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LEC. 6

Odontogenic Tumors

Odontogenic tumors comprise a complex group of lesions of diverse histopathologic types and clinical behavior. Some of these lesions are true neoplasms and may rarely exhibit malignant behavior. Others may represent tumor like malformations (hamartomas).

Odontogenic tumors, like normal odontogenesis, demonstrate varying inductive interactions between odontogenic epithelium and odontogenic ectomesenchyme.

Tumors of odontogenic epithelium are composed only of odontogenic epithelium without any participation of odontogenic ectomesenchyme. Other odontogenic neoplasms, sometimes referred to as **mixed odontogenic tumors**, are composed of odontogenic epithelium and ectomesenchymal elements. A third group, **tumors of odontogenic ectomesenchyme**, is composed principally of ectomesenchymal elements.

Classification of Odontogenic Tumors

I. Tumors of odontogenic epithelium

A. Ameloblastoma

1. Malignant ameloblastoma

2. Ameloblastic carcinoma

B. Clear cell odontogenic carcinoma

C. Adenomatoid odontogenic tumor

D. Calcifying epithelial odontogenic tumor

E. Squamous odontogenic tumor

II. Mixed odontogenic tumors

A. Ameloblastic fibroma

B. Ameloblastic fibro-odontoma

C. Ameloblastic fibrosarcoma

D. Odontoameloblastoma

E. Compound odontoma

F. Complex odontoma

III. Tumors of odontogenic ectomesenchyme

A. Odontogenic fibroma

B. Granular cell odontogenic tumor

C. Odontogenic myxoma

D. Cementoblastoma

Tumors of Odontogenic Epithelium

Epithelial odontogenic tumors are composed of odontogenic epithelium without participation of odontogenic ectomesenchyme. Several distinctly different tumors are included in the group; ameloblastoma is the most important and common of them.

AMELOBLASTOMA

The **ameloblastoma** is the most common clinically significant odontogenic tumor. Its relative frequency equals the combined frequency of all other odontogenic tumors, excluding odontomas .

Ameloblastomas are tumors of odontogenic epithelial origin. Theoretically, they may arise from rests of dental lamina, from a developing enamel organ, from the epithelial lining of an odontogenic cyst, or from the basal cells of the oral mucosa. Ameloblastomas are slow-growing, locally invasive tumors that run a benign course in most cases. They occur in three different clinicoradiographic situations, situations, which deserve separate consideration because of differing therapeutic considerations and prognosis:

1. Conventional solid or multicystic (about 86% of all cases)

2. Unicystic (about 13% of all cases)

3. Peripheral (extraosseous) (about 1% of all cases)

CONVENTIONAL SOLID OR MULTICYSTIC INTRAOSSEOUS AMELOBLASTOMA

CLINICAL AND RADIOGRAPHIC FEATURES

Conventional solid or **multicystic intraosseous ameloblastoma** is encountered in patients across a wide age range. It is rare in children younger than age 10 . The tumor shows an approximately equal prevalence in the third to seventh decades of life. About 80% to 85% of conventional ameloblastomas occur in the mandible, most often in the molar-ascending ramus area.

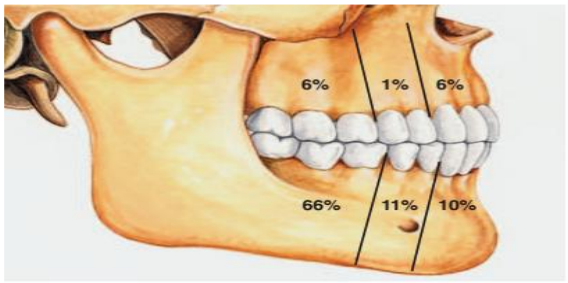
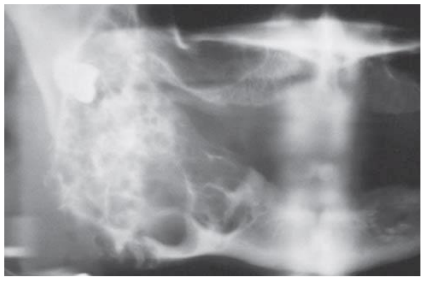


Fig. 15-55 Ameloblastoma. Relative distribution of ameloblastomas in the jaws.

The tumor is often asymptomatic, and smaller lesions are detected only during a radiographic examination. A painless swelling or expansion of the jaw is the usual clinical presentation**.** If untreated, then the lesion may grow slowly to massive proportions **.**

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)



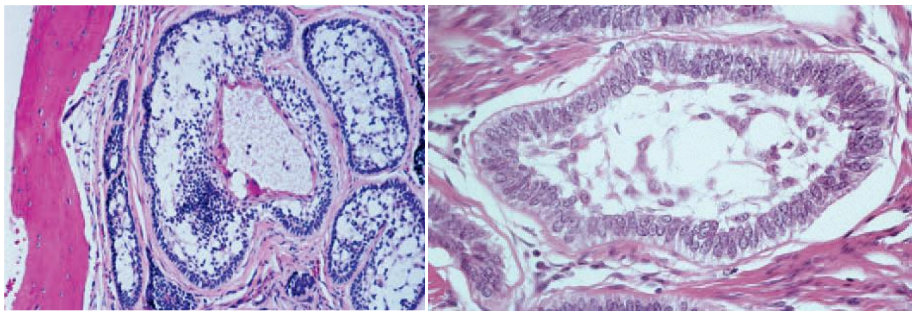
Buccal and lingual cortical expansion is frequently present. Resorption of the roots of teeth adjacent to the tumor is common. In many cases an unerupted tooth, most often a mandibular third molar, is associated with the radiolucent defect. Solid ameloblastomas may radiographically appear as unilocular radiolucent defects, which may resemble almost any type of cystic lesion .

HISTOPATHOLOGIC FEATURES

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.

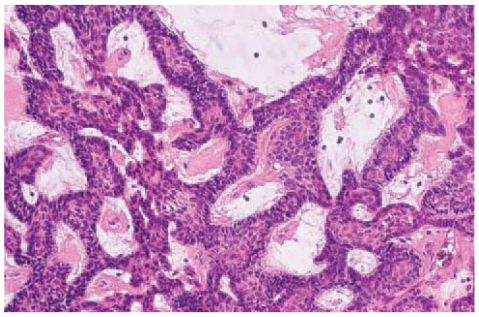
FOLLICULAR PATTERN

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)**.



PLEXIFORM PATTERN

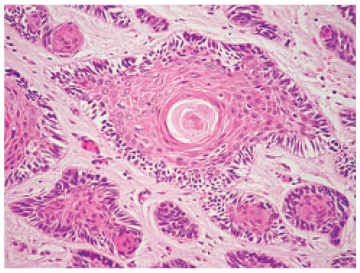
The plexiform type of ameloblastoma consists of long, anastomosing cords or larger sheets of odontogenic epithelium. The cords or sheets of epithelium are bounded by columnar or cuboidal ameloblast-like cells surrounding more loosely arranged epithelial cells. The supporting stroma tends to be loosely arranged and vascular. Cyst formation is relatively uncommon in this variety.



ACANTHOMATOUS PATTERN

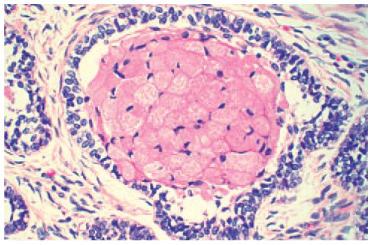
When extensive squamous metaplasia, often associated with keratin formation, occurs in the central portions of the epithelial islands of a follicular ameloblastoma, the term acanthomatous ameloblastoma is sometimes applied.

histopathologically, however, such a lesion may be confused with squamous cell carcinoma or squamous odontogenic tumor .



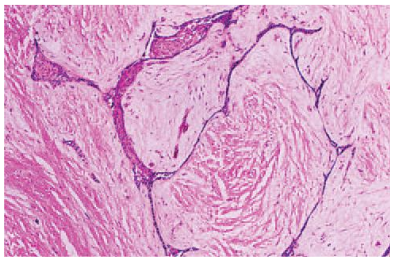
GRANULAR CELL PATTERN

Ameloblastomas may sometimes show transformation of groups of lesional epithelial cells to granular cells. These cells have abundant cytoplasm filled with eosinophilic granules that resemble lysosomes ultrastructurally and histochemically.



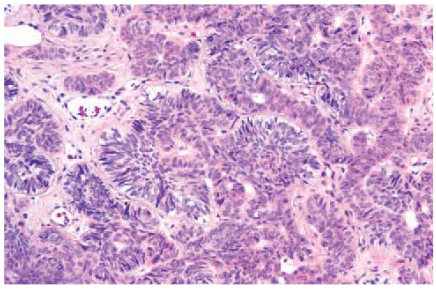
DESMOPLASTIC PATTERN

This type of ameloblastoma contains small islands and cords of odontogenic epithelium in a densely collagenized stroma.



BASAL CELL PATTERN

The basal cell variant of ameloblastoma is the least common type. These lesions are composed of nests of uniform basaloid cells, and they histopathologically are very similar to basal cell carcinoma of the skin. No stellate reticulum is present in the central portions of the nests. The peripheral cells about the nests tend to be cuboidal rather than columnar .



TREATMENT

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.

UNICYSTIC AMELOBLASTOMA

CLINICAL AND RADIOGRAPHIC FEATURES

Unicystic ameloblastomas are most often seen in younger patients, with about 50% of all such tumors diagnosed during the second decade of life. The average age in one large series was 23 years. More than 90% of unicystic ameloblastomas are found in the mandible, usually in the posterior regions. The lesion is often asymptomatic, although large lesions may cause a painless swelling of the jaws. In many patients, this lesion typically appears as a circumscribed radiolucency that surrounds the crown of an unerupted mandibular third molar , clinically resembling a dentigerous cyst.

HISTOPATHOLOGIC FEATURES

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 and basilar cytoplasmic vacuolization .

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 the third variant, known as **mural unicystic ameloblastoma**, the fibrous wall of the cyst is infiltrated by typical follicular or plexiform ameloblastoma.

TREATMENT

The clinical and radiographic findings in most cases of unicystic ameloblastoma suggest that the lesion is an odontogenic cyst. These tumors are usually treated as cysts by enucleation.

PERIPHERAL (EXTRAOSSEOUS) 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.

CLINICAL FEATURES

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, The tumor has been found in average age of 52 years on the posterior gingival and alveolar mucosa, and they are somewhat more common in mandibular than in maxillary areas.

HISTOPATHOLOGIC FEATURES

islands of ameloblastic epithelium that occupy the lamina propria underneath the surface epithelium . The proliferating epithelium may show any of the features described for the intraosseous ameloblastoma; plexiform or follicular patterns are the most common. Connection of the tumor with the basal layer of the surface epithelium is seen in about 50% of cases.

TREATMENT

Unlike the intraosseous ameloblastoma, the peripheral ameloblastoma respond well to local surgical excision.

MALIGNANT AMELOBLASTOMA AND AMELOBLASTIC CARCINOMA

Rarely, an ameloblastoma exhibits frank malignant behavior with development of metastases. The frequency of malignant behavior in ameloblastomas is difficult to determine but probably occurs in far less than 1% of all ameloblastomas.

CLINICAL AND RADIOGRAPHIC FEATURES

Malignant ameloblastomas have been observed in patients who range in age from 4 to 75 years (mean age, 30 years). For patients with documented metastases, the interval between the initial treatment of the ameloblastoma and first evidence of metastasis varies from 1 to 30 years. Ameloblastic carcinomas, in contrast, tend to develop later in life, with the mean age at diagnosis typically being in the sixth decade of life.

The radiographic findings of malignant ameloblastomas may be essentially the same as those in typical non metastasizing ameloblastomas. Ameloblastic carcinomas are often more aggressive lesions, with ill-defined margins and cortical destruction .

HISTOPATHOLOGIC FEATURES

With malignant ameloblastomas, the primary jaw tumor and the metastatic deposits show no microscopic features that differ from those of ameloblastomas with a completely benign local course. With ameloblastic carcinomas, the metastatic deposits or primary tumor shows the microscopic pattern of ameloblastoma in addition to cytologic features of malignancy.

TREATMENT

surgical excision