Chapter 78: Odontogenesis and Odontogenic Cysts and Tumors

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Cysts and tumors derived from the odontogenic tissues constitute an unusually diverse group of lesions. This diversity reflects the complex development of the dental structures, since these lesions all originate through some alteration from the normal pattern of odontogenesis. Some lesions included in this category may in fact not represent neoplasia at all, but are only minor alterations in the normal process of tooth development. Lesions such as cysts are also tumors only in the broadest sense of the word and do not represent true neoplasms.

The purpose of this chapter is to review the process of odontogenesis with emphasis on the potential origin of cells that produce the most common odontogenic cysts and tumors. Diagnostic modalities will be discussed with emphasis on the practical approach to development of a differential diagnosis. Common odontogenic cysts and tumors will be presented, and treatment approaches to various odontogenic cysts and tumors outlined.

Odontogenesis (Melfi, 1988; Ten Cate, 1989)

The primitive oral cavity, also called the *stomodeum,* is lined with ectoderm. At its deepest portion it contacts the blind superior aspect of the foregut, which is lined with endoderm. The union of these ectodermal and endodermal layers is called the *buccopharyngeal membrane.* At approximately the twenty-seventh day of development, this membrane ruptures and the stomodeum becomes connected with the foregut. The primitive oral cavity is therefore an ectodermal lined structure beneath which lies the ectomesenchyme. The mesenchyme of the head is termed *ectomesenchyme* because of its ectodermal origin from neural crest cells. This is distinct from the mesenchyme of the rest of the body, which is of mesodermal origin.

Each tooth develops from a tooth bud that forms from the lining of the oral cavity. The tooth bud consists of three separate parts: (1) the enamel organ, which is derived from the oral ectoderm; (2) the dental papilla, which is derived from the ectomesenchyme; and (3) the dental sac, which is also derived from the ectomesenchyme. The portion of the developing tooth known as the *enamel organ* produces tooth enamel while the dental papilla produces the tooth pulp and dentin; the dental sac is the precursor of the cementum and periodontal ligament. Tooth development has been broken down into stages primarily based on morphologic changes of the tooth bud. These stages include: (1) the dental lamina stage, (2) the bud stage, (3) the cap stage, and (4) the bell stage.

Dental lamina and bud stage

About 2 or 3 weeks after the rupture of the buccopharyngeal membrane, when the embryo is about 6 weeks old, the first sign of tooth development is seen. Along the oral ectoderm a ridge of basal cells begin proliferation at a more rapid rate than those cells adjacent to them. This leads to formation of a band of epithelium that runs along the crest of what will be the future dental arches. This band of proliferating epithelium is called the *dental lamina.*

At certain points along the dental lamina, each representing a location of the 10 mandibular and 10 maxillary deciduous teeth, an even more rapid cellular proliferation is seen. This further epithelial downgrowth and proliferation forms invaginations into the underlying ectomesenchymal tissue. These invaginations or buds represent the beginning of tooth development (Fig. 78-1).

Cap stage (proliferation)

As cellular proliferation continues, unequal growth occurs at various parts of the bud. A shallow depression occurs on the deep surface of the bud. Concomitant to this morphologic change in the developing bud, histologic differentiation occurs within the cells of the enamel organ. The single layer of the cuboidal cells lining the convexity of the enamel organ are known as the *outer enamel epithelium* and the more columnar cells along the concavity of the organ are called the *inner enamel epithelium.* As cells within the center of this epithelial enamel organ begin to separate because of an increase in intercellular fluid, they assume a more branched or reticular form. This area is now called the *stellate reticulum.* These changes within the enamel organ produce a figure reminiscent of a cap, hence the name given to this stage of development (Fig. 78-2).

While this differentiation is occurring within the enamel organ itself, changes within the associated ectomesenchyme also occur. The portion of the ectomesenchyme that is partially surrounded by the proliferating inner enamel epithelium begins to condense, forming the dental papilla.

At the same time there is marginal condensation in the ectomesenchyme surrounding the enamel organ and the dental papilla. In this zone a denser and more fibrous layer gradually develops, forming the dental sac.

Bell stage (histodifferentiation and morphodifferentiation)

The differential growth of the epithelium into the underlying ectomesenchymal tissue continues, changing the form from a cap shape to a bell shape.

At this stage, the inner enamel epithelium consists of a single layer of columnar cells that further differentiates into tall columnar cells with oval nuclei that are polarized away from the basal lamina. These cells will become ameloblasts and be responsible for amelogenesis (enamel formation). These preameloblasts exert an organizing influence on the underlying ectomesenchymal cells of the dental papilla, causing them to begin differentiation into odontoblasts, which will be responsible for forming the dentin of the developing tooth (Fig. 78-3).

By the time the primary tooth germ has reached the bell stage, other changes are also taking place. Lingual to the enamel organ of this primary tooth germ, the dental lamina is giving rise to the beginning of the enamel organ of the permanent tooth (Fig. 78-4). Concurrently, the dental lamina of the primary tooth begins to disintegrate. As the lamina breaks up, the cells that compose it either disappear or remain as small islands known as the *rests of Serres.*

Apposition

After differentiation of ameloblasts and odontoblasts is complete, apposition of the dental hard tissues begins. Dentinogenesis begins at approximately the fifth month in utero. Dention formation begins as odontoblasts elaborate an eosinophilic materal along the interface between the odontoblasts and ameloblasts. This material is predenting, which upon calcification forms dentin, the principal hard structure of the tooth. This dentin formation begins at the incisal region of the tooth and progresses towards the root.

Amelogenesis or enamel formation begins immediately adjacent to the forming dentin and progresses toward the incisal edge. Similar to dentin formation, the first stage of enamel formation is the laying down of enamel as an organic material that subsequently undergoes calcification (Fig. 78-5). Normal amelogenesis relies on prior occurrence of normal dentinogenesis, hence the lack of enamel formation in odontogenic tumors such as ameloblastoma, where although normal-appearing ameloblasts are present, no odontoblasts or dentin formation is present to initiate enamel formation.

Root formation

After much of the crown formation has occurred, development of the root begins. The outer and inner enamel epithelia lie immediately adjacent to each other without an intervening stellate reticulum at the periphery of the developing tooth. These two layers together form Hertwig's epithelial root sheath, which grows into the surrounding connective tissue. Dentin formation continues towards the root of the tooth and is guided by this rooth sheath (Fig. 78-6). Root formation typically is complete 2 to 3 years after tooth eruption, at which time the root sheath breaks down and disappears with the exception of occasional epithelial remnants called the *rests of Malassez* Fig. 78-7).

As the dentin grows downward into what will be the root of the tooth, the surrounding dental sac forms a thin calcified layer called *cementum* around the developing root; the dental sac also develops into connective tissue fibers that will become the periodontal ligament. On a radiograph of an unerupted and incompletely formed tooth, the dental sac is also called the *dental follicle.*

Eruption

As amelogenesis occurs, the layer of ameloblastic cells moves progressively closer to the outer enamel epithelium, squeezing the stellate reticulum in between. At the termination of crown development, the ameloblastic layer, the stellate reticulum, and the outer enamel epithelium will all essentially be fused and become known as the *reduced enamel epithelium.* This reduced enamel epithelium will remain around the developed crown until eruption of the tooth.

The Formation of Odontogenic Cysts and Tumors

The epithelium associated with odontogenic cysts and tumors is derived from one of the following sources (Fig. 78-8): (1) the reduced enamel epithelium of the tooth crown; (2) epithelial rests of Malassez, which are remnants of the Hertwig root sheath; (3) epithelial rests

of Serres, which are remnants of the dental lamina; and (4) the tooth germ itself, which includes the enamel organ, dental papilla, and dental sac. For example, an increased space between the crown of an unerupted tooth and the surrounding reduced enamel epithelium is frequently an early indication of cyst or tumor formation (Fig. 78-9).

In addition, some odontogenic tumors develop partially or entirely from cells of ectomesenchymal origin. The cellular origin for these lesions is either the dental papilla, the follicle, or the periodontal ligament.

Classification of Cysts and Tumors of the Jaws

Several systems have been devised for classifying cysts and tumors of the jaws. Some of these systems rely on morphologic differences between different lesions, either based on clinical or radiographic parameters, whereas others rely on histologic differences. Other systems categorize lesions totally on their etiology (Main, 1985; Shear, 1985). A modification of the WHO classification (Main, 1985), which divides cysts according to etiology is quite effective (see box on p. 1419). In this classification, the basic division is etiologic. A distinction is drawn between the autonomous development of the majority of cyst varieties and the inflammatory pathogenesis of the most common, the radicular cyst. This is preferable to classification based on histogenesis because opinions differ regarding the histology of several categories of cysts.

Box (p. 1419) **Classification of jaw cysts**

A. Developmental

1. Odontogenic

a. Follicular cyst

b. Odontogenic keratocyst

c. Eruption cyst

d. Alveolar cyst of infants

e. Gingival cyst of adults

f. Developmental lateral periodontal cyst

2. Nonodontogenic

a. Nasopalatine duct cyst

b. Midpalatal cyst of infants

c. Nasolabial cyst

d. Globulomaxillary cyst, median mandibular cyst, and median alveolar cyst&

B. Inflammatory

a. Radicular cyst

1) Periapical cyst

2) Inflammatory lateral periodontal cyst

C. Nonepithelial&&

a. Idiopathic bone cavity (traumatic, solitary, hemorrhagic bone cyst)

b. Aneurysmal bone cyst

c. Stafne's mandibular lingual cortical defect.

& These cysts were previously regarded as developmental nonodontogenic cysts. This is no longer believed to be true and this category is no longer used; it is of historical interest only.

&& These lesions are often classified as cysts but do not have a distinct epithelial lining.

Unlike cysts, odontogenic tumors are most often classified according to their histogenesis. Tumors are either of epithelial, mesenchymal, or unknown origin. Tumors of epithelial origin are further classified into those that have a minimal inductive change on the surrounding connective tissue and those that produce extensive inductive changes within the surrounding connective tissue. Lesions within the last group have in the past been called *mixed epithelial mesenchymal tumors* (Shafer et al, 1984). This is incorrect because it is now believed that the neoplastic portion of the lesion is epithelial in nature and that this epithelium produces significant inductive changes within the connective tissue; this leads to the misconception that the connective tissue is also neoplastic. The box on p. 1420 outlines the classification of odontogenic tumors.

Box (p. 1420) **Classification of odontogenic tumors**

A. Benign epithelial odontogenic tumors

1. Tumors producing minimal inductive change in the connective tissue

a. Ameloblastoma

b. Calcifying epithelial odontogenic tumor (Pindborg tumor)

c. Odontogenic adenomatoid tumor (adenoameloblastoma, adenomatoidodontogenic tumor)

d. Calcifying odontogenic cyst (Gorlin's cyst)

2. Tumors producing extensive inductive change in the connective tissue

a. Ameloblastic fibroma

b. Ameloblastic fibroodontoma

c. Ameloblastic odontoma (odontoameloblastoma)

d. Odontoma

1) Compound-composite odontoma

2) Complex odontoma

B. Mesenchymal odontogenic tumors

1. Odontogenic fibroma

2. Odontogenic myxoma

3. Cementoma

a. Periapical cemental dysplasia

b. Cementifying fibroma

c. Benign cementoblastoma

4. Dentinoma

C. Tumors of unknown origin

1. Melanotic neuroectodermal tumor of infancy

D. Malignant odontogenic tumors

1. Primary intraosseous carcinoma

2. Ameloblastic fibrosarcoma

3. Ameloblastic dentinosarcoma

4. Ameloblastic odontosarcoma.

Diagnosis

Clinical findings

The physical signs and symptoms of odontogenic cysts and tumors will depend to a certain extent on the dimensions of the lesion. A small lesion is unlikely to be diagnosed on a routine examination of the mouth because signs will not be demonstrable. Such lesions are only likely to be detected at an early stage as the result of routine radiographic examination. Exceptions are some early lesions that may present in conjunction with a devitalized tooth, which is detectable on clinical examination. Some cystic lesions may become secondarily infected, leading to their diagnosis. Clinical absence of one or more teeth without the history of extraction may also be a clinical indicator of an undiagnosed odontogenic cyst or tumor because many of these lesions are associated with impacted or congenitally missing teeth (Fig. 78-10).

As the lesion grows, other indirect changes may occur. An enlarging lesion between two teeth can cause the crowns to converge and the roots to diverge (Fig. 78-11). Growth that is nearly undetectable visually may lead to difficulty with denture retention.

As the lesion enlarges even further, expansion of the bone may be seen directly. This is usually toward the buccal surface of the alveolar bone because this is the thinnest area and expansion occurs here most easily (Fig. 78-12). Clinically evident expansion is often a late finding, especially in lesions developing within the ramus or angle of the mandible or within the maxillary sinus. Lesions in these areas may become extremely large before expansion is observed clinically (Fig. 78-13).

Radiologic examination

Radiographic evidence of various odontogenic cysts and tumors varies considerably from lesion to lesion, with certain lesions having almost a pathognomonic appearance whereas the only consistent thing about the appearance of other lesions is the total lack of any consistent radiographic presentation. The common lack of of physical findings and the development of the majority of these lesions within the confines of bone makes the radiologic investigation and interpretation uniquely important. The radiographic appearance of various lesions will be discussed more completely as each individual lesion is described. There are some general principles that can be applied to the radiographic diagnosis of cysts and tumors of the jaws. This section will address these principles.

In order for a radiograph to provide useful information, it must be of good quality. Artifacts caused by inappropriate placement of film labels, patient jewelry, movement, or poor technique can mask the presence of a lesion.

Radiographs are also important in treatment planning for surgical removal of odontogenic lesions. Encroachment on vital structures, extent into soft tissue, size of the lesion, and requirements for reconstruction can all be evaluated. This will be more completely discussed in the section on treatment of cysts and tumors.

Various radiographic modalities are available for use in evaluating cysts and tumors of the jaws. Many lesions are detected on routine intraoral radiographs taken for screening purposes or for evaluating of other dental disease such as caries. Because intraoral radiographs such as the periapical or bite-wing radiograph are most often used, this screening may be the first opportunity to evaluate an asymptomatic lesion. Intraoral radiographs have the advantage of providing excellent detail because the film-to-object distance is small and the film is placed intraorally immediately adjacent to the involved bone. This eliminates the overlying osseous structures often seen in routine extraoral head and neck radiographs. The major disadvantage is the lack of availability of these radiographs outside a dental office, their size limitations, and their inability to evaluate the areas that are not accessible to film placement; these are the ramus, condyle, and inferior border of the mandible. Fig. 78-14 demonstrates how a subtle change on an intraoral radiograph can lead to diagnosis of a large intraosseous lesion. Perhaps the most useful intraoral radiograph, one for which there is no good extraoral substitute, is the occlusal radiograph. This is taken with a larger film than the standard periapical or bite-wing. For lesions in the anterior maxillary alveolus and palate, there is no adequate substitute (Fig. 78-15).

Extraoral radiographs are often necessary for delineation of the entire lesion. They are useful in showing the extent of the lesion and its effect on adjacent structures. These radiographs also serve as a screening mechanism should multiple lesions be present. The panoramic radiograph is the most useful examination. It effectively shows the entire mandible including the ramus and condyle, the maxilla and maxillary sinuses, and the dentition. To reproduce this with other types of radiographs may require multiple views. The panoramic radiograph is actually a pantomogram. It makes use of the radiographic technique of tomography in which a collimated radiation source and the film are rotated around the patient's head in opposite directions. This results in blurring of structures on either side of a predetermined focal trough and has the advantage of eliminating much of the surrounding osseous interference that is seen with plain radiographs. However, this advantage is also a source of one of the major problems of the panoramic radiograph, which is that only structures within the focal trough are registered. This area of optimal focus is minimally adjustable and varies from machine to machine. It is possible that a lesion, even a rather large one, be incompletely visualized if it is outside the focal trough (Fig. 78-16). If a lesion is suspected on panoramic radiograph, an additional radiograph should be taken in the sagittal plane to help delineate any lateral and medial expansion that may be present. The panoramic radiograph also has the disadvantage of requiring the patient to remain still and in a sitting position for several seconds. This may not be possible in the debilitated, mentally retarded, or very young patient.

Other extraoral films may be used to evaluate the lesion further or as a substitute when the panoramic radiograph is not possible. For mandibular lesions, the lateral oblique radiograph will allow the body, angle, ramus, and condyle to be seen. The parasymphysis and symphysis is often difficult to see because of overlying dentition and bone from the contralateral side. The teeth are also difficult to identify because teeth from the ipsilateral maxilla and the contralateral mandible and maxilla may overlie each other. For examination of maxillary cysts and tumors, the Waters view is very helpful as a screening tool. Maxillary lesions are difficult to delineate even on the best plain films; computerized tomography may be necessary.

Computerized tomography (CT) is not a cost-effective method of evaluating most routine odontogenic cysts and tumors. There are certain indications for the use of CT scans. For example, CT is helpful in defining the extent of very large lesions, especially if there is significant distortion of normal anatomy, which makes plain radiographs difficult to evaluate. CT can also be used to determine if the lesion is contained entirely within the involved bone, or if it has eroded through the cortex. Although malignant odontogenic tumors are extremely rare, some bening cysts and tumors behave aggressively and tend to have high recurrence rates, especially if they have perforated through cortical bone and into adjacent soft tissue; CT is extremely valuable in treatment planning for the removal of such lesions (Fig. 78-17).

Differential diagnosis

Once adequate clinical and radiographic examination is complete and before a biopsy has been performed, a differential diagnosis should be formulated. This should take into account information from both the clinical and radiologic examination, but because of the limited clinical findings associated with most odontogenic cysts and tumors, this is primarily a radiographic differential.

Much can be learned about the behavior of an odontogenic cyst or tumor by paying close attention to the radiograph. Benign, slow-growing lesions tend to cause tooth movement, blunting of the roots of teeth, displacement of vital structures such as nerves and vessels, and bony hard expansion with intact cortical bone. These lesions are usually well circumscribed by a radiopaque border (Fig. 78-18). Aggressive, fast-growing, or malignant lesions tend to resorb bone around the roots of teeth and not cause tooth movement. The teeth themselves are of such increased hardness in comparison to the surrounding bone that fast-growing lesions do not have time to cause resorption of the roots before the lesion is detected. Fast-growing and aggressive lesions also tend to resorb cortical bone and spread into soft tissue without significant cortical expansion. They also are poorly delineated radiographically without much cortical bone along the periphery of the lesion (Fig. 78-19).

From a radiographic approach, a simplified method of grouping these lesions is by some common radiographic findings. Each specific radiographic appearance may be associated with several different lesions, some of which are odontogenic in origin, some of which are not, but significant reduction in the number of different lesions that must be considered in a given differential is possible by grouping lesions according to their radiographic appearance. Other clinical findings such as age, sex, symptoms if any, etc, can be used to further narrow the differential beyond what is possible radiographically. The classic radiographic presentations are radiolucent lesions, radiopaque lesions, and mixed radiopaque/radiolucent lesions. Within the purely radiolucent group, there are several subclassifications according to location and appearance of the lesion. In an attempt to facilitate the development of a differential diagnosis, the most common lesions occurring in the jaws have been categorized according to their radiographic apearance in the box below. This list includes some non-odontogenic processes, which must also be considered in the differential.

Biopsy

Perhaps the simplest biopsy technique for intraosseous lesions of the jaws is aspiration. Although no tissue is obtained with aspiration, it is a biopsy in the broadest sense of the word. Aspiration can yield extremely valuable information about the nature of the lesion, yet it causes little patient discomfort. It may be done as a procedure itself or as a prelude to incisional or excisional biopsy. A radiolucent lesion that yields straw-colored fluid on aspiration is most likely to be a cystic lesion. If pus is aspirated, an inflammatory or infectious process should be considered. White keratin-containing fluid is indicative of an odontogenic keratocyst, whereas air may indicate a traumatic bone cavty. The inability to aspirate (vacuum within the syringe) is usually indicative of a solid process such as a neoplasm. Blood on aspiration could represent several lesions, the most important of which is the vascular malformation. The most common presentation of high-flow vascular lesions of the jaws is exsanguination or near-exsanguinating hemorrhage associated with a simple procedure such as biopsy of an innocuous asymptomatic radiolucency. For this reason, all radiolucent lesions should be aspirated before a biopsy is performed.

Whether to perform an incisional or excisional biopsy initially is not always clear. Most odnotogenic cysts and tumors are benign and definitive treatment may be as simple as obtaining an excisional biopsy. However, there are some lesions that are more aggressive and require extensive curettage or marginal resection and it is desirable to know this before the onset of surgery.

Indications for excisional biopsy include small, radiographically benign lesions that are readily accessible and can be removed without encroachment on adjacent structures. In cases where the access is more invasive than the surgical removal of the lesion itself, complete excision at the initial biopsy is preferred, even if the lesion is relatively large. This would include lesions in the mandibular condyle and lesions in very young patients for whom a general anesthetic is required for any surgical intervention, even for incisional biopsy. Incisional biopsy is indicated in very large lesions and in lesions suspected of aggressive behavior so that a definitive diagnosis can be made before treatment is instituted.

Clinical, Radiographic, and Histoogic Features of Common Odontogenic Cysts

The more classic description of a cyst is that it is an epithelial-lined sac filled with fluid or semifluid material (Killey and Kay, 1966). All odontogenic cysts fit this description, but certain nonepithelial cysts commonly included with odontogenic cysts do not. Because of this, a broader definition has been accepted. A cyst is a pathologic cavity having fluid, semifluid, or gaseous contents that are not created by the accumulation of pus; frequently, but not always, it is lined by epithelium (Kramer, 1974).

For the purpose of discussion and to simplify development of a differential diagnosis, this section will present a brief overview of each common cyst, its clinical and radiographic presentation, and recommended treatment. Because the histopathology of jaw cysts is confusing and the terminology is not always used consistently by general pathologists, these lesions are frequently misdiagnosed or incorrectly named, which can lead to improper treatment. Lesions that are frequently misdiagnosed will have special mention with regard to histopathology.

Lesions will be categorized according to the classification outlined previously. The more common inflammatory cysts will be described first, followed by the developmental odontogenic and nonodontogenic cysts, and lastly, the nonepithelial cysts.

Inflammatory cysts

Inflammatory cysts make up 85% of the cysts found in the jaws (Kay and Laskin, 1985). This high frequency is the result of the prevalence of dental disease, which frequently initiates the process. Inflammatory cysts result after bacterial invasion of the dental pulp leads to a chronic low-grade infection that results in a periapical granuloma around the root of the tooth. The normally quiescent epithelial cell rests of Malassez, which are within the periodontal ligament, are activated and proliferate to surround the granuloma, leading to cyst formation. These cysts then grow by mechanisms that are not clearly defined to produce the inflammatory cyst associated with the root of the involved tooth, hence appropriately named *radicular cyst.* When the cyst is associated with the apex of the tooth root, it is called a *periapical cyst;* when it is along the side of the root, it is called a *lateral periodontal cyst.*

Periapical cyst

The periapical cyst is wel recognized as the cystic lesion most likely encountered.

Clinical features. The majority of these lesions are asymptomatic. The associated tooth is nonvital and may have evidence of the source of the initial offending infection such as large unrestored decay, a very large filling, or a history of pain in the tooth in the past.

Radiographic features. The radiographic presentation is fairly consistent. It is usually a radiolucent area of variable size attached to the root apex. The radiolucency is usually rounded or oval and surrounded by a radiopaque sclerotic bony periphery (Fig. 78-20).

Histology. The cyst is lined by stratified squamous epithelium. Pseudostratified ciliated columnar epithelium may also be seen in lesions occurring near the maxillary sinus. The thickness of the lining usually varies and it seldom exhibits keratin formation.

Treatment. Treatment requires only that the source of infection be treated. This involves endodontic therapy (root canal) of the tooth to remove the necrotic pulpal tissue or extraction of the tooth. The lesion will generally resolve after this but should be followed radiographically and enucleated if it enlarges or fails to resolve.

Lateral inflammatory periodontal cyst

The lateral inflammatory periodontal cyst is much less common than the periapical cyst. When the radicular cyst forms around an opening between the pulp and the periodontal ligament that is along the lateral aspect of the tooth instead of at the apex, a lateral inflammatory periodontal cyst is produced.

Clinical features. These lesions are clinically asymptomatic except for the associated nonvital tooth.

Radiographic features. These lesions are usually small, well-circumscribed radiolucencies associated with the lateral aspect of the tooth root. There may be signs of previous tooth injury such as dental decay, or large restoration of the tooth.

Treatment. Treatment is the same as for periapical cysts.

Developmental odontogenic cysts

Developmental odontogenic cysts account for 10% of all cysts of the jaws. The odontogenic keratocyst and the follicular cyst account for 8% whereas all other developmental odontogenic cysts accound for the other 2% (Trimble, 1986). Emphasis will be placed on the two most common lesions in this group.

Follicular cyst

This cyst has also been called a *dentigerous cyst* because of its association with the crown of unerupted teeth, but as other types of cysts can also be found around the crowns of unerupted teeth, the term *follicular cyst* is more correct because of the suspected development of the lesion from the follicle of the tooth. This cyst is most frequently identified in the mandible and is associated with the completed crown of an unerupted or impacted tooth. The cyst is thought to originate from accumulation of fluid between the reduced enamel epithelium and the completed tooth crown (see Fig. 78-9). Its incidence is the same in both sexes and is most common in childhood and adolescence. The growth rate may be quite rapid, with lesions growing up to 5 cm in diameter in 3 to 4 years (Livingstone, 1927).

Clinical features. Follicular cysts may get quite large before they are diagnosed and facial swelling may be the first clinical sign. They are always associated with an unerupted or impacted tooth, most commonly the mandibular or maxillary third molar or maxillary canine.

Radiographic features. The follicular cyst appears as a radiolucent area within the jaw and is in some way associated with an unerupted tooth crown. It is usually a well-circumscribed lesion that is unilocular and has a sclerotic bony border. There may be significant expansion of the cortical plate and displacement of tooth roots or the inferior alveolar canal. The cyst may cause displacement of the associated tooth, sometimes for quite a distance (Fig. 78-21).

Histology. The follicular cyst typically has a thin connective tissue wall that is lined by a thin layer of stratified squamous epithelium within the lumen (Fig. 78-22). Hyaline bodies are frequently found within the epithelial lining, as well as clefts from cholesterol crystals. Some cysts may also have mucous cells within the lining, which is of importance when considering the possibility that other lesions may develop within the wall or lumen of a follicular cyst. The lesions that may develop within the cyst arise from the actual epithelial lining or from rests of odontogenic epithelium that are in the connective tissue around the cyst. Odontogenic tumors such as the ameloblastoma frequently arise from the lining of the follicular cyst. Less common but of importance is the occurrence of epidermoid carcinoma arising from the lining of a follicular cyst, or of mucoepidermoid carcinoma arising from the mucous glands within the wall of the cyst. This is the suspected origin of most intraosseous

mucoepidermoid carcinomas.

Treatment. Treatment is dependent upon the size of the lesion, but enucleation and curettage is adequate. For very large lesions, decompression to allow some decrease in size is possible but the lesion should ultimately be treated with enucleation and curettage to allow histologic evaluation of the entire lesion and to prevent recurrence.

Odontogenic keratocyst

The odontogenic keratocyst is so named because of its characteristic biologic appearance. Clinically and radiographically it may appear as a follicular cyst associated with the crown of an unerupted tooth. It may also appear associated with the root of a tooth as a radicular cyst or by itself. When it occurs by itself, it is frequently called a *primordial cyst*, which is a cyst that arises where there is no apparent tooth formation associated. Virtually all primordial cysts have the histologic appearance of an odontogenic keratocyst. The odontogenic keratocyst is thought to develop from the remnants of the dental lamina, which are the rests of Seres (Harris, 1974; Main, 1970).

Clinical features. The odontogenic keratocyst may occur in many clinical situations. It may appear at any age but is rare below 10 years of age. The peak incidence is in the second and third decades and there is a predilection for males of 1.8 to 1. Anatomically the mandible is affected more frequently than the maxilla with 75% of the lesions occurring there (Browne, 1970). The third molar and ramus areas are the most frequently involved, with the first and second molar areas following. No clinical presentation is specific. The patient may be asymptomatic until gradual expansion is noted or until a secondary infection occurs.

Radiographic features. The odontogenic keratocyst may be identified in a variety of anatomic locations. The majority of the lesions are present in the third molar and ramus area of the mandible. The cyst itself may have several radiologic variations. Approximately 50% of these cysts have a unilocular appearance but a multilocular appearance can occur. As already mentioned, the diagnosis of odontogenic keratocyst is a histologic one and the cyst may have several radiographic appearances such as of a follicular or radicular cyst. The lesion is slow growing and will usually be well circumscribed with a sclerotic border. Teeth may be displaced and cortical perforation may be more common than in other cystic lesions.

Histology. Specific histologic criteria exist for diagnosis of a keratocyst (Fig. 78-23). These include: (1) a thin, stratified squamous epithelium that is a uniform 6 to 8 cells thick, without rete ridge formation; (2) prominent columnar or cuboidal basal cell layer with dense nuclear staining; (3) a corrugated surface layer of parakeratin or orthokeratin; and (4) a thin connective tissue wall. The lumenal material may vary from a straw-colored clear substance to a creamy white keratin-filled material. The importance of the histologic diagnosis is that an untrained pathologist may mistake the odontogenic keratocyst for a simple follicular cyst. The recurrence rate for these lesions is much higher and this misdiagnosis may lead to improper treatment.

Treatment. As mentioned previously, distinguishing the odontogenic keratocyst from other odontogenic cysts is important because of its higher recurrence rate. Recurrence rates from 10% to 60% have been reported. The large difference in rate of recurrence may be

related to inconsistencies in reporting of the data and inadequate follow-up because these lesions can take many years to recur. In a recent review of 426 patients, a composite recurrence rate of 34% was seen (Williams, 1991). This is consistent with other well-documented reports citing recurrence rates of 33% (Pindborg and Hansen, 1963) and 44% (Toller, 1972) respectively. The high incidence of recurrence is possible due to several factors: (1) some lesions are multilocular, making complete removal difficult; (2) the cyst lining is quite thin and friable, which makes it easy to leave fragments behind during enucleation; (3) odontogenic keratocysts have been shown to have a higher mitotic rate than other cysts, which may make any residual epithelium more likely to proliferate and lead to recurrence; (4) the cyst itself may have areas of epithelial budding that are left behind during enucleation; (5) as these lesions tend to perforate cortical bone more frequently, cystic epithelium may be located in the soft tissue from where it is more difficult to completely remove it; and (6) the lesion is often clinically mistaken for more benign cysts and treated less aggressively, only to have the diagnosis of keratocyst rendered after treatment is complete (Trimble, 1986).

Treatment consists of enucleation and curettage. If the lesion is quite large, decompression with subsequent enucleation is advantageous (Waldron, 1941). In the case of the odontogenic keratocyst, this is particularly useful because even after a short period of exposure to the oral cavity by temporary decompression, the lining epithelium of the cyst undergoes a thickening and the fibrous connective tissue thickens as well. This makes removal easier and decreases the chance of leaving remnants of epithelium behind. It has been shown that if the cystic lining can be removed in one piece, the incidence of recurrence is very small; however, if the lining is removed in fragments, the incidence of recurrence is greater than 50% (Forsell, 1980). If a wedge of tissue is removed from the mucosa overlying the cyst, the potential source of other lesions may be removed (Stoelings and Peters, 1973). Once the lesion has been enucleated, aggressive curettage of the bony walls with rotary instruments is recommended. Others have used chemical or cryotherapy modalities to further remove any remaining epithelium, but data proving that this is helpful is inadequate. If the lesion recurs, especially if it is located in the posterior mandible or maxilla, more aggressive treatment, such as marginal resection, may be indicated.

Special considerations. If multiple keratocysts are found, suspicion should be high for basal cell nevus syndrome (Gorlin's syndrome) (Gorlin, 1987; Gorlin and Golz, 1960). Patients with this syndrome also exhibit skeletal anomalies such as calcified falx, bifid ribs, fused vertebrae, and scoliosis. They also have soft-tissue aberrations such as multiple basal cell carcinomas, even in young patients and in non-sun-exposed areas; palmar pitting; and epidermoid cysts. Patients have a characteristic facies of frontal and temporoparietal bossing of the skull, well-developed supraorbital ridges, and ocular hypertelorism.

The remaining developmental odontogenic cysts only account for 2% of all jaw cysts, making their clinical significance relatively low. Some of them can present an unusual clinical picture and they deserve some review.

Eruption cyst

The eruption cyst is generally recognized as the soft tissue analogue of the follicular cyst. It is entirely within the soft tissue and does not usually produce any radiographic change. The cyst will generally undergo spontaneous rupture and resolution without any treatment. The clinical appearance is that of a smooth, tense, dark blue or purple swelling over the crown of an erupting tooth (Fig. 78-24). The lesion may cause some alarm for the parent and may also be symptomatically painful. If symptomatic, a portion of the sac can be excised leading to resolution of the symptoms.

Alveolar cyst of infants

These small cysts are also known as *dental lamina cysts of the newborn.* The correct term is *alveolar cyst of infants* and they are small discrete white swellings located on the crest of the alveolus in newborns. These small soft-tissue cystic lesions arise from the dental lamina. They are asymptomatic, require no treatment, and will resolve spontaneously.

Gingival cyst of adults

The gingival cyst (called the *gingival cyst of adults* to distinguish it from the alveolar cyst of infants) is an uncommon soft-tissue lesion. The term is used to identify a cyst that occurs in either the attached or unattached gingival tissue. It is believed to originate in the dental lamina. It occurs at any age but is much more common in patients over the age of 40. It occurs most commonly in the cuspid-bicuspid region of the mandible. The cyst is typically asymptomatic and presents as a localized swelling in the involved area. There is usually no radiographic evidence and not tendency for recurrence after excisional biopsy, which is the treatment of choice.

Developmental lateral periodontal cyst

This is the developmental counterpart of the inflammatory periodontal cyst. The cyst develops from epithelial rests of Malassez, but pulpal necrosis of the tooth is not the initiating factor. These lesions occur primarily in adults and local excision is the treatment of choice. Recurrence is rare.

Developmental nonodontogenic cysts

The remaining 5% of cysts within the jaws are made up of nonodontogenic cysts. Developmental nonodontogenic cysts within the jaws must arise from epithelium not associated with tooth development. In the past, several entities were described as fissural cysts because they were suspected to have arisen from tissue entrapped during fusion across various fissures during facial development. It has now been clearly shown that the only true fissural cyst is the midpalatal cyst of infants. Other so-called fissural cysts such as the globulomaxillary cyst, the median alveolar cyst, and the median mandibular cyst are most likely to be developmental or inflammatory odontogenic cysts (Main, 1985; Shear, 1985; Wysocki, 1981). The other two lesions within this category are the nasopalatine duct cyst, which arises from the cystic degeneration of the vestigule bilateral oronasal ducts, and the nasolabial cyst, which is a soft-tissue cyst arising from dystopic rests of the nasolacrimal

ducts.

Nasopalatine duct cyst

The nasopalatine duct cyst, also known as the *incisive canal cyst* is formed from cystic degeneration of the oronasal ducts that connect the nasal cavity to the oral cavity during development. It is the most common nonodontogenic developmental cyst. The nasopalatine duct cyst is usually asymptomatic and is discovered on routine radiographic investigation. It may also become secondarily infected leading to rapid enlargement and pain. The cyst appears as a round to ovoid or heart-shaped radiolucency lying between the maxillary central incisors (see Fig. 78-15). It may be difficult to distinguish a small lesion from the nasopalatine duct itself. The nasopalatine duct cyst is lined by stratified squamous epithelium, pseudostratified ciliated columnar epithelium, cuboidal epithelium, or a combination. The cyst has a connective tissue wall that may have mucous glands, nerves, and blood vessels within it. Small cysts may just be observed with regular radiographic examination whereas larger lesions or those that become secondarily infected should be treated with enucleation and curettage.

Midpalatal cyst of infants

The midpalatal cyst of infants arises from epithelium entrapped along the line of fusion of the palatal processes of the maxilla. The reason for induction of this epithelium to cyst formation is unknown. It is located at the midline of the hard palate of the maxilla. It is usually asymptomatic unless secondarily infected. It may produce a midpalatal swelling. An intraoral occlusal radiograph will often show the lesion, which is a well-circumscribed radiolucency with a sclerotic border, located in the midline of the hard palate. The histology shows stratified squamous or pseudostratified columnar epithelium. Treatment is access through a full-thickness palatal flap and enucleation and curettage. Recurrence is low.

Nasolabial cyst

The nasolabial cyst is a soft-tissue lesion developing within the labial vestibule just below the attachment of the nasal ala in the maxilla. It has been identified incorrectly as a fissural cyst in the past and does not originate from fissular epithelium but from remnants of the nasolacrimal ducts (Main, 1985). The clinical presentation is one of upper lip swelling or of swelling within the floor of the nose. Nearly 75% of these lesions occur in women. The cyst rarely produces a radiographic appearance. Histologically, it is a true cyst with an epithelial lining with stratified squamous, or cuboidal, or respiratory epithelium. The cyst should be treated with local excision from an intraoral approach.

Nonepithelial cysts

The nonepithelial cysts are a group of unrelated lesions that typically have been termed cysts although they do not have an epithelial lining. These lesions must be included in any discussion of cysts of the jaws because they must be considered in the differential diagnosis. They are best included here because they do not fall into the group of odontogenic tumors or benign diseases of bone.

Idiopathic bone cavity

The idiopathic bone cavity is also known as *solitaty cyst, hemorrhagic cyst, extravasation cyst,* and *simple bone cyst.* The lesion is not a true cyst and its etiology is unclear, so names that imply an etiolofy of true cyst formation should be avoided. Of the several theories regarding the development of this cavity, trauma that initiates an intramedullary hemorrhage with subsequent degeneration of the clot without bony filling, leading to an empty bone cavity, is the favored explanation. The idiopathic bone cavity occurs most frequently in the second decade of life and is most often identified in the region of the posterior body and ascending ramus of the mandible. The patient is usually asymptomatic and the defect is discovered on routine radiographic investigation (Fig. 78-25). The idiopathic bone cavity is usually a radiolucent lesion of variable size. It may become very large and surround the mandibular posterior teeth. This is different from other lesions that tend to push the teeth roots apart. The lesion may also extend to the inferior border but does not usually displace the inferior alveolar nerve, which is also unusual because other lesions tend to displace the nerve. Histologically, the lesion shows bony walls without a lining. It is fluid- or air-filled. Treatment requires biopsy to rule out other lesions, but the process of opening the lesion and causing hemorrhage within the cavity is usually enough to lead to resolution of the lesion.

Stafne's mandibular lingual cortical defect

This entity is also known by several other terms including *lingual mandibula bone cavity, static bone cavity,* and *lingual salivary gland defect.* This is an asymptomatic lesion. It usually occurs in adults over the age of 25. Radiographically, the lesion is a solitary radiolucency below the inferior alveolar canal near the angle region of the mandible (Fig. 78-26). The lesion is usually oval and exhibits no growth over long periods of time. The periphery is smooth and symmetric. The lesion is benign, has not growth potential, and a classic radiographic appearance that makes diagnosis without biopsy possible and observation without active intervention the treatment of choice.

Aneurysmal bone cyst

The aneurysmal bone cyst is not unique to the maxillofacial complex. It is a lesion that has a predilection for females and occurs most commonly in the vertebral column and the long bones. In the craniofacial skeleton, the mandible is affected most often. The patient often complains of swelling over a localized area and pain. It is felt that this lesion may develop from some other primary lesion such as the central giant-cell granuloma that undergoes alteration in local hemodynamics. Radiographically, the lesion is a radiolucency with a multilocular or honeycombed appearance. The sclerotic bone margins seen in most cysts may be absent. Histologically, the lesion is made up of a fibrous connective tissue stroma with multiple cavernous and sinusoidal spaces with multinucleated giant cells within the stroma (Fig. 78-27). Enucleation is the treatment of choice with hemorrhage during the removal being brisk because of the large number of vascular spaces within the lesion. Hemorrhage is usually controlled easily once the entire lesion is removed; when treating these lesions, the goal should be rapid enucleation to prevent blood loss with control of bony bleeding with bone wax or packing. The lesion does not recur after simple enucleation.

Clinical, Radiographic, and Histologic Features of Common Odontogenic Tumors

Benign epithelial odontogenic tumors

Ameloblastoma

Ameloblastoma is a benign neoplasm that may be locally invasive. This tumor is composed entirely of soft tissue. It may show periods of relative dormancy as well as fairly rapid expansion. It probably has a variable origin, arising from epithelial rests of Serres or Malassez, the enamel organ, the epithelial lining of dentigerous cyst, or the basal layer of the oral mucosal epithelium. Approximately 17% of ameloblastomas have been found to be associated with an impacted tooth or dentigerous cyst. Although this phenomenon is observed frequently in individuals below the age of 30, it occurs far less frequently in individuals in the fourth decade or older (Stanley and Diehl, 1965). This finding leads some to propose that microscopic examination of all dentigerous cysts or impacted tooth follicles is warranted.

Ameloblastomas have been classified as follows:

Peripheral (extraosseous)

Central (intraosseous)

Unicystic

Intraluminal
Mural
Intramural

Plexiorm unicystic

Multicystic or solid.

Because the various types of ameloblastoma exhibit differences in behavior that necessitate various methods of treatment, they will be discussed separately.

Peripheral ameloblastoma

Clinical features. Peripheral ameloblastoma occurs in soft tissue outside and overlying the alveolar bone. This neoplasm arises from the basal layer of the surface epithelium or remnants of the dental lamina. It occurs most frequently in the fourth to sixth decade and has a slight male predilection. The mandible is affected twice as frequently as the maxilla.

Radiographic features. Seldom does this neoplasm exhibit any radiographic findings. Superficial erosion in the alveolar region is occasionally observed.

Histology. The microscopic pattern of peripheral ameloblastoma is similar to that of central ameloblastoma; however, it lacks the invasiveness of its central counterpart. Most peripheral ameloblastomas are acanthomatous. Electron-microscopic studies show similarities

to both central ameloblastoma and basal cell carcinoma (Greer and Hammond, 1978). This lesion can be confused with peripheral odontogenic fibroma because features of both lesions may be present. However, in the hands of an experienced oral pathologist the diagnosis is generally not difficult.

Treatment. Because this lesion is relatively innocuous, noninvasive, and displays little tendency for recurrence, it is treated by local excision. Despite its behavior, 1-year follow-up examinations are recommended (Gold, 1991; Simpson, 1974).

Central ameloblastoma

Clinical features. Although this neoplasm may occur at any age, it is most frequently diagnosed in the third and fourth decades. Approximately four out of five tumors occur in the mandible, with 75% of these occurring in the molar region. Maxillary involvement is usually in the molar or antral region. The tumor generally grows slowly and causes expansion of adjacent cortical bone, usually without erosion through mucosa. It is rarely painful. Resorption of roots of adjacent teeth is a relatively common finding. Metastasis to the lung can occur as a result of implantation of tumor cells, but this is very rare.

Radiographic features. The central ameloblastoma is frequently multilocular and cystic, especially in advanced cases (Fig. 78-28). The neoplasm may be unilocular with smooth borders. Thinning of the cortex may be present. The desmplastic variant usually exhibits radiopacity.

Histology. There are six basic histologic patterns that occur in the ameloblastoma:

1. Follicular. This is the most common pattern. It is composed of multiple islands of epithelium with a peripheral layer of columnar cells, which bear a strong resemblance to ameloblasts, surrounding a central mass o polyhedral, loosely arranged cells, resembling stellate reticulum. The central region often undergoes cystic change. The lesion is unencapsulated, with infiltrating epithelial islands supported by a mature collagenous stroma (Fig. 78-29).

2. Plexiform. In this form the epithelial component is arranged in strands, bounded by strands of columnar cells between which may be found stellate reticulumlike cells. Occasionally, double rows of columnar cells are present. The stellate reticulumlike component is less prominent in this histologic pattern. Cystic degeneration is often present. The supporting stroma is usually loose and vascular (Fig. 78-30). Follicular and plexiform types frequently exist within the same tumor.

3. Basal cell. In this infrequently occurring variant, the cells are more cuboidal and may be arranged in sheets. This form bears resemblance to basal cell carcinoma of the skin.

4. Acanthomatous. The stellate reticulumlike cells undergo squamous metaplasia and sometimes produce keratin in this form. This type is usually present within the follicular ameloblastoma.

5. Granular cell. The stellate reticulumlike cells and frequently the peripheral layer undergo cytoplasmic transformation and take on a granular, eosinophilic appearance. The granules represent lysosomal aggregates. This form appears to be an aggressive lesion with numerous recurrences and reports of metastasis.

6. Desmoplastic. This form generally occurs in the anterior mandible and bears resemblance to fibroosseous disease. There is an abundant hyalinized stroma with distorted epithelial islands.

Treatment

Central unicystic. These lesions are limited to a cyst wall, cyst lumen, or connective tissue wall. This variant occurs in younger patients in the second or third decade and is usually associated with an unerupted third molar. As this is not an infiltrative lesion, it can be treated by enucleation and curettage of the associated cyst.

Plexiform unicystic. This lesion occurs in follicular cyst lining but does not exhibit the microscopic characteristics usually attributed to an ameloblastoma. Typical ameloblastoma patterns are often found elsewhere in the lesion. Because this lesion may exhibit more aggressive behavior, peripheral resection with a 3- to 5-mm margin is recommended (Gold, 1991; Himmelfarb, 1972; Sachs, 1991).

Multicystic or solid. As the recurrence rate for those lesions is greater than 55% after enucleation and curettage, it is recommended that the proper treatment should be resection with 1-cm margins. I the lesion has perforated bone, the periosteum or overlying mucosa is sacrificed. If the lesion has extended beyond the confines of periosteum, surgical margins should be assessed using frozen sections. Care should be taken to avoid entering the tumor mass during resection and rotary burrs should not be used for curettage because neoplastic cells may be seeded to other areas (MacIntosh, 1991).

Calcifying epithelial odontogenic tumor (Pindborg tumor)

Clinical features. This tumor occurs in patients over a broad age distribution with most cases occurring in middle age. Lesions occur twice as frequently in the mandible as the maxilla, with the premolar-molar area most often affected. Most cases present as an asymptomatic swelling. Although reported, extraosseous cases are rare. Usually the tumor occurs at an intraosseous site and it is associated with an impacted tooth in approximately half of the patients.

Radiographic features. This lesion may appear as either a difuse or well-circumscribed unilocular radiolucency. In some cases there will be a mixed pattern of radiolucency and radiopacity. Scattered flecks of calcification are sometimes seen within a radiolucent field.

Histology. Polyhedral epithelial cells arranged in sheets or small islands are seen. These cells have distinct borders and a granular eosinophilic cytoplasm. In some regions a homogeneous eosinophilic material that stains positive for amyloid may be present. Calcification, often in the form of concentric Liesegang rings may be produced within the

amyloidlike material (Fig. 78-31). Intercellular bridges are frequently prominent. The nuclei are often pleomorphic with multinucleated forms being relatively common, but mitotic figures are rare. The morphologic variability that is sometimes present may mimic adenocarcinoma or other malignancies.

A clear-cell variant that exhibitis a vacuolated cytoplasm is seen occasionally. This cell types may easily be confued with mucoepidermoid carcinoma or metastatic malignancy.

Treatment. This tumor is slow growing and locally invasive. However, because it has certain features in common with ameloblastoma some surgeons choose to treat it in the same fashion. There have been relatively few cases reported that have been followed up in the long term. Controversy exists as to whether to treat this lesion as aggressively as ameloblastoma; however, current therapy consists of either resection with a 1-cm margin or surgical excision with peripheral resection. The peripheral form of the lesion may be treated by local excision.

Odontogenic adenomatoid tumor (adenomatoid odontogenic tumor, adenoameloblastoma)

It is speculated that this lesion arises because of a late disturbance in odontogenesis. Some investigators consider it to be a benign neoplasm, whereas others believe it is a hamartoma or an odontogenic cyst. As the name implies, the structure contains ductlike elements.

Clinical features. Patients are typically under 20 years old and approximately two out of three lesions occur in females. About two thirds of these tumors are found in the anterior maxilla, usually associated with an impacted tooth, most frequently the maxillary canine. This tumor generally presents as a painless swelling. In rare instance, it has been reported as an extraosseous lesion.

Radiographic features. The lesion generally presents as a unilocular radiolucency that resembles a dentigerous cyst, but it extends further apically than the cementoenamel junction and may contain focal radiopacities. Although separation of roots and displacement of teeth is a frequent finding, root resorption is rare (Fig. 78-32).

Histology. This tumor is encapsulated and often appears contained within a cystic structure. It is composed of columnar or cuboidal epithelial cells in a ductlike arrangement. The ducts may contain an eosinophilic coagulum and calcifications may be present. There is a sparse connective tissue stroma. In other areas the epithelial cells are spindle-shaped or polyhedral and are arranged in nests, whorls, or cords. The tumor is structurally similar to the enamel organ (Fig. 78-33).

Treatment. Because the lesion is encapsulated and noninvasive, the treatment of choice is enucleation. Recurrence is extremely rare.

Calcifying odontogenic cyst (Gorlin's cyst: keratinizing and calcifying odontogenic cyst)

Although this lesion is called a cyst, it is now considered by most investigators to be an odontogenic tumor. This remains an area of debate because the lesion frequently contains both tumor and cystic elements.

Clinical features. Most lesions are found in the mandible and occur over a wide age range. There is generally no associated pain and the leion can occur extraosseously.

Radiographic features. A well-circumscribed radiolucency is present in the central intrabony lesion. Calcified radiopaque material is usually present within the radiolucency (Fig. 78-34). The tumor is usually small, but can become quite large, occupying much of the jaw.

Histology. The lesion may be cystic or may appear solid. A squamous, cuboidal, or columnar epithelial lining is present, within which is contained pale eosinophilic cells and "ghost" cells, which may keratinize and calcify. The eosinophilic cells and "ghost" cells may penetrate the surrounding connective tissue to stimulate a giant-cell reaction and dystrophic calcification. Stellate reticulumlike formations as well as sheets and ductlike configurations of epithelial cells may be seen. On occasion, melanin may be found.

Praetorius et al (1981) have classified the calcifying odontogenic cyst as follows:

Type I (Cystic)

A. Simple unicystic

Thin epithelial ling. Focal areas of stellate reticulum and "ghost" cells. Scant dysplastic dentin.

B. Odontoma type

Same as above, but also contains calcified tissue in its wall.

C. Ameloblastic type

Contains ameloblastomalike proliferation in cyst capsule and lumen.

Type II (Neoplastic)

Ameloblastomalike odontogenic epithelium invading connective tissue is characteristic. Ghost cells and dentinoid are present.

Treatment. All type I lesions are treated by enucleation and curettage. Extraosseous lesions are treated by local excision. Type II lesions are very rare and present an invasive and infiltrative behavior. Treatment should consist of marginal or en bloc resection, depending on the size of the lesion.

Ameloblastic fibroma

This is an uncommon neoplasm arising from simultaneous proliferation of both epithelial and mesenchymal tissue without formation of calcified tissue.

Clinical features. This lesion most commonly arises in the molar region of the mandible. Forty percent of the patients are under the age of 10, with a mean age at diagnosis of 14.6 years (Small, 1955). It is a slow-growing lesion that does not tend to infiltrate and rarely causes pain.

Radiographic features. The appearance of ameloblastic fibroma is very similar to that of ameloblastoma. It is of variable size, is usually unilocular but may be multilocular, and has a smooth outline. It is frequently associated with unerupted teeth.

Histology. Scattered islands of columnar or cuboidal epithelial cells are arranged in nests, cords, and strands similar to primitive odontogenic epithelium. Occasionally, stellate reticulumlike tissue is present. The surrounding connective tissue resembles dental papilla.

Treatment. Treatment of this lesion consists of enucleation and curettage. There is little tendency for recurrence. However, patients should continue to be followed on an annual basis because causes of ameloblastic fibrosarcoma have originated from recurrent ameloblastic fibroma (Leider et al, 1972).

Ameloblastic fibrodontoma

This lesion represents an immature form of complex odontoma and therefore represents a hamartoma rather than a neoplastic process.

Clinical features. Most patients are under the age of 15 and male. The maxilla and mandible are affected with equal frequency. The lesion is most often associated with an impacted tooth in the molar region. The most common complaints are swelling and failure of tooth eruption.

Radiographic features. This is a well-circumscribed expansile radiolucency containing a single or multiple radiopaque masses. The lesion's size is extremely variable.

Histology. The microscopic picture is a combination of that found in ameloblastic fibroma and composite odontoma.

Treatment. This lesion is treated by enucleation and curettage because it has little tendency for recurrence.

Ameloblastic odontoma (odontoameloblastoma)

This is an extremely rare lesion that consists of ameloblastoma and composite odontoma. It is unusual in that it consists of a relatively undifferentiated neoplastic tissue associated with a highly differentiated tissue. Recurrence may occur with inadequate removal.

Clinical features. This lesion tends to occur at any age but is more frequent in children. It is more often found in the mandible than the maxilla. Presenting symptoms include mild pain and delayed eruption of teeth.

Radiographic features. Central destruction of bone with expansion of the cortical plates is common. Many small radiopaque masses are present within the lesion.

Histology. There is a great variety of cells in a complex distribution. There are many structures resembling normal or atypical tooth germ. Sheets of typical ameloblastoma are present.

Treatment. Management of this lesion is controversial because there are so few cases. The general behavior is the same as for ameloblastoma so marginal or en bloc resection is suggested, depending on the size of the lesion.

Odontoma

Odontoma is a hamartoma derived from functional ameloblasts and odontoblasts and forms enamel and dentin in an abnormal pattern. It is frequently called *composite* because it contains more than one type of tissue. If the enamel and dentin form structures that bear resemblance to normal teeth, it is called a *compound composite odontoma*. If the dental hard tissues form a more disorganized mass, it is termed *complex composite odontoma.* The compound type is more common.

Clinical features. Odontomas are found in patients of all ages and in all locations within the jaws. These lesions are frequently associated with dentigerous cysts.

Radiographic features. This lesion appears as an irregular radiopaque mass surrounded by a thin radiolucent region. It is often located between tooth roots. Frequently, odontomas are associated with unerupted teeth. Compound composite odontomas may have the appearance of a mass of toothlike structures (Fig. 78-35).

Histology. The microscopic appearance is that of enamel, dentin, pulp, and connective tissue, similar to a dental follicle (Fig. 78-36). "Ghost" cells may also appear in this lesion (as seen in the calcifying odontogenic cyst). This lesion should not be confused with ameloblastic fibroodontoma.

Treatment. Because there is no tendency for recurrence, odontomas are treated by simple enucleation.

Mesenchymal odontogenic tumors

Odontogenic fibroma

This is a rare lesion composed of connective tissue and odontogenic islands and resembles a dental follicle. It appears to arise from periodontal ligament, dental papilla, or dental follicle. It occurs around the crown of unerupted teeth and appears much like a follicular cyst.

Clinical features. This tumor occurs most frequently in the mandibles of children, adolescents, and young adults. It exhibits slow, expansile growth and rarely produces pain.

Radiographic features. A unilocular or multilocular radiolucency with smooth borders is common.

Histology. The lesion contains collagen fibers and fibroblasts with occasional osteoid, dysplastic dentin, or cemental tissue. Its appearance must be distinguished from the Antony B type configuration found in neurofibromas.

Treatment. Enucleation is the treatment of choice, since recurrence has not been reported.

Odontogenic myxoma

This tumor also has a mesenchymal origin, arising from periodontal ligament, dental follicle, or dental papilla.

Clinical features. It occurs most often in the second and third decades and slightly more frequently in the mandible than in the maxilla. It is often associated with missing or impacted teeth. This is a slow-growing, expansile lesion.

Radiographic features. A multilocular radiolucency is often present although it may present as a mottled unilocular radiolucency. Displacement of teeth is common, but root resorption is rare. The lesion tends to erode through cortical bone and frequently involves the maxillary sinus if it occurs in the upper jaw.

Histology. There are few cells present in this tumor, but there is a prominent mucoid intercellular substance. The cells are stellate or spindle shaped with long fibrils. Occasional nests of odontogenic epithelium or collagen fibers may be seen. Hyaluronic acid and chondroitin sulfate are produced by this lesion (Fig. 78-37).

Treatment. Although this tumor is benign, it behaves rather aggressively. Because of its invasive nature, this lesion is frequently quite large by the time it is diagnosed. The treatment of choice is either marginal or en bloc resection with 1-cm margin. It is not responsive to radiotherapy.

Cementoma

Cementoma is the broad classification for lesions that have in common the production of cementum. All cementomas are benign; however, treatment will vary depending on the specific type.

Periapical cementoosseous dysplasia

This lesion appears to arise from the periodontal ligament and contains various amounts of fibrous tissue, cementum, and bone. Although it occurs quite frequently, its etiology has not been fully explained.

Clinical features. Patients are predominantly black females over the age of 20. In most cases, multiple lesions are present that are asymptomatic and usually involve the mandibular incisor root apices.

Radiographic features. This lesion passes through three tages in its maturation. The osteolytic stage occurs first and is characterized by localized dental periapical radiolucencies similar in appearance to those that occur with a dental abscess. The next period is termed the *cementoblastic stage.* During this time cementoblasts become more active and produce spicules of cementum, which produce a mixed radiolucent/radiopaque appearance. The final or mature stage consists of an abnormally large amount of calcification that appears as a dense periapical radiopacity surrounded by a thin radiolucent border (Fig. 78-38).

Histologic description. This lesion contains varying amounts of fibrous connective tissue, cementoblasts, and cemental tissue depending on the stage of the lesion.

Treatment. Periodic radiographic observation is appropriate. The teeth are vital and should not be treated by extraction or endodontic therapy. Electrical, thermal, and mechanical stimulation of the teeth can aid the clinician who is attempting to rule out dental infection during the osteolytic or cementoblastic stages.

Cementifying fibroma or cementoosiffying fibroma

This lesion closely resembles the ossifying fibroma and it probably arises from the same progenitor cell.

Clinical features. It occurs most often in young and middle-aged adults, more frequently in the mandible, and there is a marked femaled predilection. This is a slow-growing expansile tumor that can displace teeth and produce swelling, but rarely causes pain.

Radiographic features. The radiographic appearance of this lesion changes depending on its state of maturation. Independent of the stage of development, the lesion will always appear well circumscribed. Initially it will appear as a radiolucency and will later pass through a stage in which flecks of calcification are present. Finally it will become densely radiopaque. This tumor exhibits a centrifugal growth pattern, such that the lesion is always found to be round.

Histology. Collagen fibers and fibroblasts or cementoblasts as well as small masses of calcified tissue are present. The name of the lesion is dependent on the type(s) of calcified tissue present.

Treatment. This lesion is benign, noninvasive, and has demonstrated no propensity for recurrence. It is best treated by simple enucleation.

Benign cementoblastoma (true cementoma)

This relatively uncommon lesion probably represents a benign neoplasm of cementoblasts that produces a large mass of cemental tissue at the tooth root.

Clinical features. Patients are usually under 25 years old, but the age range extends into the eight decade. The mandible is more often affected than the maxilla, with the mandibular permanent first molar being the most frequent site of involvement. The involved tooth is vital and usually pain free. The slow growth of this lesion may result in cortical expansion.

Radiographic features. A well-circumscribed dense radiopacity is attached to the tooth root and is often surrounded by a narrow radiolucent line. The affected root is frequently obscured by the mass secondary to root resorption and fusion of the tumor to the rooth.

Histology. Sheets of cementumlike tissue surrounded by cementoblasts are a prominent feature. Reversal lines are sometimes present, as are cementoblasts. Fibrovascular tissue is present to a varying degree. The periodontal ligament is obliterated and the tooth root may show resorption. A soft-tissue capsule may be seen at the periphery with cemental trabeculae positioned at right angles. The lesion may resemble a benign osteoblastoma, osteoid osteoma, or osteosarcoma.

Treatment. Removal of the affected tooth is indicated. The dental extraction can usually be performed easily because there is cortical thinning secondary to expansion of the lesion. This tumor must be distinguished from hypercementosis or chronic sclerosing osteomyelitis. Benign cementoblastoma do not recur after treatment.

Dentinoma

This is an extremely rare tumor composed of connective tissue, odontogenic epithelium, and abnormal dentin.

Clinical features. This lesion occurs in the mandibular molar region in young adults. It is often associated with an impacted tooth; however, extraosseous cases can occur. Pain, swelling, and mucosal perforation have been reported.

Radiographic features. The radiographic picture may be extremely variable. It may appear as a radiolucency, a radiolucency with small radiopaque flecks, or a solitary radiopaque mass.

Histology. The connective tissue stroma resembles dental papilla. Masses of irregular dentin with demonstrable dentinal tubules are present. Undifferentiated odontogenic epithelium is present and enamel is absent. If enamel were present the lesion would be called a *complex composite odontoma.*

Treatment. This lesion is treated by enucleation and curettage.

Tumors of unknown origin

Melanotic neuroectodermal tumor of infancy

This appears to be a tumor of neural crest origin. High urinary excretion of vanillylmandelic acid is consistent with other neural crest tumors.

Clinical features. This lesion usually arises in the anterior maxillary region in children under 6 months of age. The lesion is not present at birth. The tumor is benign and has exhibited little tendency to recur; however, it does grow rapidly. It has a darkly pigmented appearance and does not ulcerate (Fig. 78-39).

Radiographic features. It presents as a ragged radiolucent lesion that resembles a malignancy. The primary central incisor is carried with the lesion when it occurs in the anterior maxilla.

Histology. The tumor is nonencapsulated and infiltrative. Cuboidal cells, many with melanin pigmentation, form alveoli that contain neuroblastlike cells. These cells have prominent round nuclei with little cytoplasm. The stroma consists of vascular and fibrous tissue.

Treatment. This lesion is treated by local excision with curettage. It may be confused with a neuroblastoma or congenital epulis of the newborn.

Malignant odontogenic tumors

Primary intraosseous carcinoma

This is an extremely rare tumor. The diagnosis is often made after metastasis has occurred. Primary intraosseous carcinomas may be of three different types (Elzay, 1982):

1. Arising from an odontogenic cyst.

2. Developing from an ameloblastoma.

Well differentiated (malignant ameloblastoma).

Poorly differentiated (ameloblastic carcinoma).

3. Arising from odontogenic epithelium.

Clinical features. Aggressive local invasion with destruction of adjacent tissues is the major feature. Regional and distant metastasis will occur.

Radiographic features. The lesion will present as a radiolucency with poorly defined margins. Destruction of bone and tooth root structure may occur.

Histology. This is highly dependent upon the tissue from which the tumor arises. However, invasiveness, cellular pleomorphism, atypical mitotic figures, and large hyperchromatic nuclei will be present.

Treatment. Resection of the tumor with 1-cm margins and frozen sections is generally indicated. Radiation therapy is usually recommended. Treatment of metastases may involve surgical removal, radiation, and/or chemotherapy.

Ameloblastic fibrosarcoma (ameloblastic sarcoma)

This is a very rare tumor and is the malignant counterpart of the ameloblastic fibroma.

Clinical features. It occurs more frequently in young adults and is more common in the mandible than in the maxilla. The tumor is generally painful, exhibits rapid growth and destroys bone. Teeth may become loose and there may be ulceration of the overlying mucosa with associated bleeding.

Radiographic features. The borders of the lesion are poorly defined and local bone destruction is present.

Histology. Most cases have occurred through malignant transformation of a preexisting ameloblastic fibroma. The odontogenic epithelium does not undergo malignant conversion. However, the mesenchymal portion displays increased cellularity, cellular pleomorphism, hyperchromatic nuclei, and atypical mitotic figures.

Treatment. Wide surgical resection is indicated. The recurrence rate is very high. Radiation therapy is not recommended because it has not proven to be effective in management of this tumor.

Ameloblastic dentinosarcoma

This tumor is similar to the ameloblastic fibrosarcoma and ameloblastic dentinosarcoma except that it contains both dysplastic enamel and dentin.

Treatment. Wide urgical resection is necessary because recurrences are frequent. The lesion is poorly responsive to radiation.

Surgical Management of Odontogenic Cysts and Tumors

Surgical goals

The vast majority of odontogenic lesions are benign and most are only minimally aggressive. This makes it crucial to define carefully the ultimate goal of surgery. Unlike malignant lesions where the primary goal is preservation of life, with other factors being of secondary importance, the treatment of odontogenic cysts and tumors must not be accomplished at the expense of function or esthetics.

The goals of management of benign odontogenic cysts and tumors are (Cherrick, 1985): (1) remove all abnormal tissue, (2) conserve healthy bone and dental structures, (3) preserve adjacent structures such as the inferior alveolar nerve, (4) restore the surgical defect to its presurgical state of anatomic form and function, and (5) prevent recurrence of the lesion.

Principles in surgical management of odontogenic cysts and tumors

Cysts and tumors of the jaws are treated in a variety of ways depending on several factors. Virtually all cysts are treated with either enucleation or enucleation and curettage. Occasionally, decompression or marsupialization can be done first to allow the lesion to decrease in size before it is removed definitively. Odontogenic tumors are treated by enucleation with or without curettage, by marginal or partial resection, and occasionally by composite resection (see box).

Box (p. 1439) **Surgical modalities of treatment of odontogenic cysts and tumors**

Enucleation and curettage

Odontogenic cysts:

Virtually all unless recurrent

Odontogenic tumors:

Odontoma
Ameloblastic fibroma
Ameloblastic fibroodontoma
Adenomatoid odontogenic tumor
Calcifying odontogenic cyst
Cementoblastoma
Central cementifying fibroma

Marginal or partial resection

Odontogenic cysts:

Recurrent odontogenic keratocyst

Odontogenic tumors:

Ameloblastoma
Calcifying epithelial odontogenic tumor (Pindborg tumor)
Odontogenic myxoma
Ameloblastic odontoma
Squamous odontogenic tumor&

Composite resection

Odontogenic tumors:

Malignant ameloblastoma
Ameloblastic fibrosarcoma
Ameloblastic odontosarcoma
Primary intraosseous carcinoma.

& These lesions are malignancies and may be treated variably and with additional modalities such as radiation or chemotherapy.

A definition of terms is necessary before proceeding. Enucleation of a lesion involves local removal of the lesion by instrumentation in direct contact with the lesion (Fig. 78-40). If curettage is also used, 1 to 2 mm of the bony wall is removed after the lesion is enucleated (Fig. 78-41). This is accomplished with aggressive curettage by hand or by the use of rotary instrumentation. Resection involves incision or osteotomy through uninvolved tissue adjacent to the lesion without disruption of the lesion. Marginal resection does not produce a continuity defect of the involved bone, for example, leaving a portion of the inferior border of the mandible intact (Fig. 78-42). Partial resection removes a full-thickness portion of the involved bone. In the maxilla this may not differ much from a marginal resection; however, in the mandible, it produces a continuity defect (Fig. 78-43). Composite resection involves removal of tumor, adjacent bone, soft tissue, and contiguous lymph node channels. The only odontogenic lesions treated with composite resection are malignant neoplasms.

The decision as to which method of treatment to use for the management of a given odontogenic lesion is based primarily on the histologic diagnosis because it provides the most information about the lesion's aggressiveness and its potential for recurrence. Other factors may play a role in helping to decide between a more or less aggressive therapy. These factors include the anatomic location of the lesion, its proximity to adjacent vital structures, the size of the lesion, its spread to adjacent soft tissue, the duration of the lesion, and the plans for reconstruction.

Lesions in the mandible are more readily diagnosed, are usually smaller at the time of diagnosis, and are more easily followed for recurrence than their counterparts in the maxilla. Lesions in the anterior of either the maxilla or mandible are easier to diagnose, treat, and follow than their counterparts in the posterior of each respective jaw. Because of this, a more aggressive approach is often recommended for lesions in the posterior maxilla than for those in the anterior mandible.

Proximity of adjacent vital structures should not deter adequate removal of a lesion. It may be possible to decompress a cyst to allow some bony fill before enucleation in an attempt to preserve adjacent teeth and nerves (Fig. 78-44). This decompression should be distinguished from marsupialization where no follow-up enucleation is accomplished. Marsupialization as treatment alone is not recommended because it requires a very compliant patient, does not allow histopathologic diagnosis of the entire lesion, and may leave remnants of tissue behind, leading to recurrence. This type of treatment is not applicable for odontogenic tumors.

Extremely large lesions may require more aggressive treatment than smaller lesions of the same histologic type. Large lesions are more difficult to remove completely with enucleation alone. They may be large because they are more aggressive in behavior or because diagnosis was delayed because of poor patient reliability or difficulty in detection. Whichever the case, a more aggressive approach may be indicated to prevent recurrence.

Virtually all odontogenic cysts and tumors originate within bone. Aggressive lesions such as ameloblastomas and odontogenic keratocysts may perforate the cortical plate that usually surrounds the intraosseous lesion, enabling communication with the adjacent soft tissue. This is associated with increased difficulty in total removal (Fig. 78-43, B). Although these lesions do not usually spread within the soft tissue and tend to remain encapsulated by the periosteum, the likelihood of removing the lesion incompletely is greater.

Several odontogenic tumors exhibit very slow growth, eventually reaching a static point in size. These lesions should be treated less aggressively than lesions that have exhibited rapid enlargement. Regardless of the treatment used, the attention to restoration of esthetics and function should be considered before treatment is begun. Preservation of a portion of the condyle and ramus can greatly improve reconstruction, as can maintenance of the mandible's continuity and range of motion during the interval between tumor removal and reconstruction.