ODONTOGENIC TUMORS

Lect 12

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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 tumorlike malformations (hamartomas).

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 ecto-mesenchymal elements. Dental hard tissue may or may not be formed in these lesions.

A third group, tumors of odontogenic ectomesenchyme, is composed principally of ectomesenchymal elements.

TUMORS OF ODONTOGENIC EPITHELIUM: AMELOPLASTOMA

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. they may arise from 1.rests of dental lamina, 2.from a developing enamel organ,3. from the epithelial lining of an odontogenic cyst, 4.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 typically have been described as having three different clinic-radiographic presentations with differing therapeutic considerations and prognosis:

1. Conventional solid or multicystic (about 75%–86% of all cases)
2. Unicystic (about 13%–21% of all cases)
3. Peripheral (extraosseous) (about 1%–4% 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 and relatively uncommon in the 10-19 years old group.it has equal prevalence in the third to seventh decades of life.

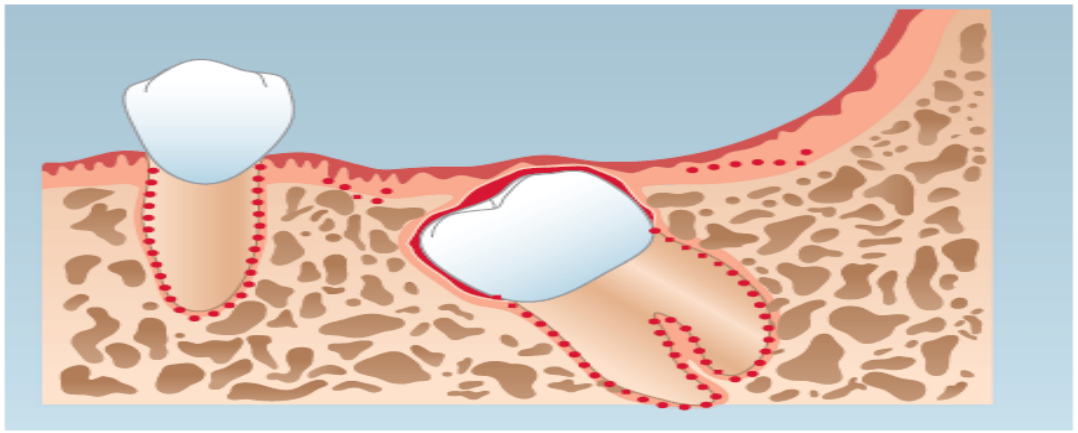


FIGURE 5-3
Possible epithelial sources of ameloblastoma *(represented by red color):* Remnants of the dental lamina *(dots above crown of molar);* reduced enamel epithelium *(on surface of molar crown);* rests of Malassez *(dots in periodontal membrane);* surface epithelium.

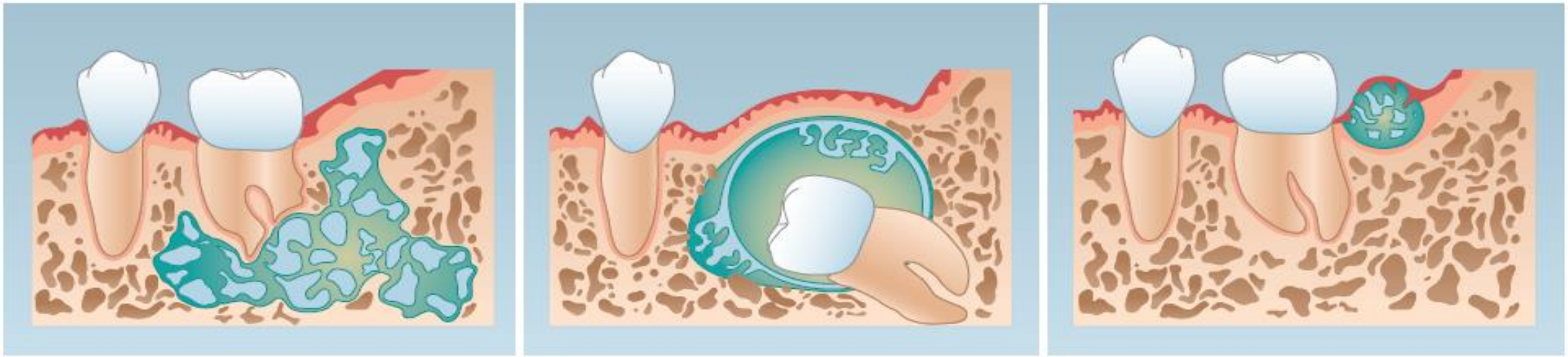
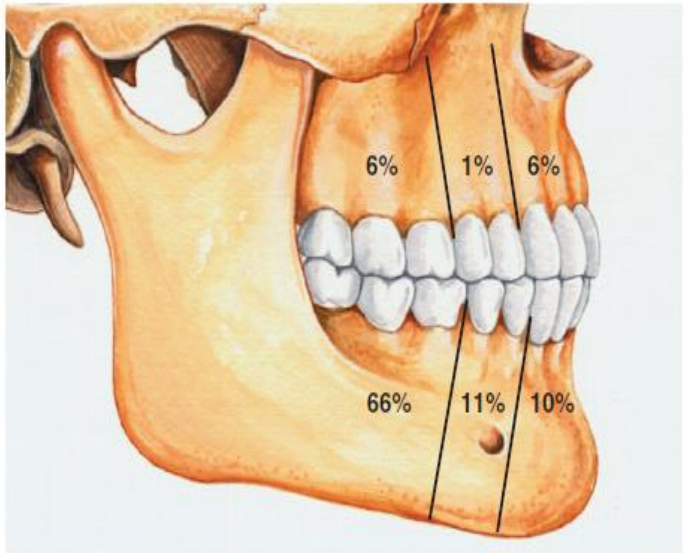
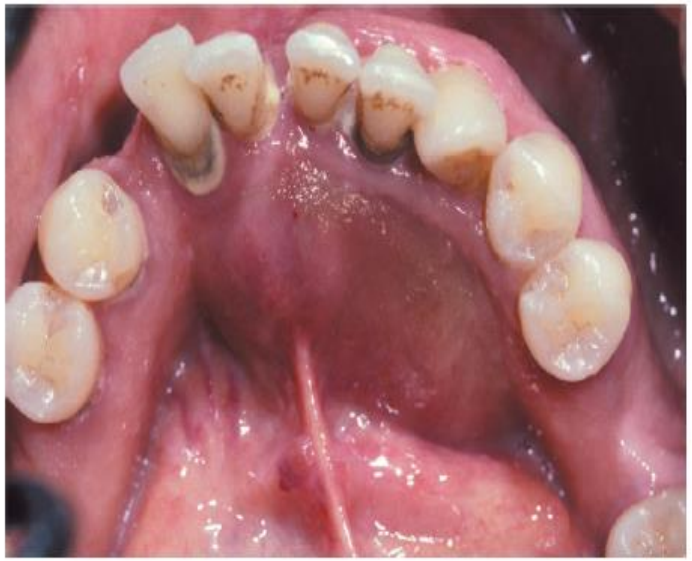


FIGURE 5-4
Ameloblastoma. Three clinical subtypes. **A,** Common (polycystic). **B,** Unicystic. **C,** Peripheral (extraosseous).

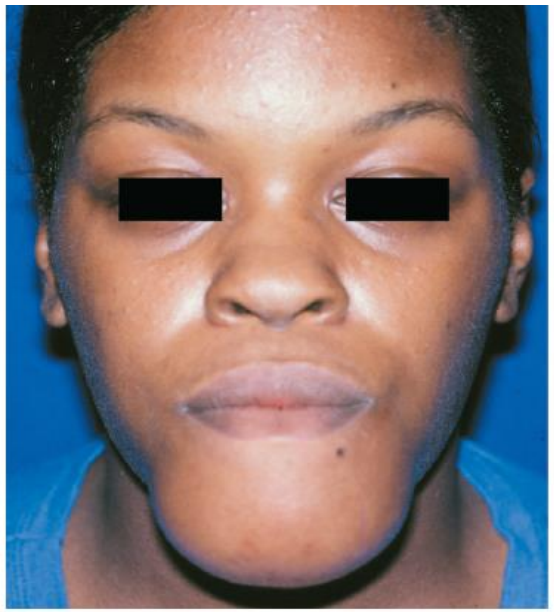
There is no significant sex predilection.Some studies indicate a greater frequency in blacks; others show no racial predilection. About 80%–85% of conventional ameloblastomas occur in the mandible, most often in the molar-ascending ramus area. About 15%–20% of ameloblastomas occur in the maxilla, usually in the posterior regions. 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.Pain and paresthesia are uncommon, even with large tumors. The most typical radiographic feature is that of a multilocular radiolucent lesion, Multilocular lesions are 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 most cases,an unerupted tooth, most often a mandibular third molar, is associated with the radiolucen defect. Solid ameloblastomas may radiographically appear as unilocular radiolucent defects, which may resemble almost any typeof cystic lesion.The margins of these radiolucent Lesions often show irregular scalloping.



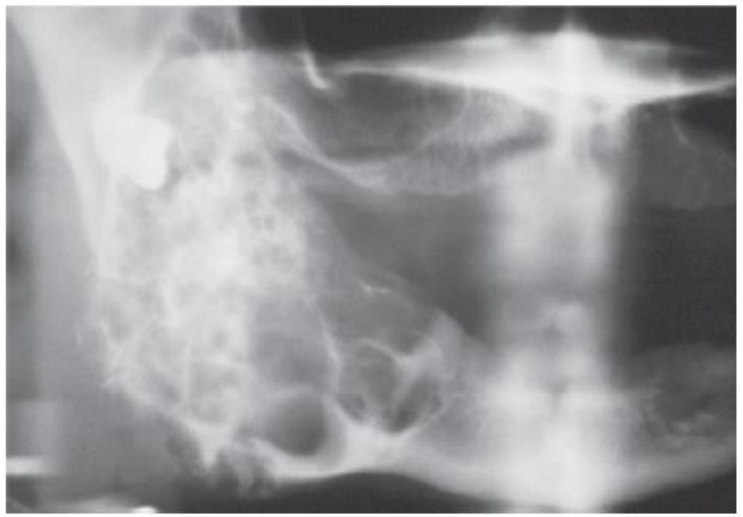
• **Fig. 15.59 Ameloblastoma.** Relative distribution of ameloblastomas in the jaws.



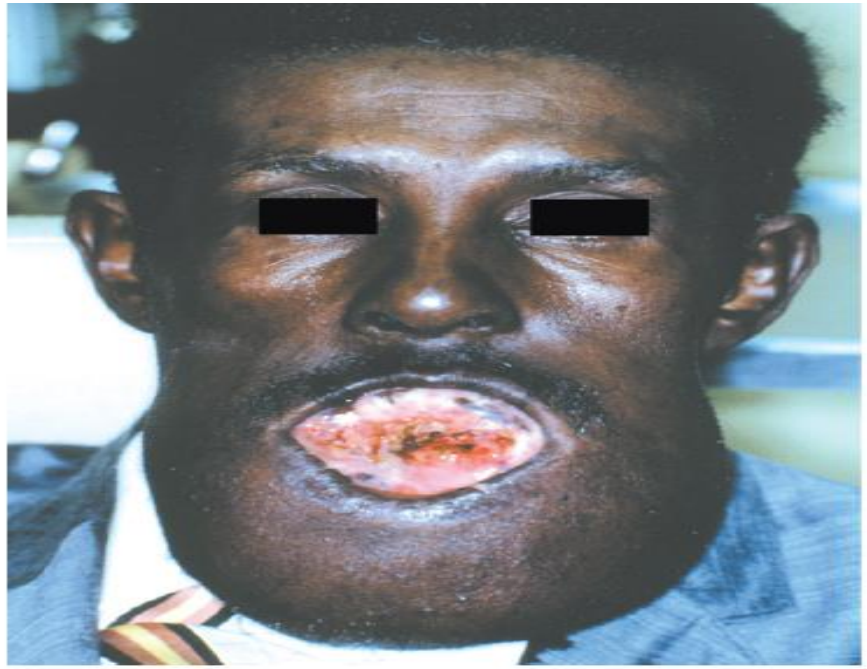
• **Fig. 15.61 Ameloblastoma.** Prominent expansion of the lingual alveolus caused by a large ameloblastoma of the mandibular symphysis.



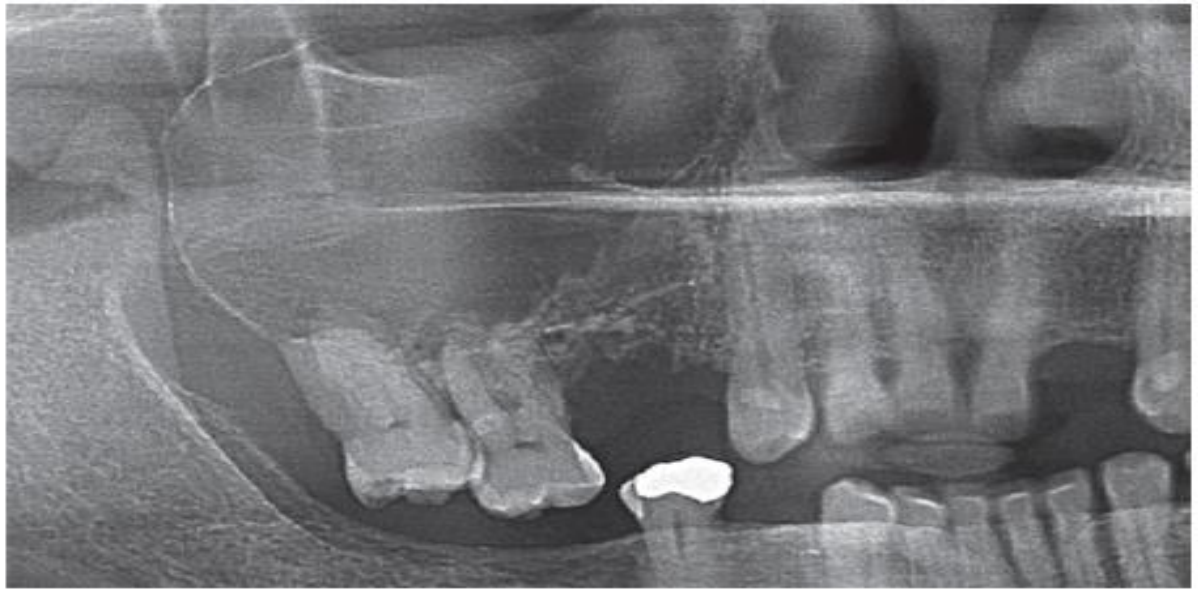
• **Fig. 15.60 Ameloblastoma.** Large expansile mass of the anterior mandible. (Courtesy of Dr. Michael Tabor.)



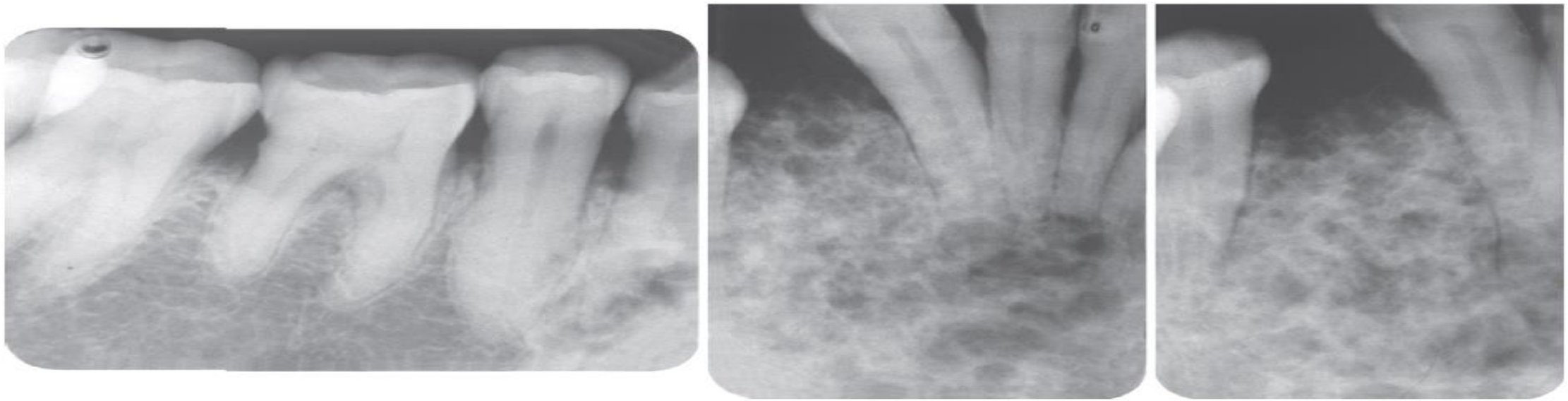
• **Fig. 15.63 Ameloblastoma.** Large multilocular lesion involving the mandibular angle and ascending ramus. The large loculations show the "soap bubble" appearance. An unerupted third molar has been displaced high into the ramus.



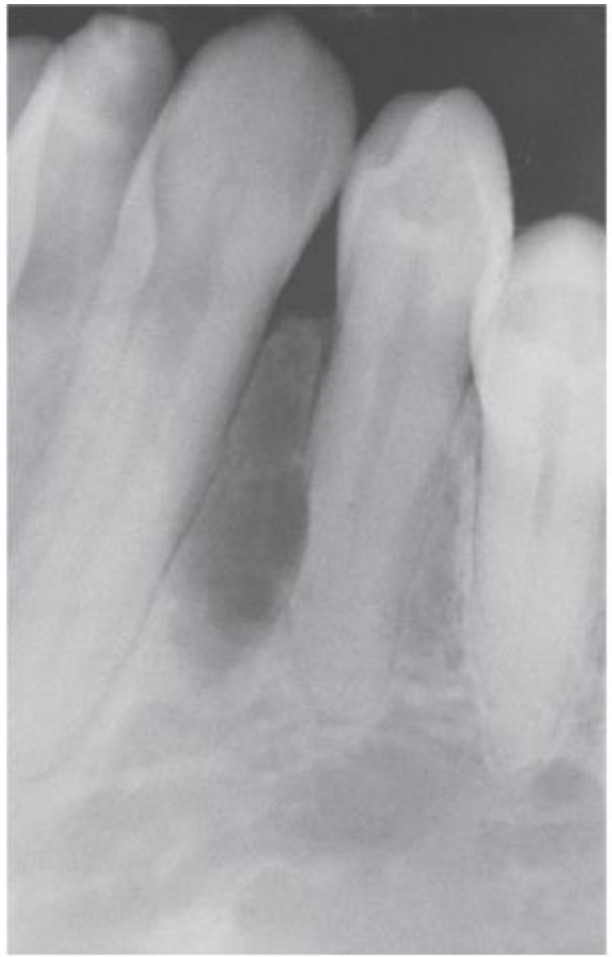
• **Fig. 15.62 Ameloblastoma.** Massive tumor of the anterior mandible. (Courtesy of Dr. Ronald Baughman.)



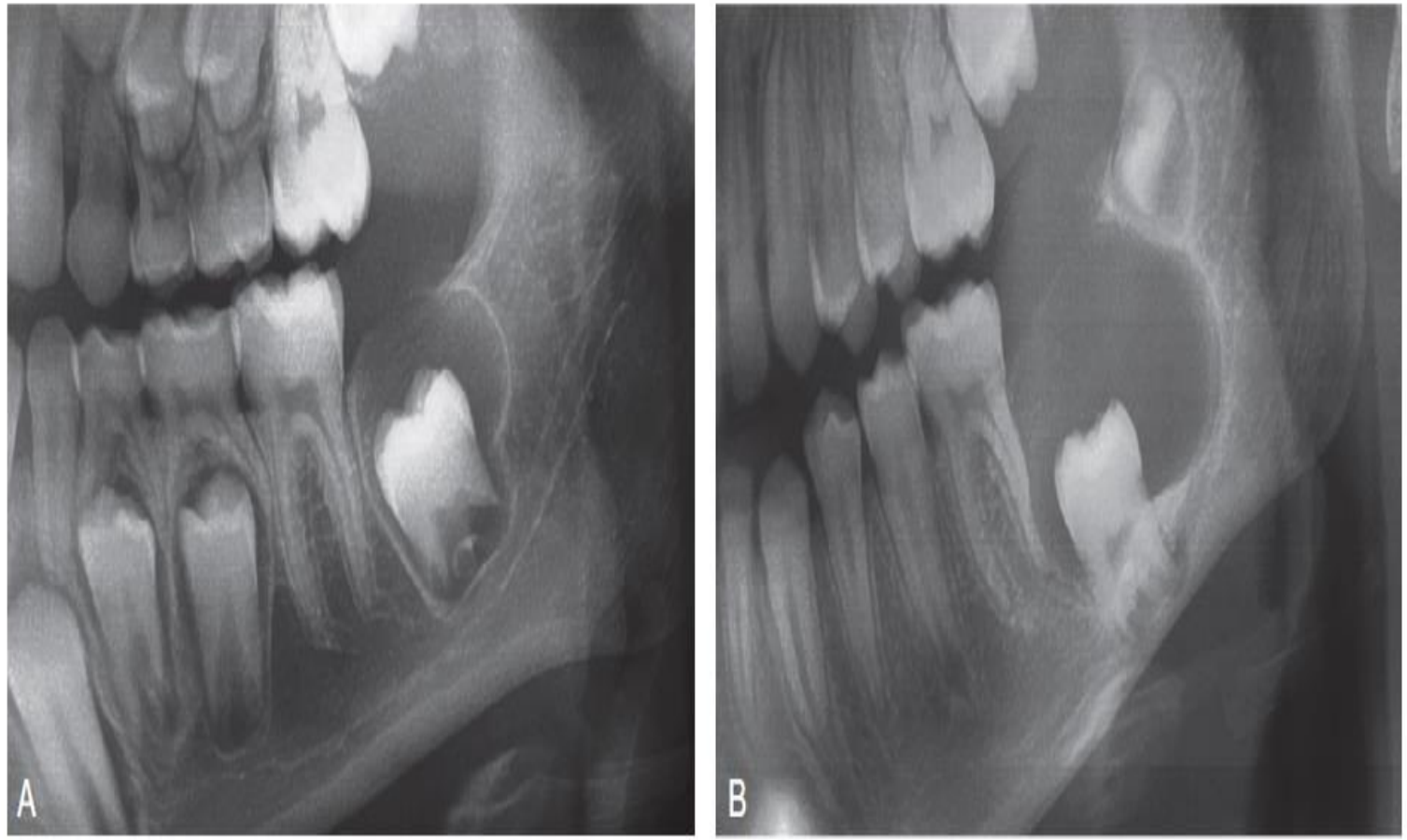
• **Fig. 15.65 Ameloblastoma.** Expansile, radiolucent mass of the right posterior maxilla, which fills the maxillary sinus. (Courtesy of Dr. Doug Oliver.)



• **Fig. 15.64 Ameloblastoma.** Periapical films showing the "honeycombed" appearance. (Courtesy of Dr. John Hann.)



• **Fig. 15.67 Ameloblastoma.** This small unilocular radiolucency lesion could easily be mistaken for a lateral periodontal cyst. (Courtesy of Dr. Tony Traynham.)



• **Fig. 15.66 Ameloblastoma. A,** Unilocular radiolucency surrounding the crown of the developing left mandibular second molar, which mimics a small dentigerous cyst. **B,** This radiograph from 3 years later shows enlargement of the lesion, including inferior displacement of the tooth. (Courtesy of Dr. Patrick Scioscia.)

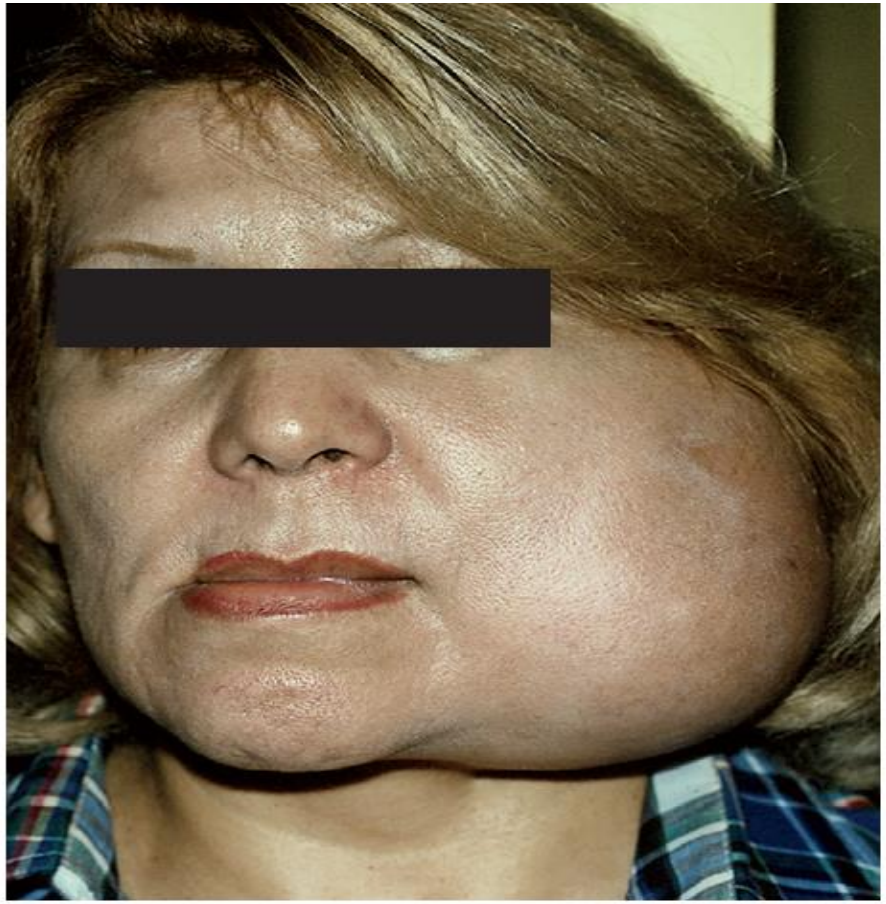
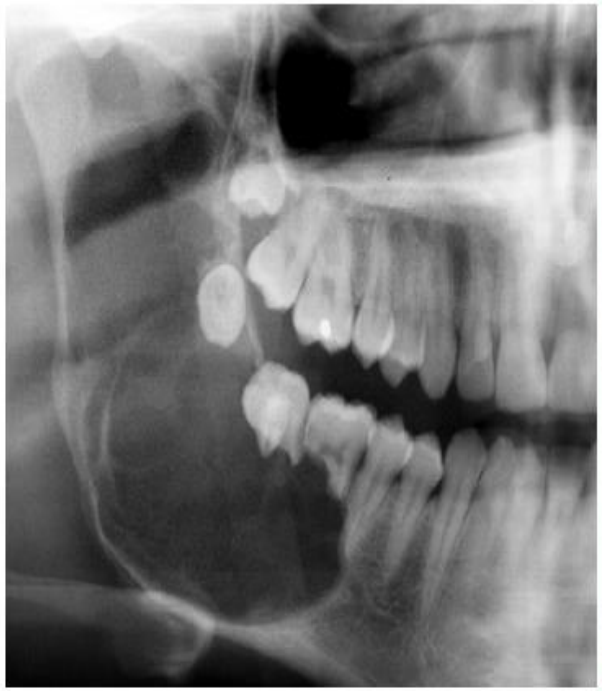
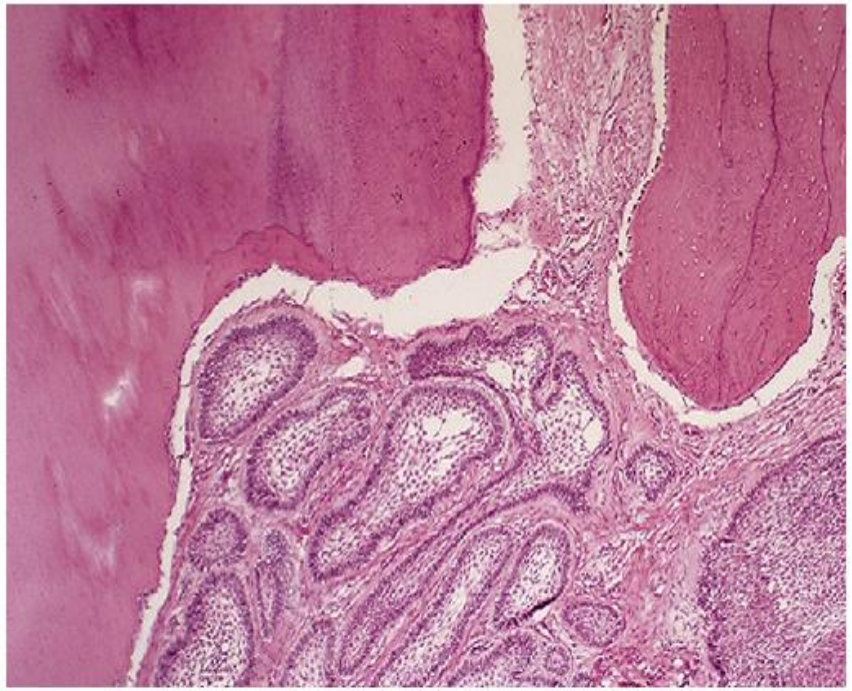


FIGURE 5-5
Ameloblastoma. Patient with large lesion of left posterior mandible.

A





B

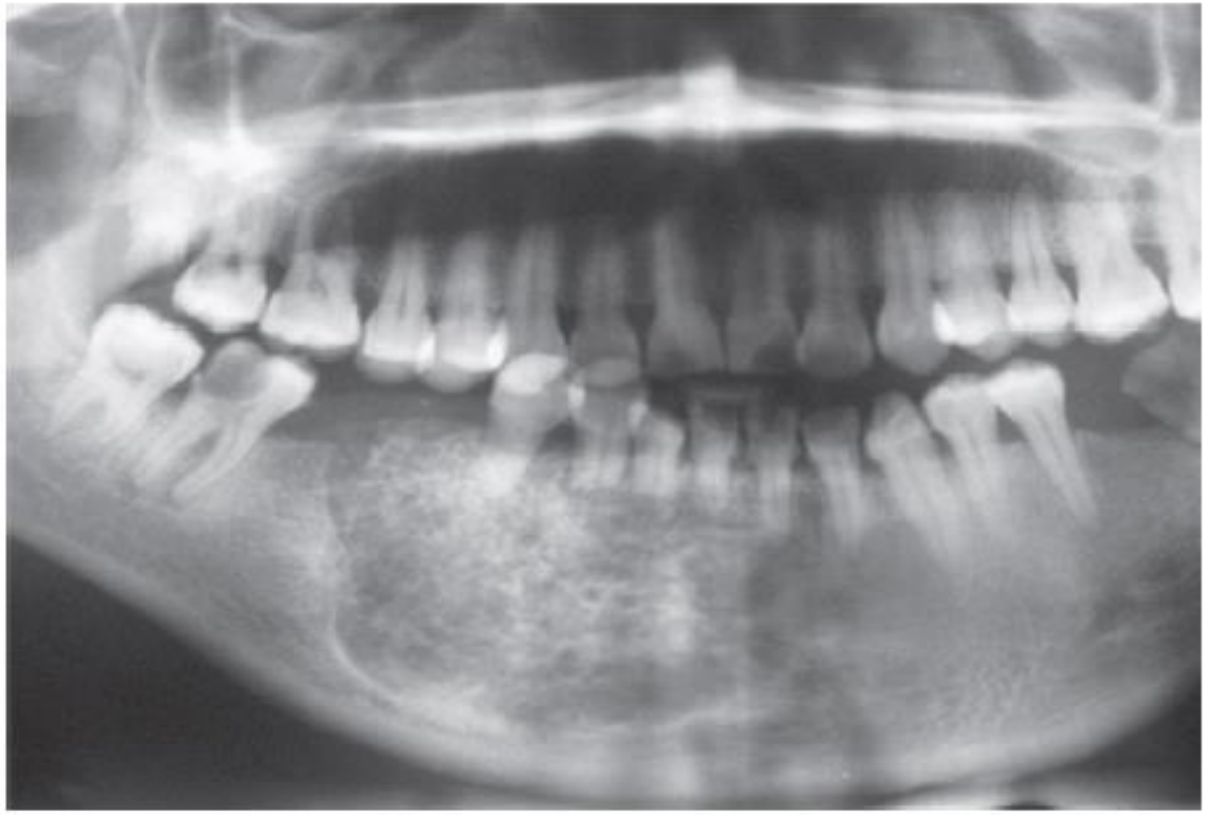
FIGURE 5-7
Ameloblastoma. A, Radiograph illustrating extensive polycystic lesion of right mandible extending into the coronoid process, with resorption of roots of the molars and premolar teeth. **B,** Photomicrograph of neoplastic cells in close proximity to resorbed roots of molar tooth *(left side)*.

One histopathologic form of ameloblastoma that does not have these characteristic features is the desmoplastic ameloblastoma which has a marked predilection to occur in the anterior regions of the jaws, with equal distribution between the mandible and the maxilla. Radiographically, the majority of these tumors resemble a fibro-osseous lesion because of their mixed radiolucent and radiopaque appearance, The mixed radiographic appearance is due to osseous metaplasia within the dense fibrous septa that characterize the lesion, not because the tumor itself is producing a mineralized product.

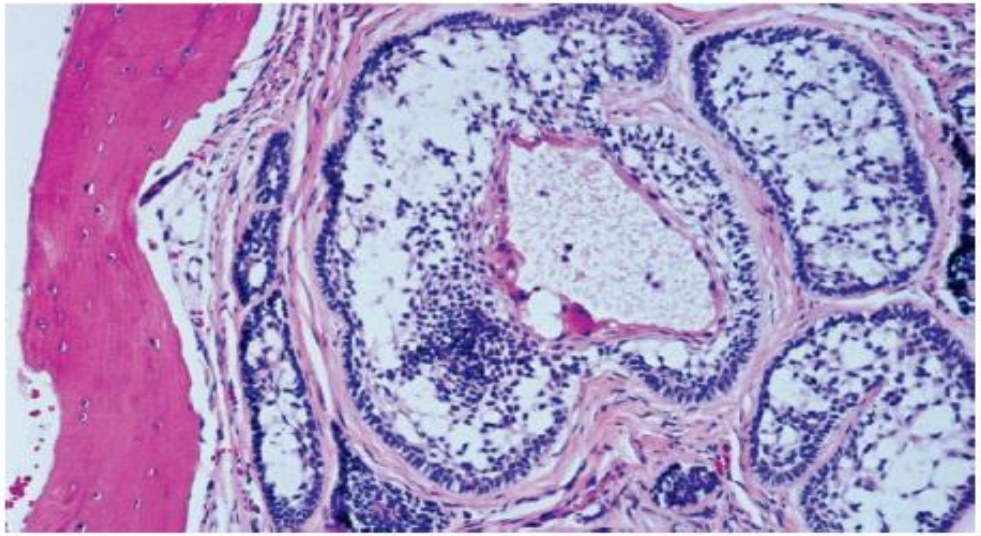
Histopathologic Features

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.

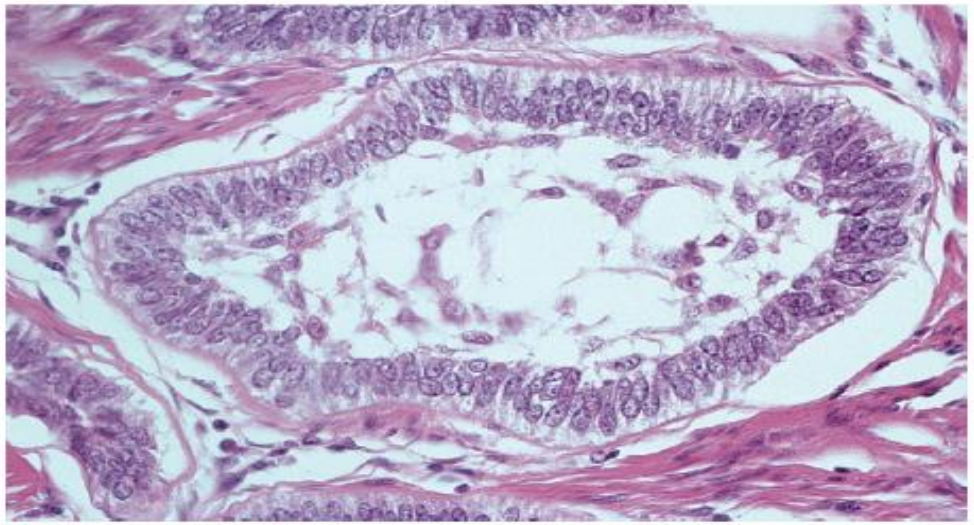
The follicular and plexiform patterns are the most common.Less common histopathologic patterns include the acanthomatous, granular cell, desmoplastic, and basal cell types.



• **Fig. 15.68 Desmoplastic Ameloblastoma.** Large mixed radiolucent and radiopaque lesion of the anterior and right body of the mandible. (Courtesy of Dr. Román Carlos.)



• **Fig. 15.69 Ameloblastoma (Follicular Pattern).** Multiple islands of odontogenic epithelium demonstrating peripheral columnar differentiation with reverse polarization. The central zones resemble stellate reticulum and exhibit foci of cystic degeneration.



• **Fig. 15.70 Ameloblastoma (Follicular Pattern).** This high-power photomicrograph highlights the peripheral columnar cells exhibiting reverse polarization.

Follicular pattern

The follicular histopathologic pattern is the most common type. 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 . If an incisional biopsy is taken from the latter area, an inappropriate diagnosis of "unicystic ameloblastoma"may be rendered by the pathologist.

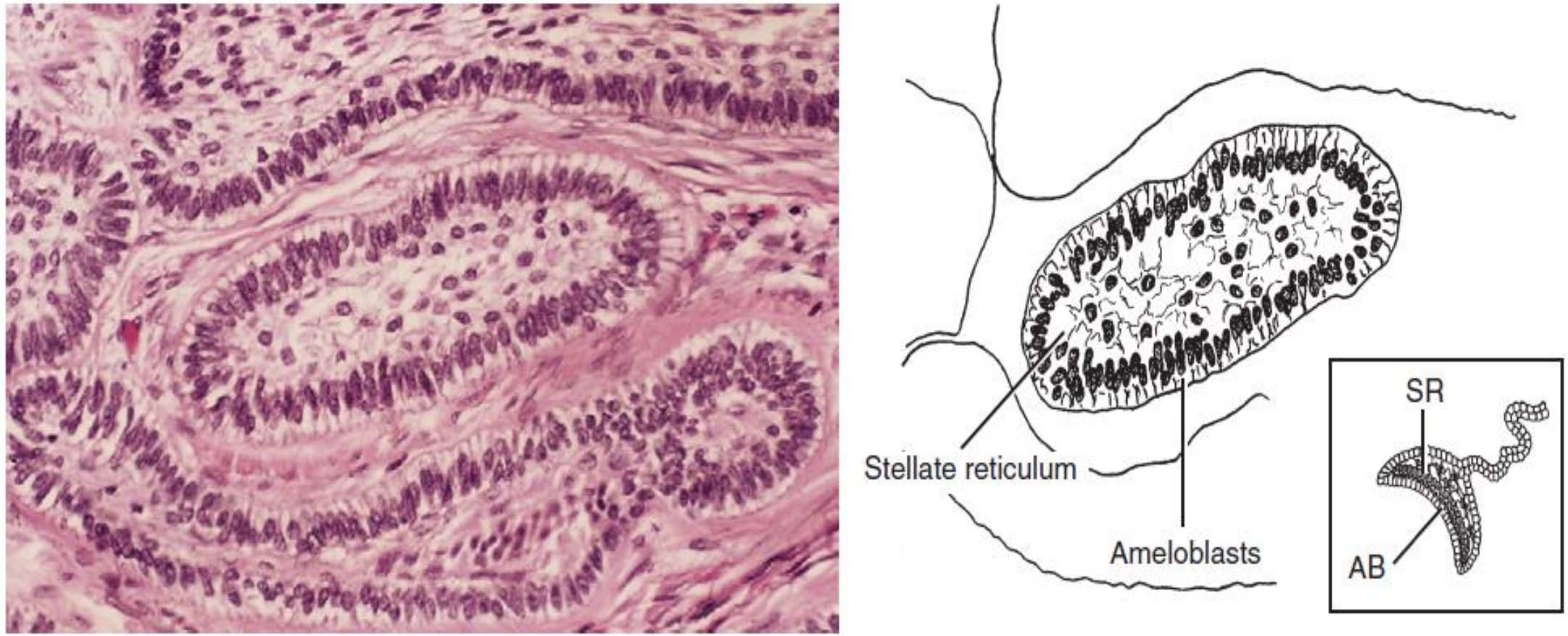


FIGURE 5-8

Ameloblastoma. Classic microscopic features of the common follicular form of ameloblastoma that demonstrate movement of the nucleus from basement membrane pole of basal cells to opposite pole (reverse polarization), resulting in outer cells resembling presecretory ameloblasts. Tumor recapitulates cap and early bell stage of odontogenesis *(inset)*. *AB,* Ameloblasts; *SR,* stellate reticulum.

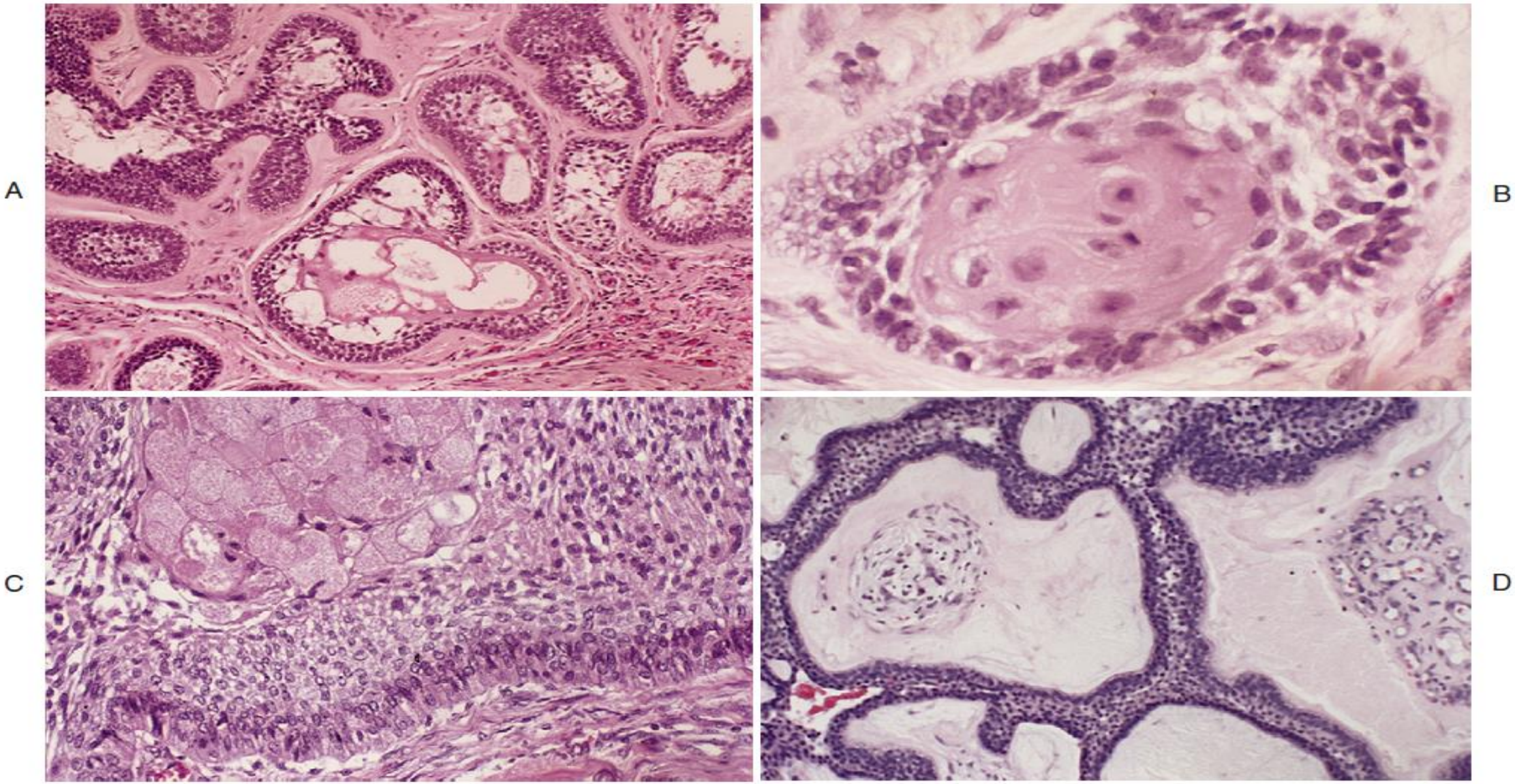
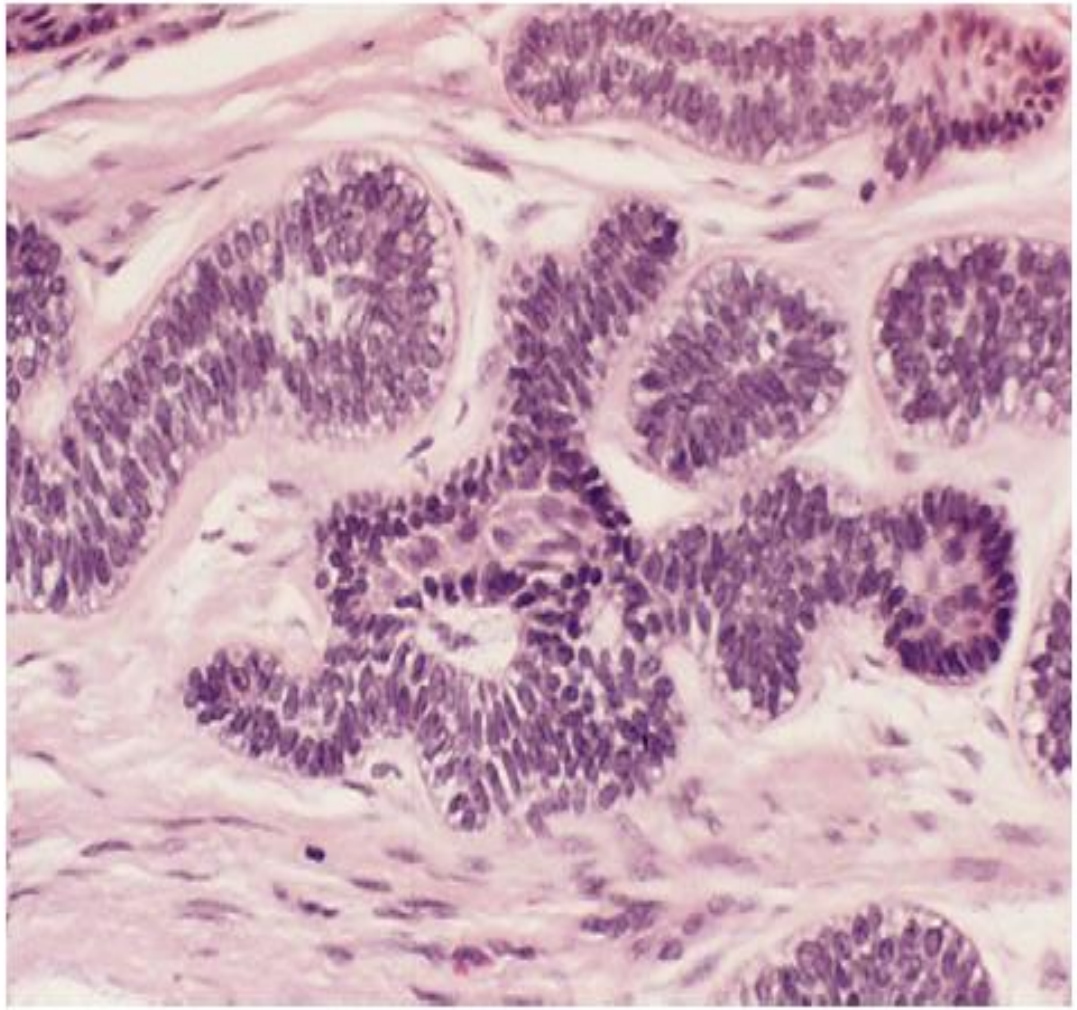
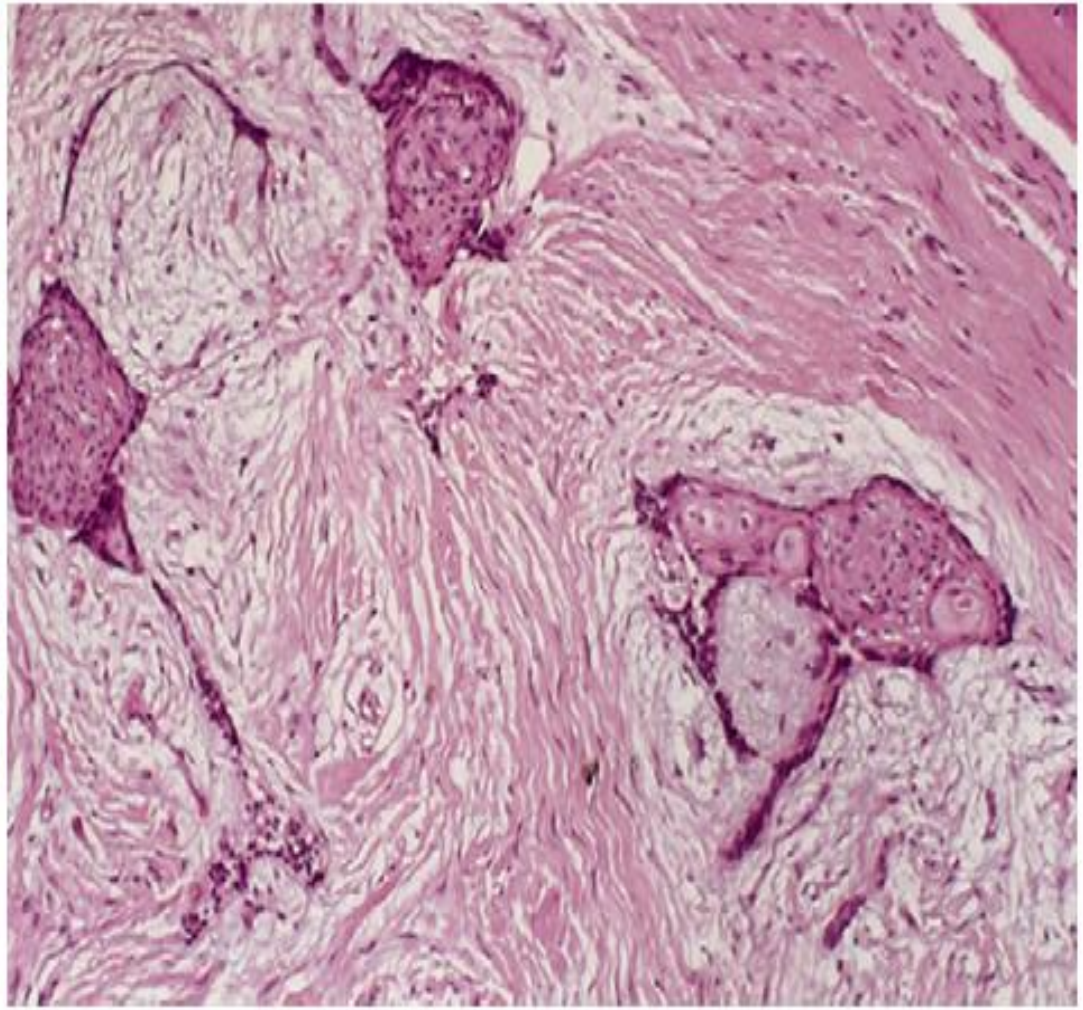


FIGURE 5-9

Ameloblastoma. Microscopic features of various histologic patterns. **A,** Follicular. **B,** Acanthomatous. **C,** Granular cell. **D,** Plexiform.

A





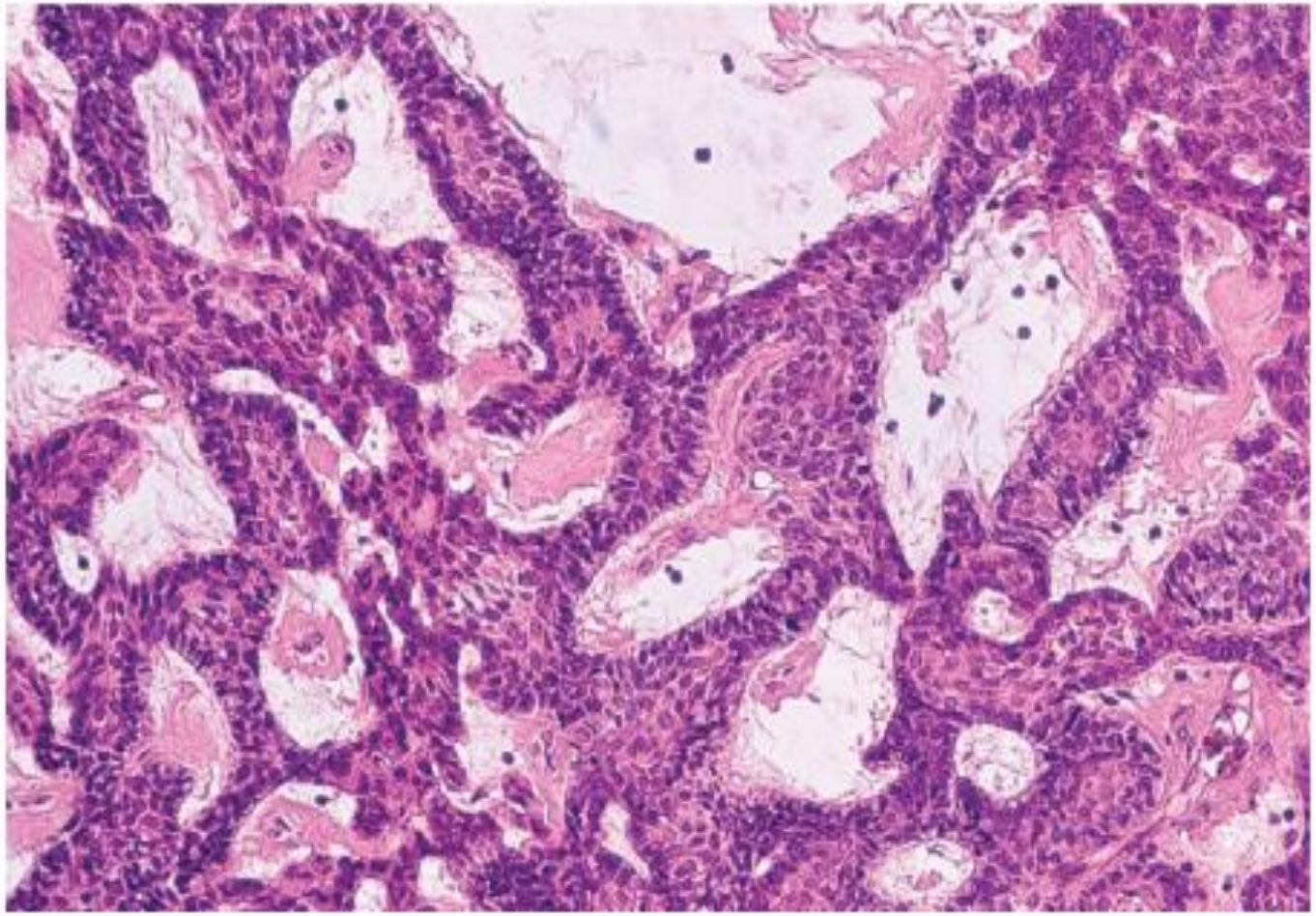
B

FIGURE 5-10

Ameloblastoma. Microscopic features of less common histologic patterns. **A,** Basal cell. **B,** Desmoplastic.

Plexiform pattern

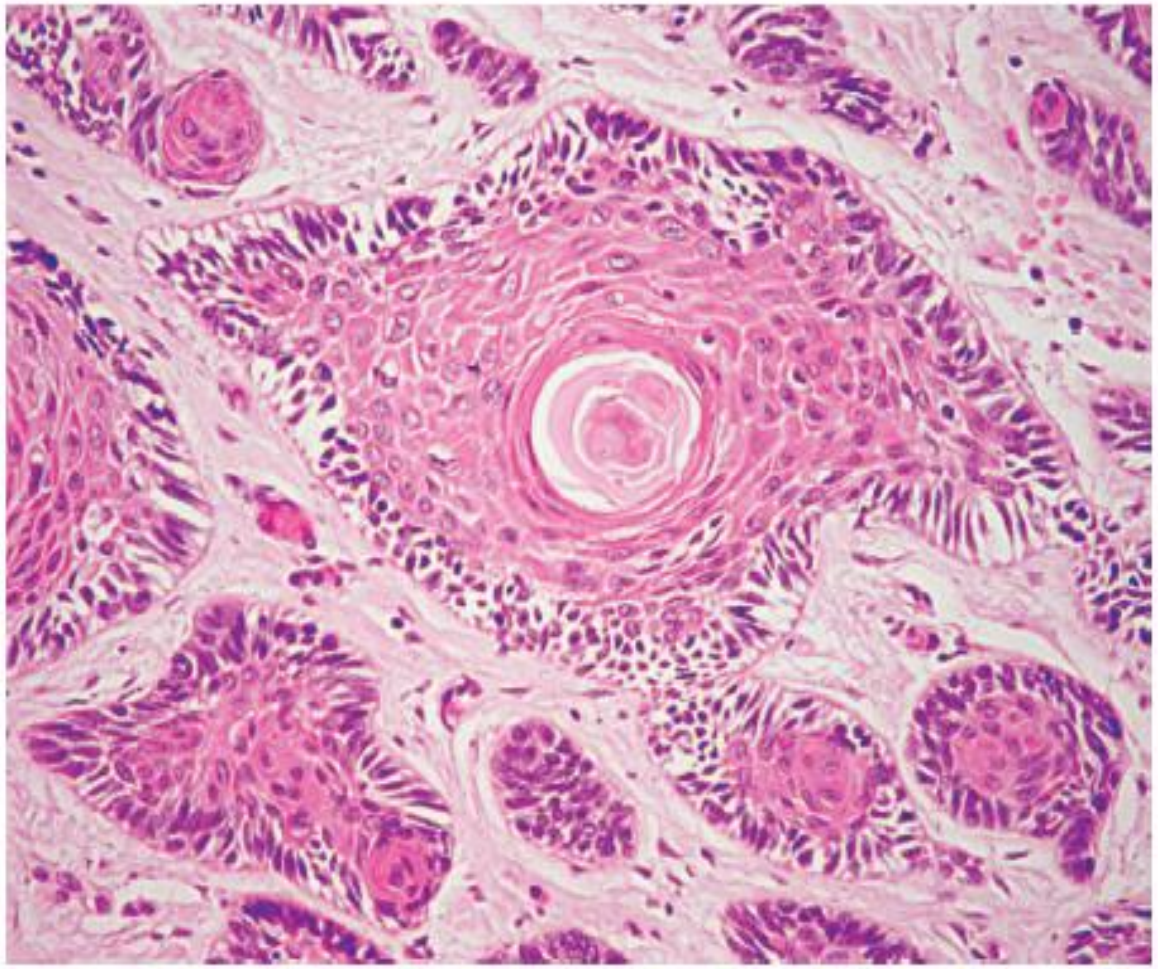
The plexiform type of ameloblastoma consists of long, anastomosing cords or larger sheets of odontogenic epithelium.The cords or sheets of epithelium are bounded by columnar or cuboidal ameloblast-like cells surrounding more loosely arranged epithelial cells. Occasionally the cuboidal cells may form structures resembling ducts, resulting in what has been termed an adenoid pattern. Such lesions may be misdiagnosed as adenomatoid odontogenic tumor or adenocarcinoma by the pathologist who is not familiar with this pattern. The supporting stroma in the plexiform pattern of ameloblastoma tends to be loosely arranged and vascular. Cyst formation is relatively uncommon in this variety. If the duct-like structures are accompanied by dentinoid deposition, then the diagnosis of a rare, relatively recently described, ameloblastoma-like tumor called adenoid ameloblastoma may be appropriate.



• **Fig. 15.71 Ameloblastoma (Plexiform Pattern).** Anastomosing cords of odontogenic epithelium.

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. This change does not indicate a more aggressive course for the lesion; histopathologically, however, such a lesion may be confused with squamous cell carcinoma or squamous odontogenicTumor.

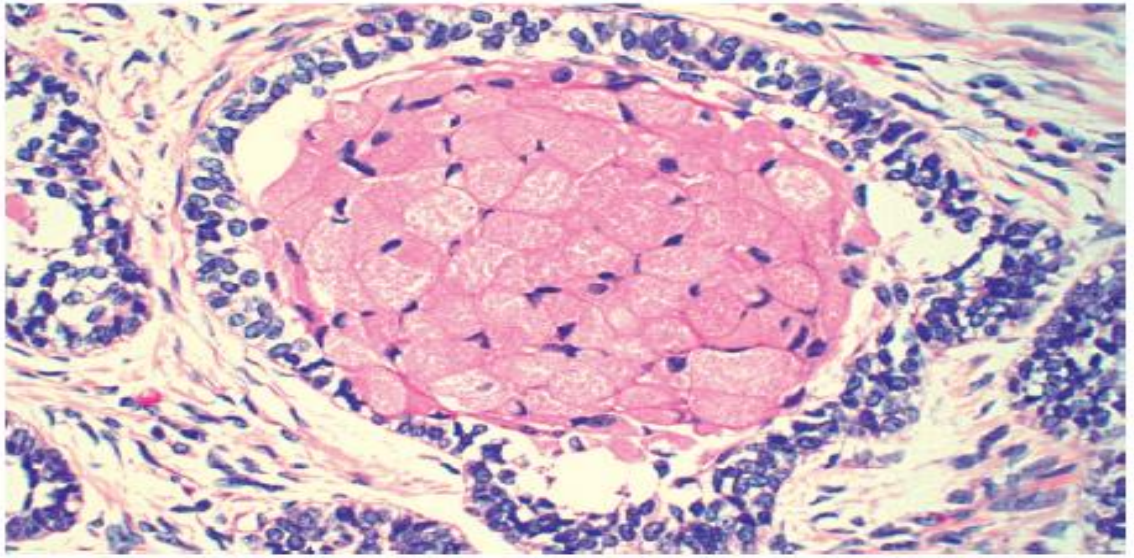


• **Fig. 15.72 Ameloblastoma (Acanthomatous Pattern).** Islands of ameloblastoma demonstrating central squamous differentiation.

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. Although originally considered to represent an aging or degenerative change in long-standing lesions,

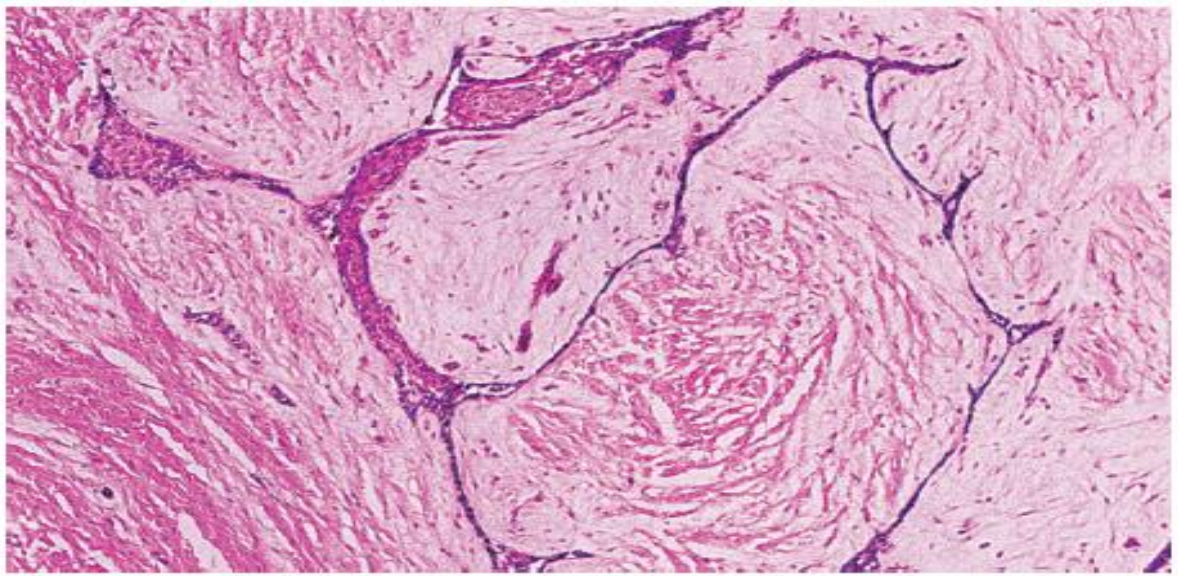
this variant has been seen in young patients. When this granular cell change is extensive in an ameloblastoma, the designation of granular cell ameloblastoma is appropriate.



• **Fig. 15.73 Ameloblastoma (Granular Cell Variant).** Tumor island exhibiting central cells with prominent granular cytoplasm.

Desmoplastic Pattern

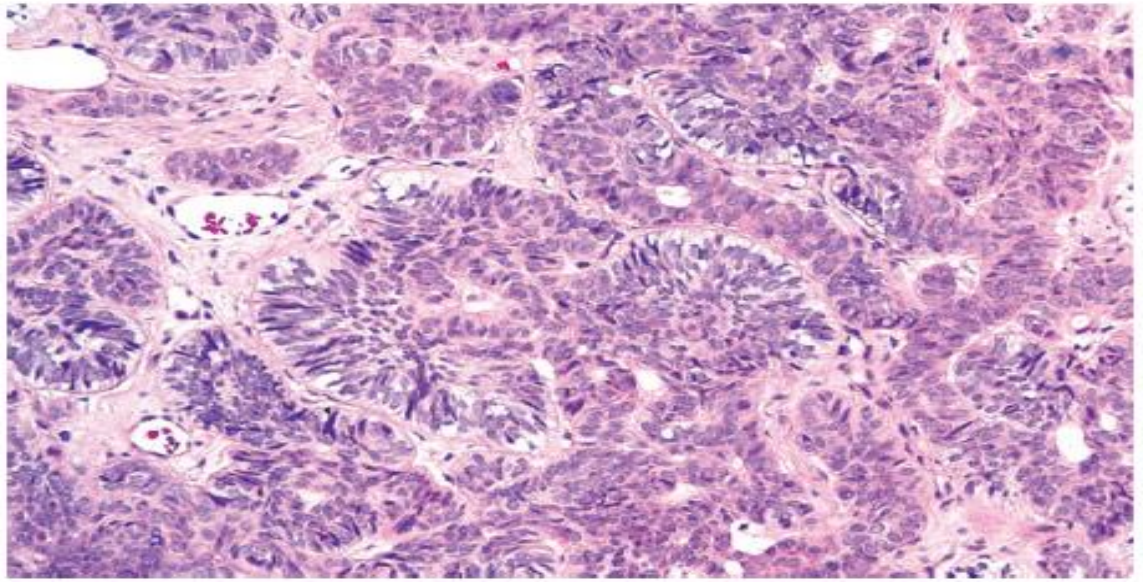
This type of ameloblastoma contains small islands and cords of odontogenic epithelium in a densely collagenized stroma. Immunohistochemical studies have shown increased production of transforming growth factor-β (TGF-β) in association with this lesion, suggesting that this may be responsible for the desmoplasia. Peripheral columnar ameloblast-like cells are inconspicuous about the epithelial islands



• **Fig. 15.74 Ameloblastoma (Desmoplastic Variant).** Thin cords of ameloblastic epithelium within a dense fibrous connective tissue 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.



• **Fig. 15.75 Ameloblastoma (Basal Cell Variant).** Islands of hyperchromatic basaloid cells with peripheral palisading.

Treatment and Prognosis

Treatment of conventional solid or multicystic intraosseous range from simple enucleation and curettage to enbloc 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%–90% have been reported in various studies after curettage. surgeons advocate that the margin of the resection should be at least 1.0–2.0 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. Orbital invasion by maxillary ameloblastomas. 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.

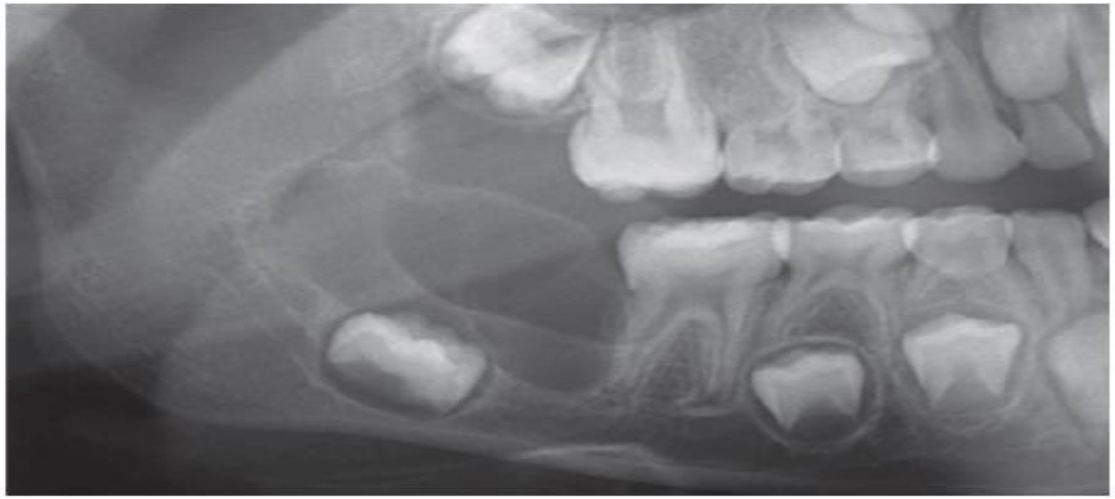
UNICYSTIC AMELOBLASTOMA

Unicystic ameloblastomas account for 10%–46% of all intraosseous ameloblastomas in various studies. Whether the unicystic ameloblastoma originates de novo as a neoplasm or whether it is the result of neoplastic transformation of nonneoplastic cyst epithelium has been long debated. Both mechanismsprobably occur.

Clinical and Radiographic Features

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

clinically resembling a dentigerous cyst. Other tumors simply appear as sharply defined radiolucent areas and are usually considered to be a primordial, radicular, or residual cyst, depending on the relationship of the lesion to teeth in the area.



• **Fig. 15.77 Unicystic Ameloblastoma.** Radiolucency associated with the crown of the developing mandibular second molar. (Courtesy of Dr. Joseph Van Sickels.)

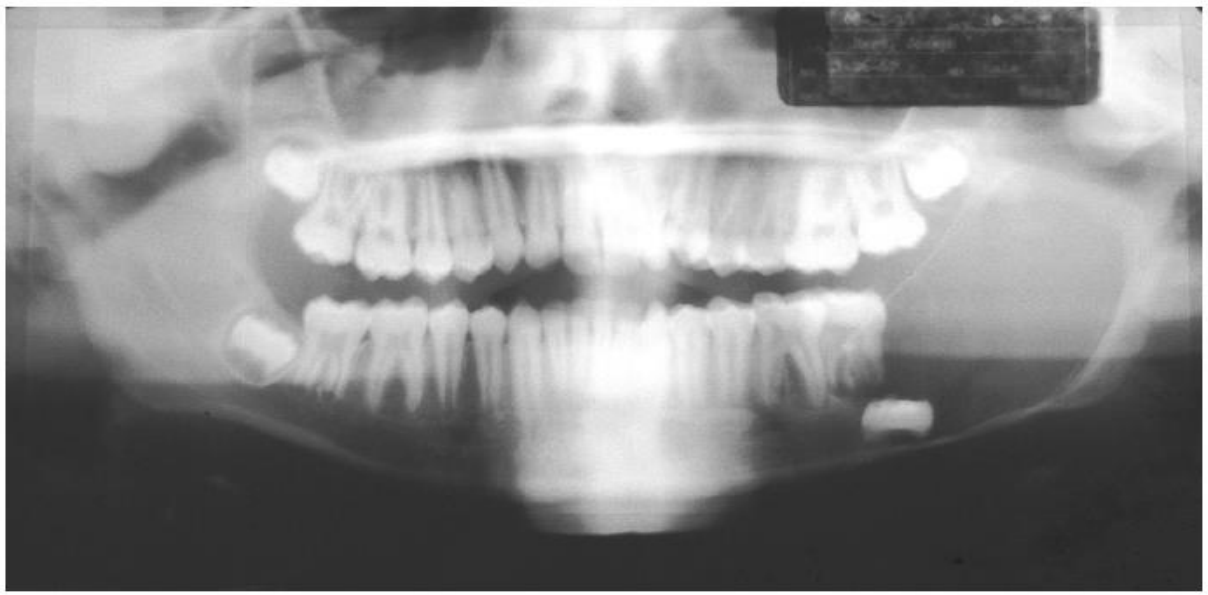


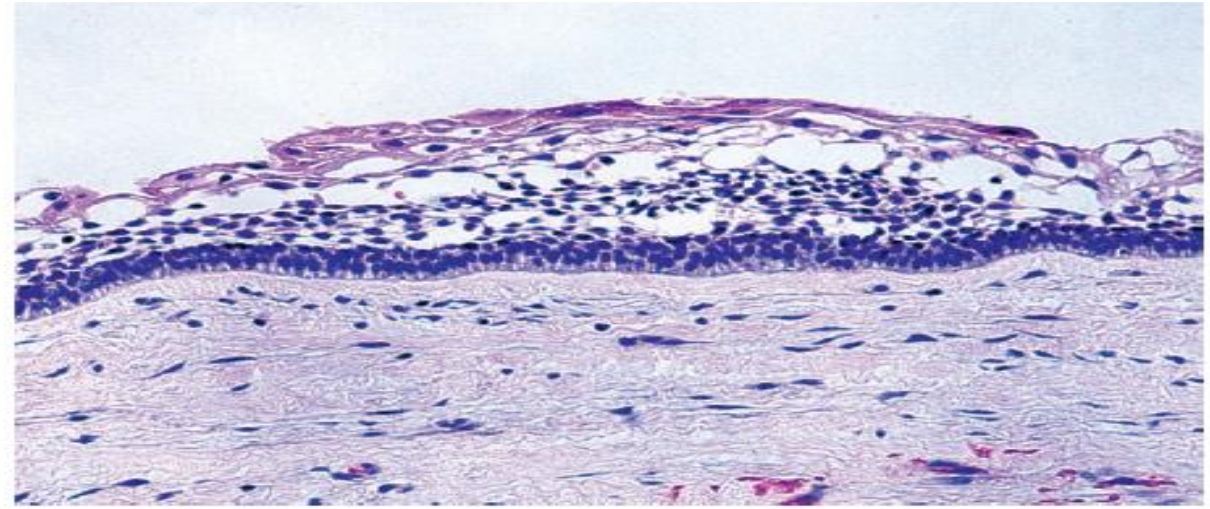
FIGURE 5-11
Unicystic ameloblastoma. Radiograph of young patient with large lesion of left mandible, exhibiting inferiorly displaced, partly formed molar and expansion of cortical plates.

Histopathologic Features

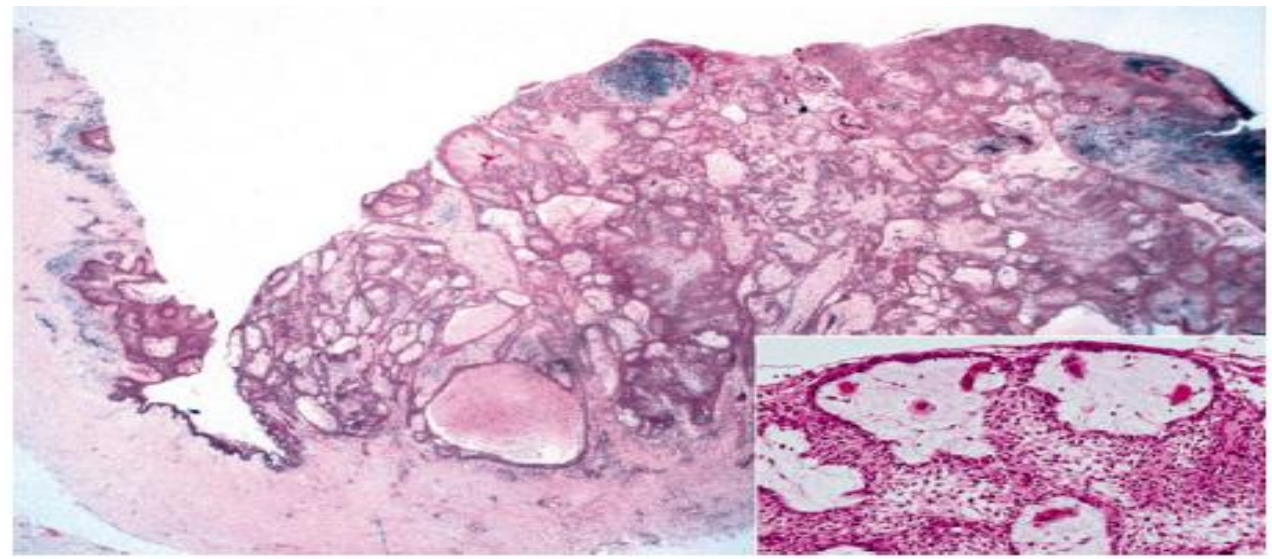
Three histopathologic variants of unicystic ameloblastoma .

In the first type (luminal unicysticameloblastoma), the tumor is confined to the luminal surface of the cyst. The lesion consists of a fibrous cyst wall with a lining composed totally or partially of ameloblastic epithelium. The lining demonstrates a basal layer of columnar or cuboidal cells with hyperchromatic nuclei that show reverse polarity and basilar cytoplasmic vacuolization.The upper epithelial cells are loosely cohesive and resemble stellate reticulum. This finding does not seem to be related to inflammatory edema.

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 . These lesions are sometimes referred to as plexiform unicystic ameloblastomas.



• **Fig. 15.79 Unicystic Ameloblastoma (Luminal Type).** The cyst is lined by ameloblastic epithelium showing a hyperchromatic, polarized basal layer. The overlying epithelial cells are loosely cohesive and resemble stellate reticulum.



• **Fig. 15.80 Unicystic Ameloblastoma (Intraluminal Plexiform Type).** Photomicrograph of an intraluminal mass arising from the cyst wall. The *inset* shows the intraluminal mass at higher magnification.

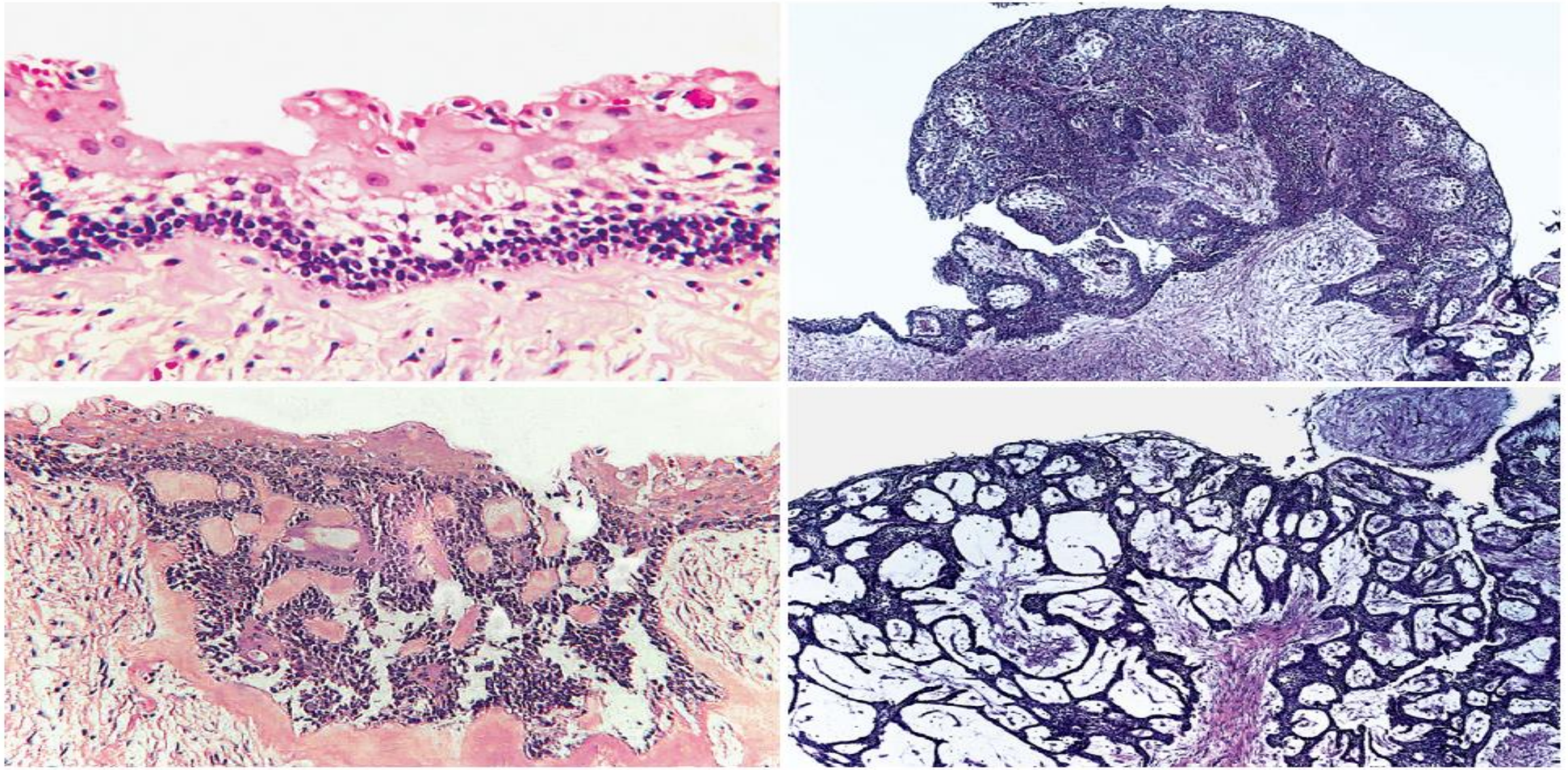


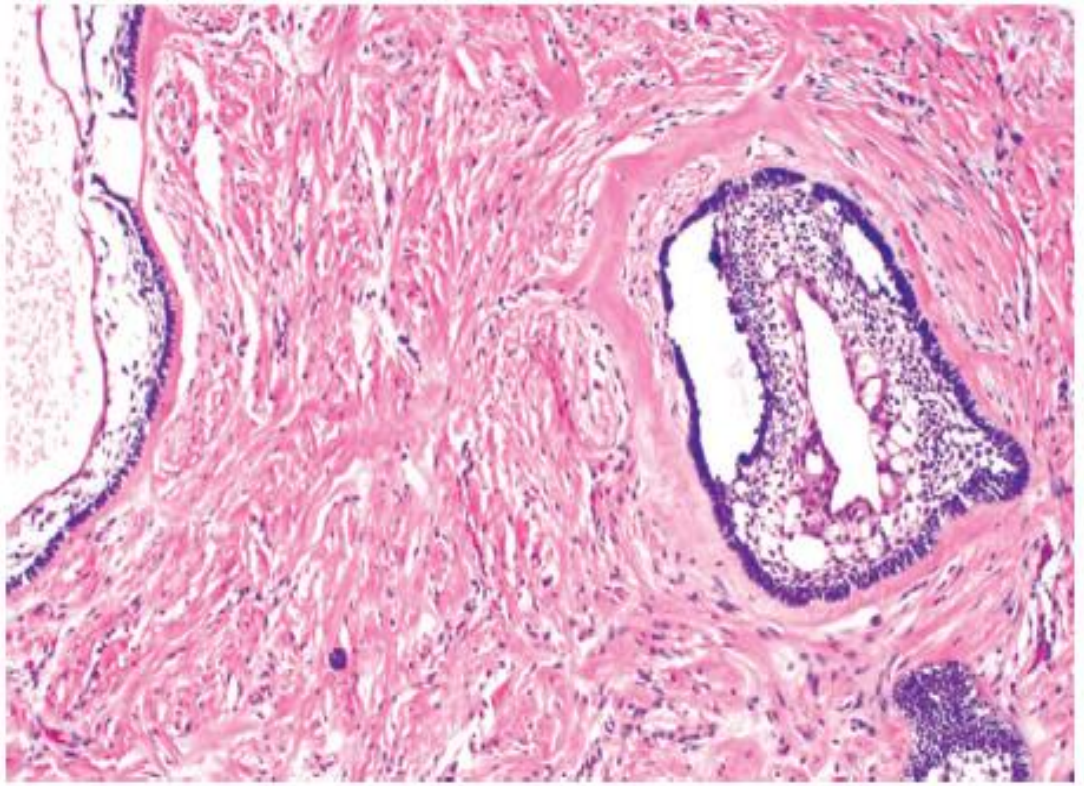
FIGURE 5-12

Unicystic ameloblastoma. Microscopic features of typical lining exhibit features of ameloblastoma **(A)**, intraluminal papillary projection **(B)**, intracapsular mural penetration **(C)**, and the plexiform pattern sometimes found **(D)**.

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.

Treatment and Prognosis

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. The diagnosis of ameloblastoma is made only after microscopic examination of the presumed cyst. If the ameloblastic elements are confined to the lumen of the cyst with or without intraluminal tumor extension, then the cyst enucleation has been adequate treatment. The patient should be kept under long-term follow up.



• **Fig. 15.81 Unicystic Ameloblastoma (Mural Type).** The epithelial lining of the cystic component can be seen on the left edge of the photomicrograph. Islands of follicular ameloblastoma are infiltrating into the fibrous connective tissue wall on the right.

PERIPHERAL (EXTRAOSSEOUS) AMELOBLASTOMA

The peripheral ameloblastoma is uncommon and accounts for about 1%–4% 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

The peripheral ameloblastoma is usually a painless, nonulcerated sessile or pedunculated gingival or alveolar mucosal lesion. The clinical features are non-specific, and most lesions are clinically considered to represent a fibroma or pyogenic granuloma. Most examples are smaller than 1.5 cm ,large lesions can occur . most are seen in middle-aged persons, with an average reported age of 52 years. Peripheral ameloblastomas are most commonly found on the posterior gingival and alveolar mucosa, and they are more common in mandibular than in maxillary areas. In some cases, the superficial alveolar bone becomes slightly eroded, but significant bone involvement does not occur.

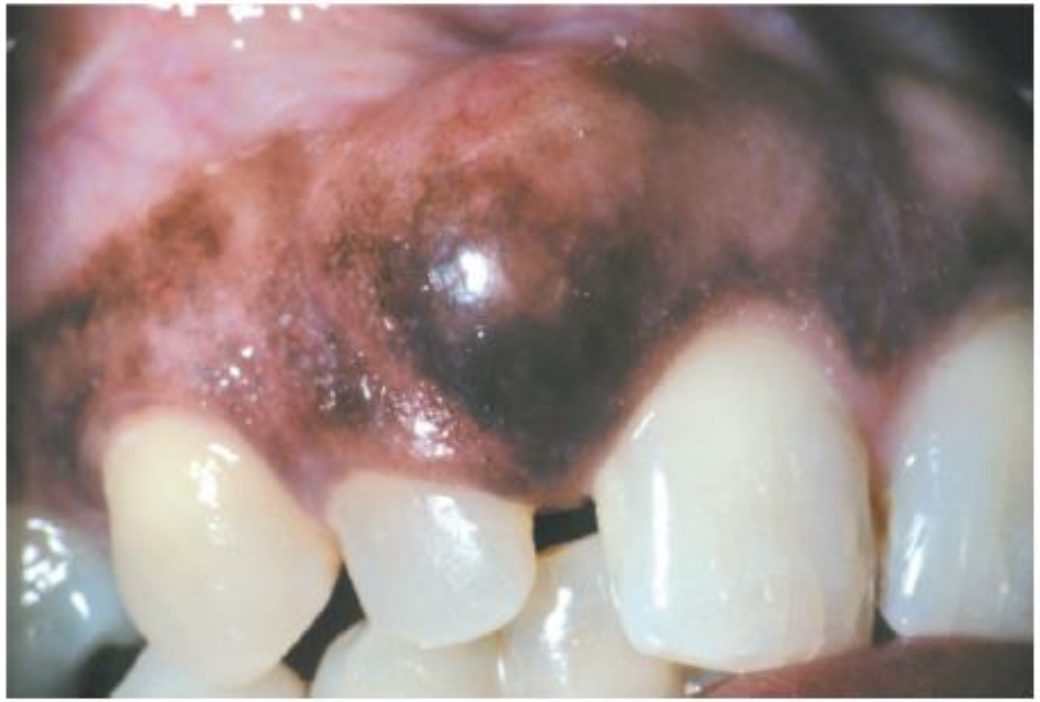
Histopathologic Features

Peripheral ameloblastomas have 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. This may represent origin of the tumor from the basal layer of the epithelium in some cases, but in other instances the tumor could develop in the gingival connective tissue and merge with the surface epithelium. A peripheral odontogenic fibroma may be confused microscopically with a peripheral ameloblastoma, particularly if a prominent epithelial component is present in the former.

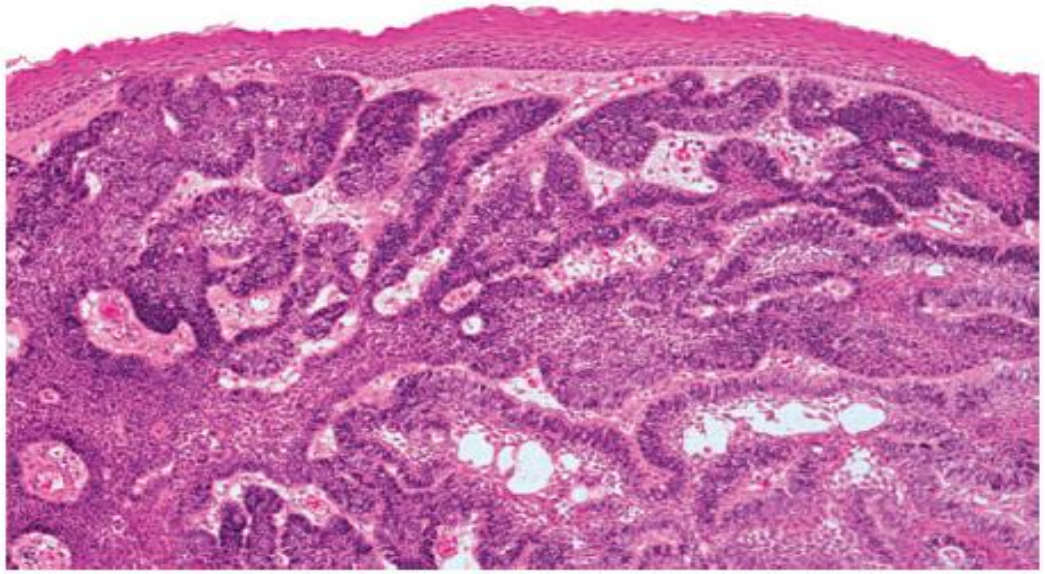
The presence of dysplastic dentin or cementum-like elements in the peripheral odontogenic fibroma and the lack of peripheral columnar epithelial cells showing reverse polarity of their nuclei should serve to distinguish the two lesions.

Treatment and Prognosis

the peripheral ameloblastoma shows an innocuous clinical behavior. Patients respond well to local surgical excision. Although local recurrence has been noted in 15%–20% of cases, further local excision almost always results in a cure.



• **Fig. 15.82 Peripheral Ameloblastoma.** Sessile gingival mass. (Courtesy of Dr. Dean K. White.)



• **Fig. 15.83 Peripheral Ameloblastoma.** Interconnecting cords of ameloblastic epithelium filling the lamina propria.

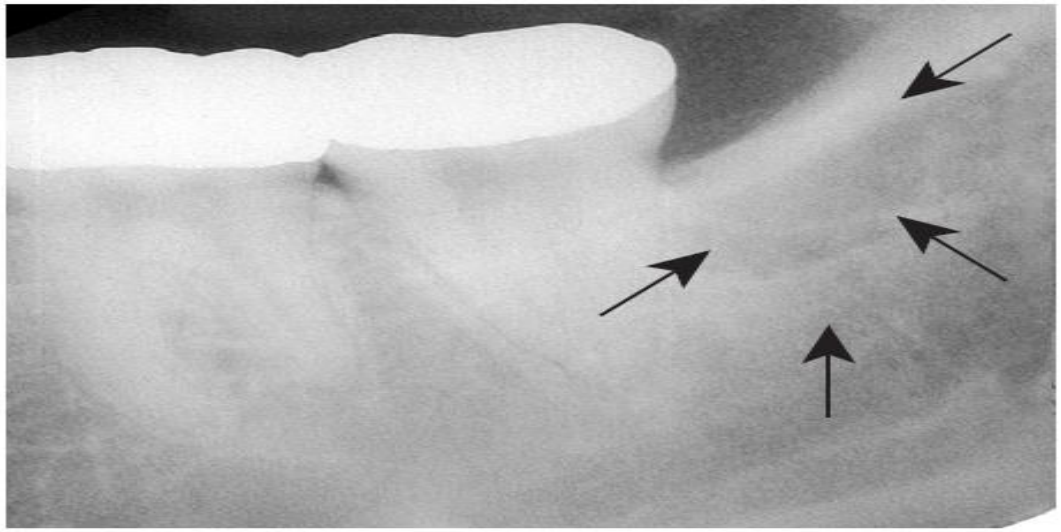


FIGURE 5-13
Peripheral ameloblastoma. Radiograph of lesion of retromolar pad exhibiting saucerization *(arrows)* of cortical bone.

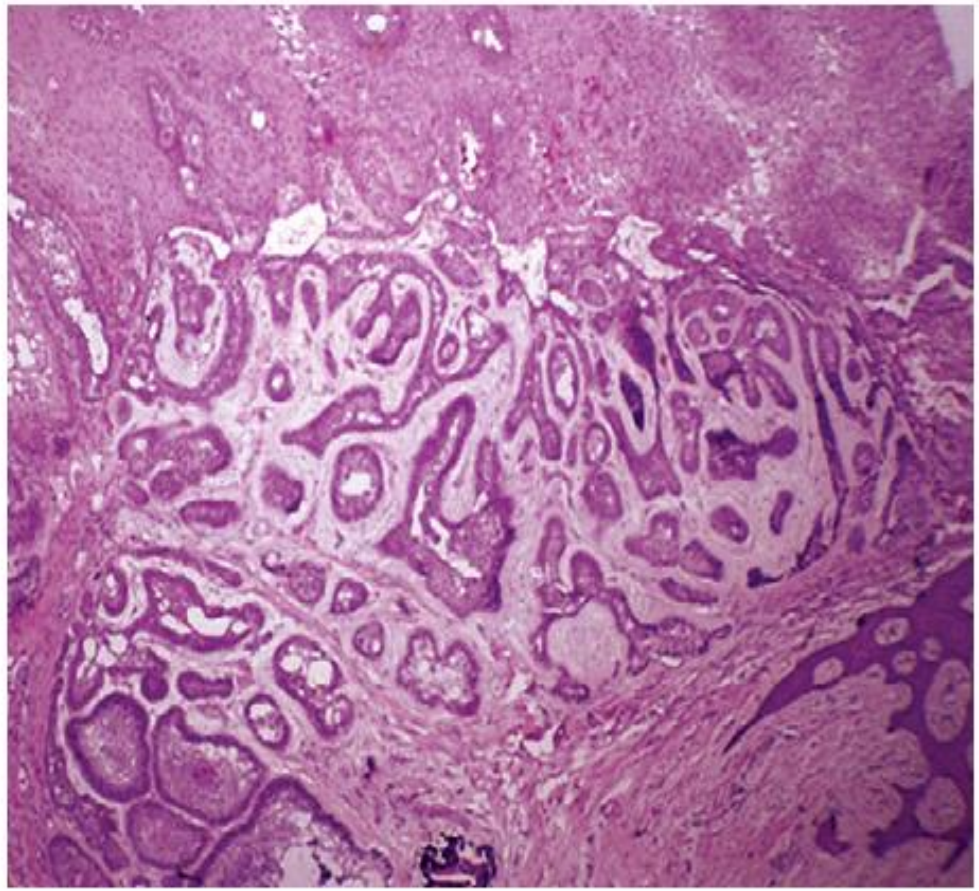


FIGURE 5-14

Peripheral ameloblastoma. Microscopic appearance exhibiting islands of follicular ameloblastoma in close proximity to surface epithelium.

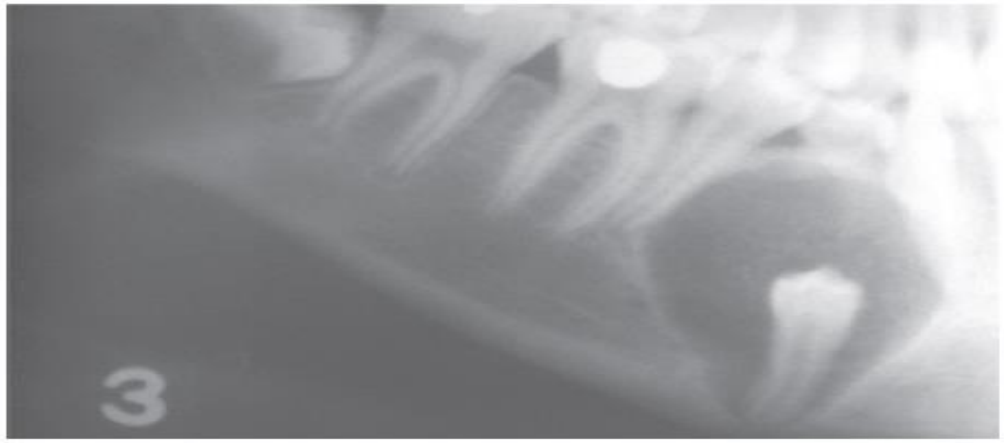
ADENOMATOID ODONTOGENIC TUMOR

The adenomatoid odontogenic tumor represents 2%–7%nof all odontogenic tumors, Postulated histogenetic sources of the tumor cellshave included enamel organ epithelium, reduced enamel epithelium, and rests of Malassez; adenomatoid odontogenic tumor is perhaps better thought as a hamartoma, rather than a neoplasm

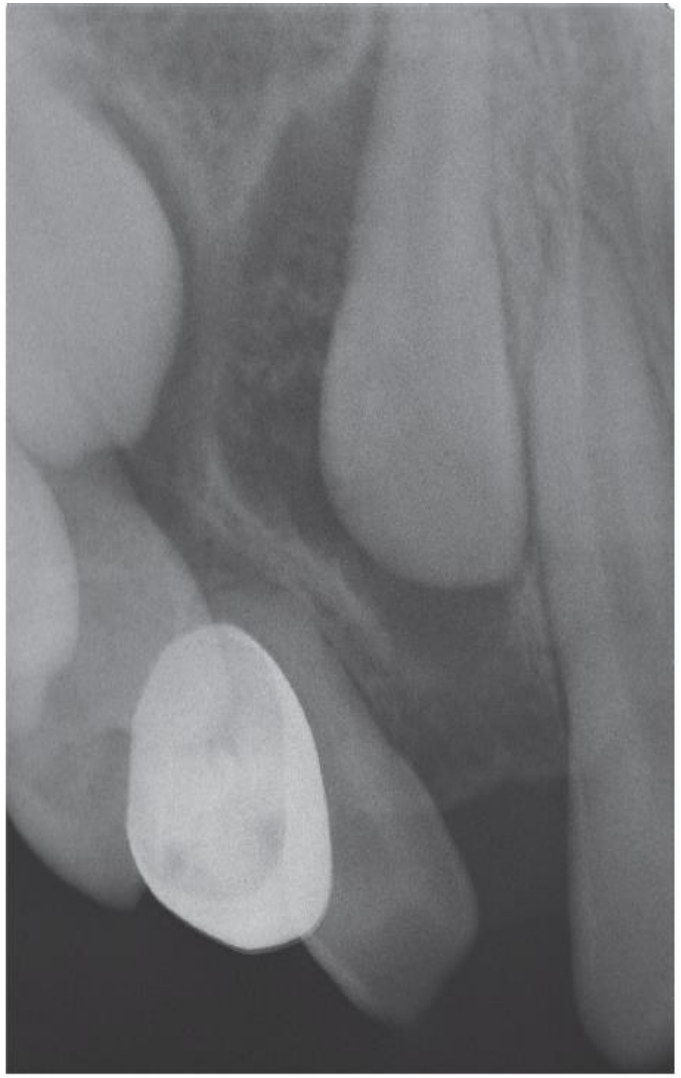
Clinical and Radiographic Features

Adenomatoid odontogenic tumors are largely limited to younger patients, and two-thirds of all cases are diagnosed when patients are 10–19 years of age. It is uncommon in a patient older than age 30. It has a striking tendency to occur in the anterior portions of the jaws and is found twice as often in the maxilla as in the mandible. Females are affected twice than males. Most adenomatoid odontogenic tumors are relatively small. They seldom exceed 3 cm in greatest diameter. Peripheral (extraosseous) forms appear as small, sessile masses on the facial gingiva of the maxilla. Clinically, these lesions cannot be differentiated from the common gingival fibrous lesions. Adenomatoid odontogenic tumors are frequently asymptomatic and are discovered during the course of a routine radiographic examination. Larger lesions cause a painless expansion of the bone. In about 75% of cases, the tumor appears as a circumscribed, unilocular radiolucency that involves the crown of an unerupted tooth, most often a canine. This follicular type of adenomatoid odontogenic tumor may be impossible to differentiate radiographically from the more common dentigerous cyst.

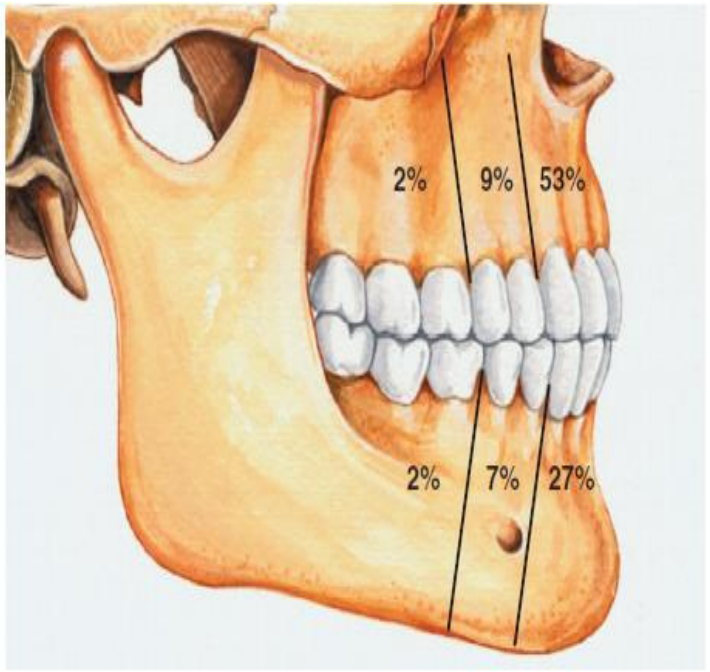
The radiolucency associated with the follicular type of adenomatoid odontogenic tumor sometimes extends apically along the root past the cementoenamel junction. This feature may help to distinguish an adenomatoid odontogenic tumor from a dentigerous cyst. The lesion may appear completely radiolucent,however, it contains fine (snowflake) calcifications .This feature may be helpful in differentiating the adenomatoid odontogenic tumor from a dentigerous cyst.



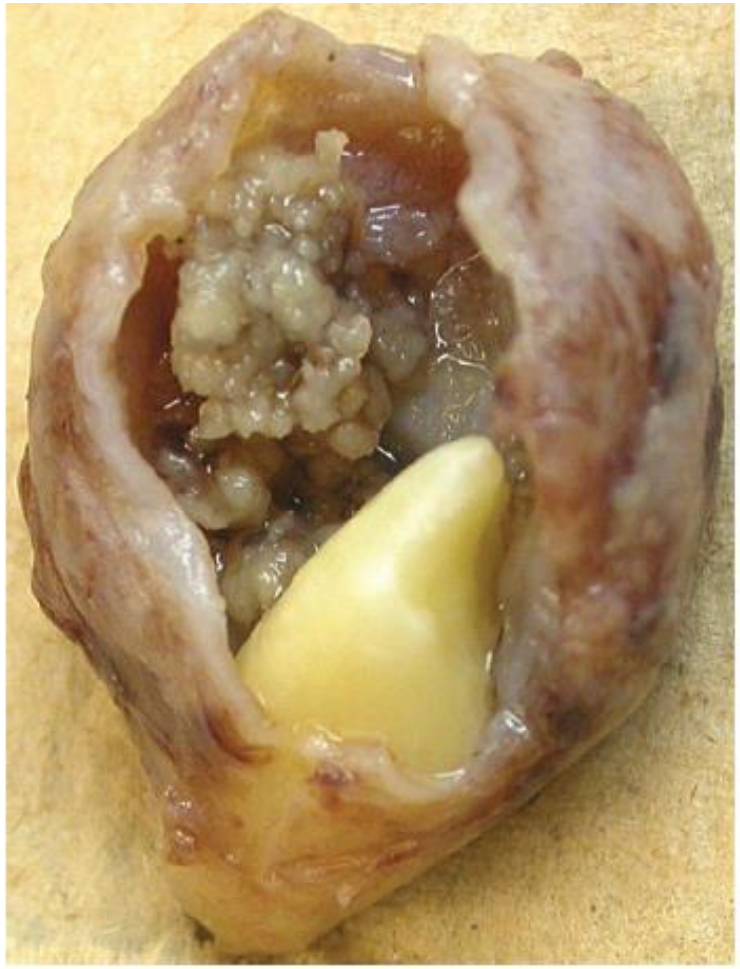
• **Fig. 15.90 Adenomatoid Odontogenic Tumor.** Radiolucent lesion involving an unerupted mandibular first premolar. In contrast to the usual dentigerous cyst, the radiolucency extends almost to the apex of the tooth. (Courtesy of Dr. Tony Traynham.)



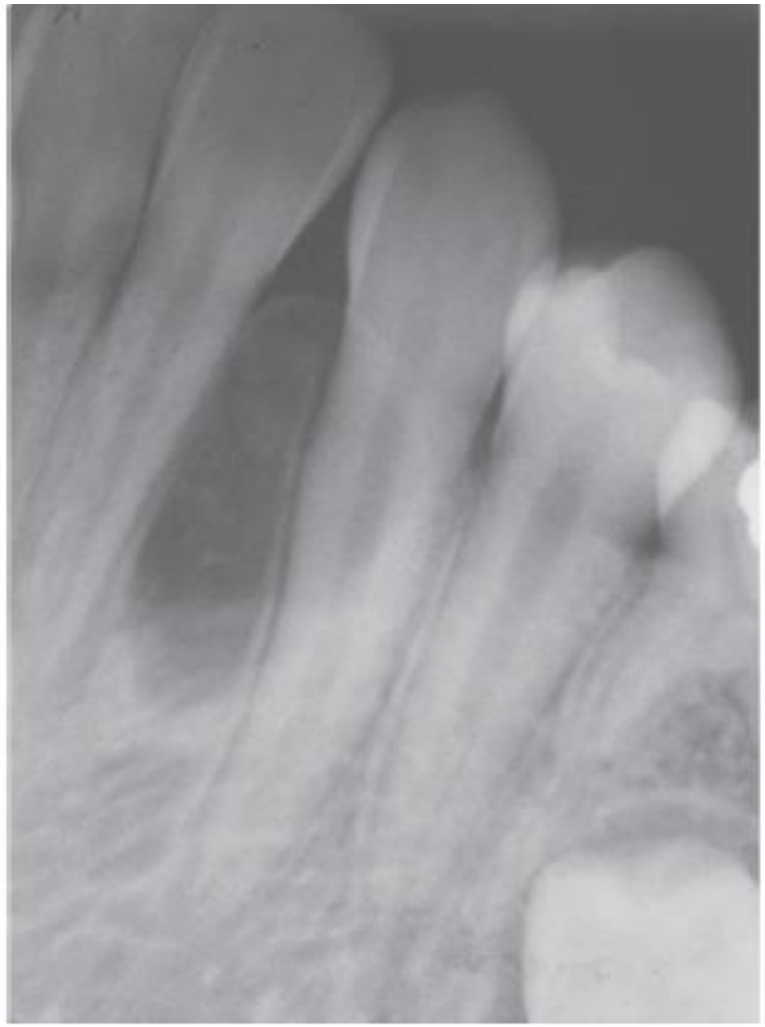
• **Fig. 15.92 Adenomatoid Odontogenic Tumor.** Well-defined pericoronal radiolucency enveloping the maxillary right lateral incisor in a 14-year-old male. Note the subtle snowflake-like calcifications within the lesion. (Courtesy of Dr. Jason Barker.)



• **Fig. 15.89 Adenomatoid Odontogenic Tumor.** Relative distribution of adenomatoid odontogenic tumor in the jaws.



• **Fig. 15.93 Adenomatoid Odontogenic Tumor.** A well-circumscribed cystlike mass can be seen enveloping the crown of a maxillary cuspid. Note the intraluminal vegetations, which represent nodular tumor growth.



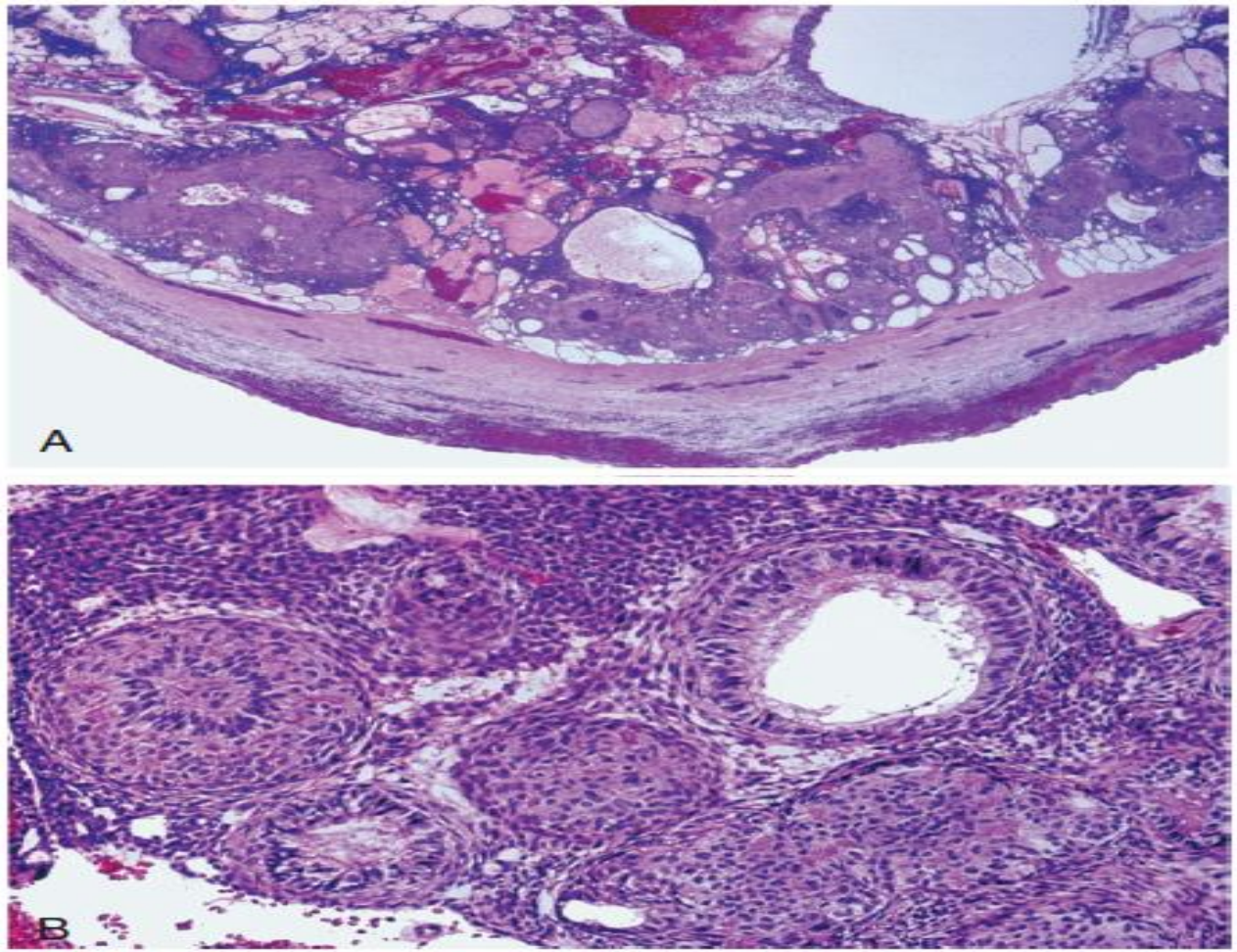
• **Fig. 15.91 Adenomatoid Odontogenic Tumor.** A small radiolucency is present between the roots of the lateral incisor and canine. (Courtesy of Dr. Ramesh Narang.)

Histopathologic Features

The adenomatoid odontogenic tumor is a well-defined lesion that is usually surrounded by a thick, fibrous capsule. When the lesion is bisected, the central portion of the tumor may be essentially solid or may show varying degrees of cystic change. Microscopically, the tumor is composed of spindle shaped epithelial cells that form sheets, strands, or whorled masses of cells in a scant fibrous stroma. The epithelial cells may form rosettelike structures about a central space, which may be empty or contain small amounts of eosinophilic material. This material may stain for amyloid.The tubular or ductlike structures, which are the characteristic feature of the adenomatoid odontogenic tumor, may be prominent, scanty, or even absent in a given lesion. The mechanism of formation of these tubular structures is likely the result of the secretory activity of the tumor cells, which appear to be preameloblasts. these structures are not true ducts, and no glandular elements are present in the tumor. Small foci of calcification may also be scattered throughout the tumor.

Treatment and Prognosis

The adenomatoid odontogenic tumor is completely benign; because of its capsule, it enucleates easily from the bone.



• **Fig. 15.94 Adenomatoid Odontogenic Tumor. A,** Low-power view demonstrating a thick capsule surrounding the tumor. **B,** Higher magnification showing the ductlike epithelial structures. The nuclei of the columnar cells are polarized away from the central spaces.

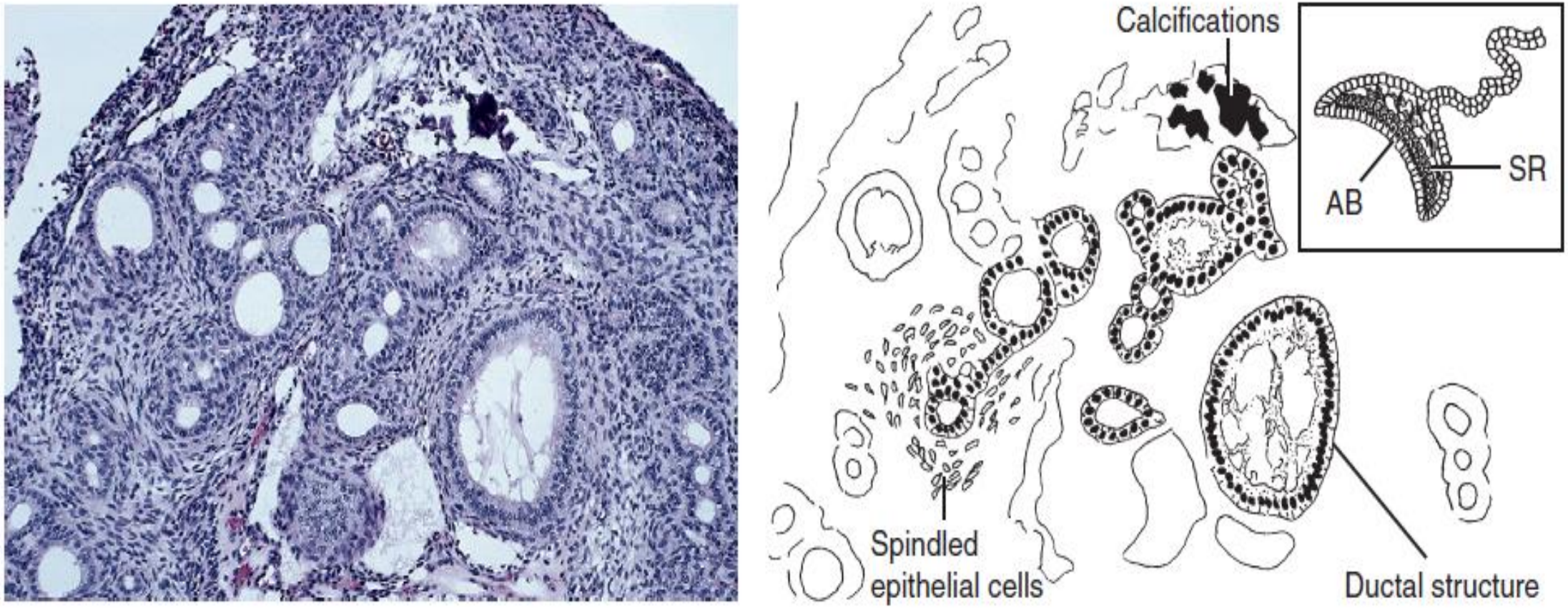


FIGURE 5-19

Adenomatoid odontogenic tumor. Microscopic features reveal the nodular pattern of spindled cells, ductal structures, and a focal area of calcification *(upper center)*, which is characteristic of this lesion. Tumor cells recapitulate presecretory ameloblasts and stellate reticulum of the cap and early bell stage of odontogenesis *(inset)*. *AB,* Ameloblasts; *SR,* stellate reticulum.

CALCIFYING EPITHELIAL ODONTOGENIC TUMOR (PINDBORG TUMOR)

The calcifying epithelial odontogenic tumor, also widely known as the Pindborg tumor, is an uncommon lesion that accounts for less than 1% of all odontogenic tumors.

Clinical and Radiographic Features:

it is most often encountered in patients between 30 and 50 years of age. There is no sex predilection.About two-thirds of all reported cases have been found in the mandible, most often in the posterior areas .A painless, slow-growing swelling is the most common presenting sign.

Radiographically, the tumor exhibits either a unilocular or a multilocular radiolucent defect, with the unilocular pattern encountered more commonly in the maxilla.The margins of the lytic defect are often scalloped and usually relatively well defined. The tumor is frequently associated with an impacted tooth, most often a mandibular molar. The lesion may be entirely radiolucent, but calcified structures of varying size and density are commonly seen.

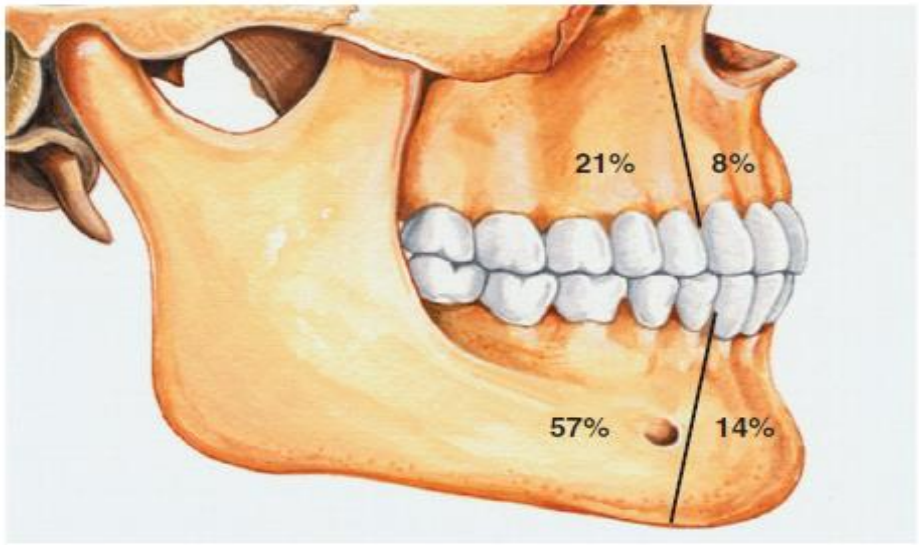
peripheral (extraosseous) calcifying epithelial odontogenic tumor have been reported. These appear as nonspecific, sessile gingival masses, most often on the anterior gingiva. Some of these have been associated with cupped-out erosion of the underlying bone.

Histopathologic Features

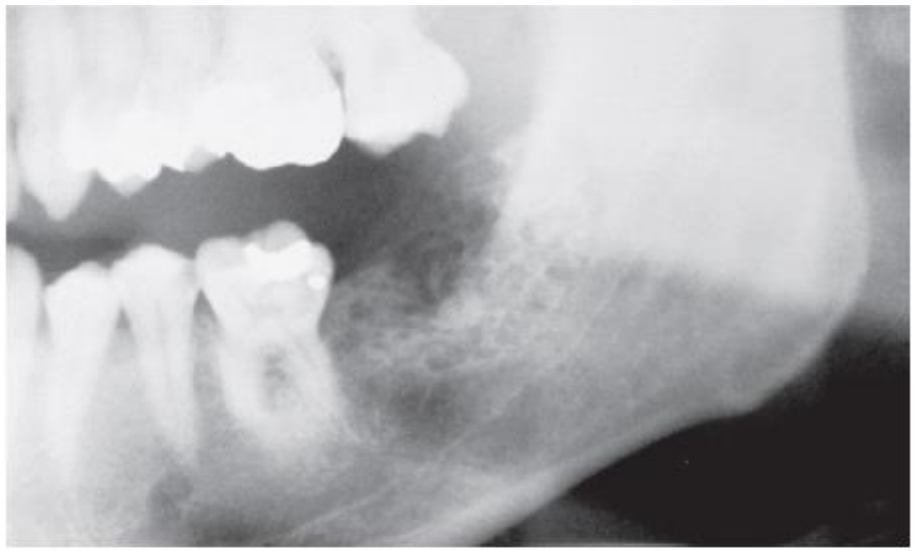
- The calcifying epithelial odontogenic tumor has discrete islands, strands, or sheets of polyhedral epithelial cells in a fibrous stroma .The cellular outlines of the epithelial cells are distinct, and intercellular bridges may be noted. The nuclei show considerable variation, and giant nuclei may be seen. Large areas of amorphous, eosinophilic, hyalinized(amyloid-like) extracellular material are also often present.The tumor islands frequently enclose masses of this hyaline material, resulting in a cribriform appearance. Calcifications, which are a distinctive feature of the tumor, develop within the amyloid-like material and form concentric rings (Liesegang ring calcifications). These tend to fuse and form large, complex masses. The amyloid-like material in the Pindborg tumor stains positive with Congo red). After Congo red staining, the amyloid will exhibit apple-green birefringence when viewed with polarized light. Conservative

Treatment and Prognosis

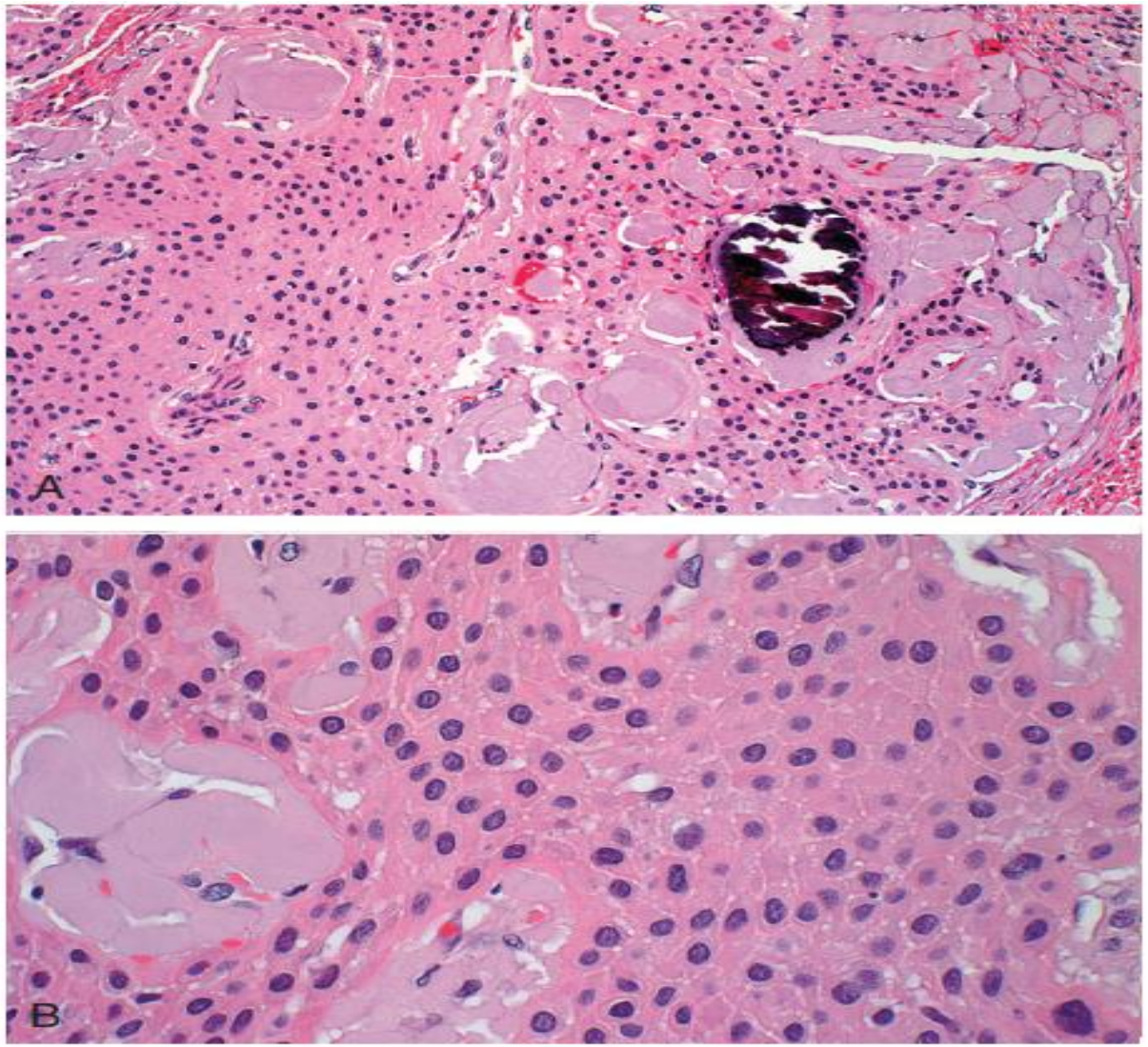
local resection to include a narrow rim of surrounding bone appears to be the treatment of choice, although lesions in the posterior maxilla should probably be treated more aggressively. tumors treated by curettage have the highest frequency of recurrence. The overall prognosis appears good.



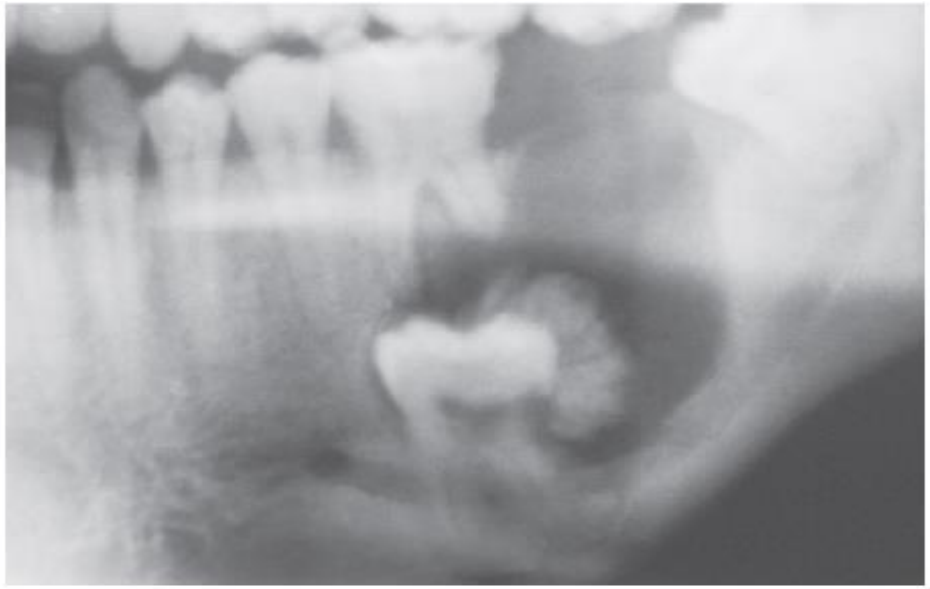
• **Fig. 15.95 Calcifying Epithelial Odontogenic Tumor.** Relative distribution of calcifying epithelial odontogenic tumor in the jaws.



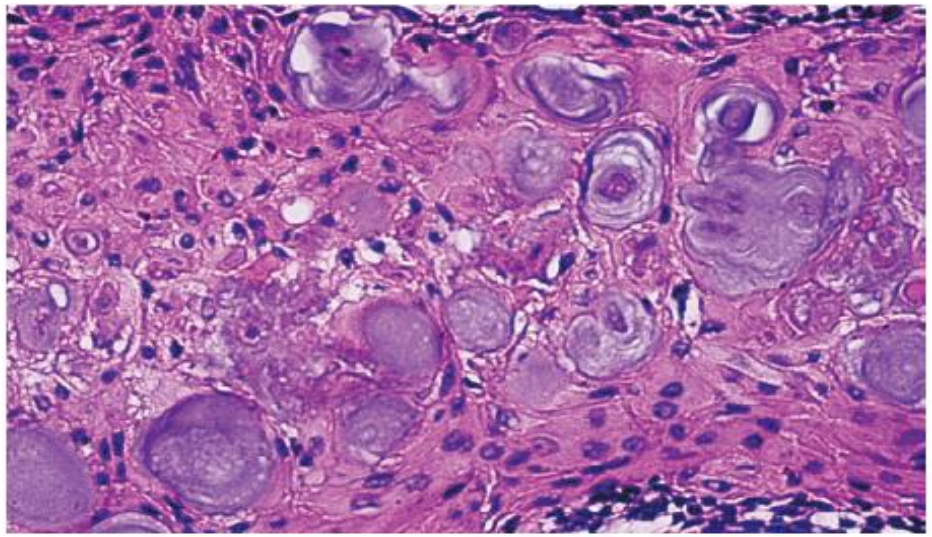
• **Fig. 15.96 Calcifying Epithelial Odontogenic Tumor.** Honeycombed multilocular radiolucency containing fine calcifications.



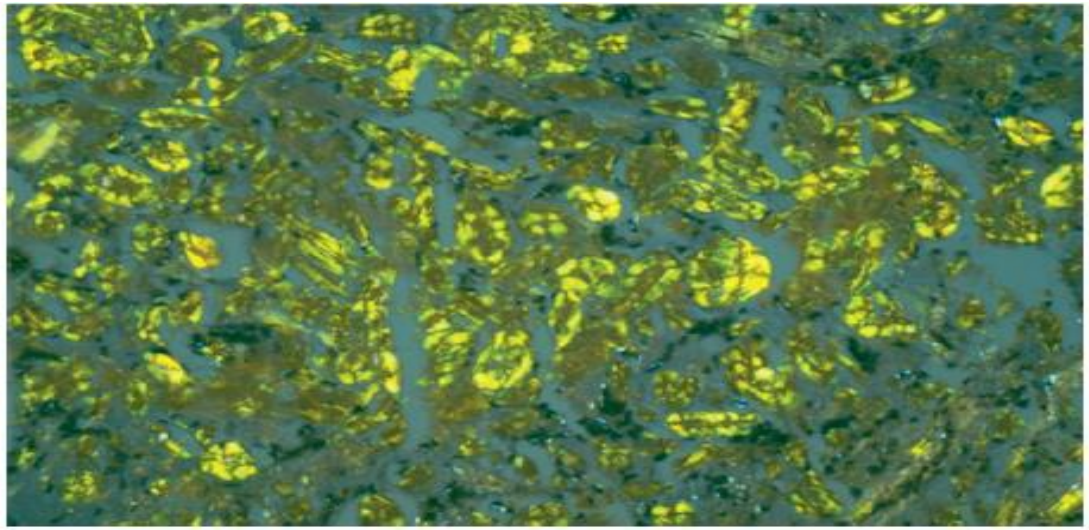
• **Fig. 15.98 Calcifying Epithelial Odontogenic Tumor. A,** Sheets of epithelial tumor cells that surround pools of amorphous, eosinophilic amyloid with focal calcification. **B,** Higher-power view showing polyhedral cells with eosinophilic cytoplasm and intercellular bridging. Adjacent amyloid deposits can be seen.



• **Fig. 15.97 Calcifying Epithelial Odontogenic Tumor.** Prominent calcification around the crown of an impacted second molar that is involved in the tumor. (Courtesy of Dr. Harold Peacock.)



• **Fig. 15.99 Calcifying Epithelial Odontogenic Tumor.** Multiple concentric Liesegang ring calcifications.



• **Fig. 15.100 Calcifying Epithelial Odontogenic Tumor.** With Congo red staining, the pools of amyloid exhibit an apple-green birefringence when viewed with polarized light.

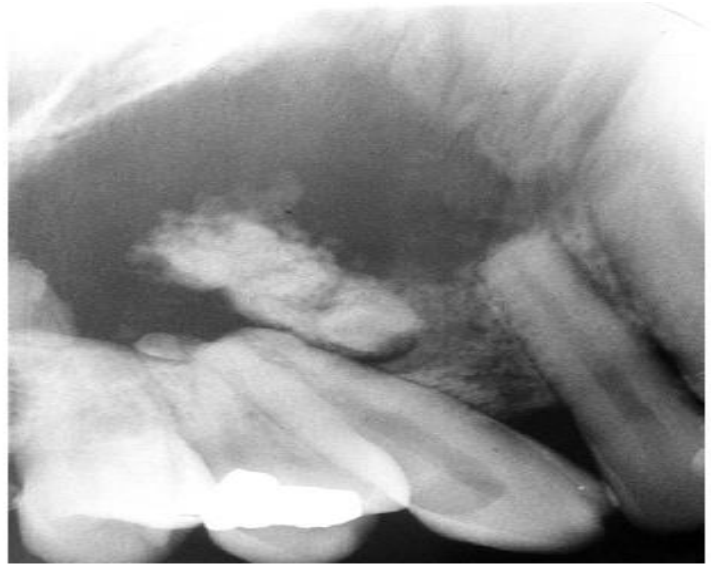
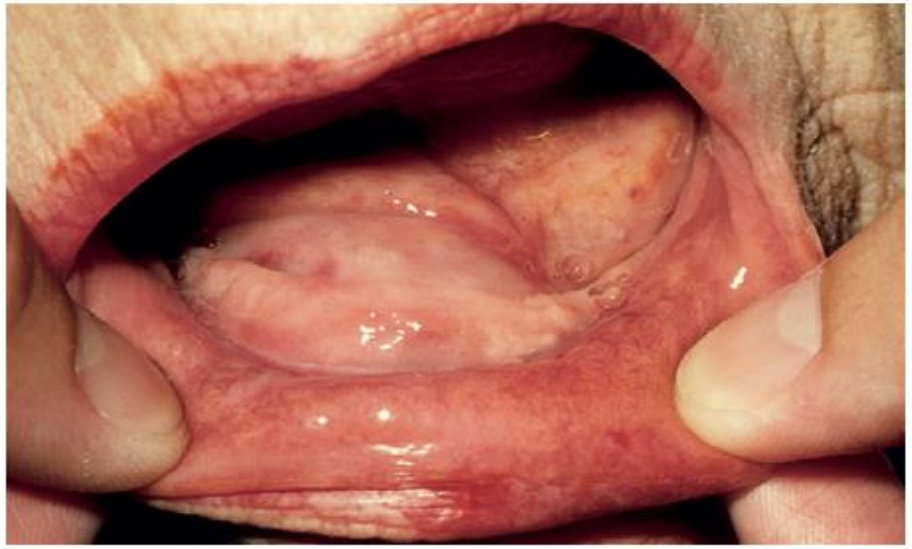


FIGURE 5-21
Calcifying odontogenic cyst. Radiograph of large lesion of maxilla with odontoma included within the lesion.

A



B

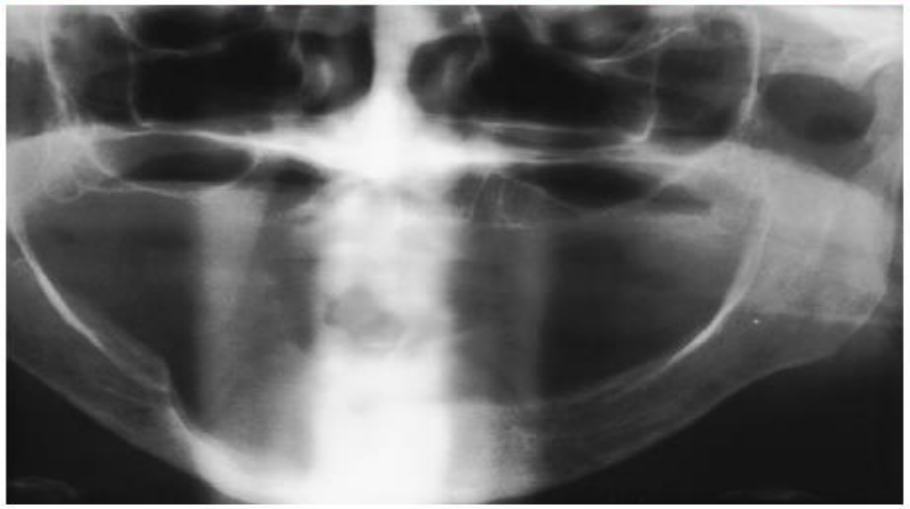


FIGURE 5-20
Calcifying odontogenic cyst. A, Patient exhibits extensive buccal and lingual expansion of the cortical plates of the right mandible. **B**, Radiograph of same patient that demonstrates a large radiolucent cup-shaped defect of the anterior right mandible with sharp line of demarcation with surrounding bone.

MIXED ODONTOGENIC TUMORS

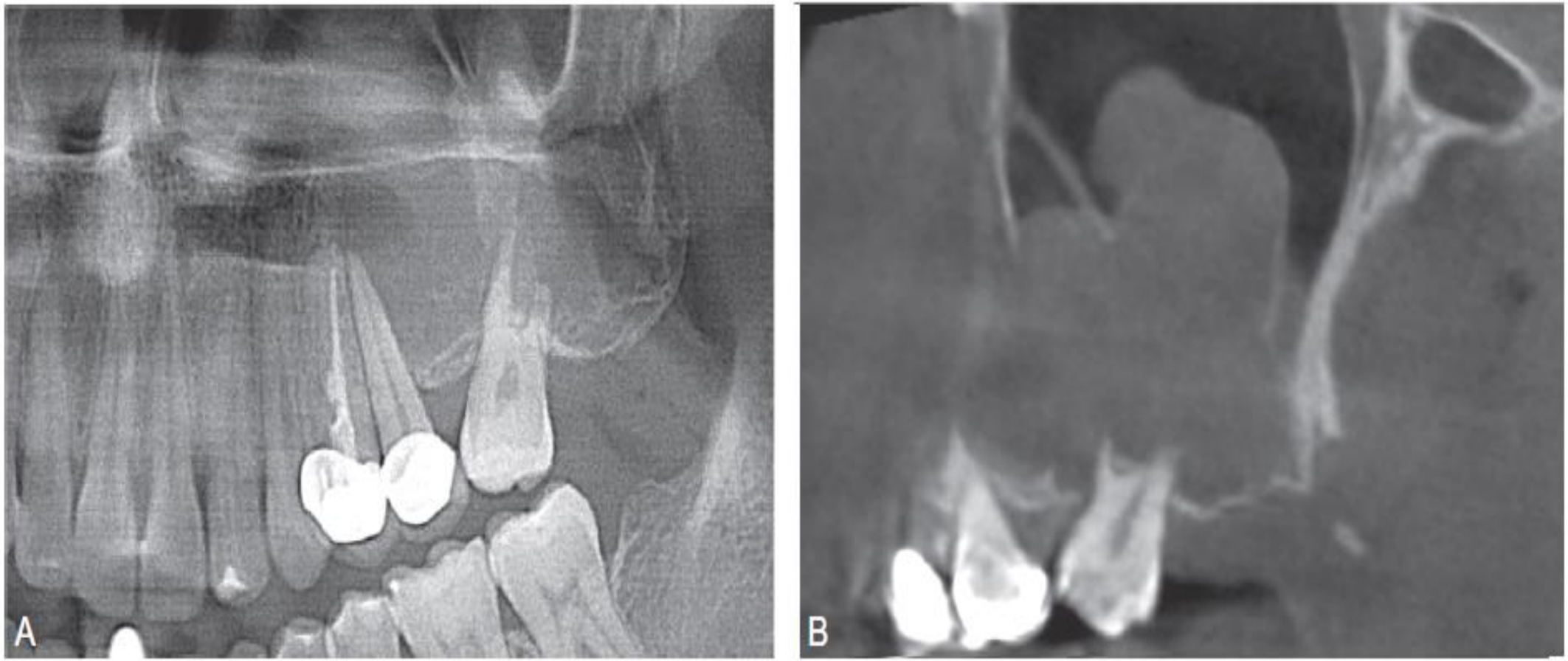
AMELOBLASTIC FIBROMA

The ameloblastic fibroma is considered to be a true mixed tumor in which the epithelial and mesenchymal tissues areboth neoplastic.

Clinical and Radiographic Features

Ameloblastic fibromas tend to occur in younger patients,most lesions are diagnosed in the first two decades of life,with an average age of 15 years. This lesion, however, is occasionally encountered in middle-aged patients. The tumor is slightly more common in males than in females. Small ameloblastic fibromas are asymptomatic; larger tumors are associated with swelling of the jaws. The posterior mandible is the most common site.

Radiographically, either a unilocular or multilocular radiolucent lesion is seen, with the smaller lesions tending to be unilocular. The radiographic margins tend to be well defined, and they may be corticated. An unerupted tooth is associated with the lesion in about 75% of cases.The ameloblastic fibroma may grow to a large size, and cases that involve a considerable portion of the body and ascending ramus of the mandible or the posterior maxilla have been reported.

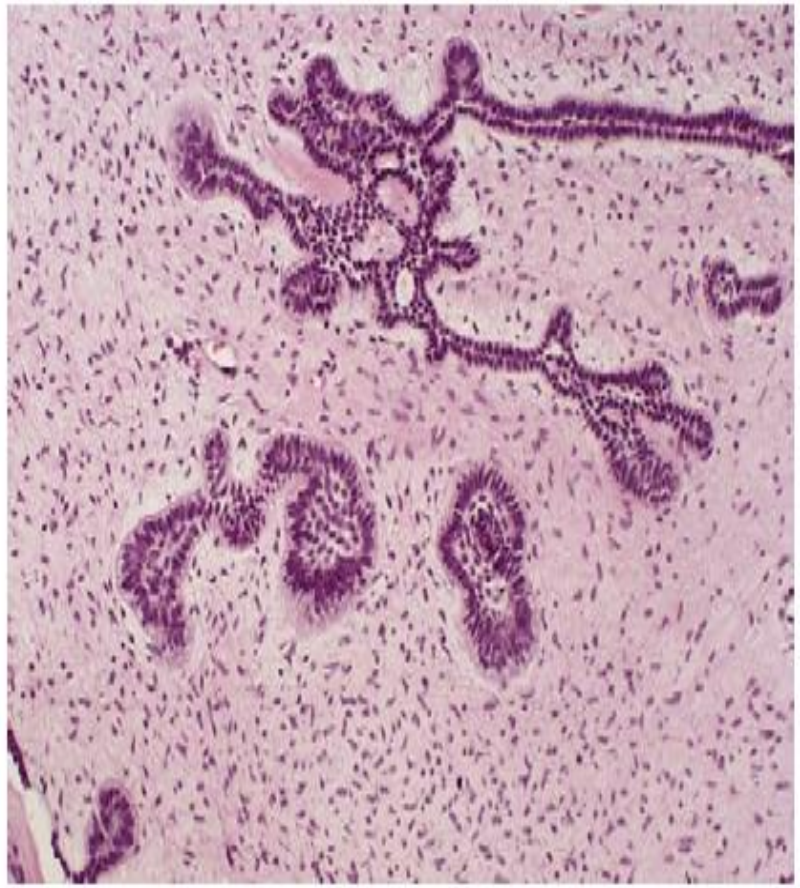


• **Fig. 15.104 Ameloblastic Fibroma. A,** Panoramic radiograph showing a destructive radiolucent lesion of the left posterior maxilla. **B,** Cone-beam computed tomography (CBCT) image demonstrating extension of the tumor into the maxillary sinus. (Courtesy of Dr. Michael Menis.)

Treatment and Prognosis

approximately19%of ameloblastic fibromas were reported to recur after enucleation or marginal resection and an adequate follow-up period. Based on these data, recent recommendations have emphasized conservative initial therapy for ameloblastic fibroma, followed by longterm monitoring.

More aggressive surgical excision should probably be reserved for recurrent lesions. Approximately 26%of the cases of the rare ameloblastic fibrosarcoma develop in the setting of a recurrent ameloblastic fibroma.



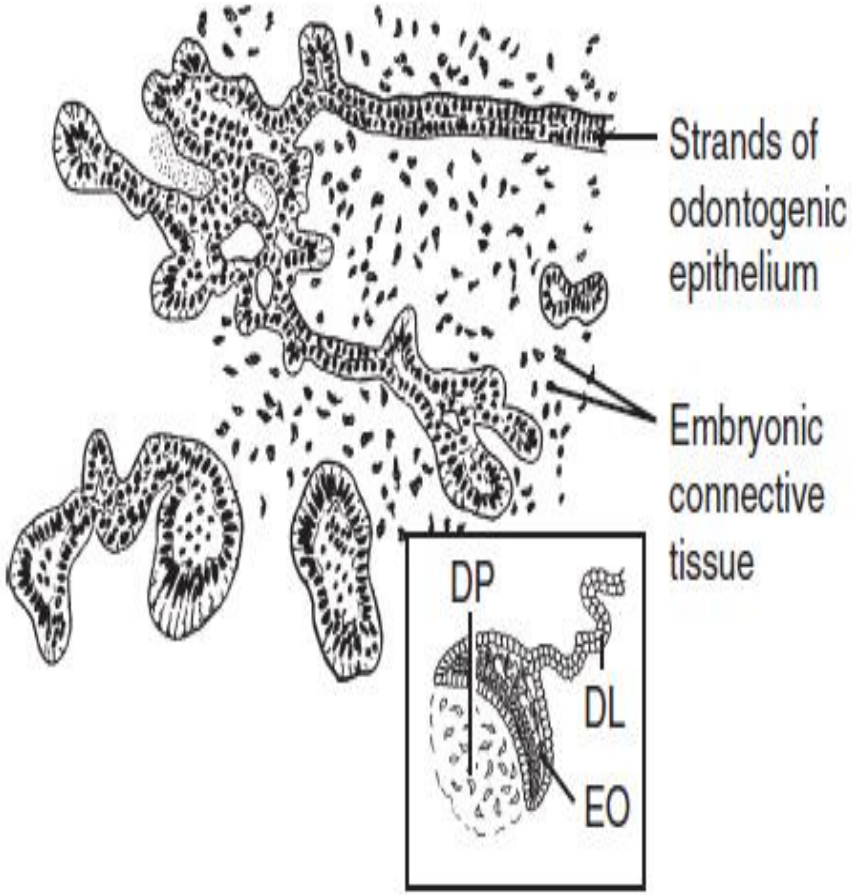


FIGURE 5-34

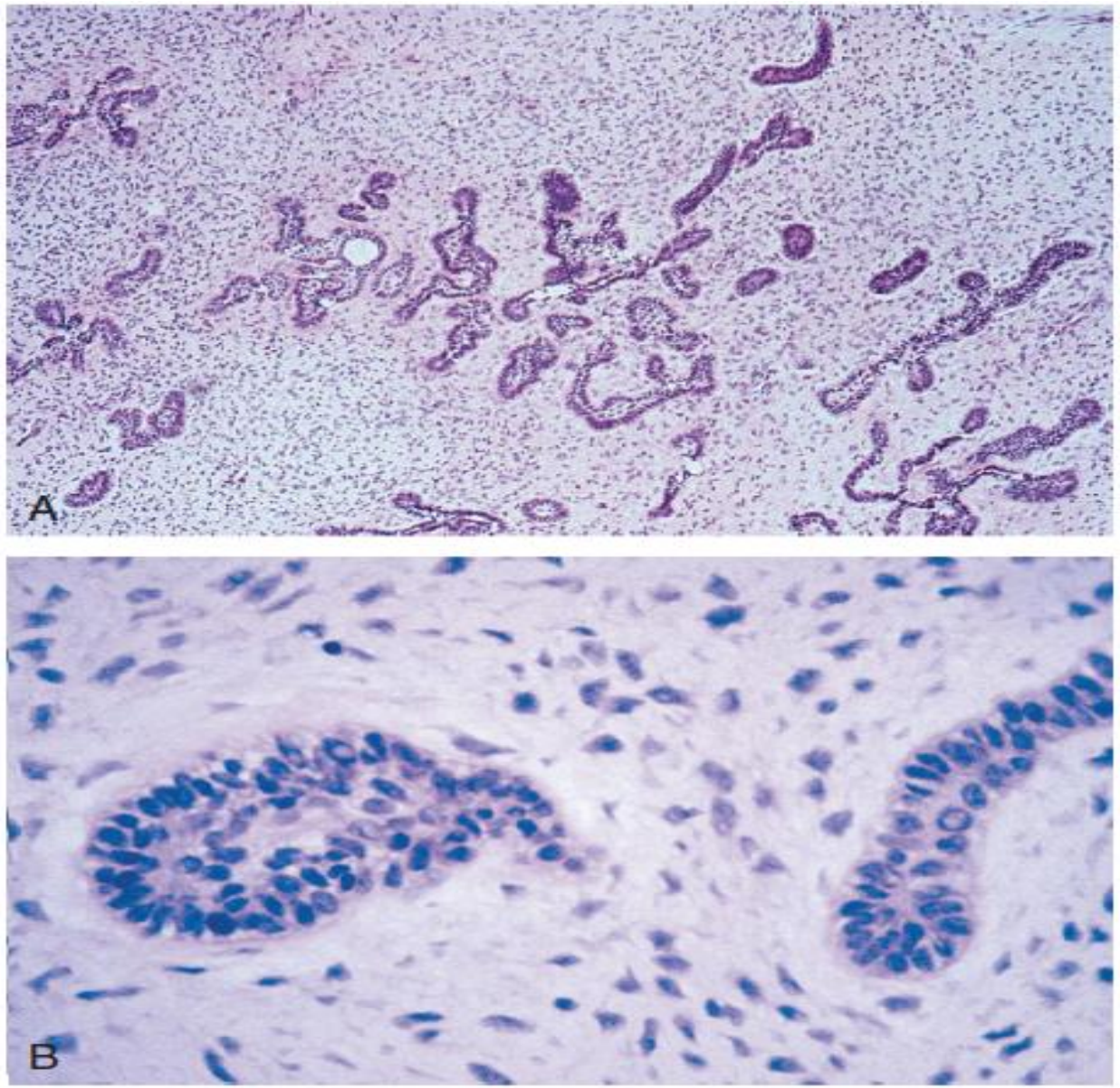
Ameloblastic fibroma. Microscopic features consist of strands and islands of odontogenic epithelium resembling the dental lamina and cap stage of odontogenesis. Tumor recapitulates the invagination of the dental lamina and cap stage of the earliest phases of odontogenesis *(inset)*. *DL*, Dental lamina; *DP*, dental papilla; *EO*, enamel organ.

Histopathologic Features:

The ameloblastic fibroma appears as a solid, soft tissue mass with a smooth outer surface. A definite capsule may or may not be present. Microscopically, the tumor is composed of a cell-rich mesenchymal tissue resembling the primitive dental papilla admixed with proliferating odontogenic epithelium.

The epithelium may have one of two patterns,The most common epithelial pattern consists of long, narrow cords of odontogenic epithelium, often in an anastomosing arrangement. These cords are usually only two cells in thickness and are composed of cuboidal or columnar cells . In the other pattern, the epithelial cells form small, discrete islands that resemble the follicular stage of the developing enamel organ. These show peripheral columnar cells, which surround a mass of loosely arranged epithelial cells that resemble stellate reticulum. In contrast to the follicular type of ameloblastoma, these follicular islands in the ameloblastic fibroma seldom demonstrate microcyst formation.

The mesenchymal portion of the ameloblastic fibroma consists of plump stellate and ovoid cells in a loose matrix, which closely resembles the developing dental papilla. Juxta epithelial hyalinization of the mesenchymal portion of the tumor is sometimes seen, and occasionally diffuse areas of hyalinized acellular lesional tissue are evident.



• **Fig. 15.105 Ameloblastic Fibroma. A,** Long, narrow cords of odontogenic epithelium supported by richly cellular, primitive connective tissue. **B,** Basophilic epithelial islands with peripheral nuclear palisading.

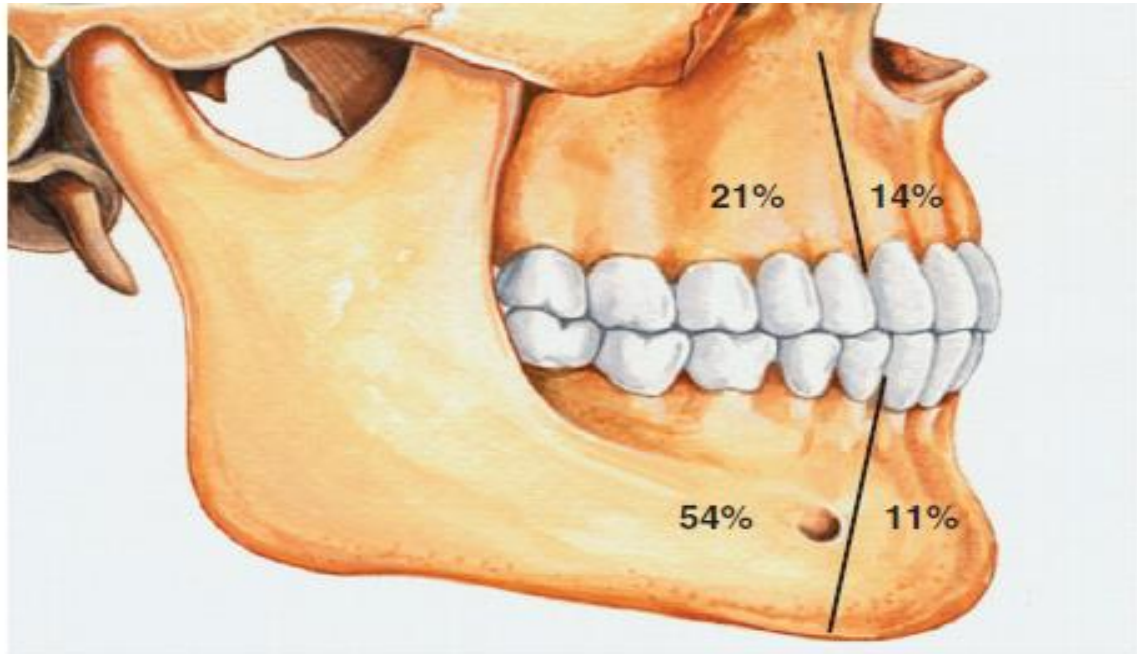
AMELOBLASTIC FIBRO-ODONTOMA

The ameloblastic fibro-odontoma is defined as a tumor with the general features of an ameloblastic fibroma but that also contains enamel and dentin. Some investigators believe that the ameloblastic fibro-odontoma is only a stage in the development of an odontoma and do not consider it to be a separate entity. however, this tumor exhibiting progressive growth and causing considerable deformity and bone destruction.Such lesions appear to be true neoplasms.

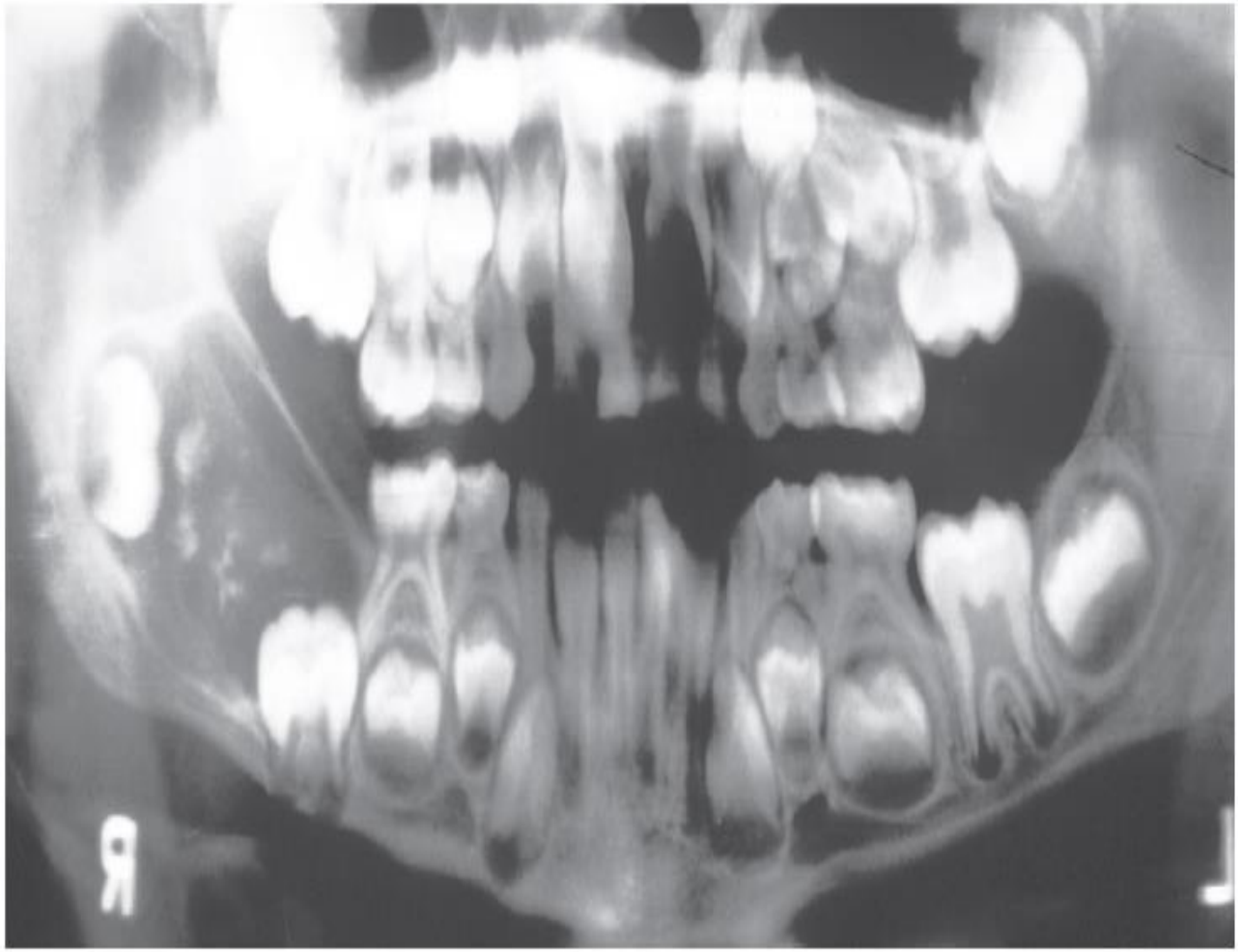
Clinical and Radiographic Features

The ameloblastic fibro-odontoma is usually seen in children with an average age of 10 years. Like the ameloblastic fibroma, ameloblastic fibroodontomas occur more frequently in the posterior regions of the jaws, and the majority involves the mandible. Males are affected somewhat more often than females, with a 3:2 ratio. The lesion is commonly asymptomatic and is discovered when radiographs are taken to determine the reason for failure of a tooth to erupt. Large examples may be associated with a painless swelling of the affected bone.

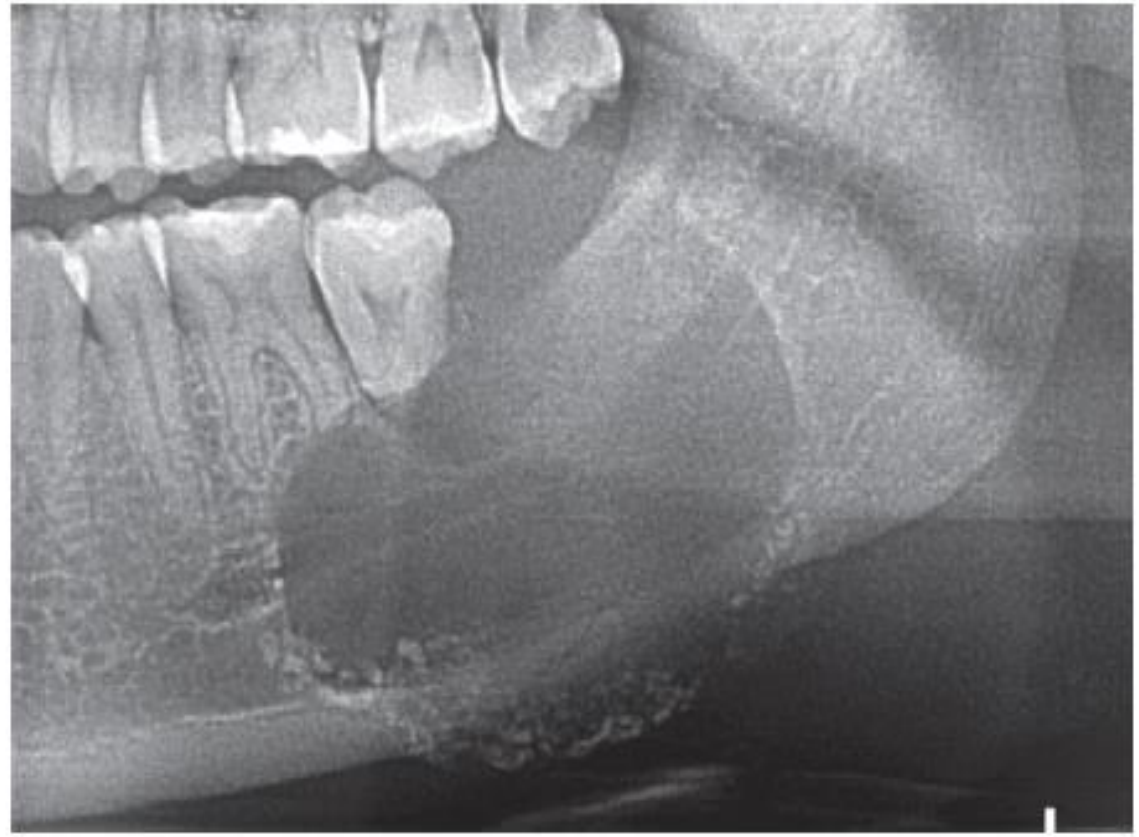
Radiographically, the tumor shows a well-circumscribed unilocular or, infrequently, multilocular radiolucent defect that contains a variable amount of calcified material with the radiodensity of tooth structure. The calcified material within the lesion may appear as multiple, small radiopacities or as a solid conglomerate mass . In most instances, an unerupted tooth is present at the margin of the lesion, or the crown of the unerupted tooth may be included within the defect. Some ameloblastic fibro-odontomas appear as largely calcified masses with only a narrow rim of radiolucency about the periphery of the lesion, which would be a pattern that certainly suggests a maturing odontoma.



• **Fig. 15.106 Ameloblastic Fibro-Odontoma.** Relative distribution of ameloblastic fibro-odontoma in the jaws.



• **Fig. 15.107 Ameloblastic Fibro-Odontoma.** Radiolucent defect in the ramus containing small calcifications having the radiodensity of tooth structure.



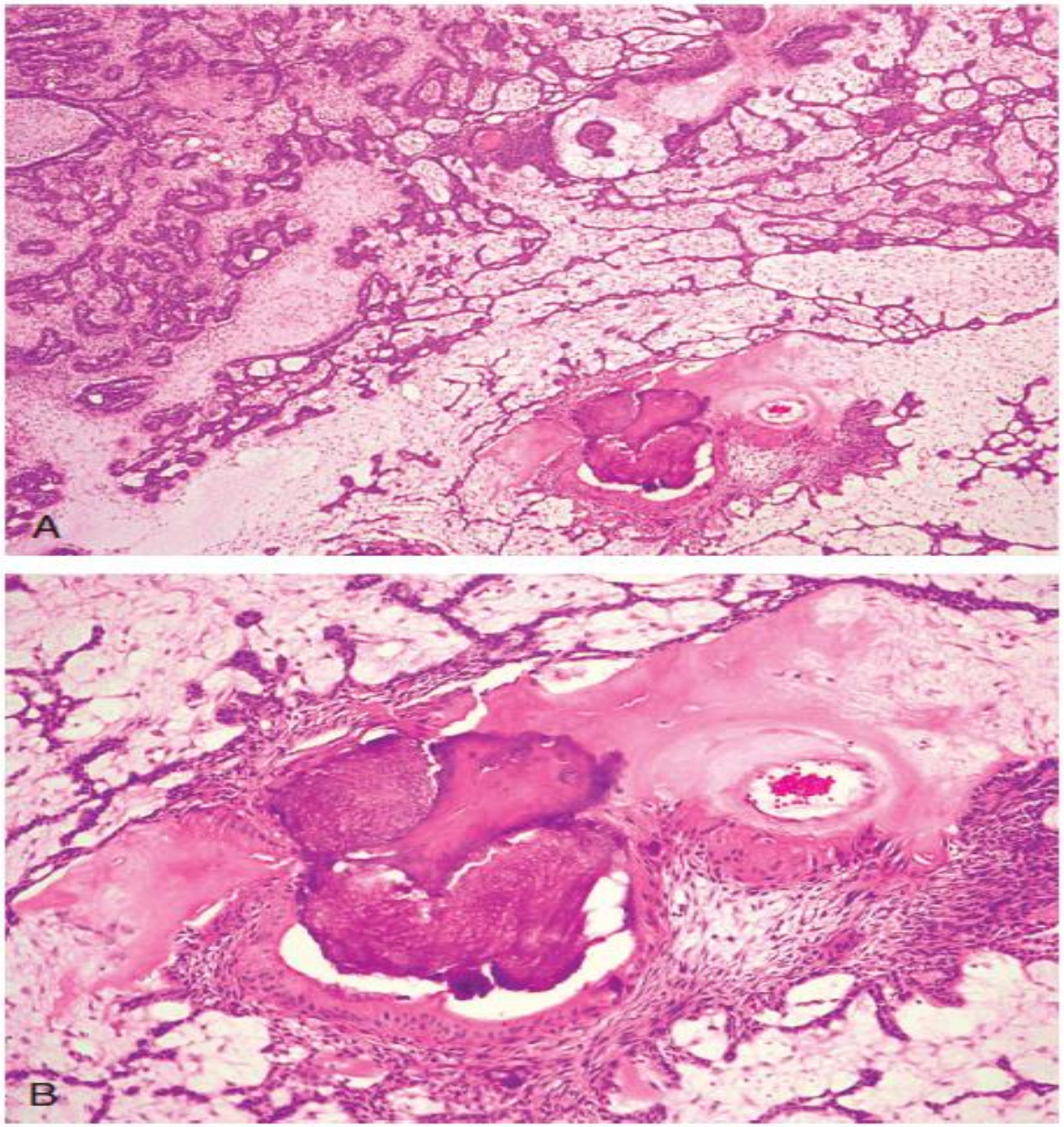
• **Fig. 15.108 Ameloblastic Fibro-Odontoma.** Large, expansile radiolucent defect that is associated with root resorption of the overlying molars. Flecks of mineralized material are present at the periphery. (Courtesy of Dr. Mark Spinazze.)

Histopathologic Features

The soft tissue component of the ameloblastic fibro-odontoma is microscopically identical to the ameloblastic fibroma and has narrow cords and small islands of odontogenic epithelium in a loose primitive-appearing connective tissue that resembles the dental papilla. The calcifying element consists of foci of enamel and dentin matrix formation in close relationship to the epithelial structures . The more calcified lesions show mature dental structures in the form of rudimentary small teeth or conglomerate masses of enamel and dentin. Some researchers have designated a similar tumor in which the calcifying component consists only of dentin matrix and dentinoid material as ameloblastic fibro-dentinoma.

Treatment and Prognosis

ameloblastic fibro-odontoma is generally treated by conservative curettage, and the lesion usually separates easily from its bony bed. The tumor is well circumscribed and does not invade the surrounding bone. The prognosis is excellent. Development of an ameloblastic fibrosarcoma after curettage has been reported, but this is exceedingly rare.



• **Fig. 15.109 Ameloblastic Fibro-Odontoma. A,** The soft tissue component of the tumor is indistinguishable from an ameloblastic fibroma. **B,** Formation of disorganized tooth structure can be seen.

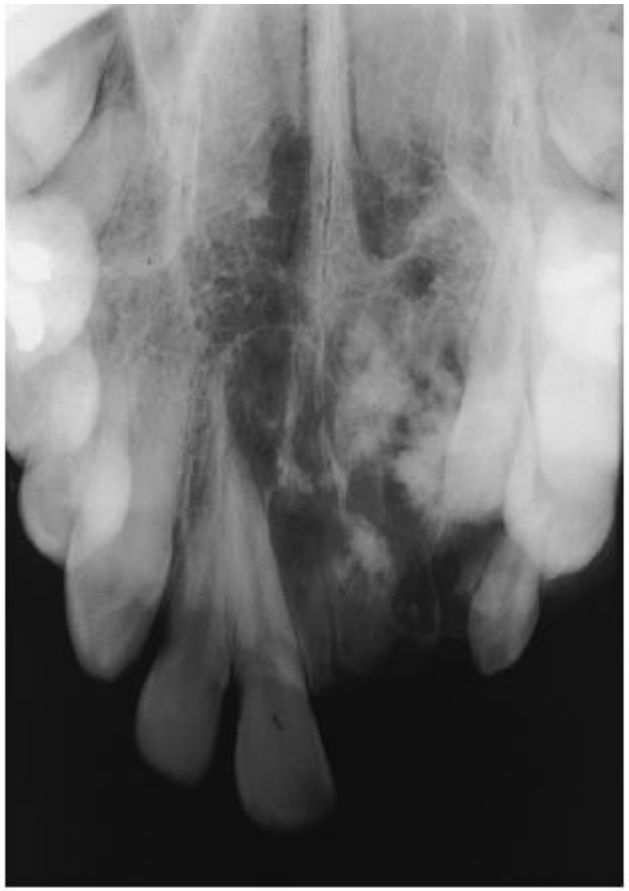


FIGURE 5-39

Ameloblastic fibro-odontoma. Radiograph of mixed radiolucent and radiopaque lesion of anterior maxilla preventing the eruption of the central incisor.

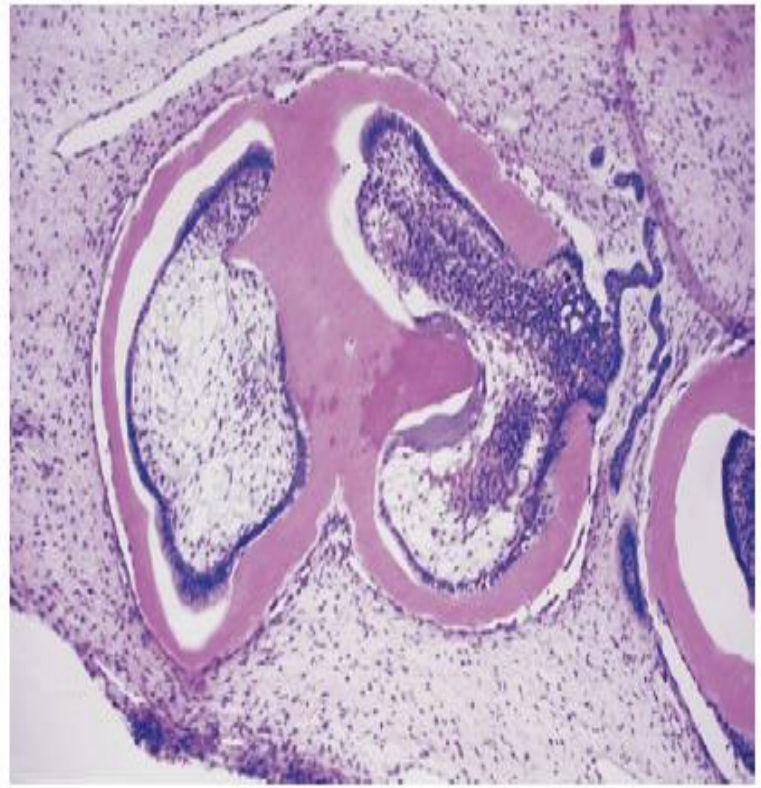


FIGURE 5-40

Ameloblastic fibro-odontoma. Photomicrograph of the irregularly shaped dentin deposits, enamel matrix, and odontogenic epithelium against a background of fibrillar connective tissue containing randomly oriented spindled cells. Tumor recapitulates cells from all phases of tooth development, including enamel and dentin production as found in the late bell stage of odontogenesis *(inset)*. *D,* Dentin; *DP,* dental papilla; *E,* enamel.

ODONTOMA

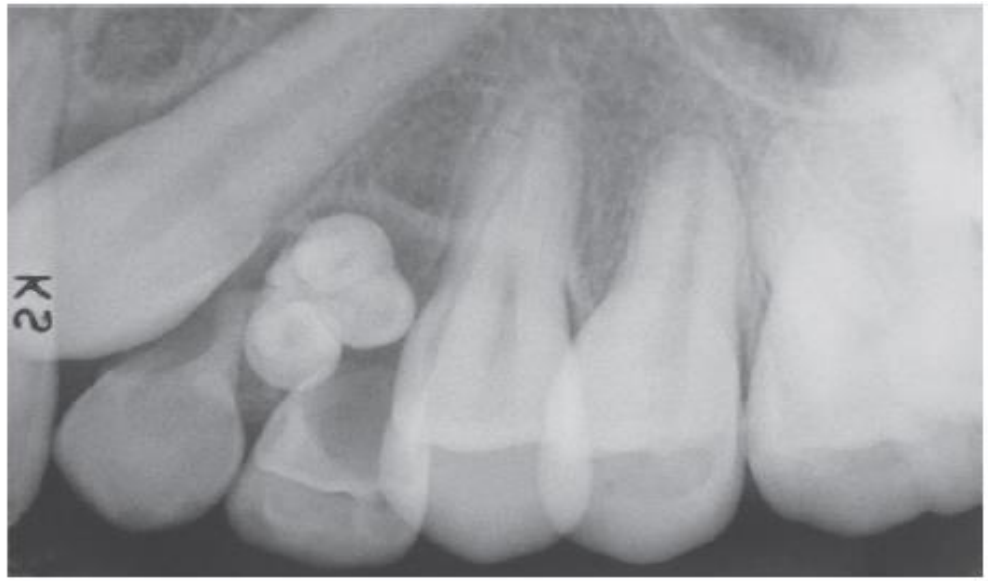
Odontomas are the most common types ofodontogenic tumors. Their prevalence exceeds that of all other odontogenictumors combined. Odontomas are considered to be developmental anomalies (hamartomas), rather than true neoplasms. When fully developed, odontomas consist chiefly of enamel and dentin, with variable amounts of pulp and cementum. Odontomas are further subdivided into compound and complex types. The compound odontoma is composed of multiple, small toothlike structures. The complex odontoma consists of a conglomerate mass of enamel and dentin, which bears no anatomic resemblance to a tooth. In most series, compound odontomas are more frequently diagnosed than complex, and it is possible that some compound odontomas are not submitted for microscopic examination because the clinician is comfortable with the clinical and radiographic diagnosis.

Clinical and Radiographic Features

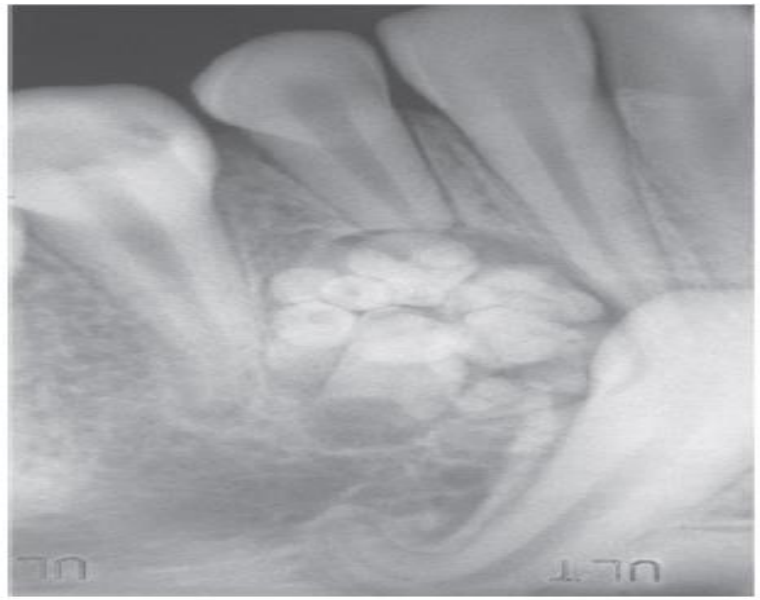
Most odontomas are detected during the first two decades of life, and the mean age at the time of diagnosis is 14 years. The majority of these lesions are completely asymptomatic, being discovered on a routine radiographic examination or when films are taken to determine the reason for failure of a tooth to erupt. Odontomas are typically relatively small ,However, large odontomas up to 6 cm or more in diameter are occasionally seen. These large odontomas can cause expansion of the jaw. Odontomas occur more frequently in the maxilla than in the mandible. Although compound and complex odontomas may be found in any site, the compound type is more often seen in the anterior maxilla; complex odontomas occur more often in the molar regions of either jaw.

Radiographically,

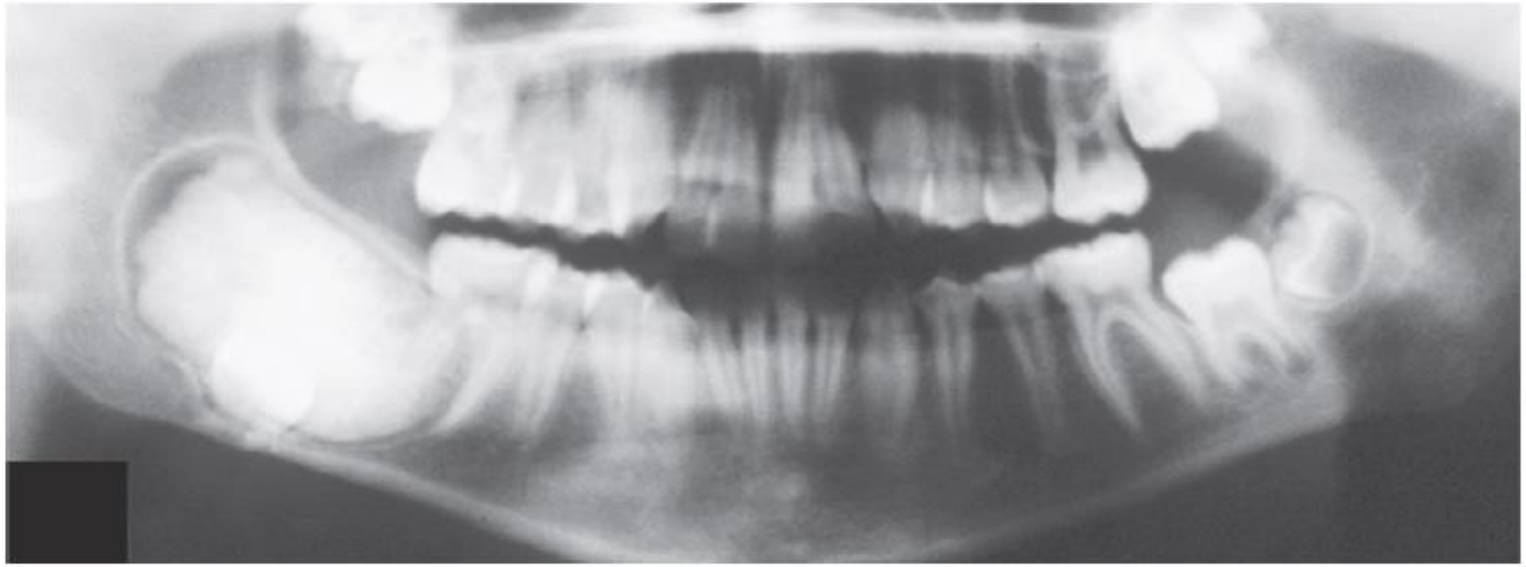
the compound odontoma appears as a collection of toothlike structures of varying size and shape surrounded by a narrow radiolucent zone . The complex odontoma appears as a calcified mass with the radiodensity of tooth structure, which is also surrounded by a narrow radiolucent rim. An unerupted tooth is frequently associated with the odontoma, and the odontoma prevents eruption of the tooth .Some small odontomas are present between the roots of erupted teeth and are not associated with disturbance in eruption. The radiographic findings are usually diagnostic.



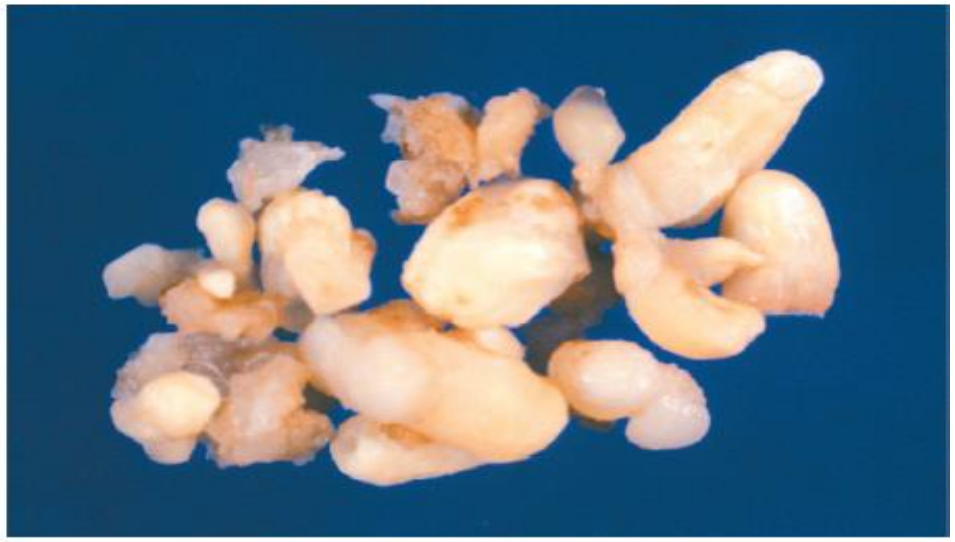
• **Fig. 15.112 Compound Odontoma.** A small cluster of toothlike structures is preventing the eruption of the maxillary canine. (Courtesy of Dr. Robert J. Powers.)



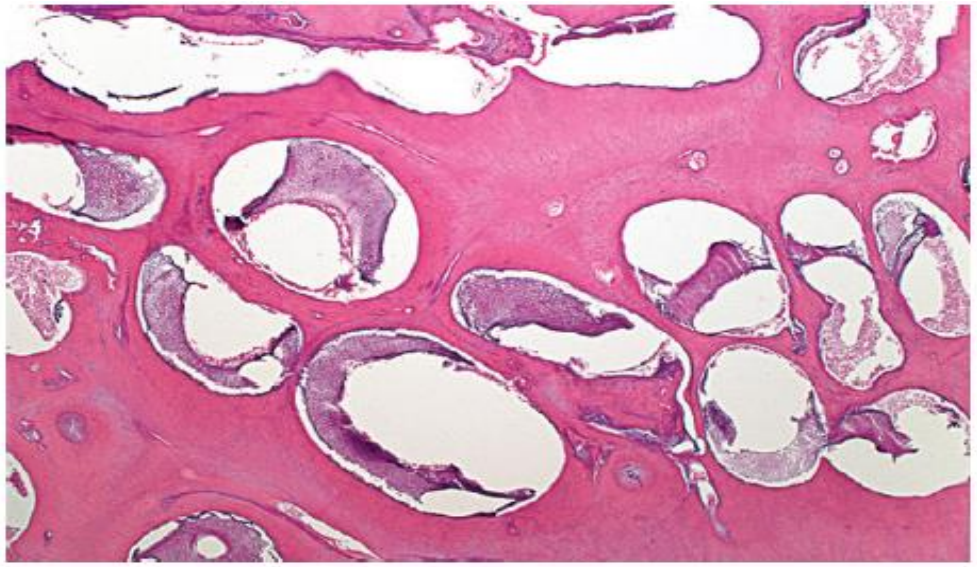
• **Fig. 15.113 Compound Odontoma.** Multiple toothlets preventing the eruption of the mandibular cuspid. (Courtesy of Dr. Brent Bernard.)



• **Fig. 15.114 Complex Odontoma.** A large radiopaque mass is overlying the crown of the mandibular right second molar, which has been displaced to the inferior border of the mandible.



• **Fig. 15.115 Compound Odontoma.** Surgical specimen consisting of more than 20 malformed toothlike structures.



• **Fig. 15.116 Complex Odontoma.** This decalcified section shows a disorganized mass of dentin intermixed with small pools of enamel matrix.

Histopathologic Features

The compound odontoma consists of multiple structures resembling small, single-rooted teeth, contained in a loose fibrous matrix . The mature enamel caps of the toothlike structures are lost during decalcification for preparation of the microscopic section, but varying amounts of enamel matrix are often present. Pulp tissue may be seen in the coronal and root portions of the toothlike structures.

Complex odontomas consist largely of mature tubular dentin. This dentin encloses clefts or hollow circular structures that contained the mature enamel that was removed during decalcification. The spaces may contain small amounts of enamel matrix or immature enamel. Small islands of eosinophilic-staining epithelial ghost cells are present in about 20% of complex odontomas. These may represent remnants of odontogenic epithelium that have undergone keratinization and cell death from the local anoxia. A thin layer of cementum is often present about the periphery of the mass.

Treatment and Prognosis

Odontomas are treated by simple local excision, and the prognosis is excellent.

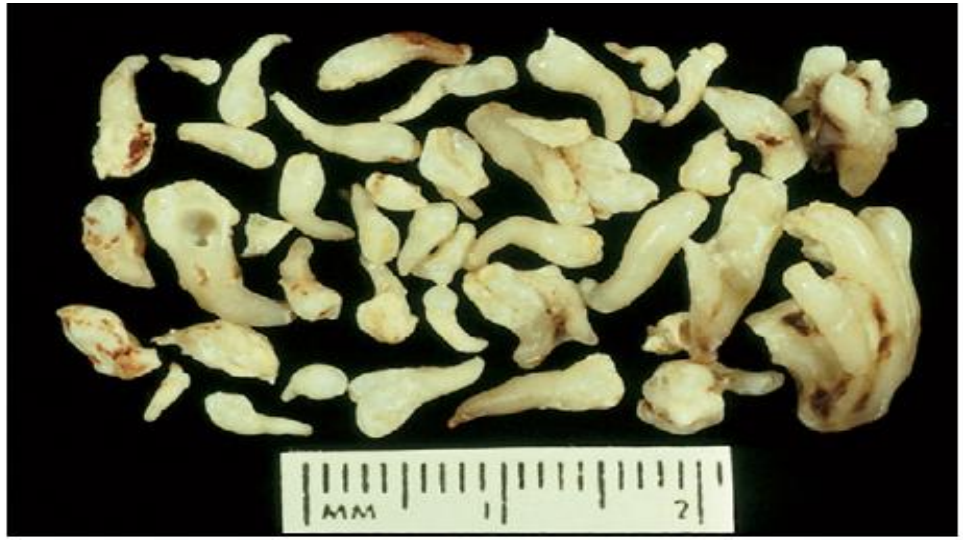


FIGURE 5-36
Compound odontoma. Multiple conical and irregularly shaped miniature teeth removed from within the capsule of a large lesion.

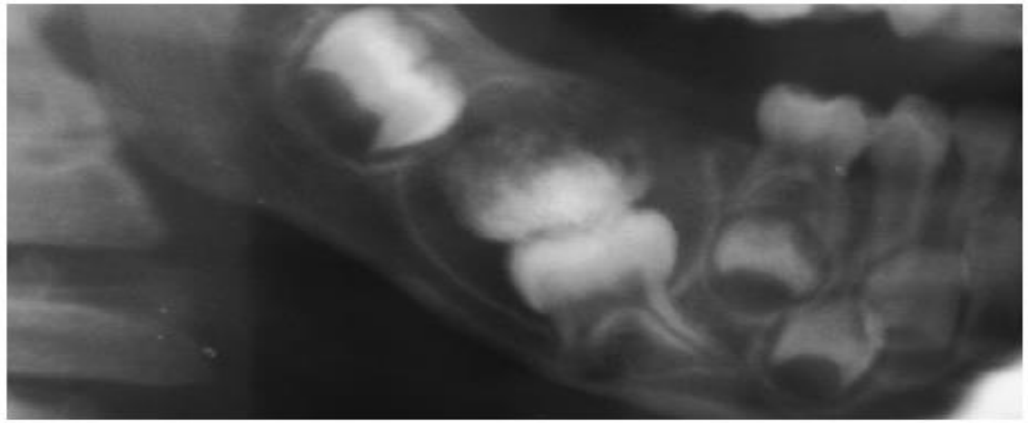


FIGURE 5-37
Complex odontoma. Radiograph of posterior mandible containing a dense structureless radiopacity preventing the eruption of an impacted molar in a young patient.

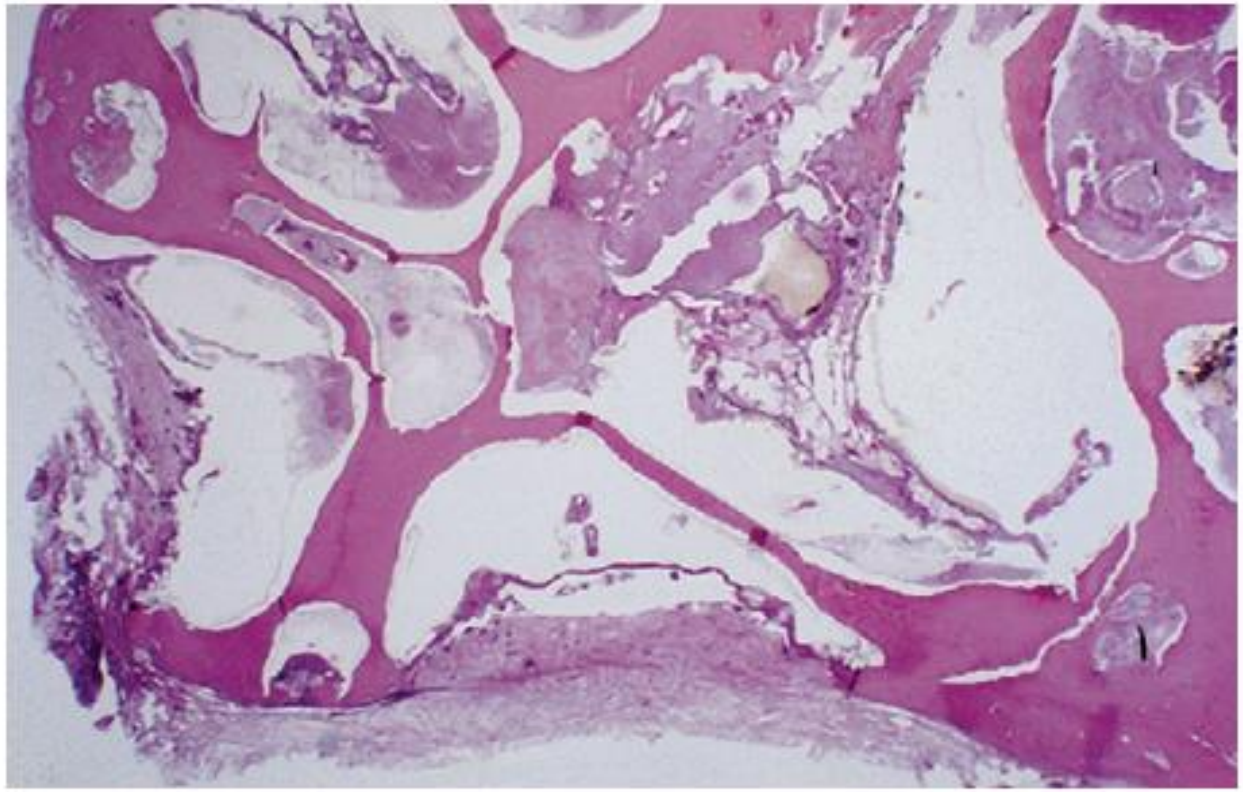
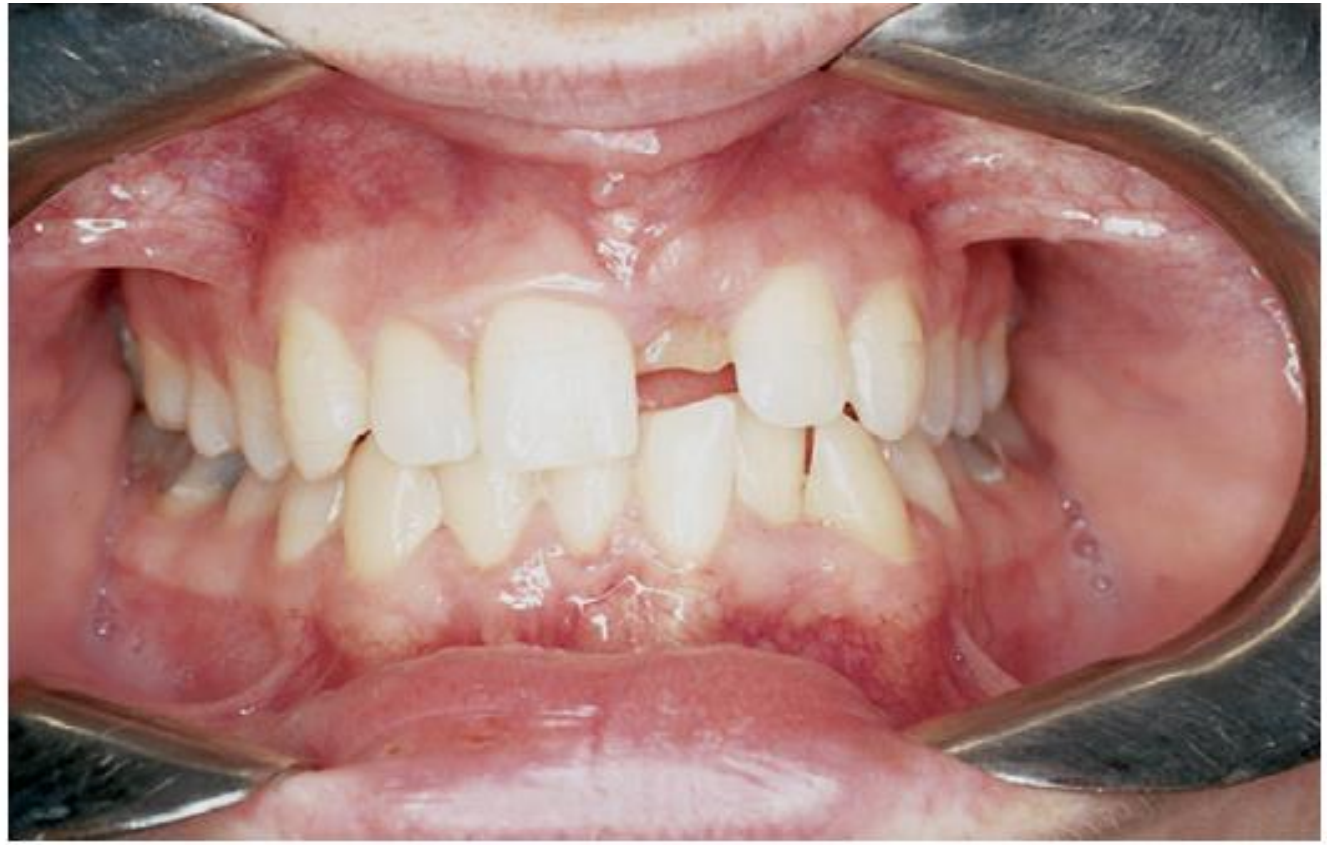
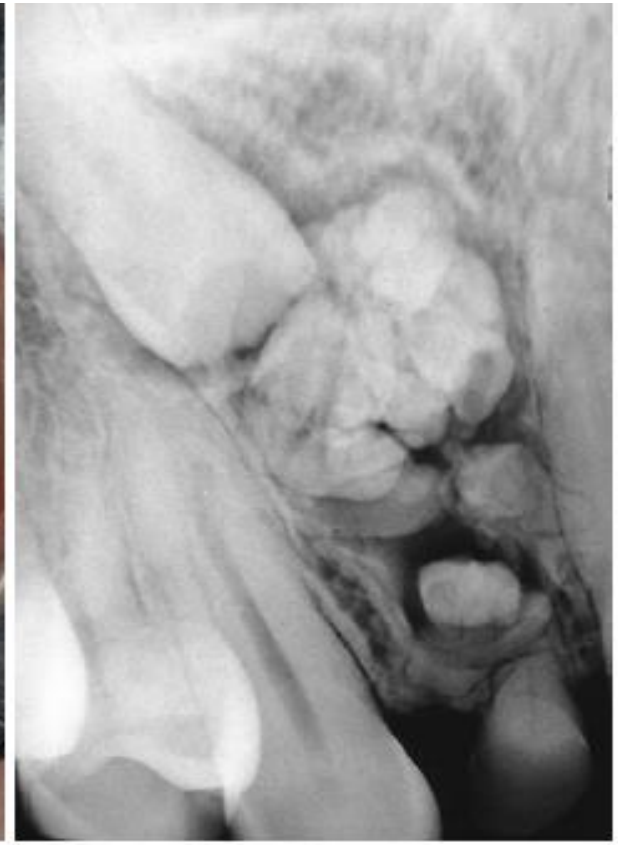


FIGURE 5-38
Complex odontoma. Microscopic appearance reveals irregular spaces containing residual elements of enamel matrix surrounded by septae of dentin interspersed with occasional areas of pulpal tissue. A fibrous connective tissue capsule is present in the periphery.

A





B

FIGURE 5-35

Odontoma. A, Common clinical finding in patients with an odontoma is retention of a deciduous tooth and failure of the permanent tooth to erupt. **B,** Radiograph of same patient revealing the presence of multiple individual toothlike structures in a well-demarcated bone cavity impeding the eruption of the permanent central incisor.