

chapter 11

DISORDERS OF BONE

Figs. 11.1, 11.2, and 11.3

Osteopetrosis is a rare disorder characterized by a marked increase in bone density resulting from a defect in the bone-remodeling mechanism. Although the pattern of bone sclerosis is very distinctive, other diseases such as pyknodysostosis and dysosteosclerosis may produce similar changes and must be considered prior to definitive diagnosis. Several hereditary patterns of osteopetrosis are seen that are associated with variations in clinical severity.

The most severe form is autosomal recessive and noted at birth or early in life. Most of the skeleton shows extensive replacement of the bone marrow by dense bone, with resultant severe normocytic anemia, granulocytopenia, and significant hepatosplenomegaly caused by extramedullary hematopoiesis. Optic atrophy, loss of hearing, facial paralysis, and pathologic fractures are common complications; poor pneumatization of the paranasal sinuses usually is seen. Most patients with this form of the disease die before age 20 years as a result of anemia or infection caused by marrow replacement by sclerotic bone.

Clinically, the benign autosomal-dominant form is less severe and may not be detected until later in life. The extent of bone involvement and symptoms vary. Some cases are detected incidentally during a radiographic examination. These patients do not exhibit anemia or hepatosplenomegaly; blindness and deafness also usually are not present. Although variable, the life span usually is normal.

The jaws may be involved in both forms of the disease. Extensive osteosclerosis often obscures the outline of the roots of the teeth and blurs the separation between cortical and cancellous bone. Teeth abnormalities such as hypodontia, malformed crowns and roots, defective enamel, and extensive caries are common. Retardation of eruption caused by the densely sclerotic bone is common, particularly in the recessive form of the disease. In some cases, only a few teeth ever erupt. Osteomyelitis following extraction of teeth is a common complication. Excellent oral hygiene, use of topical fluorides, and regular professional care are important to decrease the necessity for tooth removal. When extraction of teeth is mandated, antibiotic coverage and primary closure of extraction sites have been recommended to decrease the prevalence of infection.

► **Figure 11.1**

OSTEOPETROSIS

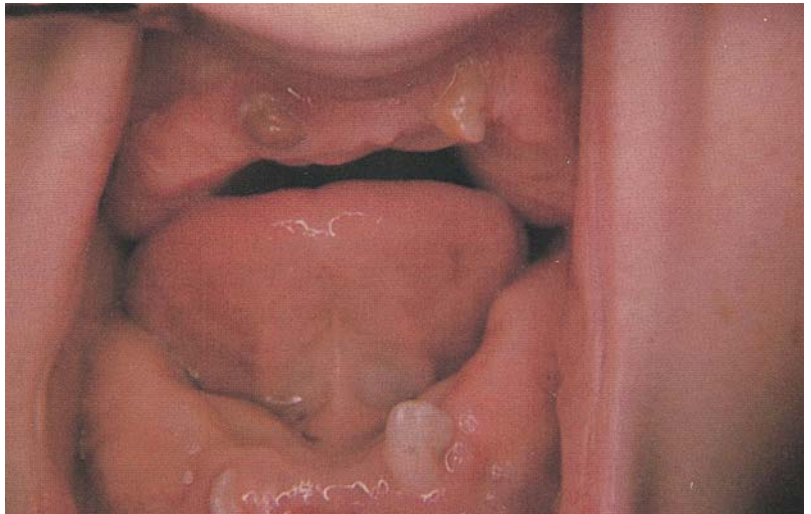
Photograph of the skin overlying the right mandible. Note numerous cutaneous fistulas that are draining purulent material from areas of necrosis within the underlying mandible.



► **Figure 11.2**

OSTEOPETROSIS

Intraoral photograph of an 8-year-old boy with severe recessive osteopetrosis. No permanent teeth have erupted.



► **Figure 11.3**

OSTEOPETROSIS

Radiograph of a mandible removed at autopsy from a 10-year-old girl who had severe childhood osteopetrosis showing the densely sclerotic bone and multiple malformed and unerupted teeth. (Courtesy of Younai F, Eisenbud L, Sciubba J. Osteopetrosis: a case report including gross and microscopic findings in the mandible at autopsy. *Oral Surg Oral Med Oral Pathol* 1988;65:214.)



Figs. 11.4, 11.5, and 11.6

Cleidocranial dysplasia is a skeletal disease often inherited as an autosomal-dominant trait. Up to 40% of the cases, however, seem to be spontaneous mutations. Although a variety of anomalies may be found in other bones, the defects chiefly involve membranous bones, with the skull and clavicles being the chief sites of the disorder. The patients tend to have short stature and a large head with frontal and parietal bossing and ocular hypertelorism. The sutures and fontanels show delayed closure or may remain open, and wormian bones commonly are observed on radiographic examination of the skull. Most patients also reveal hypoplasia or absence of nasal bones, diminished paranasal sinuses, and a broad base of the nose with a depressed nasal bridge. The midfacial skeleton may be hypoplastic, resulting in a relative mandibular prognathism.

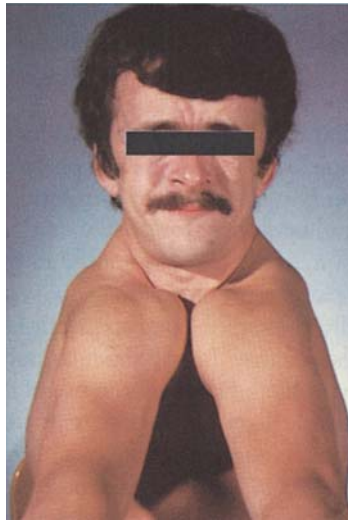
The shoulder girdle defects vary from complete absence of the clavicles to a partial absence or marked thinning of one or both clavicles. The clavicular abnormalities result in a long-appearing neck, drooping shoulders, and an unusual mobility of the shoulders, with some patients being able to move their shoulders forward until they meet. Although the clavicular defects result in variations of the muscles related to the clavicles, function generally is not impaired.

Patients with cleidocranial dysplasia often show a narrow, high-arched palate and exhibit an increased prevalence of cleft palate. Multiple supernumerary teeth, prolonged retention of deciduous teeth and delay or failure of eruption of the permanent teeth are characteristic findings. Radiographic examination reveals multiple unerupted teeth, many of which show root dilaceration or root shortening.

No treatment is directed toward the skull and clavicular anomalies, but the dental problems represent a major source of morbidity. Without treatment, the retained deciduous dentition begins to rapidly deteriorate in late youth and early adulthood, leading to a premature aged facial appearance. In the past, total extraction of the dentition followed by full dentures was performed, but this has been abandoned. Removal of the deciduous and supernumerary teeth with overdentures placed on top of the impacted permanent teeth was associated with significant discomfort secondary to subsequent eruption of the remaining dentition and is in disfavor. In such cases, eruption has been noted as late as the sixth decade. Although no one method of approach is accepted universally, significant success has been obtained through removal of all deciduous and supernumerary teeth followed by surgical exposure and orthodontic or surgical repositioning of impacted permanent teeth. In many cases, the therapeutic protocol extends over many years and is timed to coincide with the developmental stages of the succedaneous dentition.

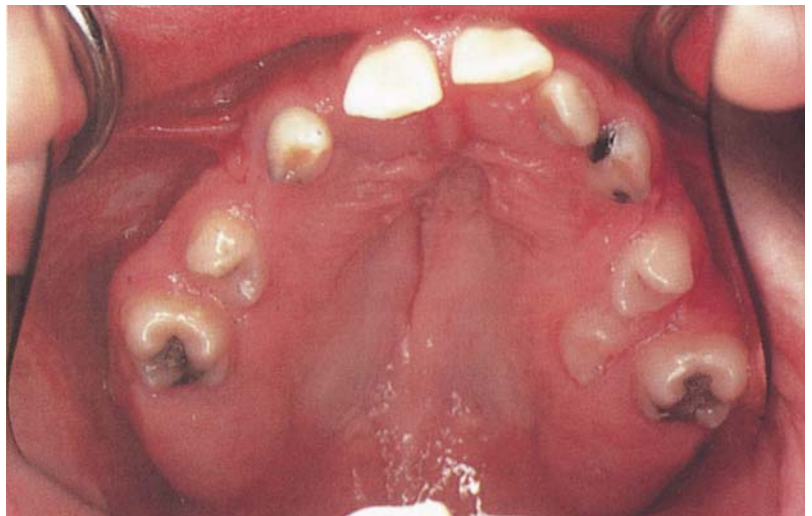
► **Figure 11.4**
CLEIDOCRANIAL DYSPLASIA

This patient can approximate his shoulders. The enlarged skull with prominent frontal bossing is apparent. (Courtesy of Halstead CL, Blozis GG, Drinnan AJ, et al. Physical evaluation of the dental patient. St Louis: CV Mosby, 1982.)



► **Figure 11.5**
CLEIDOCRANIAL DYSPLASIA

Maxillary dentition of a young adult with cleidocranial dysplasia. Several deciduous teeth are still present, and several permanent teeth have not erupted. (Courtesy of Halstead CL, Blozis GG, Drinnan AJ, et al. Physical evaluation of the dental patient. St Louis: CV Mosby, 1982.)



► **Figure 11.6**
CLEIDOCRANIAL DYSPLASIA

Panographic radiograph of a 28-year-old male showing multiple impacted and supernumerary teeth and retention of deciduous teeth. (Courtesy of Dr. J. R. Cramer.)



Fig. 11.7

An osteoporotic marrow defect is a normal anatomic variation that may be confused with a pathologic alteration. This nonneoplastic process presents as an asymptomatic radiolucent area that contains either hematopoietic or fibrofatty marrow. The lesion almost always is detected during a routine radiographic examination and is most commonly noted in mandibular edentulous areas of women. Many cases are noted in the site of a previous extraction, and rare patients report associated pain or swelling.

Radiographically, the radiolucency typically is ill-defined, seldom exceeds 1.0 cm in diameter, and usually reveals a central, faint trabecular pattern. In some cases, the fatty nature of the marrow defect can be predicted through the use of computed tomography and magnetic resonance imaging scans, possibly negating the necessity for biopsy. In many instances, the radiographic features are not diagnostic, simulate those of a number of different inflammatory or neoplastic lesions, and mandate biopsy. After the diagnosis is confirmed, no treatment is necessary. Although focal marrow defects have not been correlated to any systemic disease, the possibility of concurrent disorders associated with accelerated hematopoiesis, such as the hemolytic anemias, should be investigated in patients with widespread lucency associated with marrow enlargement.

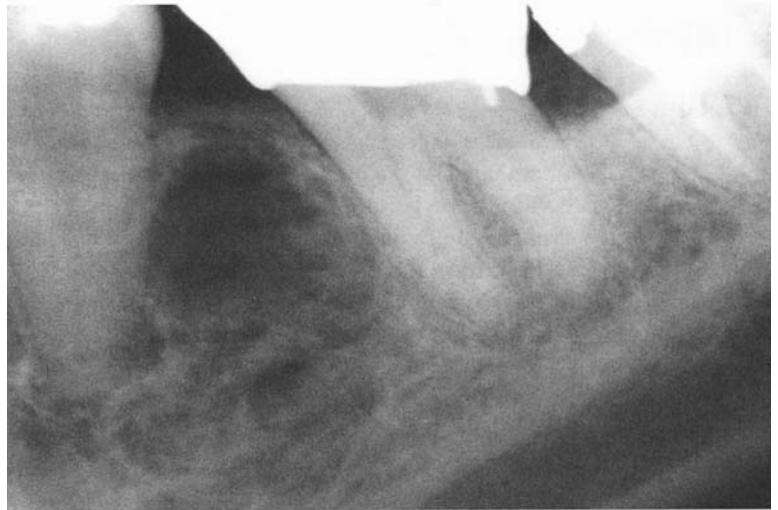
IDIOPATHIC OSTEOSCLEROSIS (DENSE BONE ISLAND, ENOSTOSIS)**Figs. 11.8 and 11.9**

Focal areas of increased radiodensity within the jaws unrelated to foci of infection are the most common causes of radiographic consultations received by oral and maxillofacial pathologists. These areas, which cannot be related to any inflammatory, dysplastic, or neoplastic process, are thought to occur in up to 10% of the population. Similar lesions have been seen in other bones.

Although idiopathic osteosclerosis occurs over a wide age range, most lesions are noted before 40 years of age, are asymptomatic, and occur in the premolar-molar areas of the mandible. Because the prevalence of the sclerotic zones does not seem to increase with age, these opacities most likely do not arise in later age or previous lesions are resorbed over time. The typical radiographic pattern is a well-defined or irregular area of radiopacity that does not demonstrate a radiolucent rim and that varies in size from 0.3 to 2.0 cm. Rare examples approach 7.0 cm in diameter. Although resorption and displacement of adjacent teeth are seen rarely, most cases do not reveal enlargement over time or buccolingual bony expansion. Bone adjacent to root apices is affected most frequently, but involvement of interradicular bone is not uncommon. Rarely, these islands of dense bone seem to surround the crown of impacted teeth in a distinctly pericoronal fashion. On occasion, the sclerotic area exhibits no relationship to a tooth.

Biopsy usually is not necessary because other radiographically similar processes such as superimposed soft-tissue calcifications, condensing osteitis, and overlying exostoses can be ruled out from clinical and radiographic features. No treatment is required because most demonstrate no tendency to enlarge or adversely affect adjacent structures.

- **Figure 11.7**
FOCAL OSTEOPOROTIC MARROW DEFECT
Irregular lytic area that was discovered during a routine radiographic examination of a female, age 49 years. Biopsy results revealed that the defect contained hematopoietic marrow. (Courtesy of Dr. Michael Sokolosky.)



- **Figure 11.8**
IDIOPATHIC OSTEOSCLEROSIS
Localized zone of increased radiodensity between the maxillary premolars.



- **Figure 11.9**
IDIOPATHIC OSTEOSCLEROSIS
Localized zone of increased radiodensity associated with the distal root of the mandibular first molar. Note resorption of the distal root. (Courtesy of Dr. Lon Doles.)



Fig. 11.10

Skeletal osteomas are benign neoplasms of mature bone that essentially are restricted to the craniofacial skeleton. These lesions consist of mature compact or cancellous bone, and differentiation from exostoses or areas of idiopathic osteosclerosis often is difficult and arbitrary. Although most commonly found in the skull, osteomas do occur within the jaws, and they most frequently arise from the posterior lingual surface of the mandible in young adults. For a definitive diagnosis of osteoma, a history of sustained growth must be demonstrated. In many instances, the tumors seem to arise in close proximity to areas of muscle attachment; it has been suggested that these lesions represent nonneoplastic, but progressive, bony hyperplasia secondary to muscle traction in sites of previous trauma.

Osteomas of the jaws most frequently arise on the surface of the bone and present as slowly growing sessile or pedunculated masses. Endosteal osteomas generally produce no symptoms until they reach a large size and cause cortical expansion. Occasional osteomas arise in the paranasal sinuses and may be associated with symptoms such as sinusitis, headache, and ophthalmologic manifestations. Radiographically, most examples show a well-circumscribed lesion that is either densely sclerotic or has a trabecular pattern. Large osteomas associated with facial distortion or symptoms such as dysphagia or difficulties in mastication are treated by conservative surgical removal.

GARDNER SYNDROME**Figs. 11.11 and 11.12**

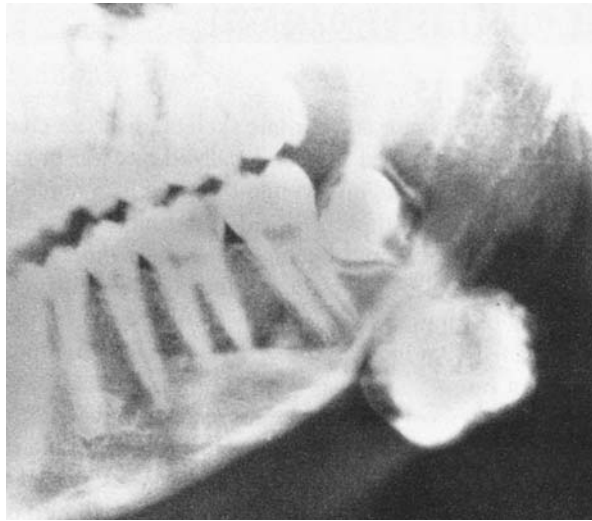
Gardner syndrome is an autosomal-dominant condition characterized by the association of multiple osteomas, fibromatoses of the soft tissues and mesentery, epidermoid cysts of the skin, and adenomatous polyps of the gastrointestinal tract, usually in the colon. The intestinal polyps have a marked tendency to undergo rapid malignant transformation. Women with the syndrome also are predisposed dramatically to development of thyroid carcinoma and should be followed closely for this complication. Multiple osteomas, most frequently involving the mandible, maxilla, and frontal bone, appear during puberty, usually preceding the appearance of the gastrointestinal polyps. Supernumerary teeth, impacted teeth, and compound odontomas of the jaws also have been described in these patients, but the reported prevalence is variable.

Although some of the jaw osteomas may cause facial asymmetry, many merely represent areas of increased radiodensity similar to that seen in idiopathic osteosclerosis and often are discovered with panoramic radiographs or computed tomography. Some investigators have used panoramic radiographs as screening tools in first-degree relatives of affected individuals; those that demonstrate significant areas of osteosclerosis or clinically evident osteomas are evaluated further with fecal occult blood tests, flexible sigmoidoscopy, or colonoscopy. Because of the high prevalence of idiopathic osteosclerosis in the general population, its presence without a family history of Gardner syndrome is insufficient justification for further evaluation.

Therapy centers around prevention of carcinoma from the adenomatous polyps, with prophylactic colectomy performed soon after confirmation of the diagnosis. Without intervention, close to 50% of affected patients develop colorectal adenocarcinoma before age 30 years, with the frequency of carcinomatous transformation approaching 100% in older patients. The fibrous neoplasms are surgically removed as they are encountered, along with any jaw osteomas and epidermoid cysts that create aesthetic concerns.

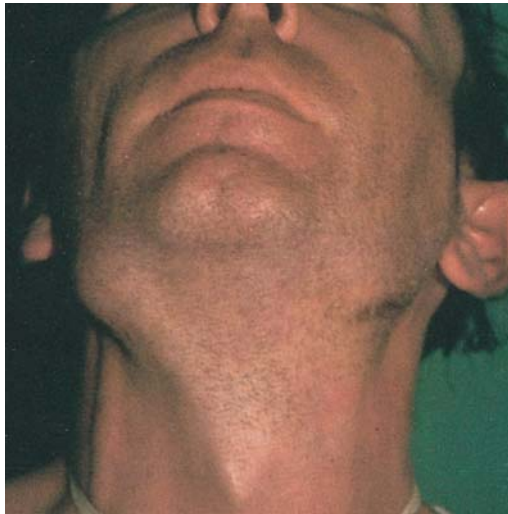
► **Figure 11.10**
OSTEOMA

A 16-year-old boy with a large, pedunculated osteoma arising from the mandibular cortex at the angle of the mandible.



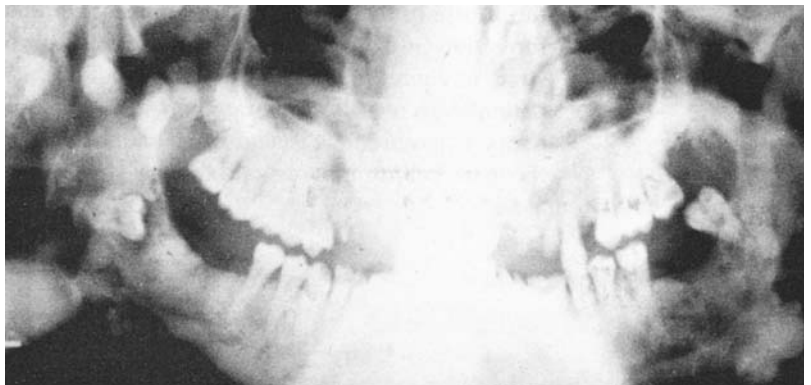
► **Figure 11.11**
GARDNER SYNDROME

Clinical photograph of a 33-year-old male. The asymmetry beneath the mandible is caused by a large osteoma. (Courtesy of Dr. David Haddox.)



► **Figure 11.12**
GARDNER SYNDROME

Panoramic radiograph of the patient depicted in Figure 11.11 showing multiple osteomas of the mandible. (Courtesy of Dr. David Haddox.)



Figs. 11.13, 11.14, and 11.15

Paget's disease is a chronic skeletal disorder characterized by abnormal resorption and deposition of bone that may be localized or widespread throughout the skeleton. Although most cases are polyostotic, involvement limited to a single bone may be seen. The jaws are affected in about 10 to 15% of the cases, with a significant maxillary predominance.

Affected bones become thickened, enlarged, and weakened. Severely involved weight-bearing long bones often develop a monkeylike bowing. Involvement of the skull with resultant narrowing of foramina occasionally results in neurologic complications such as deafness or visual disturbances. An increased hat size secondary to a progressive enlargement of head circumference is not rare.

The most common oral presentation is a progressive, symmetric maxillary enlargement that may reach massive proportions. Severe maxillary enlargement may result in a "lionlike" facies that has been termed "leontiasis ossea." With significant jaw enlargement, dentures often don't fit and remaining teeth become widely spaced or flared. Extensive root hypercementosis occasionally is detected.

Radiographically, Paget's disease demonstrates similarities to diffuse osseous dysplasia with early lucent alterations followed by the development of intermixed radiopacity. With maturation, the affected bone becomes predominantly radiopaque and exhibits the classic "cotton wool" appearance. Any patient demonstrating bony enlargements and multifocal mixed radiolucencies of the jaws should be evaluated further for the possibility of the disorder. In affected patients, serum alkaline phosphatase and urinary hydroxyproline levels usually are elevated, but blood calcium and phosphorus levels are normal.

In cases of Paget's disease with significant jaw enlargement, aesthetics and function may be improved by surgical reduction. Similar to end-stage osseous dysplasia, advanced osseous lesions of Paget's disease are sensitive to inflammation and develop osteomyelitis with little provocation. Elective surgery is not recommended in these patients, and all sources of bony inflammation must be kept to a minimum. When osteomyelitis develops, conservative surgical debridement of the affected area with appropriate antibiotic coverage is necessary.

Paget's disease rarely is a cause of death. Uncommonly, the hypervascularity of the pagetic bone acts as an arteriovenous shunt leading to high output heart failure or worsening of underlying cardiac disease. Parathyroid hormone antagonists such as calcitonin and biphosphonates have been used to minimize the effects of the disease by reducing bone turnover, especially early in the course of the disease. Two significant complications, giant cell tumor and sarcomatous transformation (usually osteosarcoma, malignant fibrous histiocytoma, or chondrosarcoma), demonstrate an increased prevalence in association with Paget's disease and must be considered in any patient who reveals accelerated growth of an affected bone. The most frequent associated malignancy, osteosarcoma, is aggressive and associated with a very poor prognosis, whereas most of the giant cell tumors respond to local surgical removal.

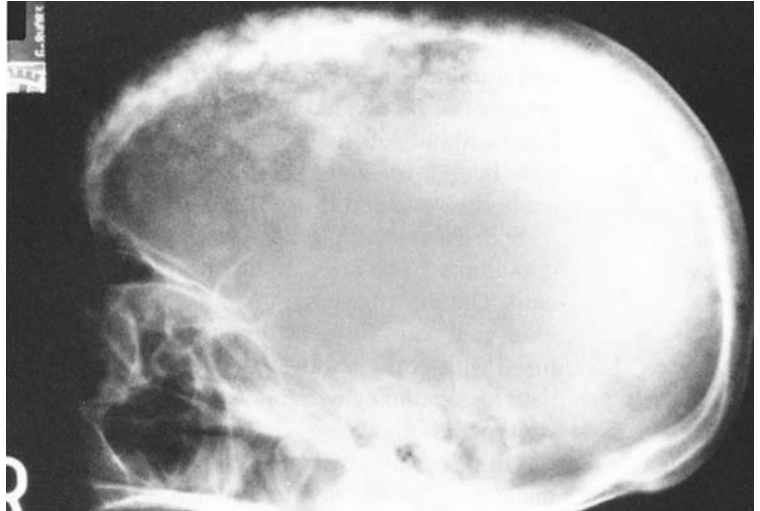
- **Figure 11.13**
PAGET'S DISEASE OF BONE
Marked symmetric maxillary enlargement
in a female, age 55. (Courtesy of Dr.
William McKenzie.)



- **Figure 11.14**
PAGET'S DISEASE OF BONE
Periapical radiograph of a patient with
Paget's disease showing irregular
radiopaque foci ("cotton wool"
appearance). (Courtesy of Dr. Mona Ellis.)



- **Figure 11.15**
PAGET'S DISEASE OF BONE
Skull film demonstrating multiple foci of
irregular increased radiodensity arranged
in the "cotton wool" pattern. (Courtesy of
Dr. Reg Munden.)



MASSIVE OSTEOLYSIS (GORHAM-STOUT DISEASE)

Figs. 11.16 and 11.17

Massive osteolysis is a rare chronic disease characterized by spontaneous and progressive destruction of one or more bones. Involvement of contiguous bones may occur. Although some investigators believe the process may represent an unusual intraosseous hemangiomatosis, the definitive cause is unknown and does not seem to be related to any known infectious, metabolic, endocrine, or neurologic disorder. Although most cases arise in children and young adults, examples have been reported in patients ranging from infancy to old age.

About one-fourth of cases have involved the craniofacial bones, with the jaws being two of the most commonly affected; occasional simultaneous involvement of both the maxilla and the mandible is reported. In the early stages, the disease is characterized by active bone lysis, often accompanied by mild to moderate pain. At this stage, radiographs show an ill-defined lysis of bone, with results of histopathologic examination demonstrating a vascular proliferation that closely resembles immature granulation tissue. With quiescence, the disease becomes painless and demonstrates no radiographic evidence of bone regeneration. Significant destruction of the jaws can lead to difficulty in mastication, swallowing, speaking, and respiratory function.

The progression of the destruction is variable, but in most cases, the process continues for months to a few years and leads to total loss of the affected bone. The diagnosis is made largely by exclusion of other diseases characterized by bone resorption and by failure to detect any biochemical abnormalities. Although there is no consistently satisfactory treatment for massive osteolysis, surgical excision of the affected area is the leading method of approach.

LANGERHANS CELL HISTIOCYTOSIS (HISTIOCYTOSIS X)

Fig. 11.18

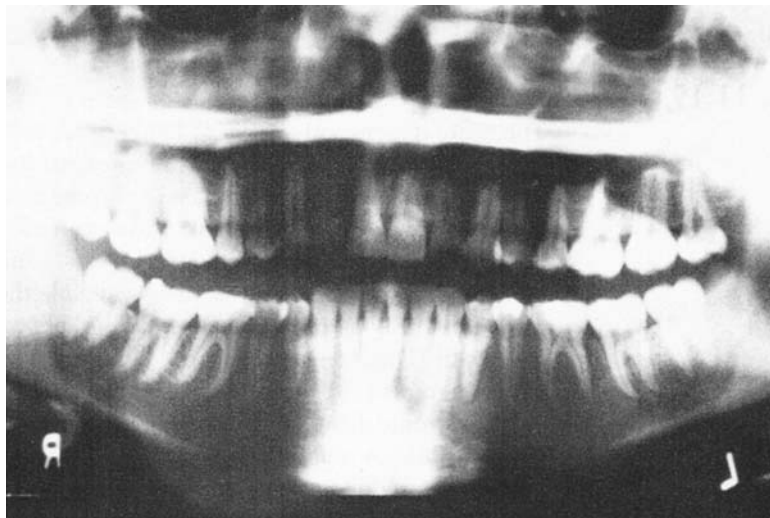
Langerhans cell histiocytosis describes a group of uncommon diseases characterized by a proliferation of differentiated histiocyte-like cells often accompanied by a variable admixture of eosinophils, multinucleated giant cells, lymphocytes, and plasma cells. The distinctive histiocytic cells have been shown to represent Langerhans cells, and they possess unique ultrastructural and immunohistochemical features. Based on the type and extent of involvement, these histiocytoses can be divided into several major categories: (1) solitary or multiple involvement of bone without soft-tissue involvement, (2) chronic multifocal histiocytosis characterized by multiple bone involvement with skin and visceral involvement (see Figs. 11.19 to 11.21), and (3) acute disseminated disease. However, many patients show overlapping clinical features.

Solitary or multiple bone involvement without visceral involvement is termed “eosinophilic granuloma.” The jaws are involved in 10 to 20% of cases and may be the only site of the disease. The most frequent gnathic site is the posterior mandible and typically is associated with mild, dull pain. In many instances, the disease mimics odontogenic infection and often is confused with periodontitis, apical inflammatory disease, and pericoronitis. Such similarities mandate histopathologic examination of supposedly inflammatory foci that do not respond to conventional therapy.

On discovery of isolated disease, thorough systemic evaluation for additional lesions is mandatory. In addition to evaluation of the soft tissues, a whole-body radiographic skeletal survey or bone scintigraphy is necessary. If no additional lesions are discovered, the focal site usually is treated surgically; intralesional injection with methylprednisolone also has been successful in a limited number of patients. Subsequent radiation therapy often is used in large or multifocal lesions.

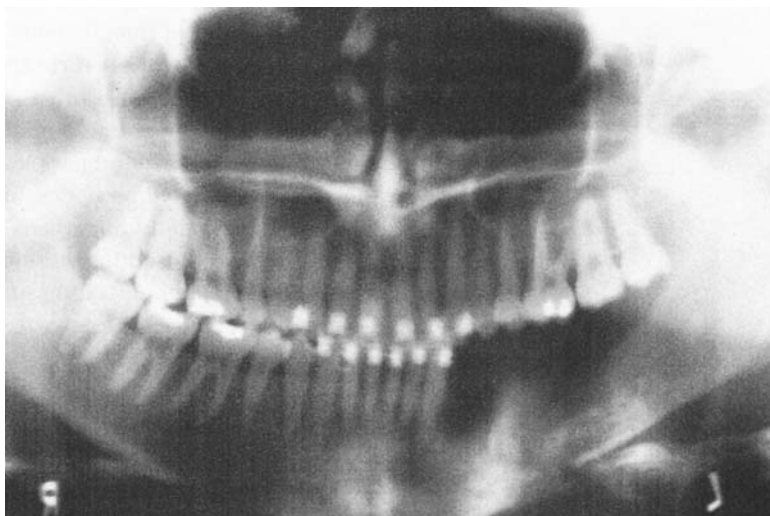
► **Figure 11.16**
MASSIVE OSTEOLYSIS

Ill-defined radiolucent area in the left mandibular premolar-molar area of a 22-year-old woman. This initially was considered to be related to pulp pathology; endodontic treatment was carried out. (Courtesy of Dr. J. R. Cramer.)



► **Figure 11.17**
MASSIVE OSTEOLYSIS

Radiograph from the patient shown in Figure 11.16 taken 11 months later showing progressive destruction of the mandible, pathologic fracture, and loss of teeth. (Courtesy of Dr. J. R. Cramer.)



► **Figure 11.18**
LANGERHANS CELL HISTIOCYTOSIS

Periapical radiograph demonstrating a lytic lesion enveloping the apices of the anterior incisors. After failure to respond to conventional endodontics, periapical surgery was performed. Results of histopathologic examination of the removed soft tissue revealed Langerhans cell histiocytosis. (Courtesy of Dr. James White.)



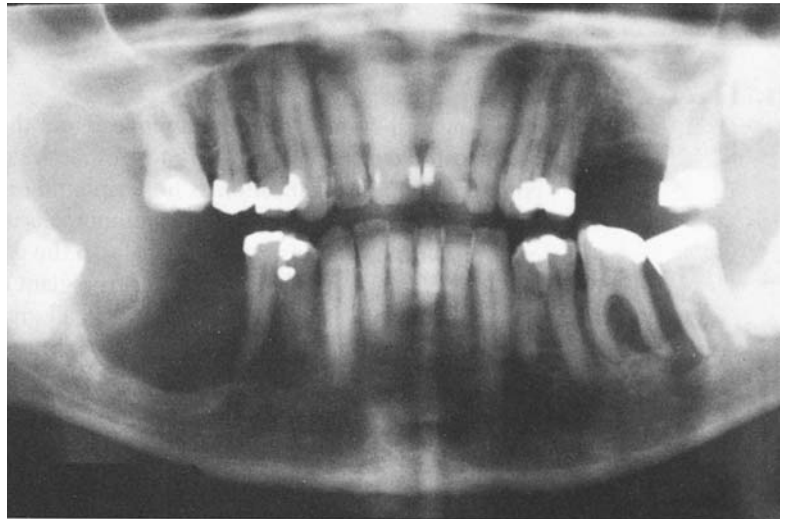
Figs. 11.19, 11.20, and 11.21

The acute disseminated form of Langerhans cell histiocytosis is termed “Letterer-Siwe disease.” Many authorities believe this form represents a separate, and most likely malignant, process whereas eosinophilic granuloma and chronic disseminated histiocytosis are variants of a single, less aggressive disease. The acute form primarily affects children before the age of 3 years and demonstrates rapid, widespread disease that exhibits both intraosseous and soft-tissue involvement. Commonly affected soft tissues include the skin, lung, liver, spleen, and lymph nodes. Although any bone may be involved, the skull, mandible, ribs, vertebrae, and femur are affected frequently. Treatment consists of multiagent chemotherapy with disappointing results and a poor prognosis.

The chronic disseminated form is nicknamed “Hand-Schüller-Christian disease” and demonstrates lesions in multiple bones with soft-tissue involvement. This pattern of histiocytosis may be encountered in patients over a wide age range, but most are children or young adults, with a definite male predilection. Bone lesions appear as sharply “punched-out” lytic defects, often with irregular margins. Infiltration of the alveolar bone often leads to severe horizontal bone loss and commonly exhibits involvement of the overlying soft tissues with ulcerative or proliferative gingival lesions. In advanced cases, the alveolar destruction gives a radiographic appearance of the teeth “floating in air.” Proliferative periostitis in the absence of a focus of infection occasionally is a sign of underlying bone involvement. Accumulations of Langerhans cells may occur in any bone, with the skull, ribs, vertebrae, and jaws leading the list. Sites of soft-tissue infiltration commonly include the skin, mucosa, lymph nodes, and viscera.

In cases of chronic disseminated Langerhans cell histiocytosis, local symptomatic sites are treated surgically, with radiation used for those that are inaccessible surgically. The prognosis is variable, with occasional cases demonstrating spontaneous resolution, whereas others are resistant to therapy, disseminate, and require use of systemic chemotherapy. The prognosis in patients with localized lesions is good, but it becomes less favorable with widespread soft-tissue and organ involvement.

- **Figure 11.19**
LANGERHANS CELL HISTIOCYTOSIS
Panoramic radiograph demonstrating multifocal areas of premature periodontal bone loss. Examination results of submitted soft tissue revealed Langerhans cell histiocytosis. (Courtesy of Dr. Robert Hobbs.)



- **Figure 11.20**
LANGERHANS CELL HISTIOCYTOSIS
Clinical photograph of a 16-year-old white male who presented with localized bone loss of the molar teeth in all four quadrants. This mirror view depicts the loss of attachment in the posterior maxilla on the right side.



- **Figure 11.21**
LANGERHANS CELL HISTIOCYTOSIS
Radiograph of the posterior maxillary dentition on the right side from the patient depicted in Figure 11.20. Note the significant loss of periodontal attachment of all three molar teeth.



Figs. 11.22, 11.23, and 11.24

The giant cell granuloma is a unique lesion of the oral cavity that occurs in gingival soft tissues or is located centrally within the jaws. Some lesions show both a soft-tissue and an intrabony component, and classification as to peripheral or central origin may be controversial. Most investigators believe that the giant cell granuloma is a nonneoplastic (reactive) lesion; however, some examples are similar histopathologically to the giant cell tumor of long bones and often behave like a true neoplasm. Whether or not true giant cell tumors occur in the jaws remains controversial, and some investigators believe giant cell granuloma and giant cell tumor represent two ends of a continuum of a single disease process. Many of the histopathologic features of giant cell granuloma suggest that the proliferation is an attempt at repair of an area of red blood cell extravasation and its contributing vessels.

Giant cell granulomas of the jaws occur in patients over a wide age range but are most common in the first three decades of life, with a slight female predilection. There is a 3:1 mandibular predominance with a predilection for occurrence in the anterior region, with occasional examples crossing the midline. Most cases involve the tooth-bearing areas, but examples may be found in the ascending ramus and tuberosity. Pain is not a common feature, and smaller lesions usually are discovered during a routine radiographic examination. Larger lesions tend to cause localized expansion of the affected bone, which is the most common presenting symptom. The radiologic findings are variable and may mimic those of several other odontogenic and nonodontogenic lesions.

Radiographically, giant cell granulomas produce a unilocular or multilocular radiolucency that may have sharp, well-defined or irregular, ragged margins. In a large radiographic review of giant cell granulomas, a direct correlation was seen between lesion size and the development of central loculations. The bone cortex may be thin and expanded or sometimes perforated by the lesion. Tooth displacement or resorption is not rare.

Most giant cell granulomas respond well to local curettage with few recurrences, and occasional reports have documented resolution following intralesional injection with corticosteroids. Some lesions, however, are locally aggressive and may require marginal resection for control. There is some degree of correlation between the clinical-radiologic-histopathologic features and behavior. Nonaggressive lesions generally show few or no symptoms, manifest slow growth, and do not demonstrate root resorption or cortical perforation. These lesions seldom recur after local curettage. The more aggressive forms of giant cell granuloma often are characterized by pain, rapid growth, root resorption, and cortical perforation. These lesions show a high prevalence of local recurrence after curettage and may require marginal resection to obtain a cure. In most instances, the histopathologic features of the giant cell granuloma are indistinguishable from those of cherubism and a brown tumor of hyperparathyroidism. Therefore, it is prudent to evaluate any patient with a central giant cell granuloma for these possibilities (see Figs. 11.25 and Figs 14.5 and 14.6).

- **Figure 11.22**
GIANT CELL GRANULOMA
Expansile mass of the left anterior maxilla.



- **Figure 11.23**
GIANT CELL GRANULOMA
Occlusal radiograph of patient in Figure 11.22, showing a destructive radiolucent lesion.



- **Figure 11.24**
GIANT CELL GRANULOMA
Multilocular radiolucent defect showing root resorption and tooth displacement in a 20-year-old female. (Courtesy of Dr. E. B. Bass.)



Figs. 11.25, 11.26, and 11.27

Cherubism is an inherited disorder that is transmitted as an autosomal-dominant trait, but it often appears as a spontaneous mutation. The chief clinical feature is a bilateral, painless expansion of the mandible, especially over the angles. The initial presentation may be apparent as early as 1 year of age, although milder cases may not become apparent clinically until age 12 years. Less commonly, symmetric maxillary lesions also may be present, usually in the tuberosities. In severe cases, the entire mandible and maxilla are involved, resulting in marked facial deformity. The term “cherubism” arose from the common facial distortion that resembles plump-cheeked little angels (cherubs). Significant involvement of the maxilla can result in stretching of the skin of the upper face to expose the sclerae below the iris of the eye, resulting in an “eyes upturned to heaven” appearance.

Radiographically, the lesions present as unilocular or, more often, multilocular lytic defects that histopathologically closely resemble giant cell granuloma. Bilateral radiolucencies with significant cortical expansion are typical; and in some cases, the lesions extend continuously from the posterior region of one jaw to the posterior area of the contralateral quadrant. Involvement of the angle and ascending ramus is present in a large percentage of those affected. Displacement and delayed eruption of teeth are seen frequently. A similar radiographic pattern has been noted in patients who demonstrate numerous clinical features of Noonan syndrome. Although many similarities exist, this latter disorder seems to be separate from both cherubism and Noonan syndrome. As with any intraosseous lesion that resembles giant cell granuloma, further evaluation should be performed to rule out a brown tumor of hyperparathyroidism.

Treatment of cherubism is not standardized and is somewhat uncertain. Some cases have responded well to early curettage; others have shown increased growth after surgical intervention. In other cases, the lesions tend to stabilize or even partially regress with advancing age, but considerable functional or cosmetic deformity may persist. Overall, in patients with obvious cosmetic problems, surgical curettage appears to accelerate return to clinical and radiographic normalcy. Radiation therapy is contraindicated.

► **Figure 11.25**
CHERUBISM

Characteristic facies of a young boy with cherubism. Extensive maxillary involvement has stretched the skin to expose the sclera, resulting in the “eyes upturned to heaven” appearance. (Courtesy of Dunlap C, Neville B, Vickers RA, et al. The Noonan syndrome/cherubism association. *Oral Surg Oral Med Oral Pathol* 1989;67:698.)



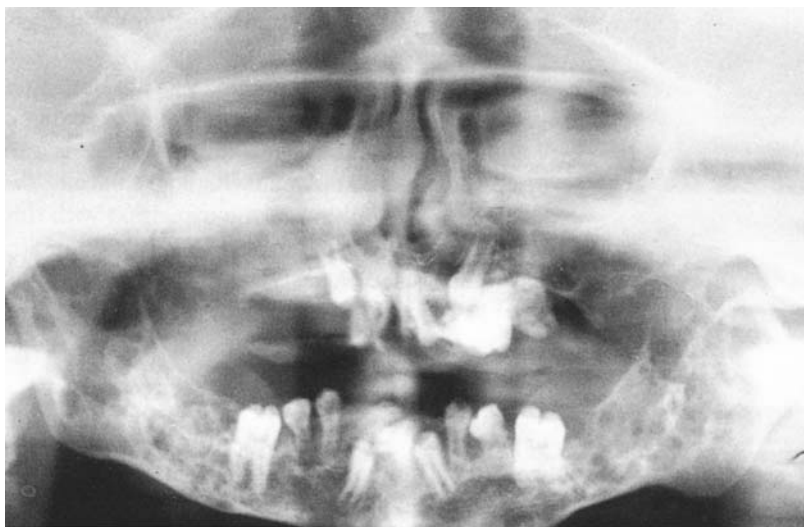
► **Figure 11.26**
CHERUBISM

Intraoral view of the patient shown in Figure 11.25 showing marked enlargement of the right maxilla and multiple missing teeth.



► **Figure 11.27**
CHERUBISM

Panographic radiograph of the patient shown in Figure 11.25 showing extensive multilocular radiolucent lesions of the mandible and maxilla. (Courtesy of Dunlap C, Neville B, Vicker RA, et al. The Noonan syndrome/cherubism association. *Oral Surg Oral Med Oral Pathol* 1989;67:698.)



SIMPLE BONE CYST (TRAUMATIC BONE CYST, HEMORRHAGIC BONE CYST)

Figs. 11.28 and 11.29

Although the name implies an epithelium-lined cavity, the simple bone cyst consists of an idiopathic cavity within bone that may be empty or that may contain only a small amount of serous or serosanguineous fluid. Although controversial, many authorities believe the process is secondary to intraosseous hemorrhage in which the hematoma liquefies, resulting in a cavity within bone. The biochemical profile and protein electrophoresis of fluid removed from simple bone cysts demonstrate significant similarity with serum. When present within the jaws, most of these cavities occur in the mandible, with most cases presenting in patients younger than 20 years of age.

Although rare cases may produce clinically obvious swelling or pain, most examples are asymptomatic and are discovered during a routine radiographic examination as a well-defined radiolucent lesion. Large, simple bone cysts frequently show a scalloped or domelike superior margin extending between the roots of the teeth. Prompt resolution typically follows surgical exposure performed for diagnostic purposes. On occasion, the lumen of true cystic lesions may resemble closely a simple bone cyst on exploration. Therefore, scraping of the lesional walls and submission of any removed soft-tissue fragments are advantageous for diagnostic confirmation.

Occurrence in association with benign fibro-osseous lesions, such as osseous dysplasia or fibrous dysplasia, is not rare. Such lesions often are more difficult to resolve via surgical exploration, presumably because of the continued presence of the associated fibro-osseous process.

ANEURYSMAL BONE CYST

Fig. 11.30

Aneurysmal bone cyst is an intrabony accumulation of blood-filled spaces surrounded by reactive fibrous connective tissue that is not lined by either epithelium or endothelium. In many cases, the connective tissue wall contains numerous giant cells and may resemble closely a giant cell granuloma. Although the cause is idiopathic, many investigators believe the process may represent a secondary reaction to an intraosseous hematoma. Some aneurysmal bone cysts seem to arise *de novo*, whereas others are the result of the development of abnormal vascular communications within a preexisting bone lesion, such as fibrous dysplasia, osseous dysplasia, ossifying fibroma, and giant cell granuloma. Aneurysmal bone cyst, traumatic bone cyst, and giant cell granuloma share many similar features and may represent portions of a spectrum related to intraosseous vascular disturbances.

Although aneurysmal bone cysts are not rare in long bones, the process rarely occurs in the jaws. Most examples present in patients younger than age 20 years, and they are somewhat more common in the mandible. Clinically, there is a sudden increase in the size of the affected bone, often accompanied by pain. Radiographically, the lesion may present as a unilocular radiolucency or, more commonly, as a lytic expansion with thinning of the cortical plate and a honeycombed or soap-bubble appearance. The loculated radiographic appearance is not caused by true septa but rather is secondary to spurs and ridges on the inner surface of the expanded cortical plate.

On surgical exposure, the lesion usually resembles a blood-filled sponge from which slow hemorrhage may be evident. Although excessive hemorrhage during surgery has been seen in extra-gnathic lesions, extensive blood loss typically is not seen in aneurysmal bone cysts within the jaws. On curettage, hemorrhage may be abundant, but it is not difficult to control. Although venous-appearing blood may well up from the spongy tissue, removal of the bulk of the lesion usually controls the bleeding. Although surgical curettage is the treatment of choice, recurrence is not rare in those that are excised incompletely. In large or persistently recurrent lesions, surgical resection often combined with cryotherapy is necessary; radiation is contraindicated. Occasional successes have been obtained with preoperative angiography and embolization of the feeder vessels. Overall, the prognosis is good.

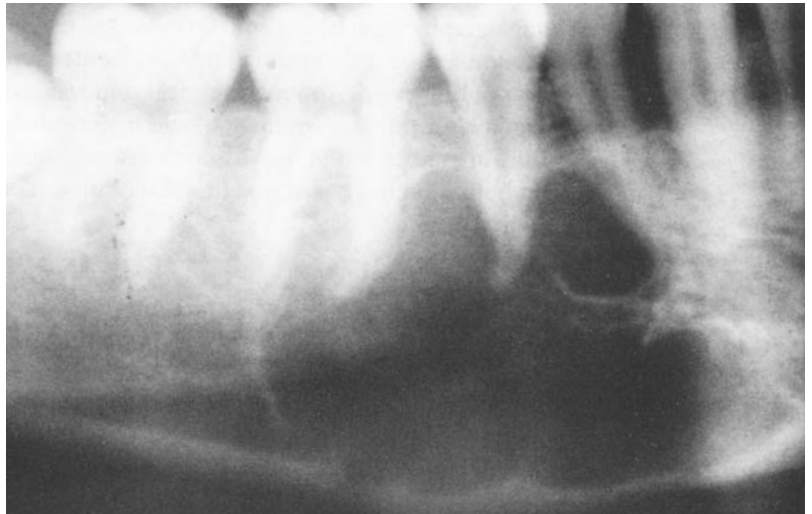
► **Figure 11.28**
SIMPLE BONE CYST

Well-defined periapical radiolucency associated with the right mandibular cuspid. Pulp vitality was within normal limits. (Courtesy of Dr. C. W. Topp.)



► **Figure 11.29**
SIMPLE BONE CYST

Large radiolucency of the right body of the mandible that scallops in between teeth and thins the inferior border of the mandible. (Courtesy of Dr. Patrick Coleman.)



► **Figure 11.30**
ANEURYSMAL BONE CYST

Large, multiloculated lesion of the posterior mandible in a 24-year-old woman. This was associated with buccal swelling.



Fig. 11.31

Intrabony hemangiomas are encountered most frequently in the craniofacial skeleton, and significant numbers of these vascular lesions involve the jaws, with the predominance occurring in the mandible. These lesions may be capillary or cavernous and represent hamartomas, reactive proliferations, or true neoplasms. Cases have been recorded in patients ranging from infancy to old age, with a peak prevalence in the third decade of life and a definite female predilection.

Although intraosseous hemangiomas often are asymptomatic, some may demonstrate signs or symptoms such as pain, paresthesia, swelling, mobile teeth, or localized gingival hemorrhage. Auscultation of a bruit is unusual without extension of the vascular lesion into the soft tissue overlying the affected bone. Although most lesions usually present as ill-defined and sometimes honeycombed lytic defects, the radiographic findings are variable and, unfortunately, nonspecific. Resorption or displacement of adjacent teeth and cortical expansion or sunburst trabeculation are not rare.

Failure to consider a central vascular lesion before intervention has resulted in severe bleeding and even exsanguination after incisional biopsy or extraction of teeth. Needle aspiration and angiography have been used as important diagnostic adjuncts before invasive procedures. Appropriate therapy is determined by the vascular supply, the age of the patient, and the size and location of the lesion. Although surgical removal is the leading therapeutic approach, cryotherapy, argon laser therapy, irradiation, and injection of sclerosing agents also have been used successfully in eradication of intraosseous hemangiomas. In significantly vascularized lesions, presurgical embolization has been used to reduce subsequent surgical hemorrhage.

ARTERIOVENOUS MALFORMATION

Figs. 11.32 and 11.33

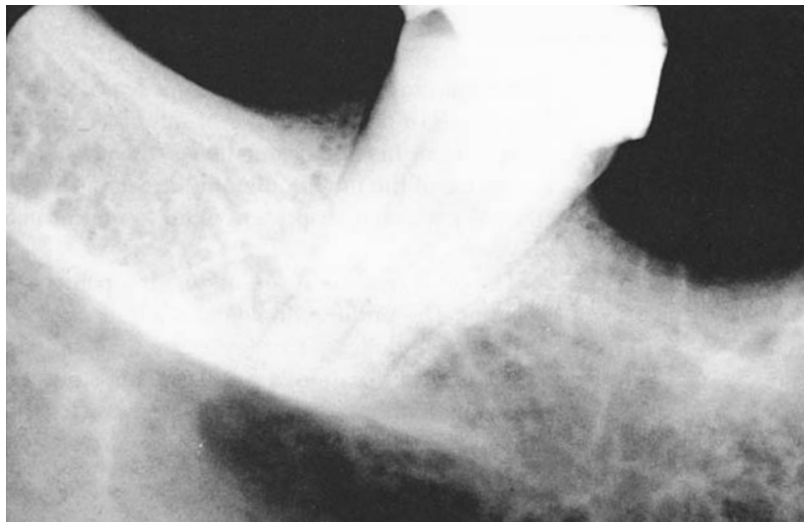
An arteriovenous malformation represents an abnormal communication between the arterial and venous circulation, with the blood flow bypassing the capillary circulation. Arteriovenous malformations may be confused with hemangiomas and have been noted in both the oral soft tissues and the jawbones. In fact, some authorities believe hemangiomas of bone are rare, and most high-flow examples likely represent arteriovenous malformations. These anomalies may be congenital or acquired, most often as a result of trauma.

Clinically, intrabony arteriovenous malformations of the jaws typically are high-flow lesions and frequently demonstrate progressive pain, swelling of the bone, spontaneous gingival hemorrhage, tooth displacement or mobility, pulsation, and discoloration or hyperthermia of the overlying mucosa. Aspiration of intrabony lytic lesions always is a good practice; but if suspicion of a vascular malformation is high, arteriography without aspiration is most appropriate. Some lesions may exhibit no significant radiographic change; in other cases, a soap-bubble or honeycomb lucency may be seen.

Most cases are treated with surgical excision after preoperative embolization of the feeder vessels. Less frequently, permanent embolic obliteration has been performed by precise placement of an occlusive material within the central vasculature of the lesion. In surgically treated cases, extensive preoperative planning with use of quality angiography is mandatory. If blood flow is blocked in the proximal portion of a feeder vessel without simultaneously blocking the flow within the malformation, the residual feeder vessels compensate and maintain significant flow. In several reports, ligation of the external carotids without controlling flow through the malformation has been followed by extensive hemorrhage and fatal exsanguination on surgical exploration. Surgical removal of the lesion preferably is performed within 24 to 48 hours after completion of successful embolization. Although not ligated, the external carotid should be exposed at the time of surgery for ready access if uncontrollable hemorrhage is encountered during the procedure. Following successful resection, the defect usually is reconstructed immediately, occasionally by replantation of the resected segment after removal of the pathologic soft tissue. A postsurgical arteriogram should be performed after 1 year to detect any possible recurrence.

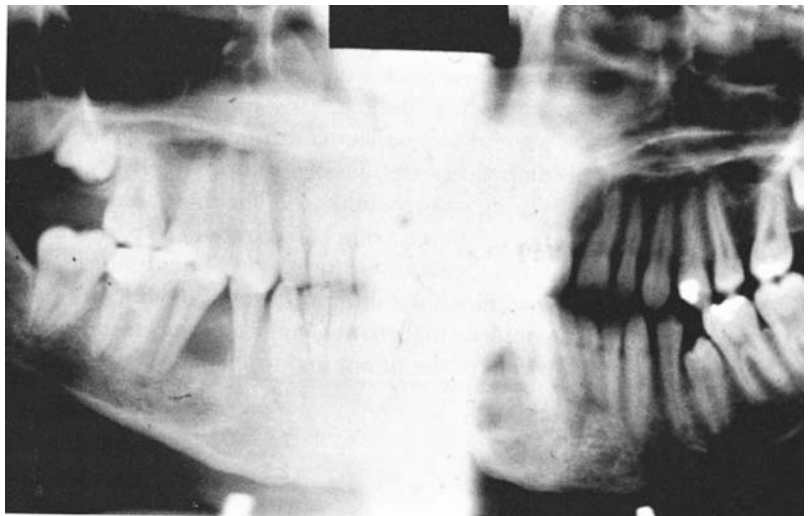
► **Figure 11.31**
HEMANGIOMA

Irregular honeycombed radiolucent lesion in the mandible of a 65-year-old male. Biopsy of this lesion resulted in severe bleeding.



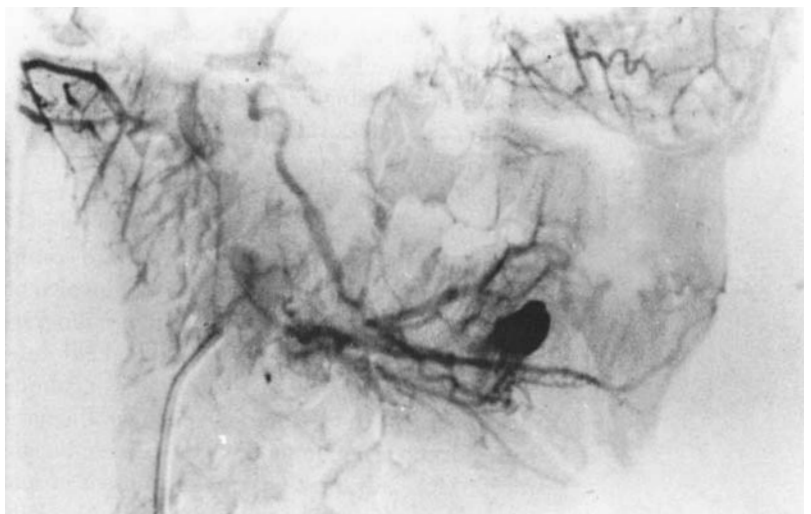
► **Figure 11.32**
ARTERIOVENOUS MALFORMATION

Panographic radiograph showing a radiolucent lesion between the mandibular premolar teeth. This lesion was believed to be a lateral periodontal cyst, but pulsation was noted on clinical examination. (Courtesy of Dr. H. W. Allsup.)



► **Figure 11.33**
ARTERIOVENOUS MALFORMATION

Subtraction angiogram of the patient shown in Figure 11.32 showing the vascular defect in the premolar area. (Courtesy of Dr. H. W. Allsup.)



DESMOPLASTIC FIBROMA (FIBROMATOSIS OF BONE)

Fig. 11.34

Desmoplastic fibroma is a neoplasm of fibroblastic cells that is histopathologically similar to the fibromatosis or desmoid tumor of soft tissues. Although most of these intraosseous fibrous neoplasms occur in various long bones, the jaws are not a rare site of involvement, with the mandible being one of the most commonly affected single bones. Although maxillary lesions may be seen, most of jaw examples present in the posterior mandible in patients with a mean age of less than 16 years.

The most common presentation is a painless swelling that may be associated with limited jaw opening. The proliferation creates a unilocular or multilocular lytic lesion that may be circumscribed or ill-defined. Cortical expansion and perforation with soft-tissue extension are not rare. On cortical perforation, a soft-tissue mass often is evident. Although metastasis is not seen, desmoplastic fibromas are locally aggressive neoplasms that tend to permeate bone and infiltrate soft tissue. Appropriate therapy is assured surgical removal.

INTRAOSSEOUS NEUROFIBROMA

Fig. 11.35

Neurofibromas are slow-growing, benign neoplasms of nerve sheath origin. Intraosseous origin is uncommon; when noted, these tumors usually are solitary, without any signs of neurofibromatosis. When intraosseous examples are seen, the most commonly affected site in the entire skeleton is the mandible. Most of these tumors originate from the inferior mandibular nerve and present as a unilocular or multilocular radiolucency that rarely may demonstrate central radiopaque areas. These tumors demonstrate a female predominance and most frequently arise during young adulthood. The most common clinical presentation is swelling of the affected bone, often accompanied by pain or paresthesia.

Malignant transformation of neurofibromas is possible, but it most frequently is noted in association with neurofibromatosis. In addition, partial removal of solitary neurofibromas does not promote malignant change and may lead to regression. The recommended therapy is surgical excision of the tumor and its extension along the nerve.

OSSEOUS DYSPLASIA (PERIAPICAL CEMENTO-OSSEOUS DYSPLASIA, CEMENTOMAS)

Fig. 11.36

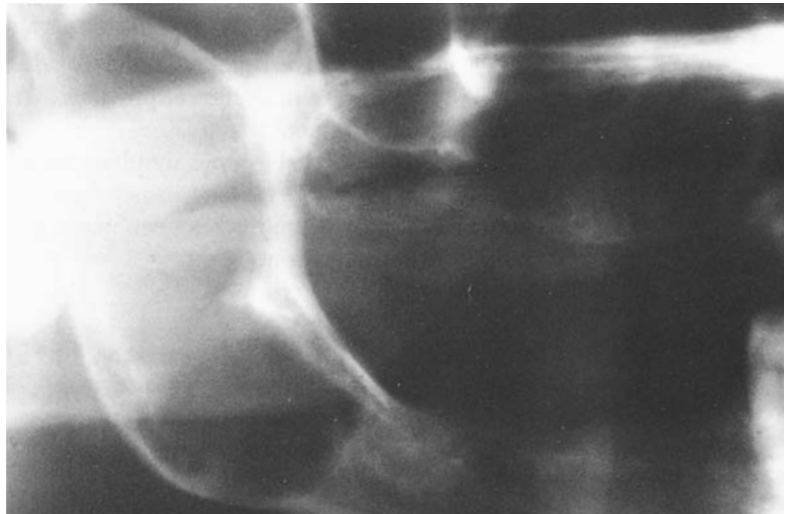
Osseous dysplasia is a common, idiopathic, and nonneoplastic fibro-osseous replacement of bone that occurs only in the tooth-bearing areas of the jaws. Although its diverse clinical presentation has been known by a variety of names in the past, it finally is being clarified as a unified disease process under a single term. Currently, there are three recognized patterns of presentation: periapical, focal, and florid. However, these variants often demonstrate overlapping features. Osseous dysplasia has a predilection for black females between the ages of 30 and 50 years. Outside the United States, a significant number of cases also are noted in middle-aged Oriental females.

The most common form of osseous dysplasia is the periapical variant (periapical cemento-osseous dysplasia). The lesions usually are confined to bone surrounding the apices of the mandibular incisor teeth, but they also can involve other teeth. Typically, the condition is asymptomatic and is detected during a routine radiographic examination. Radiographically, the initial lesion appears as a circumscribed radiolucent defect about the apex of a tooth. The process may involve multiple apices that occasionally coalesce to form linear lytic areas enveloping multiple adjacent teeth. In the radiolucent stage, the alteration cannot be differentiated radiographically from chronic apical inflammatory disease, but the associated teeth are vital and seldom are restored. More advanced lesions show areas of mineralization within the radiolucent area; in the final stage, the lesion shows a dense central calcification often surrounded by a narrow radiolucent rim. Individual lesions seldom exceed 1.5 cm in diameter, with little tendency for progressive enlargement. Because the condition is benign and usually can be diagnosed on a clinical basis, biopsy is contraindicated and no treatment is necessary.

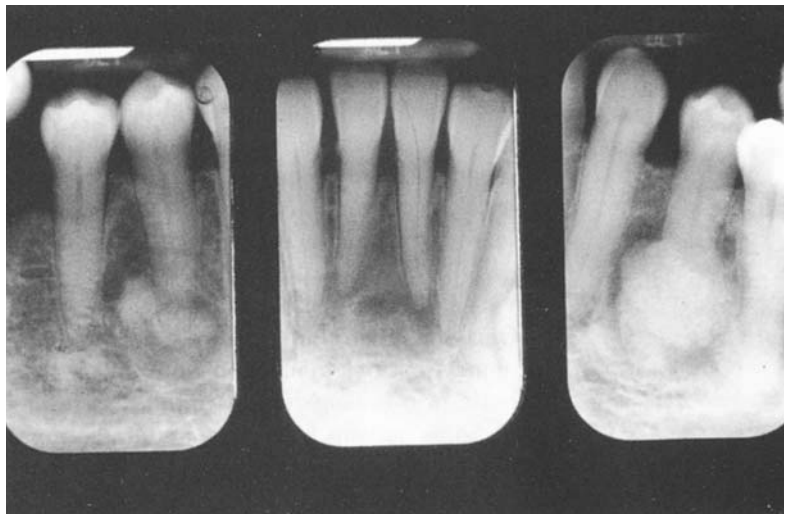
- **Figure 11.34**
DESMOPLASTIC FIBROMA
Irregular radiolucency of the anterior maxilla in a 13-year-old female. (Courtesy of Dr. T. H. Daniel.)



- **Figure 11.35**
INTRAOSSEOUS NEUROFIBROMA
Large radiolucency of the right mandibular ramus. (Courtesy of Dr. Paul Allen.)



- **Figure 11.36**
OSSEOUS DYSPLASIA
Periapical radiographs demonstrating multiple mixed radiolucencies of the anterior mandible. (Courtesy of Dr. Ken Peavy.)



FOCAL OSSEOUS DYSPLASIA (FOCAL CEMENTO-OSSEOUS DYSPLASIA)

Fig. 11.37

The second pattern of osseous dysplasia is known as the focal variant. About 80% occur in females in the fourth and fifth decades of life, and the condition has a relatively greater prevalence in blacks than whites. Of the three types of osseous dysplasia, the focal variant is the most common pattern discovered in Caucasians and often does not reveal future development of additional lesions. The affected areas are singular, generally asymptomatic, and often located in posterior regions of the tooth-bearing areas of the jaws. Frequently, the lesion develops at the site of a previous tooth extraction. Like the periapical variant, focal osseous dysplasia demonstrates several radiographic stages of development that reflect an increasing amount of mineralized product being formed within the lesion. It begins as a largely radiolucent defect, progressing to a mixed radiolucency, and finally becoming an almost totally radiopaque mass with a thin peripheral lucent border.

During its early radiolucent or mixed stage of maturation, focal osseous dysplasia may be confused easily with an ossifying fibroma because these processes demonstrate similar radiographic and histopathologic features. The most characteristic distinguishing feature is the appearance of these pathoses on surgical exploration. Focal osseous dysplasia exhibiting significant radiolucency is friable and difficult to remove intact, whereas ossifying fibroma is an adherent mass that separates cleanly from the surrounding bone. In addition, focal osseous dysplasia frequently is located in apical areas or in sites of previous extraction, whereas ossifying fibroma demonstrates no relationship to either.

The diagnosis of focal osseous dysplasia sometimes can be made on the basis of its clinical and radiographic features. Biopsy is necessary in other cases to rule out other more significant pathoses. Once the diagnosis has been made, only periodic observation is required because continued growth usually is not seen.

FLORID OSSEOUS DYSPLASIA (DIFFUSE OSSEOUS DYSPLASIA, FLORID CEMENTO-OSSEOUS DYSPLASIA)

Figs. 11.38 and 11.39

Florid osseous dysplasia exhibits demographic and radiographic features that are similar to the periapical and focal forms of the condition. However, the florid form typically affects multiple sites in the posterior quadrants of either jaw, sometimes in association with identical periapical involvement of the anterior mandible. The process may be completely asymptomatic or may be associated with pain, an alveolar-mucosal fistula, and sequestration. The radiographic changes exhibit similarities with those seen in Paget's disease (see Figs. 11.13, 11.14, and 11.15), but patients with osseous dysplasia exhibit involvement of only the tooth-bearing areas of the jaws and usually do not reveal generalized expansion of the affected bone. Although cortical expansion typically is not seen, formation of simple bone cysts or aneurysmal bone cysts within the altered bone is not rare and may be associated with enlargement of the affected area. Generally, it is agreed that the clinical and radiographic features are sufficient for diagnosis; surgical intervention is not indicated and even may be contraindicated in the final radiopaque stage.

Once the lesions have matured into the predominantly radiopaque phase, the altered bone is extremely sensitive to inflammation. Inflammation introduced by periodontal disease, periapical pathosis, or overlying denture trauma can result in necrosis and sequestration of the sclerotic bone. Some patients have presented to their oral health care practitioner with several contiguous teeth embedded in necrotic, sclerotic bone that underwent sequestration and was removed by the affected individual.

Prophylactic removal of the altered bone would be problematic and without guarantee of redevelopment. The best therapy is maintenance of the dentition in an inflammatory-free state. Without introduction of inflammation, the altered bone remains asymptomatic. If all teeth are removed and replaced with complete dentures, normal ridge resorption often results in exposure and ultimate sequestration of the sclerotic masses. If infection does develop, saucerization of the dead bone may be required.

► **Figure 11.37**

FOCAL OSSEOUS DYSPLASIA

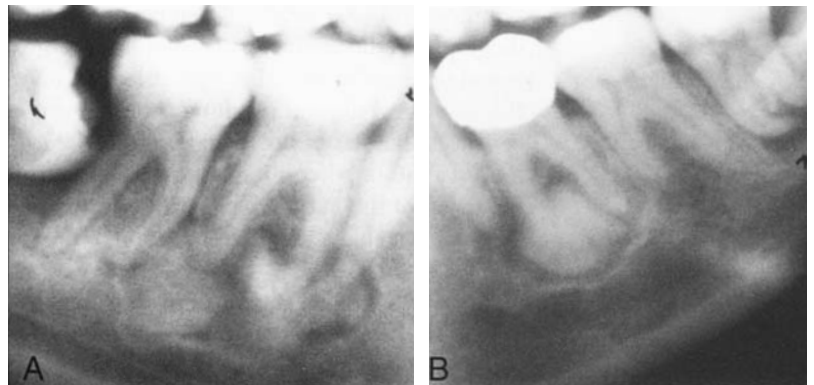
A mixed radiolucent/radiopaque lesion in the edentulous second molar area of a 50-year-old female. (Courtesy of Waldron CA. Fibro-osseous lesions of the jaws. J Oral Maxillofac Surg 1985;43:249.)



► **Figure 11.38**

FLORID OSSEOUS DYSPLASIA

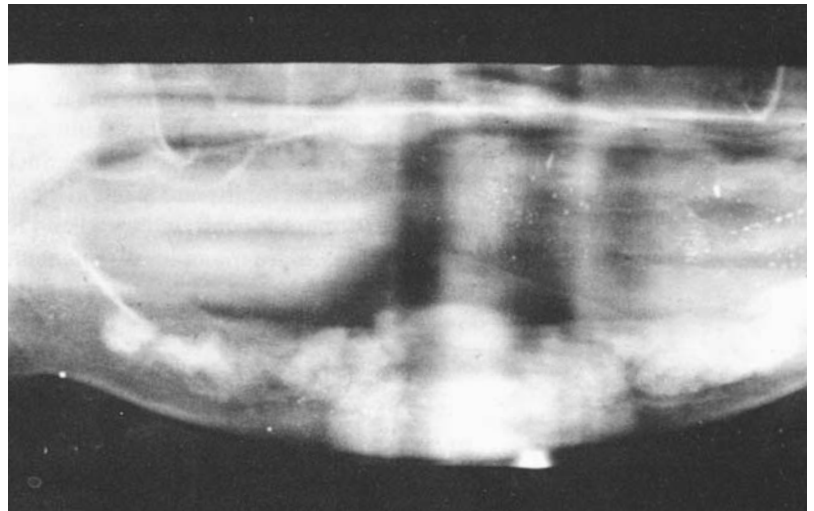
(A) Well-defined mixed radiolucency enveloping the apices of the mandibular first molar on the right side. (B) Contralateral side of the same patient exhibiting mixed radiolucency enveloping apices of the first molar and lytic lesion adjacent to the roots of the third molar.



► **Figure 11.39**

FLORID OSSEOUS DYSPLASIA

Extensive mandibular lesions in a 62-year-old black female. The lesions were asymptomatic and detected when radiographs were made before denture construction. (Courtesy of Waldron CA. Fibro-osseous lesions of the jaws. J Oral Maxillofac Surg 1985;43:249.)



Figs. 11.40, 11.41, 11.42, and 11.43

Fibrous dysplasia is considered a developmental, tumorlike (hamartomatous) lesion of bone. The disease may involve several or many bones (polyostotic fibrous dysplasia) or may be localized to a single bone (monostotic fibrous dysplasia). Polyostotic fibrous dysplasia is relatively rare and may, in some cases, involve more than 60% of the skeleton, with the mandible and craniofacial bones being some of the most commonly involved. Some affected individuals also show areas of skin pigmentation and a variety of endocrine abnormalities (McCune-Albright syndrome). Extensive polyostotic disease frequently results in pathologic fractures and severe bone deformity. Monostotic fibrous dysplasia is more commonly encountered. Maxillary fibrous dysplasia, although commonly included in this group, is not strictly monostotic because the process commonly involves a group of contiguous bones separated by sutures and is described more appropriately as craniofacial fibrous dysplasia.

A painless enlargement of the affected bone arising during childhood is the most common presenting symptom, and it may slowly increase to produce considerable deformity. Most examples are detected during the first two decades of life, although milder examples may not produce symptoms until later in life. When occurring in the jaws, there is a maxillary predominance. Radiographically, the characteristic finding is a “ground-glass” appearance best appreciated on periapical or occlusal views. Mandibular lesions may appear multilocular on lateral projections, but this is an optical illusion resulting from uneven endosteal erosion caused by the lesion, which contrasts with areas of preserved cortex. The area seldom is well-defined radiographically and tends to blend imperceptibly into adjacent normal bone. Complex maxillary lesions tend to cause obliteration of the maxillary sinus and involve the zygoma and orbit. Lateral skull films often demonstrate a characteristic thickening of the base of the skull involving the occipital, sphenoid, and frontal bones.

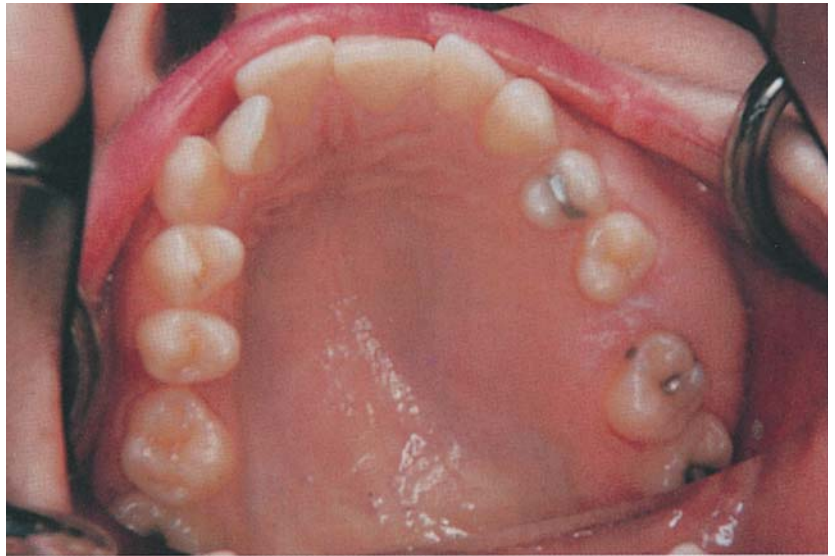
Most, but not all, examples of fibrous dysplasia of the jaws tend to stabilize and cease growing in adult life; accordingly, most therapeutic interventions are delayed until after the pubertal growth spurt and longer if possible. Milder forms of the disease may not require treatment, whereas other patients require one or more surgical reductions for correction of the cosmetic or functional deformity. The response to surgical contouring is variable, with some patients requiring multiple procedures before stabilization. Rare cases demonstrate accelerated growth and require resection for clinical improvement.

Although controversial, some surgeons recommend radical surgical excision and primary reconstruction of any cases of craniofacial fibrous dysplasia in which the surgery would not cause a greater functional or aesthetic disturbance. Such aggressive surgery is thought to prevent recurrence, stop further deformity, and avoid malignant transformation.

Radiation treatment is contraindicated and has been associated with development of postradiation bone sarcoma. Rare examples of “spontaneous” sarcomatous transformation also have been reported. The prevalence of malignant transformation is approximately 0.4%, 400 times the rate noted in normal individuals.

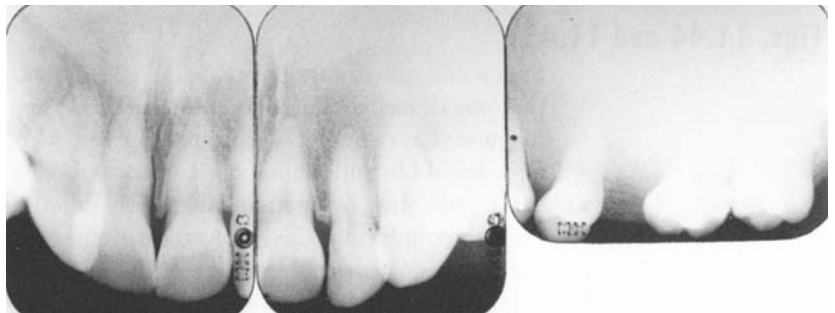
► **Figure 11.40**
FIBROUS DYSPLASIA

Painless enlargement of the maxilla in a 32-year-old female. The enlargement was first detected when she was a teenager, and it has not grown appreciably in the past 6 years.



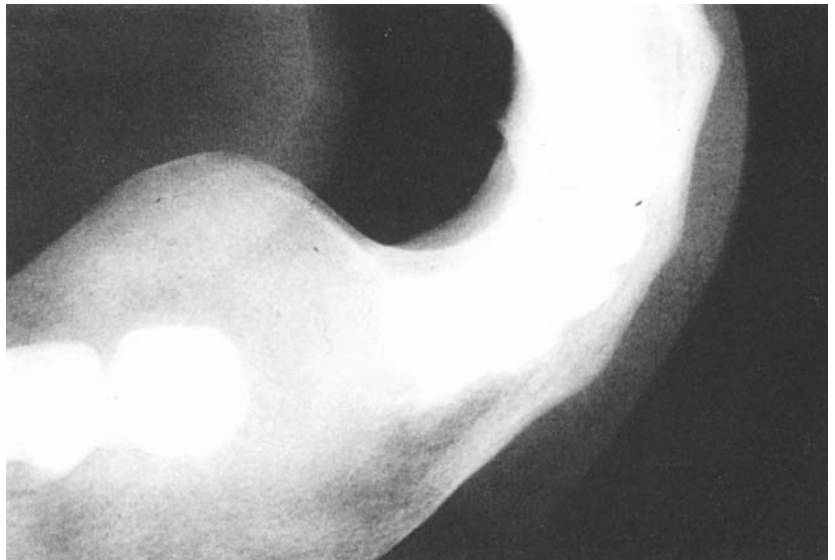
► **Figure 11.41**
FIBROUS DYSPLASIA

Periapical radiographs of the patient shown in Figure 11.40 showing the characteristic "ground-glass" appearance of the lesional area with extension into the maxillary sinus. The altered bone pattern blends into a normal bone pattern in the incisor area. (Courtesy of Waldron CA. Fibro-osseous lesions of the jaws. J Oral Maxillofac Surg 1985;43:249.)



► **Figure 11.42**
FIBROUS DYSPLASIA

Expansile lesion of the mandible in a 16-year-old boy.



SEGMENTAL ODONTOMAXILLARY DYSPLASIA (HEMIMAXILLOFACIAL DYSPLASIA)**Figs. 11.44 and 11.45**

Segmental odontomaxillary dysplasia is a unilateral developmental abnormality of the maxilla that was described initially under the term “hemimaxillofacial dysplasia” and often has been misdiagnosed as craniofacial fibrous dysplasia. Most affected patients present in the first two decades of life and demonstrate unilateral enlargement of the maxilla and the overlying gingival soft tissues. In a few patients, facial asymmetry and unilateral hypertrichosis are seen.

Radiographically, the involved quadrant exhibits an increased radiodensity with thickened bony trabeculae, decreased size of the adjacent maxillary sinus, one or more missing premolars, abnormal spacing of the erupted teeth, and delayed eruption of adjacent permanent teeth. Deciduous teeth within the zone of altered bone often demonstrate one or more developmental abnormalities, such as enlarged crowns, enlarged roots, splayed roots, or enlarged pulps.

Segmental odontomaxillary dysplasia seems to remain clinically and radiographically stable and does not require surgical intervention. Therapy is directed toward achieving adequate dental function and aesthetics in the involved quadrant.

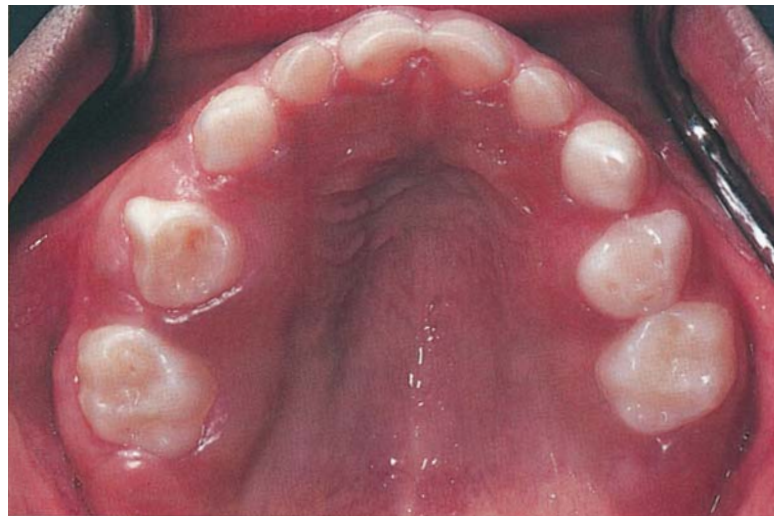
► **Figure 11.43**
FIBROUS DYSPLASIA

Computed tomography of a patient with unilateral craniofacial fibrous dysplasia of the maxilla that expands the alveolar ridge on the right side. (Courtesy of Dr. Craig Little.)



► **Figure 11.44**
SEGMENTAL ODONTOMAXILLARY DYSPLASIA

Mirror view of maxillary arch. Note the unilateral enlargement of the alveolar ridge and the widely spaced dentition.



► **Figure 11.45**
SEGMENTAL ODONTOMAXILLARY DYSPLASIA

Periapical radiograph of the left side of the maxilla from the patient depicted in Figure 11.44. Note the coarse trabecular pattern, the absence of the first premolar, and the widely spaced dentition.



Figs. 11.46, 11.47, and 11.48

Ossifying fibromas are true neoplasms of bone that demonstrate a mixture of cellular fibrous connective and cellular bone. On occasion, these tumors reveal psammomatoid differentiation in which acellular droplets of mineralized material are noted that previously have been termed “cementum.” Because this pattern is present in ossifying fibromas outside of the jaws, it is recognized that this material is a variation of bone and not odontogenic. Therefore, the use of “cementifying fibroma” as a diagnostic term is being discontinued.

True ossifying fibromas are rare, and many jaw lesions that were previously assigned that diagnosis actually represent focal osseous dysplasia. Only through recent research have definitive criteria begun to emerge. As mentioned in the discussion of osseous dysplasia, ossifying fibroma is an adherent tumor that separates well from the surrounding bone, is not restricted to the tooth-bearing areas of the jaws, and does not occur predominantly in middle-aged black females.

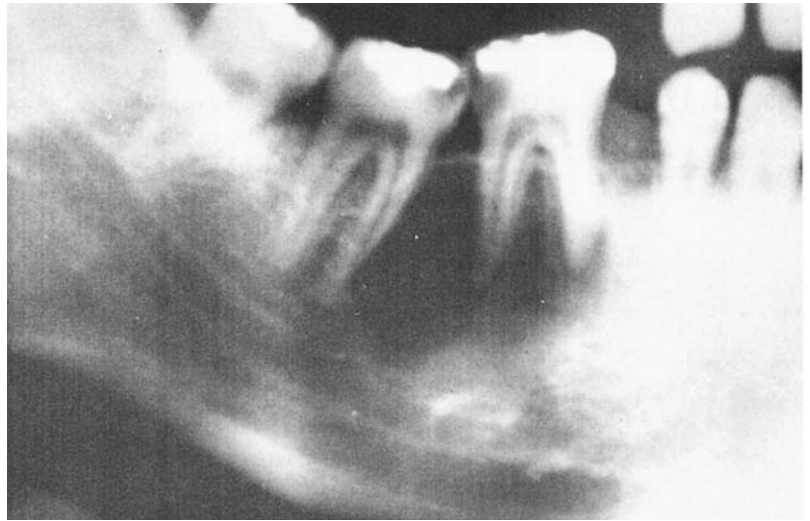
Ossifying fibromas are somewhat more common in females, exhibit a mandibular predominance, and most frequently arise before 40 years of age. Smaller lesions are asymptomatic and usually are discovered during a radiographic examination, whereas larger ossifying fibromas produce a painless enlargement of the jaw. Some examples have reached a large size, resulting in considerable facial deformity.

The tumor is well-circumscribed, may be encapsulated, and typically is well-defined radiographically. Depending on the degree of mineralization in the tumor, the lesion may vary radiographically from completely radiolucent to a mixed radiolucent/radiopaque process. Teeth adjacent to the tumor may be displaced, but root resorption seldom is noted.

Ossifying fibromas show variable clinical behavior. Some have limited growth potential, and others may be aggressive clinically and show rapid or more extensive growth. These latter types sometimes are termed “juvenile,” “active,” or “aggressive” ossifying fibromas and most commonly are seen in patients younger than 15 years. Although the histopathologic criteria for juvenile ossifying fibromas are poorly defined and controversial, many exhibit significant psammomatoid differentiation. Most ossifying fibromas respond well to enucleation or curettage, and the prognosis is good. Larger and more clinically aggressive lesions may require marginal resection and bone grafting to eliminate the disease.

► **Figure 11.46**
OSSIFYING FIBROMA

Circumscribed radiolucent defect in the mandibular molar area of a 28-year-old female. Localized expansion of the alveolus was present.



► **Figure 11.47**
OSSIFYING FIBROMA

Surgical specimen from the lesion shown in Figure 11.46. The tumor was encapsulated and separated with ease from the bony defect.



► **Figure 11.48**
JUVENILE OSSIFYING FIBROMA

A magnetic resonance image of an enlargement of the maxilla on the left side in a 6-year-old black male. Note the tumor nearly filling the maxillary sinus.



OSTEOBLASTOMA

Figs. 11.49

Osteoblastoma is a true neoplasm of osteoblasts that typically exhibits plump, pleomorphic cells intermixed with irregular bone similar to that occasionally seen in osteosarcomas. This uncommon skeletal tumor rarely is seen in the jaws and demonstrates a mandibular predominance. Most osteoblastomas arise in patients between 10 and 20 years of age and are diagnosed more frequently in males (2:1). Pain is a fairly common symptom; in small lesions, the discomfort often is nocturnal and relieved by salicylate treatment. In addition, sensitivity to percussion may be seen and has been responsible for clinical misdiagnosis as periapical inflammatory disease. Radiographic findings are variable and consist of combinations of radiolucency and radiopacity. Some osteoblastomas present as circumscribed or ill-defined radiolucent areas that may contain focal areas of calcification, whereas others have shown a predominantly radiopaque mass surrounded by a thin radiolucent rim. Although some osteoblastomas may be aggressive locally, may recur, or may be associated with sarcomatous transformation, most tumors follow a benign course and respond to local curettage or en bloc resection.

CEMENTOBLASTOMA

Fig. 11.50

Cementoblastoma is a true neoplasm of osteoblastlike cells thought to be derived from cells of the inner dental follicle that are termed “cementoblasts.” Although many consider osteoblastoma and cementoblastoma to represent one pathologic process, others believe that cementoblastoma should remain as a separate entity because of its unique ability to fuse with dental hard tissue. From a practical standpoint, the diagnosis of cementoblastoma is made when the tumor is fused to the root surface of a tooth and demonstrates histopathologic features of an osteoblastoma.

Cementoblastomas most commonly arise in patients younger than age 30 years, with most cases affecting the mandibular first molars or premolars. These slow-growing tumors typically produce cortical expansion and often are associated with pain at night that can be relieved by aspirin. Although the lesion rarely may be radiolucent or mixed, most present as a radiopaque mass with a thin radiolucent border.

Typically, therapy involves surgical removal of the affected tooth along with the attached tumor. Incomplete removal usually results in recurrence. Occasionally, endodontic therapy followed by root resection coronal to the radicular tumor has been used successfully in examples that involved surgically accessible teeth.

EWING'S SARCOMA

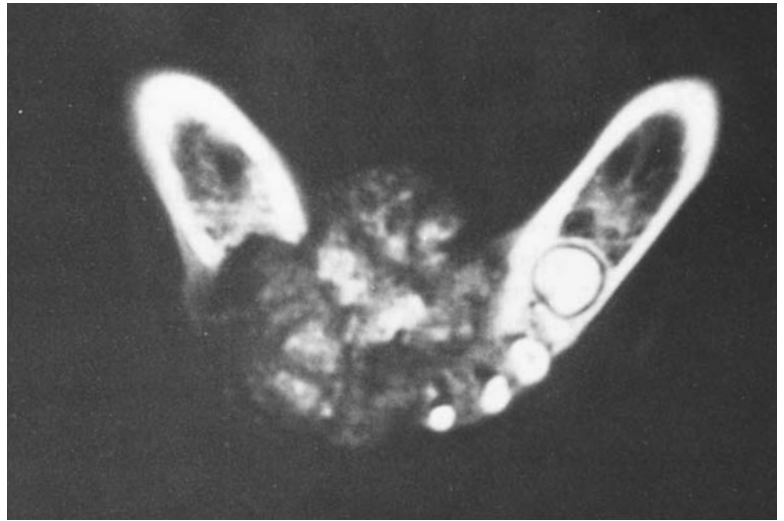
Fig. 11.51

Ewing's sarcoma is a malignancy of uncertain histogenesis that arises predominantly in bone and consists of a proliferation of undifferentiated small cells. Although these sarcomas account for 10 to 15% of all primary bone sarcomas, only about 2% occur in the jaws. When disseminated, tumor involvement of multiple bones is not rare. On discovery in the jaws, thorough evaluation should be performed to rule out a distant primary, but approximately 90% of reported cases in the jaws initially involved that location. Most of these cancers are seen in Caucasian children and adolescents, with less than 6% of cases noted in patients older than age 20 years.

The tumor often is associated with significant necrosis and may be confused with an inflammatory process. Pain and swelling often are early complaints. Radiographically, the tumor shows an irregular osteolytic lesion with or without cortical expansion. Although not commonly detected in the jaws, Ewing's sarcoma frequently produces laminated periosteal hyperplasia (onion-skinning) of the cortical bone overlying the tumor bed. When present in the jaws, widening of the periodontal ligament space; destruction of unerupted tooth follicles; and tooth displacement, mobility, and resorption may be seen; swelling and erythema of the overlying soft tissues are not rare. The prognosis of Ewing's sarcoma has shown a dramatic improvement in recent years. Radiation therapy and multiple drug chemotherapy have resulted in 90% local control and 40% long-term survival rates. When compared with mandibular lesions, primary maxillary tumors have been associated with faster growth and a greater resistance to therapy.

► **Figure 11.49**
OSTEOBLASTOMA

Computed tomography of an ill-defined mixed radiolucency of the anterior mandible in a 3-year-old Caucasian male. (Courtesy of Dr. Ed Marshall.)



► **Figure 11.50**
CEMENTOBLASTOMA

Panoramic radiograph exhibiting a radiopaque mass attached to the distal root of the left mandibular first molar. (Courtesy of Dr. John Wright.)



► **Figure 11.51**
EWING'S SARCOMA

A 15-year-old girl with an ill-defined radiolucent lesion of the tuberosity causing extrusion of the maxillary second molar. She presented with facial swelling that was believed to be caused by periapical inflammatory disease. (Courtesy of Damm DD, White DK, Drummond JF, et al. Ewing's tumor of the jaws. *Pediatr Dent* 1985;7:57.)



Figs. 11.52 to 11.57

Osteosarcoma is a primary malignancy of bone-forming cells and is the most frequent type of primary cancer of bone, but it still represents an uncommon tumor. About 7% of all osteosarcomas occur in the jaws, and they tend to occur in older patients more often than do osteosarcomas of the extragnathic skeleton. The mean age (36 years) for patients with osteosarcoma of the jaws is about a decade and a half greater than that recorded for osteosarcomas of the long bones. The disease is somewhat more common in males than in females, with the mandible and maxilla being about equally involved.

Swelling and pain are the most common initial complaints, followed by loosening of teeth, paresthesia, and nasal obstruction in the case of maxillary tumors. Some patients report symptoms for relatively long periods, suggesting slow growth of the tumor. Depending on the degree of mineralization of the neoplastic osteoid produced by the tumor, the radiographic findings may reveal a densely sclerotic process, a mixed radiolucency, or a completely radiolucent process. The margins of the lesion are ill-defined. Penetration of the cortex with growth of the tumor outside the cortex results in the radiographic appearance of trabeculae of new bone radiating out from the bone surface, the so-called “sunburst” appearance. This radiographic feature is observed in about 25% of jaw osteosarcomas and is best appreciated on an occlusal view of a mandibular lesion. Teeth involved in the area frequently show irregular (“spiking”) root resorption.

In their early stages, jaw osteosarcomas may present only subtle radiographic changes, consisting only of a slight variation in the trabecular pattern. An important radiographic change consisting of a symmetric widening of the periodontal ligament space around one or several teeth may be noted in some cases but is not specific for osteosarcoma. This has been shown to be the result of tumor infiltration in the periodontal ligament. This radiographic change, when accompanied by pain, discomfort, or minor changes in the trabecular pattern, may be of great importance in early diagnosis of osteosarcoma of the jaws.

Radical surgery is the usual treatment, with radiation and chemotherapy sometimes used adjunctively. The tumor tends to extend through intramedullary spaces beyond the apparent clinical or radiographic margins, and local recurrence after surgical removal is a major problem. Osteosarcomas are prone to develop hematogenous metastases, chiefly to the lungs. Jaw osteosarcomas do not metastasize as readily as those arising in long bones; local recurrence after inadequate initial surgery is the chief problem.

The overall survival rate for patients with osteosarcoma of the jaws is about 40%, which is considerably better than that for patients with osteosarcomas of the long bones. Recently, large and multicenter studies of patients with osteosarcoma have demonstrated improved survival through use of specific presurgical chemotherapeutic regimens. This initial therapy is followed by radical surgical removal, with thorough review of the histopathologic specimen to evaluate the response to the initial chemotherapy. In those patients with significant local residual tumor, further chemotherapy or radiation often is used. Overall, the prognosis of osteosarcoma of the jaws seems to be related to prompt diagnosis, tumor grade and size, aggressive chemotherapy, and definitive surgical resection. Because secondary malignancies are not rare in patients treated for osteosarcoma, lifelong follow-up is mandatory.

Juxtacortical osteosarcoma is an uncommon variety that occasionally involves the jaws, originates close to the surface of the bone, and presents in two different patterns. In the parosteal variant, the tumor is ill-defined and often involves the cortex and medullary bone. Histopathologically, the tumor exhibits a high degree of structural differentiation, with its bland features making separation from a benign neoplasm difficult. Although these tumors initially demonstrate a good prognosis and minimal metastatic potential, recurrence is likely after less than radical initial excision, and the lesion often then develops into a typical higher grade osteosarcoma. In the periosteal variant, the tumor grows outward from an intact periosteum and exhibits features of

malignancy that are obvious histopathologically. Although the process exhibits a better prognosis than typical osteosarcoma, local recurrence and distant metastasis is possible if the tumor is not promptly and adequately treated by wide surgical resection. Adjunctive radiation therapy or chemotherapy typically is not used.

Osteosarcoma also is a well-known late sequela of radiation therapy given for other purposes. Jaw osteosarcomas have developed 10 to 15 years after radiation therapy was administered to benign (or occasionally malignant) lesions of the jaws or overlying soft tissues. Finally, an increased prevalence of osteosarcoma is noted in patients with Paget's disease of bone.

Figure 11.52
OSTEOSARCOMA

(A) Painful expansion of the posterior mandible in a 44-year-old female. Two molar teeth had been extracted several weeks previously, and the patient had been treated with antibiotics for a presumed infection. (B) Occlusal radiograph of same patient showing destruction of the buccal cortex with "sunburst" production of tumor bone.

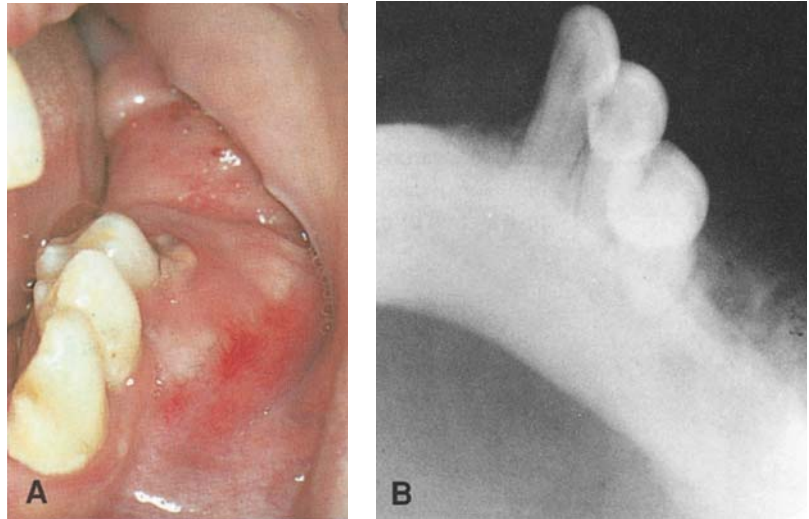


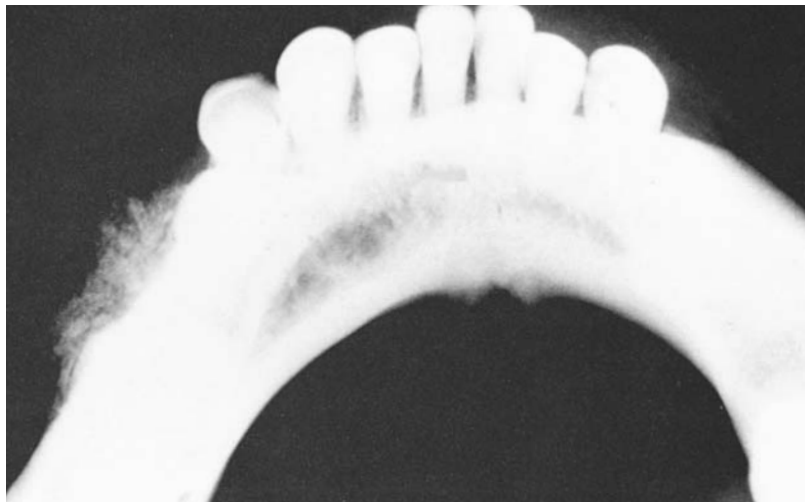
Figure 11.53
OSTEOSARCOMA

Periapical radiograph of the patient depicted in Figure 11.52 showing symmetric widening of the periodontal ligament space of the premolar teeth.



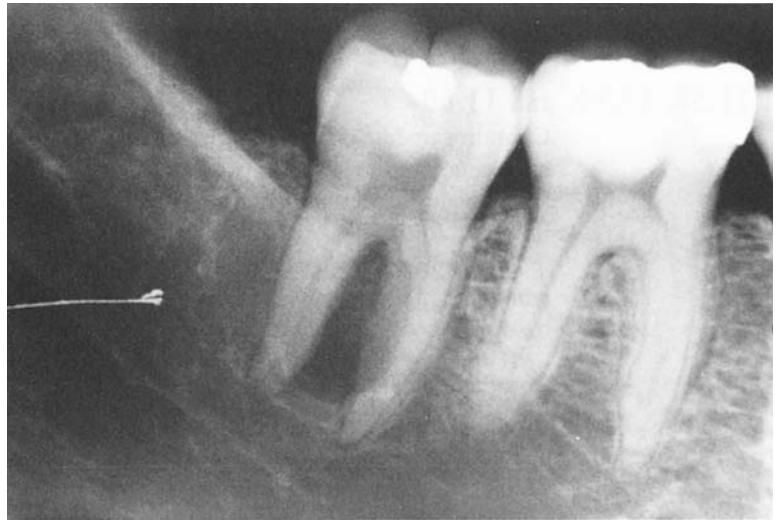
Figure 11.54
OSTEOSARCOMA

Occlusal radiograph of a 40-year-old female who presented with a firm lobular mass on the buccal aspect of the mandible. This demonstrates the "sunburst" tumor bone formation with cortical bone destruction.



► **Figure 11.55**
OSTEOSARCOMA

Periapical radiographs of a 32-year-old female who had a painful swelling over the second molar. Note the ill-defined radiolucent lesion in the furcation area. (Courtesy of Dr. E. B. Bass.)



► **Figure 11.56**
OSTEOSARCOMA

Same patient depicted in Figure 11.55. Endodontic treatment was performed, and the filling material was extruded into the furcation area. Biopsy specimen revealed an osteosarcoma. (Courtesy of Dr. E. B. Bass.)



► **Figure 11.57**
JUXTACORTICAL OSTEOSARCOMA, PAROSTEAL VARIANT

Periapical radiograph showing a low-grade osteosarcoma arising on the alveolar bone surface between the mandibular canine and lateral incisor teeth.



Figs. 11.58, 11.59, and 11.60

Chondrosarcoma is the primary malignancy of cartilage-forming cells within bone. This cancer is uncommon in the jaws and exhibits a lower prevalence than osteosarcoma. Although examples have been recorded in patients over a wide age range, the peak prevalence for jaw tumors is in the third and fourth decades of life. Chondrosarcoma occurs with equal frequency in both jaws. The most common initial complaint is a swelling or mass, although a few patients complain initially of pain. Dental complaints, such as localized periodontal bone loss and loosening or migration of teeth, are not uncommon. Maxillary tumors may cause initial symptoms of nasal obstruction.

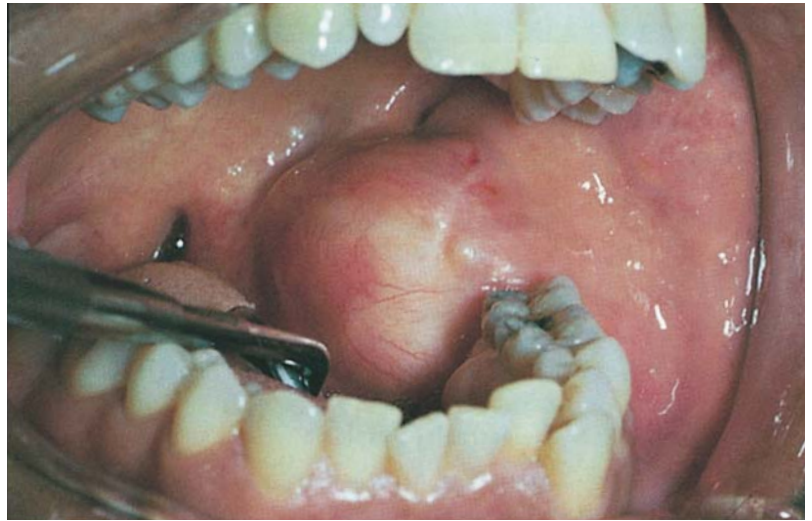
Radiographically, chondrosarcomas characteristically show a destructive radiolucent lesion with ill-defined margins. The radiolucent area frequently contains scattered radiopaque foci resulting from calcification of the neoplastic cartilage. Symmetric widening of the periodontal ligament space and irregular root resorption may be noted in teeth involved by the tumor. The radiographic features usually suggest a malignant tumor but cannot be differentiated from osteosarcoma.

Because chondrosarcomas are not radiosensitive and are less responsive to chemotherapy than are osteosarcomas, treatment typically consists of radical surgical resection, with adjuvant therapies often reserved for patients with postsurgical residual disease. Overall, metastasis of chondrosarcomas of the jaws is uncommon, and most treatment failures are caused by uncontrollable local disease secondary to inadequate removal during the initial therapy. Because recurrence may develop as late as 20 years after initial therapy, follow-up must be lifelong.

One exception to the typical pattern is mesenchymal chondrosarcoma, a variant that is not rare within the jaws. Although the aggressiveness of the tumor is variable, this pattern on occasion may be highly malignant, and it demonstrates significant recurrence along with lymphatic and hematogenous metastasis. Radical surgery, often combined with chemotherapy and radiation therapy, is the treatment of choice; overall, the prognosis is poor, with approximately 70% of those affected succumbing to their tumor.

► **Figure 11.58**
CHONDROSARCOMA

Clinical photograph of a 50-year-old woman showing a massive tumor projecting lingually in the posterior molar area. She stated that the tumor had been present for many years with slow growth.



► **Figure 11.59**
CHONDROSARCOMA

Panorgraphic radiograph from the patient depicted in Figure 11.58 showing a destructive radiolucent lesion containing numerous radiopaque foci.



► **Figure 11.60**
CHONDROSARCOMA

Same patient depicted in Figures 11.58 and 11.59. Computed tomography exhibiting a large lesion projecting lingually from the mandible.



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