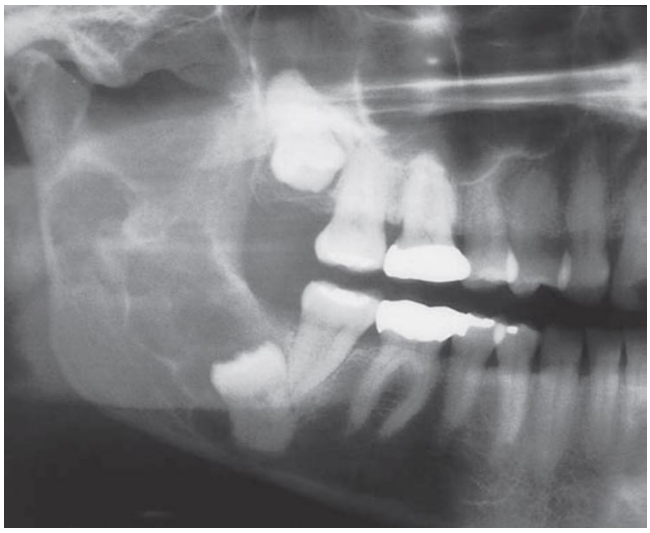
Odontogenic tumors

2. Calcifying Epithelial Odontogenic Tumor (Pindborg Tumor)

Calcifying epithelial odontogenic tumor (CEOT), also known as **Pindborg tumor**, is a benign tumor of odontogenic origin that shares many clinical features with ameloblastoma. The cells from which these tumors are derived are unknown, although dental lamina remnants and the stratum intermedium of the enamel organ have been suggested.

The lesions may be unilocular or multilocular. Small loculations in some lesions have prompted use of the term honeycomb to describe this lucent pattern. A CEOT may be completely radiolucent, or it may contain opaque foci, a reflection of the calcified amyloid seen microscopically. The lesions are usually well circumscribed radiographically.

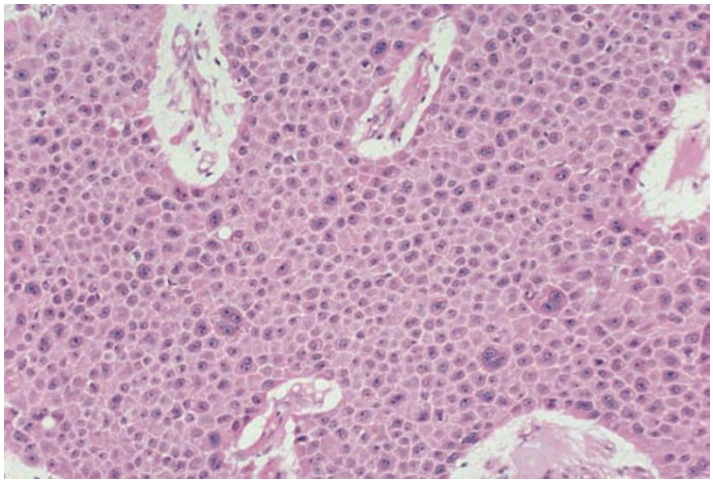


Histopathology

The CEOT has a unique and sometimes strange microscopic pattern. Large polygonal epithelial cells, arranged in sheets or islands, contain nuclei that show considerable variation in size and shape. Mitotic figures are rare. The cytoplasm is abundant and eosinophilic. Focal zones of clear cells occasionally can be seen in a so-called clear cell variant. Extracellular amyloid of epithelial origin is also typical of these tumors. This homogeneous, pale-staining eosinophilic material can be stained with Congo red or thioflavine T. Immunohistochemical staining for cytokeratins is also positive, suggesting that keratin proteins form an important component of the amyloid in this tumor. Concentric

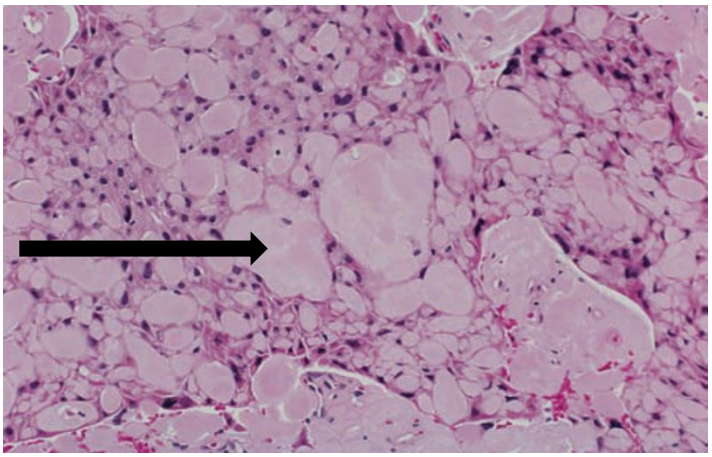
calcific deposits with a characteristic annular staining pattern (Liesegang rings), seen in the amyloid material, are responsible for radiopacities when sufficiently dense. The incidental finding of Langerhans cells in CEOTs has been reported, but their significance in this setting is undetermined.

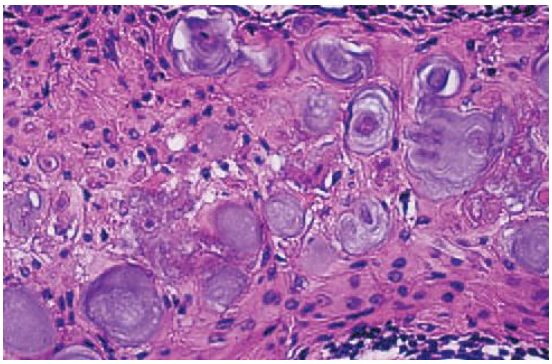
Calcifying epithelial odontogenic tumor composed of a sheet of atypical and multinucleated tumor epithelial cells.



Calcifying epithelial odontogenic tumor showing

amyloid deposits.





Calcifying epithelial odontogenic tumor. Multiple concentric Liesegang ring calcifications.

Treatment

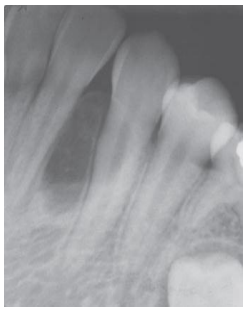
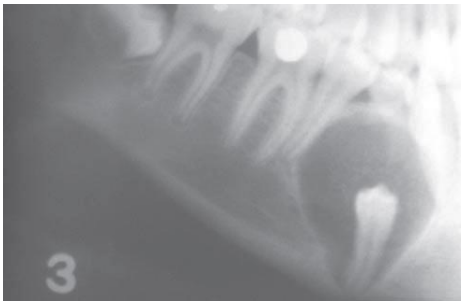
This tumor has a locally infiltrative potential but apparently not to the same extent as ameloblastoma. It is slow growing and causes morbidity through direct tumor extension. Various forms of surgery, ranging from enucleation to resection, have been used to treat CEOTs. The overall recurrence rate has been less than 20%, indicating that aggressive surgery is not indicated for the management of most of these benign neoplasms.

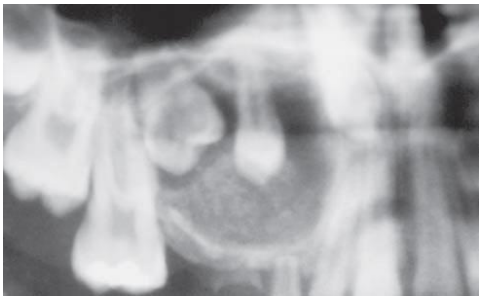
3. Adenomatoid Odontogenic Tumor

Adenomatoid odontogenic tumor (AOT) was formerly termed adeno-ameloblastoma because it was believed to be a subtype of ameloblastoma that contains ductlike or gland-like structures. Clinically, microscopically, and behaviorally, it is clearly different from ameloblastoma, and the term adeno-ameloblastoma is not used.

Clinical features:

Adenomatoid odontogenic tumors are largely limited to younger patients, and two thirds of all cases are diagnosed when patients are 10 to 19 years of age. Most adenomatoid odontogenic tumors are relatively small. They seldom exceed 3 cm in greatest diameter, Peripheral (extraosseous) forms of the tumor are also encountered but are rare. These usually 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. In about 75% of cases, the tumor appears as a

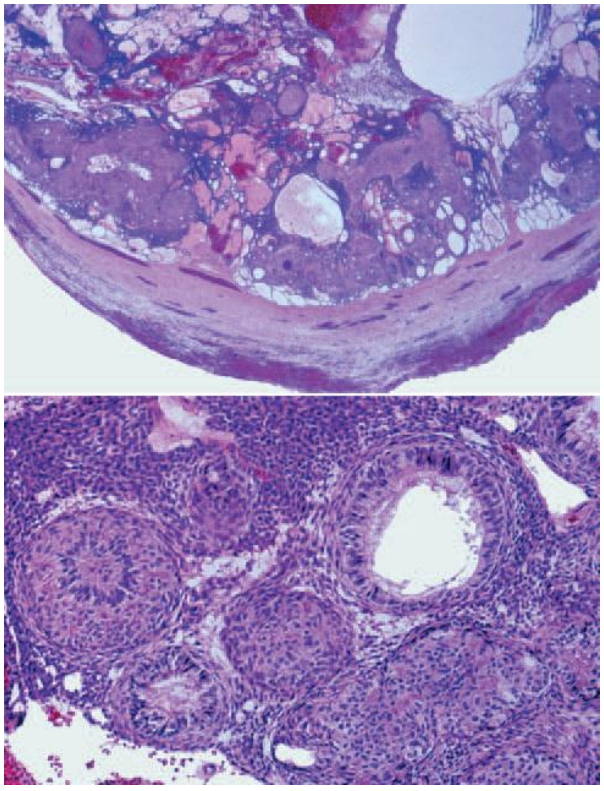


circumscribed, unilocular radiolucency that involves the crown of an unerupted tooth, most often a canine. This follicular type of adenomatoid odontogenic tumor may be impossible to differentiate radiographically from the more common dentigerous cyst. The lesion may appear completely radiolucent; often, however, it contains fi ne (snow flake) calcifications. This feature may be helpful in differentiating the adenomatoid odontogenic tumor from a dentigerous cyst.

Histopathological features:

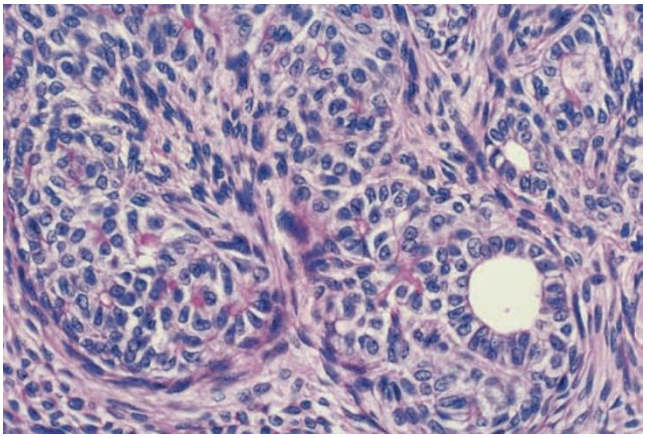
The adenomatoid odontogenic tumor is a well-defined lesion surrounded by a thick, fibrous capsule. When the lesion is bisected, the central portion of the tumor may be essentially solid or may show varying degrees of cystic change. Microscopically, the tumor is composed of spindle shaped epithelial cells that form sheets, strands, or whorled masses of cells in a scant fibrous stroma. The epithelial cells may form rosette-like 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. These consist of a central space surrounded by a layer of columnar or cuboidal epithelial cells. The nuclei of these cells tend to be polarized away from the central space. The mechanism of formation of these tubular structures is not entirely clear but is likely the result of the secretory activity of the tumor cells, which appear to be pre-ameloblasts. In any event, 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. These have been interpreted as abortive enamel formation. Some adenomatoid odontogenic tumors contain larger areas of matrix material or calcification. This material has been interpreted as dentinoid or cementum. Some lesions also have another pattern, particularly at the periphery of the tumor adjacent to the capsule. This consists of narrow, often anastomosing cords of epithelium in an eosinophilic, loosely

arranged matrix. some adenomatoid odontogenic tumors have been described with focal areas that resemble calcifying epithelial odontogenic tumor, odontoma, or calcifying odontogenic cyst.



(Adenomatoid odontogenic tumor. Low power view demonstrating a thick capsule surrounding the tumor. The higher magnification showing the ductlike epithelial structures. The nuclei of the columnar calls are polarized away from the central spaces)

Adenomatoid odontogenic tumor exhibiting pseudoducts and rosettes.



Treatment: by enucleation; no recurrences

4. Malignant ameloblastoma and ameloblastic carcinoma

Ameloblastoma rarely exhibits frank malignant behavior with development of metastases. The term malignant ameloblastoma should be used for a tumor that shows the histopathologic features of ameloblastoma, both in the primary tumor and in the metastatic deposits. The term ameloblastic carcinoma should be reserved for an ameloblastoma that has cytologic features of malignancy in the primary tumor, in a recurrence, or in any metastatic deposit. These lesions may follow a markedly aggressive local course, but metastases do not necessarily occur.

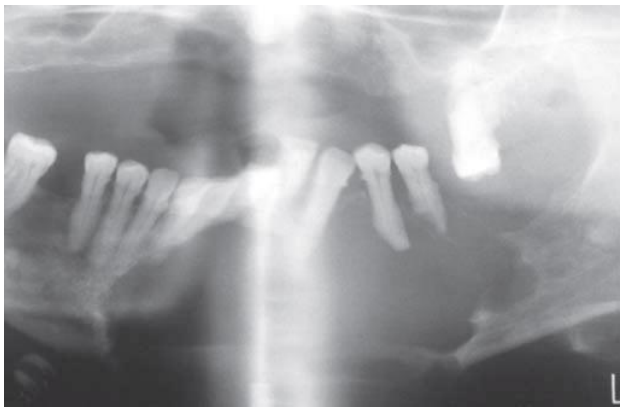
Clinical features

Malignant ameloblastomas have been observed in patients who range in age from 4 to 75 years (mean age, 30 years). in contrast, tend to develop later in life, with the mean age at diagnosis typically being in the sixth decade of life. Metastases from ameloblastomas are most often found in the lungs. Cervical lymph nodes are the second most common site for metastasis of an ameloblastoma. Spread to vertebrae, other bones, and viscera has also occasionally been confirmed. 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.

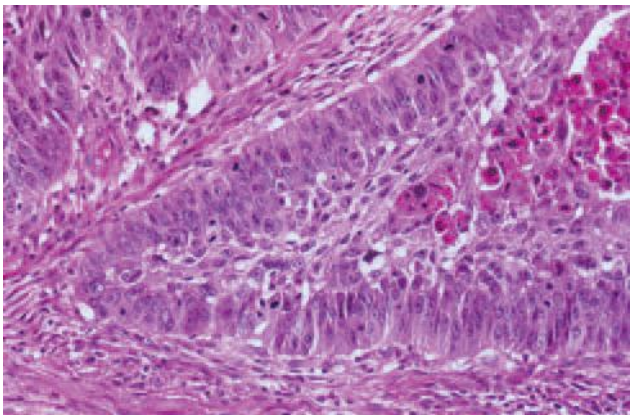
Histopathological features

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. These include an increased nuclear-to-cytoplasmic ratio, nuclear

hyperchromatism, and the presence of mitoses, Necrosis and areas of dystrophic calcification.



panoramic radiograph shows irregular destruction of the mandible.



Ameloblastic carcinoma. Ameloblastic epithelium demonstrating hyperchromatism, pleomorphism, and numerous mitotic figures

Treatment and prognosis

The prognosis of patients with malignant ameloblastomas appears to be poor. About 50% of the patients with documented metastases and long-term follow-up have died of their disease. Lesions designated as ameloblastic carcinoma have demonstrated a uniformly aggressive clinical course, with perforation of the cortical plates of the jaw and extension of the tumor into adjacent soft tissues.

Mixed odontogenic tumors

1.Ameloblastic fibroma

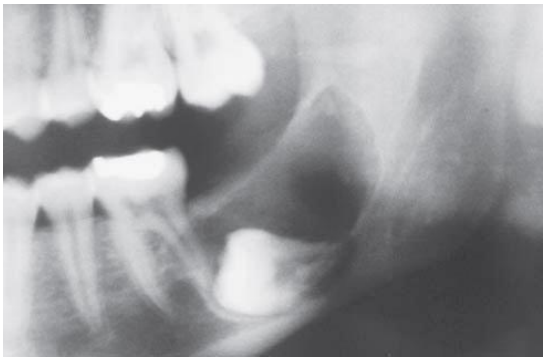
ameloblastic fibroma is considered to be a true mixed tumor in which the epithelial and mesenchymal tissues are both neoplastic.

Clinical features

Ameloblastic fibromas tend to occur in younger patients Small ameloblastic fibromas are asymptomatic; larger tumors are associated with swelling of the jaws. The posterior mandible is the most common site;

Radiographical features

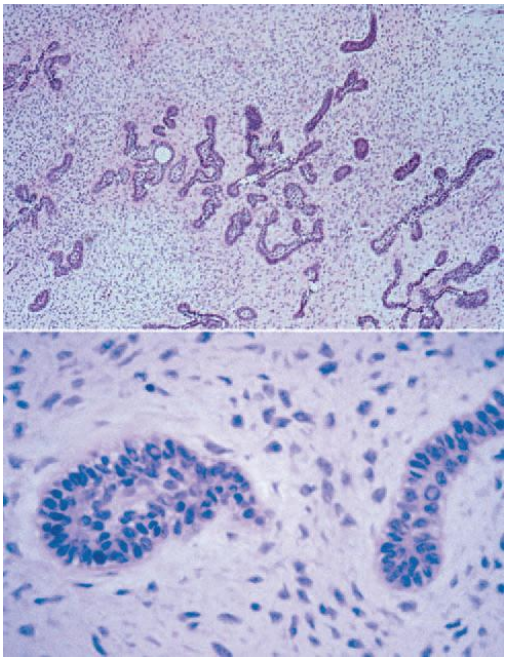
Either a unilocular or multilocular radiolucent lesion



Histopathological features

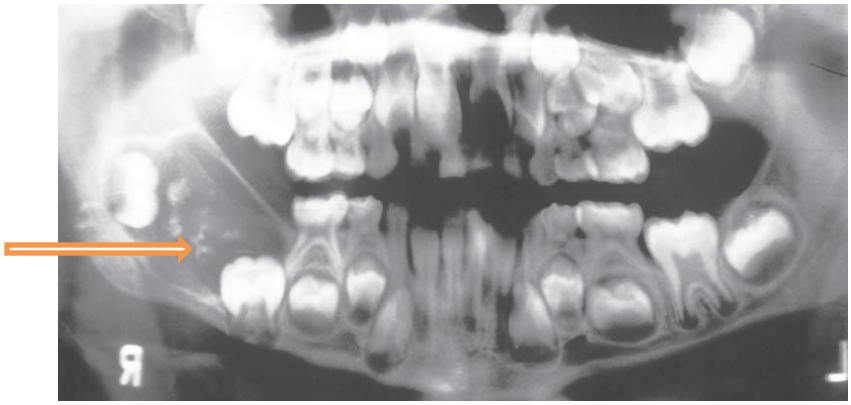
The tumor is composed of a cell-rich mesenchymal tissue resembling the primitive dental papilla admixed with proliferating odontogenic epithelium. The proliferating odontogenic epithelium may have one of two patterns present together. The most common epithelial pattern consists of long, narrow cords 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 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.



2.Ameloblastic fibro-odontoma

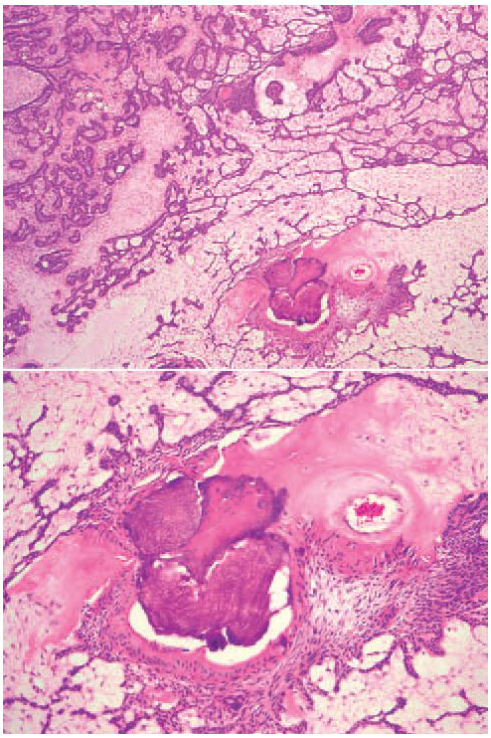
The ameloblastic fibro-odontoma is defined as a tumor with the general features of an ameloblastic fibroma but that also contains enamel and dentin. The ameloblastic fibro-odontoma is usually encountered in children with an average age of 10 years. It is rarely encountered in adults. Like the ameloblastic fibroma, ameloblastic fibro-odontomas occur more frequently in the posterior regions of the jaws. Radiographically, the tumor shows a well-circumscribed unilocular or, rarely, multilocular radiolucent defect that contains a variable amount of calcified material with the radiodensity of tooth structure.



Histopathological 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.

Ameloblastic fi bro-odontoma: The soft tissue component of the tumor is indistinguishable from an ameloblastic fibroma. With formation of disorganized tooth structure.



3.Odontoameloblastoma

The odontoameloblastoma is an extremely rare odontogenic tumor that contains an ameloblastomatous component and odontoma-like elements. Odontoameloblastoma appears to occur more often in younger patients, and either jaw can be affected. Pain, delayed eruption of teeth, and expansion of the affected bone may be noted. Radiographically, the tumor shows a radiolucent, destructive process that contains calcified structures.

Histopathological features

The histopathologic features of the odontoameloblastoma are complex. The proliferating epithelial portion of the tumor has features of an ameloblastoma, most often of the plexiform or follicular pattern. The ameloblastic component is intermingled with immature or more mature dental tissue in the form of developing rudimentary teeth, which is similar to the appearance of a compound odontoma, or conglomerate masses of enamel, dentin, and cementum, as seen in a complex odontoma.

4.Odontoma

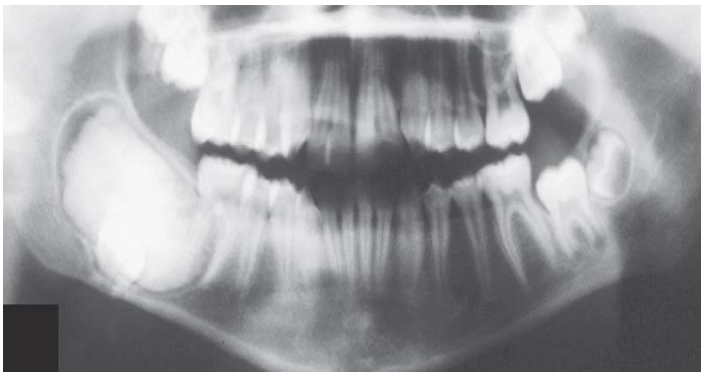
Odontomas are the most common types of odontogenic tumors. Their prevalence exceeds that of all other odontogenic tumors combined. Odontomas are considered to be developmental anomalies (hamartomas), rather than true neoplasms. odontomas consist chiefly of enamel and dentin, with variable amounts of pulp and cementum. In their earlier developmental stages, varying amounts of proliferating odontogenic epithelium and mesenchyme are present. 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.

Clinical features

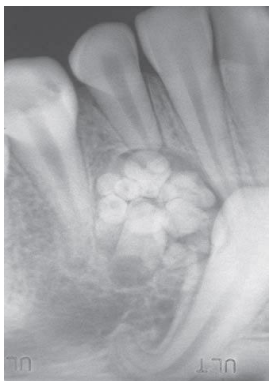
Most odontomas are detected during the first two decades of life, and the mean age at the time of diagnosis is 14 years. The majority of these lesions are completely asymptomatic, being discovered on a routine radiographic examination or when films are taken to determine the reason for failure of a tooth to erupt. Odontomas are typically relatively small and seldom exceed the size of a tooth in the area where they are located. 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 somewhat 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. Occasionally, an odontoma will develop completely within the gingival soft tissues.

Radiographical features

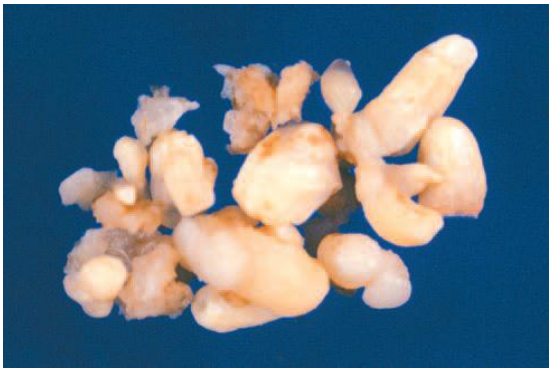
The compound odontoma appears as a collection of toothlike structures of varying size and shape surrounded by a narrow radiolucent zone. The complex odontoma appears as a calcified mass with the radiodensity of tooth structure, which is also surrounded by a narrow radiolucent rim. An unerupted tooth 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, and the compound odontoma is seldom confused with any other lesion. A developing odontoma may show little evidence of calcification and appear as a circumscribed radiolucent lesion. A complex odontoma, however, may be radiographically confused with an osteoma or some other highly calcified bone lesion.



Complex odontoma



compound odontoma



Compound odontoma

Treatment and prognosis

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

Tumors of odontogenic ectomesenchyme

1.Odontogenic Myxoma

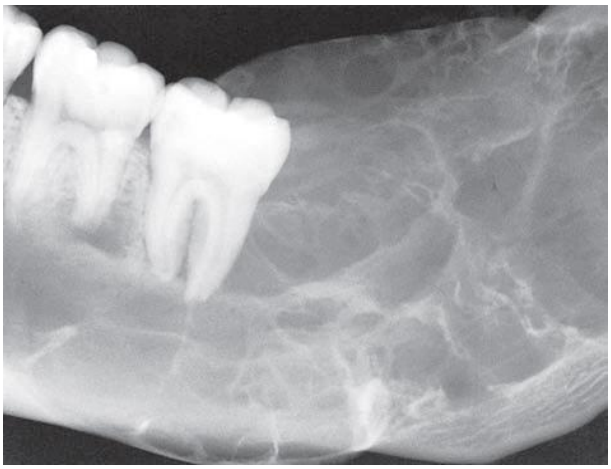
Odontogenic myxoma is a benign mesenchymal lesion that mimics microscopically the dental pulp or follicular connective tissue. It is a relatively common odontogenic tumor, representing 1% to 17% of all tumor types. Although myxomas are noted at various sites of the body, including the dermis, heart (left atrium), and other head and neck sites, only odontogenic myxoma of the jaws is derived from odontogenic ectomesenchyme. This benign neoplasm is infiltrative and may recur after inadequate treatment.

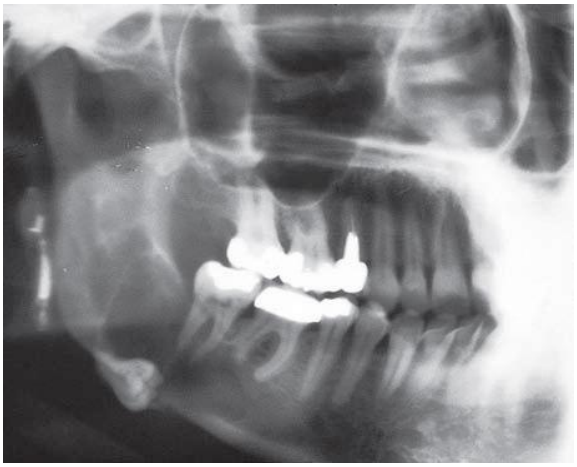
Histogenesis

Periodontal ligament or dental pulp

Clinical Features

- Adults (median age, about 30 years)
- Either jaw
- Radiographically: well-circumscribed or diffuse lesion. Often multilocular with a honeycomb pattern (Other radiographic patterns and descriptors include "honeycomb, soap bubble, or "tennis racket." Cortical expansion or perforation and root displacement or resorption may be seen.

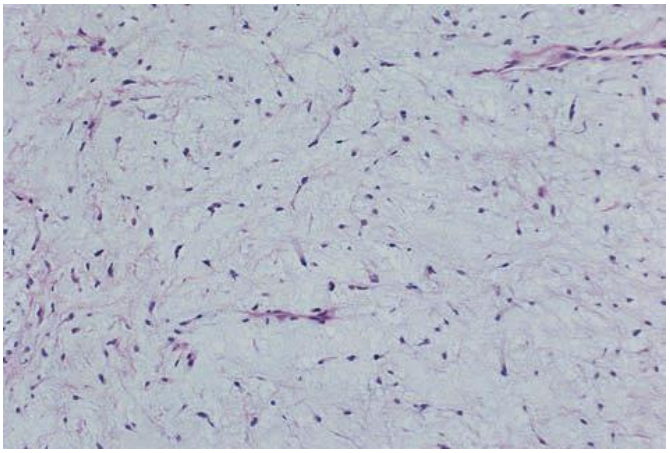




Histopathology

- This tumor is composed of bland, relatively acellular myxomatous connective tissue, benign fibroblasts and myofibroblasts with variable amounts of collagen are found in a mucopolysaccharide matrix.
- Bony islands, representing residual trabeculae, and capillaries are found scattered throughout the lesion
- Odontogenic rests typically are absent

- Odontogenic myxomas have a very low proliferation rate.



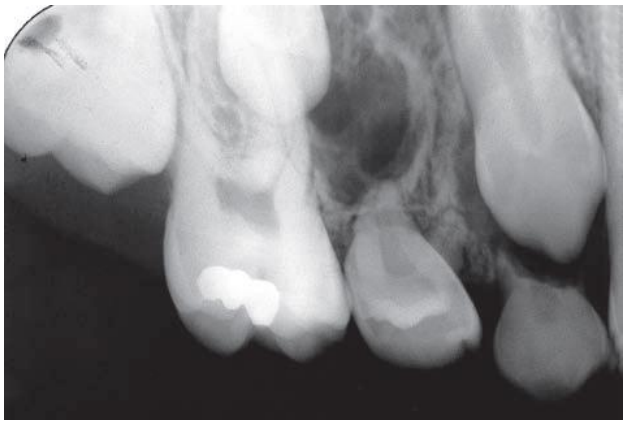
Odontogenic myxoma exhibiting typical bland myxoid appearance.

Treatment and prognosis

Surgical excision (conservative to radical) is the treatment of choice. However, because of its loose, gelatinous consistency and absence of a capsule, recurrence is more likely if the lesion is treated too conservatively. Although these lesions exhibit some aggressiveness and have a moderate recurrence rate, the prognosis is very good. Repeated surgical procedures do not appear to stimulate growth or metastasis. Follow-up examinations should be performed for a minimum of 5 years.

2. Central Odontogenic Fibroma

Central odontogenic fibroma is a rare ectomesenchymal tumor that is regarded as the central counterpart to peripheral odontogenic fibroma. It has been seen in all age groups and is found in both the mandible and the maxilla, with a 2:1 female predilection. It results in a radiolucent lesion that usually is multilocular, often causing cortical expansion. Approximately 45% of cases occur anterior to the first molar region of the maxilla, often with a cortical bony depression of the palatal contour.

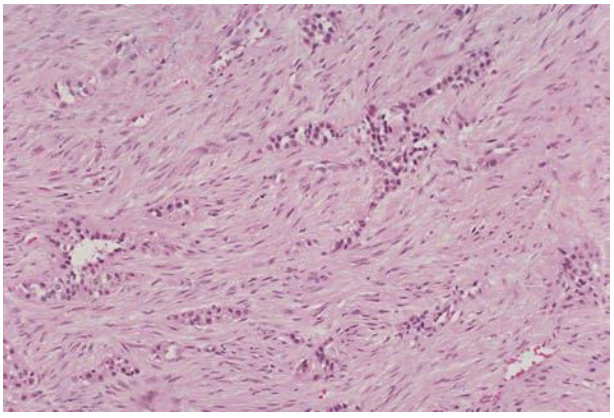


Central odontogenic fibroma of the right maxilla.

Histopathological features

Two patterns are recognized

- simple or epithelium-poor type: the lesion is composed of a mass of mature fibrous tissues containing few epithelial rests.
- complex (World Health Organization [WHO]) type) consists of mature connective tissue contains an abundant odontogenic epithelial component in the form of rests, along with calcified deposits of what is regarded as dentin or cementum.



Central odontogenic fibroma containing strands of odontogenic epithelium.

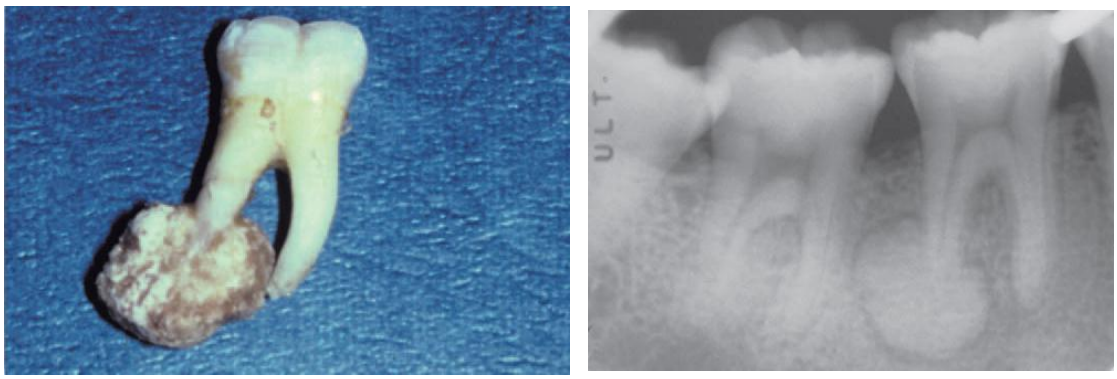
Treatment by enucleation or excision, and recurrence is very uncommon.

3. Cementoblastoma (true cementoma)

Clinical Features

Cementoblastoma, also known as true cementoma, is a rare benign neoplasm of cementoblasts that microscopically resembles an osteoblastoma but is connected or fused to the root of a tooth. It occurs predominantly in the second and third decades of life, typically before 25 years of age. There is no gender predilection. It is seen more often in the mandible than in the maxilla and more often in posterior than in anterior regions. It is intimately associated with the root of a tooth, and the tooth remains vital. Cementoblastoma may cause cortical expansion and, occasionally, low-grade intermittent pain.

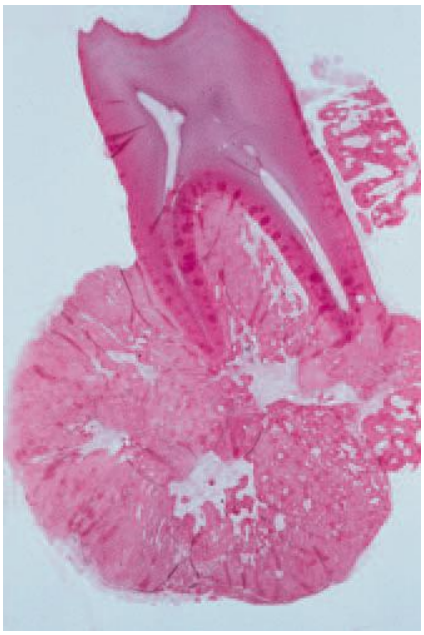
Radiographically, this neoplasm is an opaque lesion that replaces the root of the tooth. It is usually surrounded by a thick uniform radiolucent ring that is contiguous with the periodontal ligament space and the advancing front of the tumor.



Cementoblastoma

Histopathology

This lesion appears microscopically as a dense mass of mineralized cementum-like material with numerous reversal lines. Intervening well-vascularized soft tissue contains cementoblasts, often numerous, large, and hyperchromatic. The histologic features are similar if not identical to those of an osteoblastoma but with attachment to a tooth root.



Treatment

Because of the intimate association of this neoplasm with the tooth root, it cannot be removed without sacrificing the tooth by way of a surgical extraction procedure. Bone relief typically is required to remove this well-circumscribed mass. Recurrence is not seen.