Panoramic radiograph in pathology

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The panoramic radiograph is an excellent extraoral radiograph to reveal abnormalities and pathologic conditions of the jaws and to show calcification of adjacent soft tissue. A single panoramic radiograph provides a significant image of the maxillofacial skeleton and dentition and often is used as a survey or screening film because of the area it covers. The panoramic radiograph does not provide fine radiographic detail, however, and often is followed by intraoral radiographs that can provide sharper detail of an abnormal finding. Computed tomography (CT) and magnetic resonance imaging (MRI) often are utilized to further delineate the extent of a pathologic process found on panoramic radiograph.

DEVELOPMENTAL DEFECTS

Exostosis/torus

Frequency/incidence
The incidence of exostoses of the jaws is approximately 27% [1]. The prevalence and number of exostoses increase with age, and exostoses are more common in men than women. Palatal tori, however, are more common in women. Tori are large exostoses of the midline of the hard palate and the lingual surface of the mandible.

Signs and symptoms
Exostoses and tori are localized outgrowths or nodules on cortical bone, which usually are asymptomatic [2]. They may interfere with normal speech if excessively large and may become inflamed and painful if traumatized.

Etiology/pathophysiology
These outgrowths of bone are composed of dense cortical bone but may contain medullary bone. Several theories have been proposed concerning the development of exostoses that implicate genetic and environmental factors, including masticatory stress [3].

Image of choice for diagnosis
Exostoses and tori may be depicted on panoramic and periapical radiographs depending on the size and density of the structures. The diagnosis of exostosis primarily is a clinical diagnosis, which may be confirmed by the radiograph.

Image hallmark
If an exostosis is of sufficient size and density, it may appear as a well-defined radiopacity on the radiograph in the area of clinical change (Fig. 1).
Management

No treatment usually is necessary; however, exostoses and tori are removed when a prosthesis is placed on the involved mucosa, when they are chronically inflamed and painful, and when they interfere with normal speech.

Condylar hyperplasia

Frequency/incidence

Condylar hyperplasia is a developmental abnormality that results in enlargement and occasionally deformity of the condylar head. This is an extremely rare condition with an incidence of less than 1% [4].

Signs and symptoms

The clinical features include asymmetry of the lower third of the face, deviation of the mandible and chin away from the affected side, and compensatory vertical growth of the maxilla on the affected side. Patients may also have significant temporomandibular joint symptoms, such as pain, joint noises, and diminished mouth opening. The mandibular midline also may be shifted, creating an anterior lateral crossbite with class III molar and canine occlusion on the affected side [5–7].

In addition, the height of the ramus will be increased on the affected side.

Etiology/pathophysiology

The cause of condylar hyperplasia is unknown but may result from circulatory abnormalities, previous trauma, hormonal disturbances, abnormal joint loading, genetic alterations, and as representing a cartilaginous exostosis [5–7].

Image of choice for diagnosis

The panoramic radiograph is the initial screening film of choice. Further enhanced images include conventional CT and MRI.
The condyle may appear relatively normal in shape but enlarged (Fig. 2). It also may exhibit alteration in shape and be more radiopaque. The ramus and mandibular body on the affected side may also be enlarged, resulting in a characteristic depression of the inferior border of the mandible where the enlarged side joins with the contralateral normal side. In addition, the condylar neck is classically elongated on the ipsilateral side [8,9].

Management

Treatment decisions are primarily based on the age of the patient [8]. Treatment consists of a combination of orthodontics and orthognathic surgery to correct the skeletal deformity and any dental malocclusion. Successful treatment has been reported with high condylectomy [7].

Calcified stylohyoid ligament

Frequency/incidence

Approximately 2% to 18% of the general population present radiographic evidence of ossification of the stylohyoid ligament [10]. There appears to be a slight predominance for women, and symptoms are more common in patients aged 40 and older.

Signs and symptoms

Most patients are asymptomatic; however, when symptoms occur, there is no correlation between the extent of the calcification and symptoms present. Eagle syndrome is a term used to describe the cluster of symptoms experienced. The symptoms include pain in the peritonsillar region and base of the tongue area posterior to the angle of the mandible, dysphagia and otalgia. Eagle syndrome previously was reported only in patients post-tonsillectomy. Now it is applied to patients meeting the criteria of the previously described symptoms, with elongated, calcified stylohyoid ligaments [10–12]. Some patients may exhibit syncope when they quickly turn their heads because the rigid ligament compresses the carotid artery and cervical sympathetic chain (carotid artery syndrome) [12]. There also have been reports of cervical spine disease resulting in difficult intubations in patients with calcified stylohyoid ligaments [13].

Etiology/pathophysiology

The stylohyoid ligament is a fibrous sheath that persists as a part of the ceratohyal element of the second branchial arch, extending from the base of the skull to the lesser horn of the hyoid bone and passing between the internal and external carotid arteries [14]. For unknown reasons, this ligamentous structure may undergo ossification, especially in the upper part of the ligament.

Image of choice for diagnosis

The panoramic radiograph is the image of choice for visualization. The ligament also may be seen on a routine skull series and cervical spine radiographs.
Calcified stylohyoid ligament presents as a radiopaque change along the course of the structure in which the calcification may vary in density and length (Fig. 3).

Management
Treatment consists of surgical removal of the affected ligament as close to the cranial base as possible in patients with significant symptoms [11]. This can be accomplished through either an intraoral or extraoral approach, although the extraoral approach is preferred because of better access and visualization.

Nasopalatine duct cyst

Frequency Incidence
The nasopalatine duct cyst is the most common nonodontogenic cyst of the jaws with a prevalence of approximately 1% [15,16]. It is most frequently found in patients between the ages of 40 and 60 years, with a slight male predilection [17].

Signs and symptoms
Most patients are asymptomatic; however, pain, swelling, drainage, and movement of teeth may be noted. A large cyst may present with palatal swelling in which the cortical plate has been perforated and drainage of fluid or pus ensues.

Etiology/Pathophysiology
The cause of the cyst is unknown, but some authors have attributed its development to trauma, infection, and spontaneous proliferation of residual embryonal epithelium as possible causes [15–18].

Image of choice for diagnosis
The panoramic radiograph is the initial screening film of choice, followed by periapical radiographs for definitive visualization.

Image hallmarks
The classic presentation is a symmetrical, ovoid or heart-shaped radiolucency between the roots or at the apices of vital maxillary central incisors (Fig. 4) [19].

Management
Thorough surgical curettage is the treatment of choice for symptomatic lesions or lesions measuring more than 6 mm in diameter. Recurrence after surgical removal is reported to
be approximately 2% [19]. Several other cystic lesions may mimic a nasopalatine duct cyst. Differential diagnosis includes an apical periodontal cyst, lateral periodontal cyst, and odonto-genic keratocyst [20].

INFLAMMATORY DISEASES

Submandibular salivary gland cortical defect

Frequency/incidence

In 1942, Stafne first described 34 cases of this radiographic abnormality just anterior to the angle of the mandible [21]. The prevalence rate has been reported to be approximately 0.05%, with a slight male predominance [22].
Signs and symptoms
This defect usually is asymptomatic and diagnosed on routine panoramic radiograph. When symptoms occur, they consist of pain and swelling in the area of the submandibular gland. There is no cortical expansion.

Etiology/pathophysiology
The cause of this cortical defect is unknown; however, various causes have been proposed, including stress alteration, pressure atrophy from an inflamed gland, and a congenital defect [22]. The contents of the defect contain submandibular salivary gland elements, fat, and connective tissue [23].

Image of choice for diagnosis
The panoramic radiograph is the initial screening film of choice. A lateral jaw film also can detect the defect, and the diagnosis can be confirmed with CT, MRI, and a sialogram of the submandibular gland that shows salivary gland contents in the cortical depression or defect.

Image hallmarks
The classic presentation is a well-defined radiolucency near the angle of the mandible below the inferior alveolar canal (Fig. 5). Most of these defects exhibit a hyperostotic border, and most cases are unilateral. A similar lingual cortical defect can be associated with the sublingual gland, although it is quite rare [24].

Management
There is no treatment for the cortical defect; however, a symptomatic submandibular gland may require removal.

Periapical inflammatory disease

Frequency/incidence
Inflammatory disease of pulp with periapical bone involvement is common, with incidence rates of 40% to 70% [25,26]. The most common location for periapical involvement is the maxillary anterior region, followed by posterior maxilla, the posterior mandible, and the anterior mandible [27].

Signs and symptoms
Patients with periapical inflammatory disease may be asymptomatic or present with pain, swelling, drainage, and possibly fever and regional lymphadenopathy. The associated teeth also may be painful and tender to palpation and percussion. The involved teeth are nonvital.

Etiology/pathophysiology
Inflammatory periapical bone disease is initiated by the presence of micro-organisms in the apical portion of the root canal of the involved tooth [25]. The infection and resultant inflam-
matory response may spread to periapical bone, producing a variety of pathologic conditions, including inflamed granulation tissue (periapical granuloma, chronic apical periodontitis), abscess, and apical periodontal cyst (radicular cyst). The development of inflamed granulation tissue or abscess depends on the virulence of the involved organisms and extent of the infection. Periapical granulation tissue may continue to destroy bone and ultimately destroy cortical bone. Fistula formation may occur. An abscess may perforate through cortical plates and create a soft tissue space infection, which may create significant morbidity. Activation of residual odontogenic epithelium in the periodontal membrane may result in cyst formation. The cyst may expand and destroy a significant amount of bone. The cyst may be asymptomatic, or the patient may complain of pain. In a review of 256 periapical lesions, Nair et al found that the majority (55%) were granulomas and the remaining lesions (45%) were periapical cysts or abscesses [25].

*Image of choice for diagnosis*

The panoramic radiograph is the initial screening film of choice, followed by periapical radiographs for further delineation of the lesion.

*Image hallmarks*

The classic presentation is a radiolucency at the apex of a nonvital tooth (Fig. 6). The size and extent of the lesion varies considerably, with extension laterally and inferiorly from the apex. A periapical abscess may show only minimal widening of the periodontal membrane of the involved tooth.

*Management*

Treatment for periapical inflammatory disease involves either extraction of the involved tooth or root canal therapy with the appropriate use of antibiotics if the clinical situation warrants it. In a follow-up study after root canal treatment, approximately 90% of the lesions resolved and were not detectable on radiographs [28]. The treatment for unresolved lesions is surgical removal of the involved tissue and submission of the tissue for microscopic examination. The distinction between periapical granuloma and apical periodontal cyst can only be made by microscopic examination [29].
Periodontal disease

Frequency/incidence
Periodontal disease is the leading cause of tooth loss in patients over the age of 35 years. There is an increased prevalence associated with increased age, diabetes, smoking, and limited oral hygiene [30]. To some degree, periodontal disease affects most individuals at some point in their lifetime. Although periodontitis typically is a disease of adulthood, aggressive forms, termed early-onset periodontitis, may occur in children [31].

Signs and symptoms
The typical sign of periodontal disease is erythematous and edematous gingiva that bleeds with tissue manipulation. The normal stippling is not present and the gingiva also may be blunted or apically positioned, depending on the degree of tissue involvement [32]. Patients may report bleeding with brushing and gingival pain, although these are not consistent symptoms.

Etiology/pathophysiology
Periodontal disease primarily is caused by bacteria that colonize the gingival crevice. The resultant bacterial products and inflammation ultimately cause destruction of supporting alveolar bone, which leads to loss of bone support for the teeth. Gingival inflammation and periodontal bone loss may be localized to several teeth or generalized. Tissue pockets of more than 4 mm are indicative of an active disease process.

Image of choice for diagnosis
The panoramic radiograph is an excellent initial screening tool and is followed by periapical radiographs, which better define the amount of alveolar bone loss.

Image hallmarks
Gingival inflammation usually does not show alveolar bone loss. When the inflammatory response involves alveolar bone, gradual destruction of the interdental bone takes place. The amount of loss of bone that supports the teeth indicates the severity of the disease process (Fig. 7).

Management
Treatment depends on the severity of the symptoms and tissue destruction. It consists of a combination of oral hygiene procedures performed by the patient, mechanical debridement, surgical removal of diseased tissue, and topical antimicrobial agents.
Osteomyelitis

Frequency/incidence

Osteomyelitis of the jaws was once a frequently encountered disease and was difficult to treat. Now it is much less common because of the improvement in dental and medical health care and the use of antibiotics [33]. Osteomyelitis is more common in the mandible because of its relative avascularity when compared with the maxilla.

Signs and symptoms

Patients often present with fever, malaise, edema of the overlying soft tissue, lymphadenopathy, purulent discharge, pain, and leukocytosis [34].

Etiology/pathophysiology

Osteomyelitis is an infectious process within the marrow spaces of bone. In the face, the angle and body of the mandible is the most commonly infected site. Most cases are the result of a bacterial dental infection. Infected mandibular fractures are the second most common cause of osteomyelitis [35]. Hematogenous spread also is responsible for cases of facial osteomyelitis. The cases may be acute or chronic and suppurrative or nonsuppurrative in nature. Diagnosis is based on clinical, histopathologic, and laboratory studies.

Image of choice for diagnosis

The panoramic radiograph serves as an initial screening tool. In many cases, CT bone scintograms and MRI are superior to conventional radiographs in delineating the extent of the infectious process. Bone scintography studies with technetium 99 are abnormal in 90% to 95% of patients with osteomyelitis [34].

Image hallmarks

In the early stages, conventional radiographic images may fail to show any abnormalities; however, areas of ill-defined radiolucencies with or without radiopaque sequestrum and reactive bone soon develop. The radiographic changes may exhibit a mottled or “moth-eaten” pattern and mimic malignancy (Fig. 8). The diagnosis of osteomyelitis is based on clinical and radiographic findings.
Management
Treatment consists of prompt aggressive surgical debridement of all infected bone down to healthy bleeding tissue, followed by long-term intravenous or oral antibiotics for 6 to 12 weeks with high-dose aqueous penicillin G (12–20 million U/day) or clindamycin (2700 mg/day) if the patient is allergic to penicillin [36]. The patient must be closely followed clinically and radiographically for resolution of the infection. Bony defects that result from the infection may require further surgical procedures.

Proliferative periostitis (periostitis ossificans)

Frequency/incidence
Proliferative periostitis is a relatively rare, specific type of chronic osteomyelitis that occurs almost exclusively in children and young adults [37]. The average age of onset is 11 years with a male-to-female ratio of 1.4 to 1 [38].

Signs and symptoms
Patients most commonly present with symptoms related to their primary infection and a painless swelling of bone of the affected area. The most common location is the posterior body of the mandible with an associated carious molar or premolar. The swelling may be of insidious onset with only minimal pain over the involved area [39].

Etiology/pathophysiology
In most cases, the infected focus resulting in the periosteal hyperplasia represents a carious tooth, periodontal disease, or an infected extraction site. There have been reports of proliferative periostitis resulting from an infected dentigerous cyst [40].

Image of choice for diagnosis
Panoramic and lateral oblique radiographs are appropriate films to establish the diagnosis. If these images do not show the characteristic changes, occlusal and posteroanterior radiographs may be helpful.
Image hallmarks
The classic radiographic appearance shows reduplication of cortical bone with a lamellar or layering effect (“onionskin” appearance) stimulated by the underlying inflammatory process (Fig. 9) [41].

Management
Treatment involves elimination of the source of infection. The cortical enlargement usually undergoes resolution within 6 to 12 months. The presence of proliferative periostitis in the absence of an inflammatory process requires biopsy of the involved area because processes such as Ewing sarcoma, Langerhans cell histiocytosis, and metastatic disease can induce this reaction.

Antral pseudocyst

Frequency/incidence
The antral pseudocyst is a relatively common finding on routine panoramic radiographs [42]. Reports indicate that 1.5% to 10% of the population have these on routine radiographic evaluation [43].

Signs and symptoms
Patients are usually asymptomatic unless a localized infection is present [44]. These latter patients exhibit symptoms of maxillary sinusitis including fever, nasal discharge, headache, and pain.

Etiology/pathophysiology
The antral pseudocyst represents an accumulation of fluid between antral mucosa and the floor of the sinus. The cause is unknown but some cases may represent the extension of an inflammatory exudate from an infected maxillary tooth. The terms sinus mucocele and retention cyst have been used for this entity; however, there is no accumulation of mucous or true cyst lining in these lesions, and they should not be confused with the other, more aggressive lesions.

Fig. 10. Antral pseudocyst. (Courtesy of Dr. Dan Trinler, Lexington, KY.)
Image of choice for diagnosis

The panoramic radiograph is the diagnostic film of choice. A water’s view image and CT may be useful if the diagnosis is unclear on panoramic radiograph.

Image hallmarks

The classic radiographic presentation is a “domed-shaped,” finely radiopaque lesion with its base on the antral floor (Fig. 10). In rare instances, the entire sinus may be involved [45]; however, other space-occupying lesions of the sinus must be ruled out when it appears in this fashion.

Management

The lesion often regresses on its own and no treatment is necessary in most cases. If symptoms are present, either transantral or endoscopic techniques may be used for removal of the lesion [42].

Sialolith

Frequency/incidence

Salivary calculi (sialoliths) are a common cause of salivary gland dysfunction and may occur in any of the salivary glands and at almost any age. They occur most commonly in the submandibular gland duct (83%) and involve the parotid gland duct in 10% of cases. Minor salivary gland duct and sublingual gland involvement represents the remaining 7% of cases [46]. Occasionally, the salivary gland itself may be involved. The incidence of sialolithiasis is approximately 1% in the general population [47].

Signs and symptoms

Some calculi remain asymptomatic; however, most calculi obstruct salivary flow to the point that the involved gland eventually becomes enlarged and painful. The gland often becomes tense and painful during or after a meal, which has stimulated the gland to produce and secrete saliva. A palpable mass may be present depending on the location of the stone within the duct, and attempts to express saliva from the involved duct may produce minimal or no flow. These symptoms are primarily related to the submandibular and parotid gland systems. Minor gland sialolithiasis usually manifests as a small nodular mass with possible localized tenderness. If the involved gland has become secondarily infected, purulent drainage may be seen at the meatus of the duct.

Etiology/pathophysiology

The mechanism behind the development of salivary calculi is not well understood but has been attributed to mucous stasis within a duct followed by the deposition of calcium salts in

Fig. 11. Sialolith. (Courtesy of Dr. Denise Clarke, Port Angeles, WA.)
a nidus, leading to the development of a calcified mass. In most cases, the sialolith develops within the duct of the affected gland. Approximately 90% of submandibular stones are situated in the distal portion of the Wharton duct or at the hilum [48].

*Image of choice for diagnosis*

The panoramic radiograph is an excellent screening film to detect submandibular and parotid calculi. This image may be coupled with an occlusal radiograph to determine the location of the sialolith in the involved duct.

*Image hallmarks*

Sialoliths appear as radiopaque, ovoid to linear structures often superimposed on normal radiographic anatomy (Fig. 11). They rarely develop in the gland itself.

*Management*

Treatment usually involves surgical removal of the sialolith from the duct; however, other modalities of treatment include laser therapy, extracorporeal electromagnetic shock-wave lithotripsy and catheter retrieval under fluoroscopy [49]. If chronic sialadenitis is present, the entire gland usually requires removal. Appropriate antibiotic therapy is indicated if purulent drainage is present.

**LOCALIZED METABOLIC DISORDERS**

**Idiopathic osteosclerosis**

*Frequency/incidence*

The incidence of osteosclerosis of the jaws is 4% to 31% [50]. Approximately 90% of osteosclerosis cases occur in the posterior mandible, with an age predilection between 20 to 40 years. Osteosclerosis shows equal sex distribution [50–52].
Signs and symptoms
Osteosclerosis is an asymptomatic, localized area of increased density of bone without expansion, which is discovered during routine radiographic examination.

Etiology/pathophysiology
The cause of idiopathic osteosclerosis is unknown. It is not associated with inflammatory and neoplastic processes [50–52]. It is distinguished from condensing osteitis, which has an inflammatory origin.

Image of choice for diagnosis
Osteosclerosis is seen on panoramic and periapical radiographs.

Image hallmarks
Osteosclerosis usually is seen as a single, well-defined, homogeneous radiopacity that lacks a radiolucent rim (Fig. 12) [50,52,53]. It can be circular to ovoid to linear in shape, and the size ranges from several millimeters to actual diffuse areas of involvement. It typically is seen between teeth, at the apices of teeth, and below tooth-bearing areas. Root resorption occurs in approximately 10% of cases [51].

Management
Once the diagnosis of idiopathic osteosclerosis is made, no treatment is required. Care should be taken to rule out soft tissue calcification, such as a sialolith, which is superimposed on the image, and an exostosis.

Osseous dysplasia (cemeto-osseous dysplasia)

Frequency/incidence
Osseous dysplasia represents a reactive process of the jaws, which is subdivided into three, sometimes overlapping, categories [54–56]. Periapical cemental dysplasia overwhelmingly is seen in African American women aged 20 to 40 years and involves the periapical region of the anterior mandibular teeth [54,56,57]. Focal osseous dysplasia typically is recognized in women in their 20s and 40s and is found in the posterior mandibular region in tooth-bearing areas [56–58]. Florid osseous dysplasia represents a process that usually involves the tooth-bearing areas of multiple quadrants of the jaws and primarily is seen in middle-aged to elderly African American women [56,59].

Signs and symptoms
Periapical cemental dysplasia and focal osseous dysplasia generally are asymptomatic [54,57,60]. Long-term lesions may exhibit mild cortical expansion. Florid osseous dysplasia usually is asymptomatic but may show cortical expansion. In addition, some cases of florid osseous dysplasia will become secondarily inflamed and exhibit fistula formation, loose teeth, and pain.

Etiology/pathophysiology
The cause of osseous dysplasia is unknown [54,57]; however, it is theorized that it represents a reactive process in which normal bone is replaced by a poorly cellularized bonelike material and cellular fibrous connective tissue, which originate from periodontal membrane [54,57].

Image of choice for diagnosis
This process is often identified first on periapical radiographs; however, the panoramic radiograph is used to show the extent of the abnormality.

Image hallmarks
The radiographic changes depend on the subdivision of the process and the degree of mineralization of the proliferation [57]. Periapical cemental dysplasia involves from one to all of the
mandibular anterior teeth. Occasionally, it may also include other teeth. The initial changes are well-defined radiolucencies at the apices of the involved teeth. The area of bone remodeling may extend inferiorly and between teeth, and there usually is a progressive mineralization of the lesions. Thus, the changes range from radiolucent to radiolucent/radiopaque to radiopaque with a radiolucent rim in the final stage. Focal osseous dysplasia represents a solitary lesion associated with the apex or lateral surface of the root of the involved tooth, and it will demonstrate the same maturation pattern [56,57]. The florid variant exhibits the same maturation pattern as the former two categories but will involve multiple teeth and usually multiple quadrants of the jaws (Fig. 13) [55,56]. The alterations may extend beyond tooth-bearing areas and persistent radiolucencies may be present that represent idiopathic bone cavities [55].

**Management**

Once the diagnosis of osseous dysplasia is made, the patient can be observed. Florid osseous dysplasia may require antibiotic therapy and surgical debridement if the process becomes secondarily infected and undergoes sequestration. Patients with this process must practice sound oral hygiene to help prevent these complications. Focal osseous dysplasia can mimic central ossifying fibroma of bone radiographically, and these two conditions may not even be separated by microscopic examination. Radiographic changes that support focal osseous dysplasia over ossifying fibroma include contact of the lesion with the apex and/or lateral aspect of the root and mineralization of the lesion without an increase in the dimensions of the lesion. If the two cannot be separated on a radiographic basis, biopsy usually is performed. The ossifying fibroma usually is a solid structure, whereas focal osseous dysplasia usually is friable and removed in pieces [54].

**Simple bone cyst (idiopathic bone cavity, traumatic bone cyst)**

**Frequency/incidence**

Simple bone cysts primarily occur in the mandible, with a predilection for the anterior mandible between the mental foramina [61,62]. It usually affects individuals in their teens and 20s with near equal sex distribution [62–65].
**Signs and symptoms**

Simple bone cysts usually are asymptomatic [64], but expansion may occur. Some patients give a history of previous trauma to the area and may complain of mild pain and tenderness in the area [64].

**Etiology/pathophysiology**

It is theorized that simple bone cysts occur secondarily to intraosseous hemorrhage in which the normal repair mechanism is disrupted, resulting in the defect [61]. The source of hemorrhage may be trauma, focal degeneration of connective tissue, or idiopathic [61,62,64].

**Image of choice for diagnosis**

Simple bone cysts often are noted on routine panoramic radiographs, the film most utilized to identify odontogenic cysts and neoplasms of the jaws.

**Image hallmarks**

Simple bone cysts often appear as well-defined radiolucencies of the anterior and midbody of the mandible (Fig. 14) [61,62,64]. The border adjacent to teeth often extends between their roots, producing a scalloped border [61,63]. The radiographic image is not diagnostic for simple bone cyst alone, and other odontogenic lesions and neoplasms may exhibit a similar radiographic appearance.

**Management**

Management includes exploration and curettage of the bony walls to induce clot formation. Any soft tissue obtained during the procedure is examined microscopically to rule out a true odontogenic cyst [61,65]. Follow-up radiographic examinations are needed to ensure that bony fill takes place.

**SYSTEMIC METABOLIC DISORDERS**

**Osteopetrosis**

**Frequency/incidence**

Osteopetrosis is an extremely rare disorder related to a defect in normal bone remodeling. It is believed to occur in only 0.005% of the population [66,67]. It has three hereditary forms, which are variable in severity [68].
Signs and symptoms

Patients with the autosomal-recessive infantile form of the disease exhibit anemia and hepatosplenomegaly. They are susceptible to infections because of the granulocytopenia and exhibit enlargement of the jaws. A prominent sign is hypertrophy of the mandible, especially an increased width at the gonial angles [68]. Osteomyelitis is a common complication of tooth extraction. Symptoms related to cranial nerve compression, such as blindness, facial paralysis and otologic disorders may develop [69,70]; and pathologic fractures are common. Patients with the autosomal-dominant adult form may exhibit few signs and symptoms.

Etiology/pathophysiology

Osteopetrosis represents a group of hereditary disorders characterized by a defect in osteoclastic activity, which results in increased bone formation at the expense of normal marrow spaces [66,68]. The affected bone exhibits a decrease in vascularity and reduction or elimination of hematopoietic marrow. The bones become brittle and are susceptible to fracture and infection [66,68]. The severe type affects children who rarely survive past 2 years of age, whereas the milder forms primarily affect the skeleton and have a better prognosis [66,68].

Image of choice for diagnosis

The jaw changes are best demonstrated on panoramic radiograph. Other conventional facial radiographs may also be useful in evaluating the extent of the disorder throughout the cranio-maxillofacial skeleton.

Image hallmarks

Radiographic images of the process show loss of normal medullary trabecular pattern with a generalized radiopacity of the affected bone resulting from increased bone density (Fig. 15). The distinction between cortical and medullary bone becomes nonexistent as the medullary spaces are obliterated [66,68,71].

Management

Patients with osteopetrosis are highly susceptible to the development of osteomyelitis. Prevention plays an important role in managing a patient with this disease. Emphasis is placed on excellent oral hygiene and routine dental prophylaxis. Endodontic therapy is encouraged over dental extraction to avoid excessive trauma to bone. If debridement is necessary, conservative bone removal is indicated, with minimal periosteal stripping after limited incisinal access [66,69]. Although therapeutic levels of antibiotics are difficult to achieve in these patients, they are recommended [66].
Cleidocranial dysplasia

Frequency/incidence
Cleidocranial dysplasia is a rare hereditary disorder that affects the cranial bones and formation of the clavicles [72,73]. It exhibits equal sex distribution.

Signs and symptoms
Affected individuals are usually short in stature and exhibit a spectrum of hypermobility of the shoulders, which results in a characteristic appearance of a long neck with a narrow, pigeon-shaped chest [73]. These patients also may be brachycephalic and exhibit ocular hypertelorism and altered tooth eruption [72,73].

Etiology/pathophysiology
Cleidocranial dysplasia is a disorder of bone formation and ossification in which the cause is unknown. Most cases are autosomal dominant; however, 40% of cases represent mutations [72,73].

Image of choice for diagnosis
The characteristic dental abnormalities are demonstrated on the panoramic radiograph. The anteroposterior (A-P) chest radiograph will show the degree to which the clavicles are affected.

Image hallmarks
The characteristic findings on panoramic radiograph are retained deciduous teeth, multiple impacted teeth, and supernumerary teeth, which may be impacted (Fig. 16). A nasoalveolar cleft also may be observed. A-P chest films may show the degree of malformation of the clavicles.

Management
Treatment of this disorder is challenging and complex, requiring a multidisciplinary approach. The basic principles include removing any barriers to the eruption of permanent teeth [74–76]. This requires timely removal of deciduous teeth and bone with maintenance of space for future eruption [74,75]. Success is dependent on the formation of the teeth, which may be dam-

Fig. 16. Cleidocranial dysplasia.
aged during surgical exposure, root development, and retention. These patients often require orthognathic surgery to correct skeletal malformation resulting from hypoplastic maxilla and hyperplastic mandible. Alternatives for tooth replacement if the permanent teeth are not positioned and retained include tooth transplantation, dental implants, and dental prostheses [74–76].

**Paget disease**

**Frequency/incidence**

Paget disease is a rare disorder of bone metabolism, which results in the alteration of normal bone deposition and resorption [77]. The disorder predominantly affects individuals in their 40s and 50s, with an increased prevalence in men [78–80].

**Signs and symptoms**

The disease is associated with progressive enlargement of the affected bone. Enlargement of the jaws results in changes in occlusion, spacing of teeth, and facial deformity [81]. It may also cause pain in the affected bone, and multiple bones may be affected. Patients may exhibit neurologic complications, such as visual disturbances and hearing loss, when there is craniofacial bone involvement.

**Etiology/pathophysiology**

The cause of the alteration in bone remodeling is unknown [81]. Inflammation and endocrine and genetic factors have been considered but not substantiated.

**Image of choice for diagnosis**

The panoramic radiograph is an excellent imaging method to reveal involvement of the jaws with Paget disease.

**Image hallmarks**

The characteristic changes of the jaws in Paget disease are seen as areas of radiopacity surrounded by areas of radiolucency (Fig. 17). The areas tend to coalesce and become more radiopaque. They often exhibit a poorly defined hazy periphery, which is described as a “cotton wool” appearance. The teeth may exhibit hypercementosis, and there is an increase in interdental spacing [81]. Florid osseous dysplasia may appear in a similar fashion.

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Fig. 17. Paget disease. (Courtesy of Dr. Mark McIlwain, Sheffield, AL.)
Management

Medical treatment consists of the alleviation of bone pain and the use of bisphosphates to reduce progression of the disease by inhibiting osteoclastic activity [77,82]. In patients with facial disfigurement, recontouring procedures may be performed [77,83]. Patients with advanced involvement of the jaws are susceptible to the development of osteomyelitis, giant cell tumor, and sarcomas [83].

Cherubism

Frequency/incidence

Cherubism is a rare familial disorder that affects children between the ages of 14 months to 20 years, with an increased prevalence in boys [84–86]. Isolated cases have been reported, indicating the possibility of a spontaneous mutation [85,86].

Signs and symptoms

Cherubism is characterized by asymptomatic, bilateral expansion of the mandible, resulting in the classic picture of fullness of the cheeks. Involvement of the maxilla results in increased exposure of sclera, giving the appearance of an “upward gaze” to the eyes [84,86,87].

Etiology/pathophysiology

Most cases (80%) are autosomal dominant, affecting 100% of boys and 50% to 70% of girls [84–86]. The remaining 20% of cases represent spontaneous mutations [84,85,88]. The enlargement of the jaws is secondary to space-occupying lesions, which microscopically resemble central giant cell granuloma.

Image of choice for diagnosis

The maxillofacial lesions associated with cherubism are best visualized by panoramic radiograph.

Image hallmarks

The disorder is characterized by diffuse multilocular radiolucencies of the posterior mandible and maxilla, with the mandible being involved more frequently than the maxilla (Fig. 18) [84,87,89]. These lesions may alter tooth-eruption patterns, formation of teeth, and displace the inferior alveolar canal [85].

Fig. 18. Cherubism. (Courtesy of Dr. Denver Tackett, McDowell, KY.)
Management

Cherubism is a self-limiting disorder after the second decade of life. Excess deformity may require recontouring but this is best postponed as long as possible because postsurgical regrowth has been reported [84,90].

Fibrous dysplasia of bone

Frequency/incidence

Fibrous dysplasia of bone is a rare proliferative condition that affects the craniofacial skeleton and other bones of the body. There are two main patterns: monostotic, which represents single-bone involvement, and polyostotic, which involves multiple bones. The skull and the jaws represent the most commonly affected bones; however, the ribs and long bones also are involved [56,91]. This condition typically develops in the first two decades of life; the monostotic form is more common [56]. Fibrous dysplasia of bone is a component of McCune-Albright syndrome in which the affected individual exhibits polyostotic fibrous dysplasia, endocrine abnormalities, and café-au-lait skin pigmentation [56]. Ninety-five percent of individuals with McCune-Albright syndrome are female [56,91].

Signs and symptoms

Fibrous dysplasia of bone is an asymptomatic process that is characterized by expansion of the affected bone [56,92].

Etiology/pathophysiology

This disorder is believed to result from a sporadic mutation in the GNAS1 gene postzygotically in a somatic cell [93]. The resulting clinical manifestations, whether McCune-Albright syndrome or fibrous dysplasia, depend on the size of the embryonic cell mass when the mutation occurs and where in the cell mass it develops. The earlier the mutation occurs within the embryonic tissue, the more likely the chances of the multisystem disorder developing.

Image of choice for diagnosis

The changes of fibrous dysplasia of the jaws are best depicted on panoramic radiograph. Lateral skull and occlusal and periapical radiographs also are of help in demonstrating the characteristic findings. CT is helpful in determining the extent of the process, especially when multiple bones of the craniofacial complex are involved.

Fig. 19. Fibrous dysplasia of bone. (Courtesy of Dr. Richard Lee, Findlay, OH.)
Image hallmarks

The most characteristic change of fibrous dysplasia of the jaws is a finely radiopaque lesion that blends with normal surrounding bone (Fig. 19). On fine-grain films, such as periapical radiographs, the lesion exhibits a finely trabecular pattern, which has been termed “ground glass” in appearance [56,92]. Some cases, however, exhibit a multilocular or mottled appearance.

Management

The treatment of fibrous dysplasia of the jaws is variable. Most cases tend to stabilize as patients reach skeletal maturity [56], and mild cases may not require treatment. Patients with significant cosmetic and/or functional deformities may require one or more surgical reduction procedures. In some patients, there may be continued enlargement of the affected bone in adulthood. Regrowth of the lesion after recontouring is variable, with 25% to 50% of patients exhibiting some postoperative expansion [56,94].

Gardner syndrome

Frequency/incidence

Gardner syndrome is a rare adenomatous polyposis syndrome that is characterized by familial polyposis, desmoid tumors, epidermoid cysts, and dental and skeletal abnormalities [95,96]. The mean age of diagnosis is 25 years; however, onset of symptoms may range from 2 to 70 years of age [95]. Gardner syndrome has been reported to occur in between 1 to 8300 and 1 to 16,000 births [95,97].

Signs and symptoms

The extracolonic signs and symptoms are most often recognized first. These include multiple epidermoid cysts, impacted teeth, exophytic hard tissue masses of the mandible, and retinal pigmented lesions [96].

Etiology/pathophysiology

Gardner syndrome is an autosomal-dominant disorder with close to 100% penetrance [95,96,98]. The expression of the syndrome is variable. Nearly one third of cases occur spontaneously [95,96]. A gene on chromosome 5 has been identified as being associated with the development of the disorder [99].
Image of choice for diagnosis
The dental and jaw changes associated with the syndrome are best identified on a panoramic radiograph.

Image hallmarks
The changes seen on radiographs consist of multiple impacted supernumerary and regular teeth, osteosclerotic areas, and exophytic sclerotic masses of cortical bone, usually seen at the angles of the mandible and termed osteomas (Fig. 20).

Management
Gardner syndrome is a serious condition that requires early intervention because untreated patients will develop colorectal adenocarcinoma [95,96]. In women, there also is an increased risk of developing thyroid carcinoma [96]. The impacted teeth may have to be removed because of development of odontogenic cysts, and the exophytic osteomas may be removed for cosmetic and functional problems.

Hyperparathyroidism

Frequency/incidence
Hyperparathyroidism ranks third among the incidence of endocrine disorders behind diabetes mellitus and hyperthyroidism. It occurs in 30 of every 100,000 patients [100,101]. This disorder may occur at any age but it predominantly affects those in their 40s and 50s, with a female-to-male ratio of 3 to 1 [100,101].

Signs and symptoms
Hyperparathyroidism often is asymptomatic [100]; however, some patients will complain of or exhibit a variety of symptoms including the following: mental depression, confusion, lethargy, muscle weakness, nausea and vomiting, peptic ulceration, renal calculi, anorexia, and skeletal demineralization [102,103]. The signs and symptoms may be vague and nonspecific, and the disease may elude diagnosis for some time.

Etiology/pathophysiology
Hyperparathyroidism classically is divided into primary and secondary forms. The most common cause of the primary form is a parathyroid adenoma that secretes parathyroid hormone. The excess parathormone results in a derangement of calcium, phosphate, and bone metabolism. This process has been linked to rearrangements of segments of chromosome 11, thus allowing for overexpression of a regulatory protein [100]. Inactivation of tumor suppressor genes and mutations of calcium-sensing receptor genes also have been postulated [100]. The secondary form is caused by gland hyperplasia in response to low-serum calcium levels resulting from renal failure or intestinal malabsorption. This disorder may result in alterations of bone because of the increased levels of parathormone, which stimulates increased bone resorption [100].

Image of choice for diagnosis
Panoramic and periapical radiographs are the ideal imaging methods to evaluate jaw changes that may occur with the disorder.

Image hallmarks
In primary hyperparathyroidism, the jaw changes may occur as unilocular or multilocular radiolucencies referred to as “brown tumors” [100]. Microscopically, they are identical to central giant cell granuloma. In addition, the changes may consist of a generalized alteration of medullary bone that exhibits a “ground glass” appearance; however, this change is more commonly seen in secondary hyperparathyroidism (Fig. 21). This change usually is accompanied by a loss of lamina dura. Distinct lytic lesions are less common in secondary hyperparathyroidism.
Primary hyperparathyroidism is treated with parathyroidectomy with a 90% success rate [100]. Those patients refractory to or not surgical candidates are managed medically with oral-phosphate supplements, estrogen therapy, and calmodulin to decrease serum calcium [100]. The secondary form is treated by correcting the underlying medical condition [100].

DEVELOPMENTAL ODONTOGENIC CYSTS

Dentigerous cyst

Frequency/incidence
Dentigerous cysts are the second most common odontogenic cyst and account for 24% of true jaw cysts [104]. The frequency of dentigerous cysts is 1.44 per 100 unerupted teeth [105]. The mandibular third molars are the most commonly involved teeth.

Signs and symptoms
Most patients with dentigerous cysts are asymptomatic. Large cysts may cause expansion of bone in the involved area.

Etiology/pathophysiology
The dentigerous cyst represents a pathologic cystic cavity lined by follicular epithelium that develops around the crown of an impacted tooth [106]. It is capable of significant enlargement and destruction of bone. The cause of its development is unknown.

Image of choice for diagnosis
The panoramic radiograph is an ideal imaging method to detect dentigerous cysts. The cysts also may be detected with periapical radiographs.

Image hallmarks
The radiographic presentation of the dentigerous cyst is a well-circumscribed, unilocular radiolucency around the crown of an impacted tooth (Fig. 22). The cyst measures from 3 to 5 mm to many centimeters [107]. It typically affects permanent impacted teeth; but in rare incidences,
deciduous teeth may be involved. The cyst is usually an isolated lesion, but several teeth may be affected. Multiple dentigerous cysts may occur without the characteristic features of the nevoid basal cell carcinoma syndrome [108].

**Management**

The treatment of dentigerous cysts is complete enucleation of the cyst and removal of the involved impacted tooth. The cystic tissue is submitted for microscopic examination to rule out other aggressive odontogenic lesions.

**Odontogenic keratocyst**

**Frequency/incidence**

Odontogenic keratocysts make up approximately 12% of odontogenic cysts [109]. The mandible is involved in 65% of the cases, and the molar region is the most common location [109]. There is an equal sex distribution. A significant recurrence rate has been reported [110,111], and it has been reported that 8.8% to 12% of patients with odontogenic keratocysts have the nevoid basal cell carcinoma syndrome [110].

**Signs and symptoms**

Most patients with odontogenic keratocysts are asymptomatic; however, some may exhibit pain and swelling of the involved area of the jaw.

**Etiology/pathophysiology**

Odontogenic keratocysts most likely develop from remnants of dental lamina. The cause is unknown.

**Image of choice for diagnosis**

The panoramic radiograph is an excellent imaging method to demonstrate the odontogenic keratocyst, especially in the posterior regions of the jaws. Periapical radiographs also are able to detect the cyst when it develops in tooth-bearing areas.

**Image hallmarks**

Odontogenic keratocysts typically present as unilocular radiolucencies; however, they also may be multilocular, especially large keratocysts (Fig. 23). Keratocysts are often associated
with impacted teeth and resemble dentigerous cysts radiographically. They also can develop in between teeth (lateral periodontal cyst position) and in place of a tooth. In addition, keratocysts may mimic nasopalatine duct cysts [109] and apical periodontal cysts. In some cases, a curved scalloped border may be present [112].

Management
Treatments of odontogenic keratocysts is complete removal, ranging from enucleation and curettage to en bloc resection. The treatment depends on many factors, including position and extent of the lesion and patient cooperation. No single treatment is applied to all patients. Recurrent lesions often require more aggressive therapy [113].

Nevoid basal cell carcinoma syndrome

Frequency/incidence
The incidence of the syndrome is 1 in 57,000 to 1 in 164,000 births [114].

Signs and symptoms
Up to 75% of patients have odontogenic keratocysts of the jaws, and these often develop before the age of 20 [114]. These cysts are situated around the crowns of unerupted teeth and between teeth. The basal cell carcinomas usually develop during or after puberty; by the age of 40, up to 97% of affected persons exhibit these changes. Most patients have palmar and/or plantar pits. Other potential abnormalities include frontal bossing, hypertelorism, strabismus, palate abnormalities, calcified falx cerebri, and macrocephaly [115].

Etiology/pathophysiology
The nevoid basal cell carcinoma syndrome is an autosomal dominant disorder, and an abnormality of chromosome 9 has been identified in those patients who have the syndrome [116]. Its expression is thought to be caused by a modification of a tumor suppression gene.

Image of choice for diagnosis
The panoramic radiograph is the image of choice to identify the cystic lesions of the jaws.
The jaw changes represent single or multiple unilocular radiolucencies, usually situated around the crowns of unerupted teeth and between the roots of teeth (Fig. 24).

Management
The radiolucent lesions microscopically represent odontogenic keratocysts, and treatment is the same for those in the non-syndrome patient.

Lateral periodontal cyst

Frequency/incidence
Lateral periodontal cysts are relatively rare, comprising approximately 8% of cysts of the jaws [117]. The lesion typically is seen in adults and most commonly in the sixth decade of life. There is no sex predilection [118,119].

Signs and symptoms
The lateral periodontal cyst is asymptomatic and usually discovered on routine radiographs.

Etiology/pathophysiology
Lateral periodontal cysts develop from rests of dental lamina found in the periodontal membrane and interradicular bone. The cause is unknown. The cysts are not associated with an inflammatory process.

Image of choice for diagnosis
Lateral periodontal cysts are identified on panoramic and periapical radiographs.

Image hallmarks
The lesion typically is viewed as a small, well-circumscribed radiolucency between the roots of vital teeth (Fig. 25). It may have a sclerotic border [120]. Most lesions occur in the mandibular canine-premolar area and the maxillary incisor to first premolar region [121]. The odontogenic keratocyst may give a similar radiographic appearance.

Management
Lateral periodontal cysts are treated by enucleation. Recurrence is rare [122].
Calcifying odontogenic cyst

**Frequency/incidence**
Calcifying odontogenic cysts are rare and represent 1% to 2% of all odontogenic lesions [123]. They are found in patients of a broad age range, and there is equal sex distribution. The mandible and maxilla are almost equally affected, and 65% of cysts occur in the anterior regions of the jaws [124].

**Signs and symptoms**
Most calcifying odontogenic cysts are asymptomatic but enlargement of bone may occur. The cysts also may develop primarily in the gingiva with no bone involvement.

**Etiology/pathophysiology**
The cause of calcifying odontogenic cysts is unknown. The cysts most likely develop from dental lamina.

**Image of choice for diagnosis**
Calcifying odontogenic cysts are detected on panoramic and periapical radiographs.

**Image hallmarks**
The radiographic appearance of calcifying odontogenic cysts is variable (Fig. 26). They may be unilocular or multilocular. The cysts are usually well-outlined and may contain areas of calcification [125]. They may also develop in association with odontomas, and they are often associated with impacted or unerupted teeth and found in between teeth.
Management
Treatment of calcifying odontogenic cysts is enucleation but they may require more aggressive therapy depending on the extent of the lesion and the microscopic findings [126].

ODONTOGENIC NEOPLASMS

Ameloblastoma

Frequency/incidence
Ameloblastomas represent 1% of all oral tumors, and 80% to 85% occur in the mandible. There is no sex predilection [127]. The average age at diagnosis is 33 years [128].

Signs and symptoms
Ameloblastomas are infiltrating, locally aggressive neoplasms that can destroy bone, perforate cortical plates, and cause considerable expansion. They are generally considered to be slowly growing, and there usually is no associated pain [127]. In rare instances, the ameloblastoma has metastasized; and in 75% to 80% of cases, the lungs represented the metastatic focus [129]. Ameloblastomas also may develop outside of bone on the gingiva.
Etiology/pathophysiology
Ameloblastomas arise from odontogenic epithelium, principally dental lamina, and possibly dentigerous cyst lining. Peripheral ameloblastomas also may develop from the basal cell layer of gingival surface epithelium. The cause is unknown.

Image of choice for diagnosis
The image of choice for the detection of ameloblastomas is the panoramic radiograph. The periapical radiograph may also be used to detect ameloblastomas and other jaw proliferations. CT also is used to determine the extent of the lesion.

Image hallmarks
Ameloblastomas have a variable radiographic appearance. The classic description is that of a multilocular, well-defined radiolucency of the posterior mandible in the molar-ramus region (Fig. 27) [130]. Ameloblastomas also may be unilocular, and they occur in the anterior mandible and maxilla. They may be associated with impacted teeth and may occur in between teeth.

Management
Complete surgical excision is the treatment for ameloblastomas. Reported treatment ranges from curettage to radical resection depending on the type of ameloblastoma, its location, and the extent of the lesion. More radical treatment may lower the recurrence rate [131], which varies greatly in published reports [127]. Fifty percent of all recurrences occur within 5 years of surgery [127].

Calcifying epithelial odontogenic tumor

Frequency/incidence
Calcifying epithelial odontogenic tumors are rare, representing 1% of all odontogenic tumors [132]. Most of the tumors occur in men and women from the ages of 20 to 40 years. There is equal sex distribution [133]. The tumors occur twice as often in the mandible than the maxilla. Many of the reported cases have developed in association with impacted teeth.
Signs and symptoms
Most cases of calcifying epithelial odontogenic tumors are asymptomatic. They may cause cortical expansion, and may develop in the gingiva as an asymptomatic swelling.

Etiology/pathophysiology
The origin of calcifying epithelial odontogenic tumors is believed to be reduced enamel epithelium or stratum intermedium [134]. The cause is unknown.

Image of choice for diagnosis
The image of choice for the evaluation of jaw cysts and neoplasms is the panoramic radiograph. CT is helpful to determine the extent of the lesion.

Image hallmarks
The radiographic appearance of a calcifying epithelial odontogenic tumor is variable. It may be unilocular or multilocular and associated with an impacted tooth (Fig. 28). The tumor’s outline ranges from well circumscribed to irregular, and it may exhibit areas of radiopacity.

Management
The treatment of calcifying epithelial odontogenic tumors is surgical excision. Maxillary lesions may require more aggressive therapy if vital structures are involved [134]. Recurrences may develop up to several decades after initial therapy [134].

Adenomatoid odontogenic tumor

Frequency/incidence
Adenomatoid odontogenic tumors are the fifth most common odontogenic tumors, with an incidence of between 2.2% and 7.1% [135]. They occur most commonly in the second decade of life, primarily in women [136]. The tumors are most often seen in the anterior regions of the jaws, usually the maxilla.

Signs and symptoms
Most cases of adenomatoid odontogenic tumor are asymptomatic. The tumors may cause cortical expansion. They are often discovered on routine radiographic examination or when a patient is being evaluated for a delay in eruption of a permanent anterior tooth.
Etiology/pathophysiology
The tumors most likely arise from dental lamina. The cause is unknown. The tumors have been described as occurring with other odontogenic neoplasms [137].

Image of choice for diagnosis
The panoramic and periapical radiographs are the images used to detect this tumor.

Image hallmarks
Adenomatoid odontogenic tumors present as well-defined, unilocular radiolucencies (Fig. 29). Most cases are associated with the crown of an unerupted tooth, usually a maxillary cuspid. They also occur between the roots of anterior teeth. The radiolucency may show a flocculent pattern of discrete, noncoalescing radiopacities [138].

Management
Treatment of adenomatoid odontogenic tumors is surgical enucleation. The lesion usually is well encapsulated, and recurrence, even after conservative therapy, is rare to nonexistent [135].

Odontogenic myxoma

Frequency/incidence
Odontogenic myxomas are rare, accounting for 3.1% to 11.8% of all odontogenic tumors [139]. They involve the mandible more than the maxilla and are most common in men and women in their teens and 20s. There is a slight female predilection [140].

Signs and symptoms
Most cases of odontogenic myxoma are asymptomatic; however, enlargement of the involved bone may be present.

Etiology/pathophysiology
Odontogenic myxomas are benign but potentially locally aggressive neoplasms of odontogenic ectomesenchyme. Their cause is unknown.

Image of choice for diagnosis
Odontogenic myxomas are best visualized on the panoramic radiograph. Small myxomas situated between teeth also may be seen on periapical radiographs. CT is helpful in determining the extent of the lesion.
The radiographic appearance of odontogenic myxomas is variable. The classic description is a circumscribed, multilocular (“soap bubble”) radiolucency of the body and posterior mandible (Fig. 30) [141]. They can be unilocular, especially when they occur between teeth, and they may be associated with the crown of an unerupted tooth.

Management
The treatment of odontogenic myxomas varies from curettage to en bloc resection, depending on the location and extent of the lesion. Recurrence rates range from 10% to 33% [140].

Odontoma

Frequency/incidence
Odontomas are the most common odontogenic proliferations. Their frequency has been reported to be 46% to 74% of all odontogenic tumors [139,142]. Most odontomas, however, represent hamartomas rather than true neoplasms. They usually are found in the first to third decades of life, and there is an equal sex distribution.

Signs and symptoms
Odontomas usually are asymptomatic; they are found on routine dental radiographs and when a patient is being evaluated for a delay in eruption of a permanent tooth [143]. In rare instances, multiple odontomas have been described in systemic abnormalities, such as cleidocranial dysostosis and Gardner syndrome [144].

Etiology/pathophysiology
Most odontomas represent developmental anomalies and not true neoplasms [145]. The cause is unknown.

Image of choice for diagnosis
Odontomas are seen on both panoramic and periapical radiographs.

Image hallmarks
Odontomas exhibit a varying radiographic appearance. Compound odontomas are characterized by the formation of tooth-like structures. There may be a peripheral area of radiolucency. Complex odontomas are radiopaque and represent a haphazard arrangement of dental hard and soft tissue (Fig. 31). A radiolucent rim also may be present. Odontomas usually are seen adjacent to the crown of an unerupted tooth and between the roots of teeth [146].
Management

Treatment of odontomas is conservative surgical removal (enucleation). Recurrences are rare to nonexistent [147].

Ameloblastic fibroma

Frequency/incidence

Ameloblastic fibromas are rare odontogenic neoplasms, representing 2.5% of all odontogenic tumors [148]. The average age at the time of diagnosis is 15 years. More than 80% of cases occur in mandible in the premolar-molar region.
Signs and symptoms
Most ameloblastic fibromas are asymptomatic. They may cause expansion of the involved bone. They are often discovered when a patient is being evaluated for a delay in eruption of a permanent tooth.

Etiology/pathophysiology
Ameloblastic fibromas are benign, encapsulated odontogenic neoplasms of both odontogenic epithelium and mesenchyme. Their cause is unknown.

Image of choice for diagnosis
The image of choice for detection is the panoramic radiograph. Periapical radiographs may also be used to detect the lesion.

Image hallmarks
The ameloblastic fibroma is usually seen as a unilocular radiolucency associated with the crown of an unerupted posterior mandibular tooth (Fig. 32) [149]. It often exhibits a sclerotic border and, at times, may be compartmentalized.

Management
Treatment is enucleation and curettage. Recurrences occasionally have been described and follow-up is important [150].

Ameloblastic fibro-odontoma

Frequency/incidence
The incidence of ameloblastic fibro-odontomas ranges from 1.7% to 3.1% of all odontogenic neoplasms [134,151]. They occur most commonly in the posterior regions of the jaws in the first two decades of life. Eighty-three percent are associated with unerupted teeth [151].

Signs and symptoms
Most ameloblastic fibro-odontomas are asymptomatic. They may be associated with enlargement of the involved bone, and they often are detected when the patient is evaluated for an unerupted tooth.
**Etiology/pathophysiology**

Ameloblastic fibro-odontomas are neoplasms of odontogenic epithelium and mesenchyme. They have the capacity to form dental hard tissue [152]. The cause is unknown.

**Image of choice for diagnosis**

The panoramic radiograph is the image of choice for the detection and evaluation of ameloblastic fibro-odontomas.

**Image hallmarks**

Ameloblastic fibro-odontomas typically appear as mixed radiolucent-radiopaque lesions associated with the crown of one or more unerupted posterior teeth (Fig. 33). They are more common in the mandible. The affected teeth may be displaced away from the normal tooth-bearing area [153]. The radiopaque material has the radiodensity of tooth structure.

**Management**

Treatment is by conservative surgical enucleation or curettage with removal of the associated unerupted teeth. Maintenance of the teeth may lead to recurrence [151,153].

**NONODONTOGENIC NEOPLASMS**

**Central giant cell granuloma**

**Frequency/incidence**

More than 60% of all central giant cell granulomas occur before the age of 30 years. Two thirds of all cases develop in women, and the mandible is affected approximately 70% of the time. The lesion affects the anterior portions of the jaws more frequently than the posterior segments, and it is not uncommon for the lesion to cross the midline of the mandible [154].

Fig. 34. Central giant cell granuloma.
Signs and symptoms
Most central giant cell granulomas are asymptomatic. They may cause expansion and perforation of bone, and a minority of cases may be associated with pain and paresthesia [155].

Etiology/pathophysiology
The cause of central giant cell granuloma is unknown. There is no consensus that this process represents a true neoplasm; however, the lesion may exhibit aggressive growth and destroy significant bone. The microscopic features are the same as seen in the brown tumor of hyperparathyroidism [154].

Image of choice for diagnosis
The image of choice is the panoramic radiograph. Periapical radiographs may also be used to detect these lesions, especially those that develop in tooth-bearing areas of the jaws.

Image hallmarks
Most central giant cell granulomas appear as well-defined multilocular radiolucencies (60%) of the anterior mandible below the apices of involved teeth (Fig. 34). Root resorption and tooth displacement may occur. The granulomas also may appear unilocular (39%) [154]. They may develop between teeth and be associated with unerupted teeth.

Management
The usual treatment of central giant cell granuloma is aggressive curettage; the recurrence rate with this treatment ranges from 15% to 20%. More aggressive lesions may be treated by resection. Nonsurgical management has been considered recently and has included intralesional injection of corticosteroids and the systemic use of calcitonin [156].

Ossifying fibroma

Frequency incidence
Ossifying fibromas are most common during the third and fourth decades of life, primarily in women. Most cases develop in the mandible, but 20% develop in the maxilla [57,157].

Signs and symptoms
Most ossifying fibromas are asymptomatic. Some will exhibit enlargement of the involved bone [57].
Etiology/pathophysiology

Central ossifying fibromas represent true primary neoplasms of bone. The cause is unknown.

Image of choice for diagnosis

The panoramic radiograph is the image of choice for detecting the lesion. Periapical radiographs may also be used to identify ossifying fibroma.

Image hallmarks

Ossifying fibromas may be either unilocular or multilocular (Fig. 35). They usually are well defined and may have a sclerotic border. Depending on the degree of mineralization of the tumor’s connective tissue, the fibromas may appear radiolucent or radiolucent/radiopaque. Displacement of the roots of teeth also may be seen [157,158].

Management

Conservative surgical treatment is the treatment of choice for most ossifying fibromas. Aggressive tumors may need more aggressive therapy. The tumor usually is separated from surrounding normal bone and typically can be removed in a single piece. This is in contrast to focal osseous dysplasia, a reactive process with a similar radiographic picture, which blends with surrounding normal bone and is removed in small fragments [57].

Osteoblastoma/cementoblastoma

Frequency/incidence

Osteoblastomas and cementoblastomas are believed to represent the same neoplastic process. The main difference is that cementoblastomas are attached to the root of an involved tooth and originate from progenitor cells in the periodontal membrane rather than medullary bone as in osteoblastomas. Both are rare neoplasms of the jaws. Approximately 70% of osteoblastomas have been reported to occur in the mandible [159]. They show a mean age of occurrence of 20 years, with a range of 5 to 59 years [159]. Cementoblastomas typically occur before the age of 30 years, with a range of 6 to 75 years [159]. More than 90% of cases occur in the molar-premolar region of the jaws, and the mandible is more commonly affected [159].

Signs and symptoms

Most osteoblastomas and cementoblastomas exhibit enlargement of the involved bone at the time of discovery. Most patients will complain of spontaneous pain. Displacement of teeth may occur [160,161].

Etiology/pathophysiology

The lesions represent true neoplasms of osteoblasts and cementoblasts. The cause is unknown.

Image of choice for diagnosis

The panoramic radiograph is the image of choice for detection. Periapical radiographs are helpful in determining whether attachment to an involved tooth may exist, and CT is useful to determine the extent of the lesion.

Image hallmarks

The osteoblastoma usually appears as a well-circumscribed lesion that exhibits radiolucent and radiopaque areas (Fig. 36). In some cases, it will not be well delineated from surrounding bone. It is not continuous with the root surface of involved teeth. The cementoblastoma has a distinct radiographic appearance. It typically is well defined from surrounding bone, and it appears attached to the root of a tooth. The cementoblastoma usually is a mixed radiolucent/radiopaque lesion or one that is mostly radiopaque. The periodontal membrane space of the involved tooth characteristically is continuous with a radiolucent rim that surrounds the lesion [159,161,162].
Management

Osteoblastomas of the jaws are treated by complete surgical removal, often by curettage. Recurrence is uncommon. Cementoblastomas are treated by the surgical removal of the affected tooth and tumor mass. Removal of the mass and only the root portion of the tooth that is involved also has been utilized [161,162]. Follow-up information on this technique is not well established.

Osteosarcoma

Frequency/incidence

Osteosarcomas of all bones in the skeleton affect only 1 in 100,000 persons each year. Osteosarcomas of the jaws account for approximately 5% of all osteosarcomas. Lesions generally occur in the third decade of life in the jaws, but osteosarcomas of long bones usually occur in the second decade of life. Men have a slightly increased incidence, and the mandible and maxilla are equally affected [163,164].

Signs and symptoms

Cortical expansion and pain are the most common symptoms of an osteosarcoma. Depending on the location within the jaws, paresthesia and tooth mobility may be present [165,166].

Etiology/pathophysiology

An osteosarcoma is a malignancy of bone-forming cells. The cause is unknown, but there is an increased incidence in the growth plates of long bones. There also is an association of the disease with other pathologic processes that involve increased bone remodeling and growth and previous radiation therapy [163,164].

Image of choice for diagnosis

The panoramic radiograph is the image of choice for visualizing the disease. CT is helpful in determining the extent of the process.
Image hallmarks
The lesion of osteosarcoma may be radiolucent, mixed radiolucent/radiopaque, or radiopaque (Fig. 37). It may exhibit a mottled appearance, and the borders are commonly ill defined. The peripheral borders may exhibit a radiating growth pattern of tumor bone referred to as a “sun-burst” pattern, which is present in approximately 25% of the cases. Those lesions that arise in tooth-bearing areas may involve periodontal membrane and produce a widened space. This is not an exclusive feature of osteosarcoma because it may be seen with other malignancies [163,166].

Management
Surgical resection is the treatment of choice for osteosarcomas. Pre- and postoperative chemotherapy and postoperative radiation also are utilized. The survival rate does not appear to increase with the use of adjunctive chemotherapy as in long bones [163,165,166]. Up to 18% of the patients will exhibit metastasis, usually to lung. Uncontrolled local recurrence is the main cause of death in patients with osteosarcoma of the jaws. Mardinger et al [163] reported a 3-year survival rate of 72% for their patients.

Chondrosarcoma

Frequency/incidence
Chondrosarcomas are rare, malignant neoplasms. They are reported to account for less than 3% of all head and neck tumors. They occur most often in the third to sixth decades of life. Men show a very slight predilection. There is equal distribution in the mandible and maxilla [167].

Signs and symptoms
The most common symptom of chondrosarcoma of the orofacial structures is a painless swelling or mass. Other symptoms include pain, loose teeth, epistaxis, nasal obstruction, and visual or ocular changes [168].

Etiology/pathophysiology
Chondrosarcoma is a malignancy of cartilage-forming cells. The cause is unknown.
Image of choice for diagnosis

The panoramic radiograph is the image of choice for detection. CT scan is used to evaluate the extent of the tumor.

Image hallmarks

Chondrosarcomas typically appear as ill-defined lesions that may be radiolucent or mixed radiolucent/radiopaque (Fig. 38). They may also induce widening of the periodontal membrane space [169].

Management

Surgical resection is the treatment of choice for chondrosarcomas. Chemotherapy and radiation therapy normally are not effective against these neoplasms. The prognosis of chondrosarcoma of the jaws is poor [167,169].

Ewing sarcoma

Frequency/incidence

Ewing sarcomas are rare, malignant neoplasms that predominantly occur in the first two decades of life. About two thirds of all reported Ewing sarcomas occur in the lower skeleton. Less than 1% of cases occur in the facial skeleton. The mandible is more commonly involved than the maxilla. Sixty-five percent of the tumors occur in males [170].

Signs and symptoms

The most common symptoms are pain and swelling in the affected area. Loosening of teeth also may occur [170].

Etiology/pathophysiology

Ewing sarcoma is a malignancy of primitive cells, thought to be derived from neuroectoderm [170]. The cause is unknown.
The image of choice for the identification of Ewing sarcoma is the panoramic radiograph. CT is helpful to determine the extent of the neoplasm.

Image hallmarks
The typical radiographic pattern is that of an ill-defined radiolucency (Fig. 39). The tumor may stimulate replication of cortical bone identical to that seen in proliferative periostitis [171,172].

Management
Management of Ewing sarcoma consists of surgical resection and the use of radiation therapy and multiple chemotherapeutic agents. With the use of multiple modalities for treatment, the estimated 5-year survival rate for the tumor at a major treatment institution is 83% [170].

Metastatic neoplasms of the jaws

Frequency/incidence
Metastasis to the jaws is not rare. The most common location is the molar region of the mandible [173]. The most common primary sites in women are breast, adrenal gland, colorectal system, genital organs, and thyroid gland. Breast is the most common organ, representing 30% of cases [174]. The most common primary sites in men are lung, prostate, kidney, bone, and adrenal gland [173]. 30% of cases of jaw metastases represent the initial sign of the cancer. The male-to-female ratio is 1 to 2 [174].

Signs and symptoms
Signs and symptoms range from mild soreness and pain to facial deformity and numb chin syndrome [174]. Loose teeth also are a sign.

Etiology/pathophysiology
Metastatic cancer to the oral cavity is believed to result from hematogenous spread from the primary site.

Image of choice for diagnosis
The panoramic radiograph and periapical films are the images of choice for detecting metastasis to the jaws.
Image hallmarks
The typical radiographic presentation of jaw metastasis is that of a radiolucency that is either poorly defined or well outlined (Fig. 40). Certain cancers, such as breast and prostate, may induce new bone formation and create a mixed radiolucent/radiopaque lesion [175].

Management
Metastasis to the jaws indicates that the therapy for the primary cancer has failed to control the disease. Treatment consists of modalities used for disseminated primary disease. Generally, patients diagnosed with metastatic disease of the jaws do not survive more than 1 year [173].

HEMATOLOGIC/LYMPHORETICULAR DISEASES

Neutropenia

Frequency/incidence
Neutropenia is a rare, pathologic reduction of neutrophils that is subdivided into congenital (hereditary) and acquired forms. The congenital form commonly manifests in infants and adolescents. The acquired form more commonly occurs in later life [176].

Signs and symptoms
Patients with neutropenia exhibit an increased incidence of bacterial infections. They exhibit fever, malaise, weakness, and symptoms of an infection at a specific anatomic site. In the oral cavity, patients typically develop aphthous-like ulcers, gingivitis, and periodontal bone loss [176,177].

Etiology/pathophysiology
Neutropenia is defined as a circulating neutrophil count below 1500 mm$^3$. Primary or congenital neutropenia usually is the result of a genetic abnormality. Acquired neutropenia may
result from a variety of causes, including chemical and drug exposure, radiation, and bacterial and viral infections. Because the neutrophil is the primary defense against bacterial infections, the patient experiences an increased incidence of infections from these agents [176,177]. The most common organisms are gram-negative bacteria and Staphylococcus aureus.

Image of choice for diagnosis

The image of choice for detection of the radiographic changes are panoramic and periapical radiographs.

Image hallmarks

The oral radiographic changes of neutropenia are areas of significant periodontal bone loss (Fig. 41) [177].

Management

The infections that result from neutropenia are treated with appropriate antibiotics. Human granulocyte colony-stimulating factor has shown promise, and patients are encouraged to maintain meticulous oral hygiene to decrease the effects of periodontal bacterial flora [176,177].

Langerhans cell histiocytosis

Frequency/incidence

This is an uncommon disorder of Langerhans histiocytes in which 10% to 20% of all cases affect the jaws. The mean age of diagnosis of the process in the jaws is reported as 18 years. The mandible is more often affected than the maxilla, and it is more common in men [178].

Signs and symptoms

Bone lesions are the most common presenting symptoms of Langerhans cell disease [178]. Patients may present with lymphadenopathy. Infants may exhibit a skin rash and necrotizing areas of the gingiva and alveolar ridge. Older children and adults also may exhibit changes, including pain, gingival swelling, tooth mobility, parasthesia, and facial swelling [178,179].
Etiology/pathophysiology

Langerhans cell histiocytosis represents a symptom complex characterized by the pathologic accumulation of histiocytic cells that normally populate epidermis, mucosa, lymph nodes, and bone marrow and are involved with the presentation of antigens to T lymphocytes. The cause is unknown. The process is subdivided into three categories but overlap of symptoms exists [180,181]. The categories include acute disseminated, chronic multifocal, and a process that affects one or multiple bones without soft tissue involvement (eosinophilic granuloma of bone). The acute disseminated form affects soft tissue, lymph nodes, and visceral organs and generally does not affect bone. The chronic multifocal form exhibits bone, skin, and visceral involvement.

Image of choice for diagnosis

The images of choice to detect the jaw lesions of Langerhans cell histiocytosis are panoramic and periapical radiographs.

Image hallmarks

The jaw lesions of Langerhans cell histiocytosis are characterized by a radiolucency that often is present in interradicular or periapical bone (Fig. 42). The histiocytosis often mimics an inflammatory process [178] and may be poorly defined. Involved teeth may totally lack alveolar bone support and appear “floating in soft tissue.” Multiple lesions may be present.

Management

Once the microscopic diagnosis of Langerhans cell histiocytosis is established, the patient must be evaluated for multifocal involvement. Localized jaw lesions usually are treated by curettage. Low-dose radiation may be used for less accessible bone lesions [178]. The other forms require systemic therapy, including chemotherapy. Eosinophilic granuloma of the jaws has a
good prognosis, with a reported recurrence rate of 1.6% to 25% [178,179]. Death is rare in the chronic disseminated form; the acute disseminated form has a poor prognosis [180,181].

**Non-Hodgkin lymphoma**

**Frequency/incidence**

Approximately 24% of non-Hodgkin lymphomas develop in extranodal sites. Primary intra-osseous lymphoma accounts for 5% of extranodal lymphomas, and jaw involvement is rare. When the oral cavity is involved, the jaws are affected approximately 30% of the time [182, 183,184].

**Signs and symptoms**

Lymphomas of the head and neck and oral cavity primarily manifest as a soft tissue swelling. Those that involve the bone of the jaws often are associated with swelling of the involved area, pain, tooth mobility, and cervical lymphadenopathy [184,185].

**Etiology/pathophysiology**

Most lymphomas of the jaws are B-cell lymphomas. The cause is unknown. The development of lymphomas has been associated with the Epstein-Barr virus and human T-cell lymphoma virus 1 [186].

**Image of choice for diagnosis**

Panoramic and periapical radiographs are utilized to detect the change of the jaws associated with lymphoma. CT, MRI, and bone scintography also are helpful in evaluating the extent of involvement of the process.

**Image hallmarks**

Non-Hodgkin lymphoma of the jaws appears as single or multiple areas of ill-defined bone destruction, which may exhibit areas of reactive bone formation (Fig. 43) [185–187]. These areas may affect alveolar bone and mimic inflammation.

**Management**

Oral lymphoma is treated by radiation and/or chemotherapy in association with other site involvement. The survival time for patients with oral lymphoma is 11 to 38 months [185].

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Fig. 43. Non-Hodgkin lymphoma. (Courtesy of Dr. William Schiro, Lansing, MI.)
Multiple myeloma

Frequency/incidence

Multiple myeloma is a malignancy of plasma cells with variable systemic presentations. It is a rare disorder that is usually seen after the age of 40 years, with a mean age of occurrence of 60 years [188,189,190].

Signs and symptoms

Multiple myeloma may be asymptomatic [191] or associated with the signs and symptoms of anemia, leukopenia, thrombocytopenia, hypercalcemia, and renal disease [188,192]. Oral involvement may be associated with jaw and tooth pain, tooth mobility, paresthesia, soft tissue swelling, and pathologic fracture [188,192].

Etiology/pathophysiology

The cause of this plasma cell cancer is unknown.

Image of choice for diagnosis

The panoramic radiograph is the choice for detecting multiple lesions of myeloma. Periapical radiographs may also detect the changes in alveolar bone.

Image hallmarks

Multiple myeloma exhibits a varied radiographic appearance. The lesions are radiolucent and may exhibit a well-defined or poorly defined border (Fig. 44). They may be unilocular or multilocular, and there may be multiple areas of bone destruction. The lesion may lack any reactive or defined border, which gives the lesion a “punched out” appearance [192].

Management

Isolated lesions (plasmacytoma) are treated by radiation therapy [190,191]. Disseminated disease is treated by chemotherapy [190].

References


