that show reverse polarity. The overlying epithelial cells are loosely cohesive and resemble stellate reticulum. In the second microscopic variant, one or more nodules of ameloblastoma project from the cystic lining into the lumen of the cyst. This type is called an **intraluminal unicystic ameloblastoma**. 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 edematous, plexiform pattern

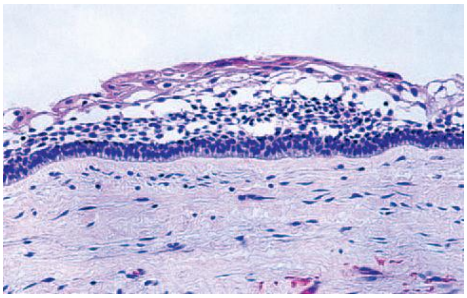
that resembles the plexiform pattern seen in conventional ameloblastomas. In the third variant, known as **mural unicystic ameloblastoma**, the fibrous wall of the cyst is infiltrated by typical follicular or plexiform ameloblastoma. The extent and depth of the ameloblastic infiltration may vary considerably. 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.



Intraluminal variant



mural variant



luminal variant

Treatment and prognosis:

Patients with conventional solid or multicystic intraosseous ameloblastomas have been treated by a variety of means. These range from simple enucleation and curettage to *en bloc* resection. The conventional ameloblastoma tends to infiltrate between intact cancellous bone trabeculae at the periphery of the lesion before bone resorption becomes radiographically evident. Therefore, the actual margin of the tumor often extends beyond its apparent radiographic or clinical margin. Attempts to remove the tumor by curettage often leave small islands of tumor within the bone, which later manifest as recurrences. Recurrence rates of 50% to 90% have been reported in various studies after curettage. Recurrence often takes many years to become clinically manifest, and 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. surgeons advocate that the margin of the resection should be at least 1.0 to 1.5 cm past the radiographic limits of the tumor. Ameloblastomas of the posterior maxilla are particularly dangerous because of the difficulty of obtaining an adequate surgical margin around the tumor. some studies suggest that the ameloblastoma may be radiosensitive, radiation therapy 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 ameloblastoma 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. Rarely, an ameloblastoma exhibits frank malignant behavior. unicystic ameloblastoma usually treated as a cyst by enucleation. If the ameloblastic elements are confined to the lumen of the cyst with or without intraluminal tumor extension, then the cyst enucleation has probably been adequate treatment. The patient, however, should be kept under long-term follow-up. If the specimen shows extension of the tumor into the fibrous cyst wall for any appreciable distance, then subsequent management of the patient is more controversial. Some surgeons believe that local resection of the area is indicated as a prophylactic measure; others prefer to keep the patient under close radiographic observation and delay further treatment until there is evidence of recurrence. Recurrence rates of 10% to 20% were described after enucleation and curettage of unicystic ameloblastomas in many literatures.

Peripheral ameloblastoma:

The **peripheral ameloblastoma** is uncommon and accounts for about 1% to 10% of all ameloblastomas. This tumor probably arises from rests of dental lamina beneath the oral mucosa or from the basal epithelial cells of the surface epithelium. Histopathologically, these lesions have the same features as the intraosseous form of the tumor. The peripheral ameloblastoma 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 1.5 cm in size. Unlike the intraosseous ameloblastoma, the peripheral ameloblastoma shows an innocuous clinical behavior. Patients respond well to local surgical excision. Although local recurrence has been noted in 15% to 20% of cases, further local excision almost always results in a cure.