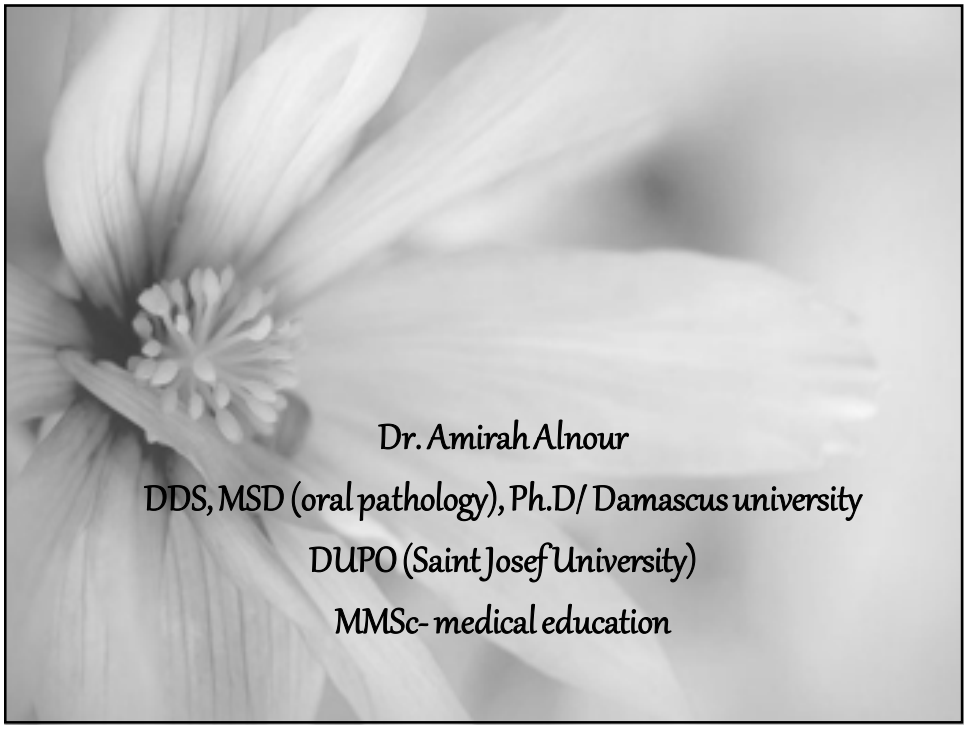
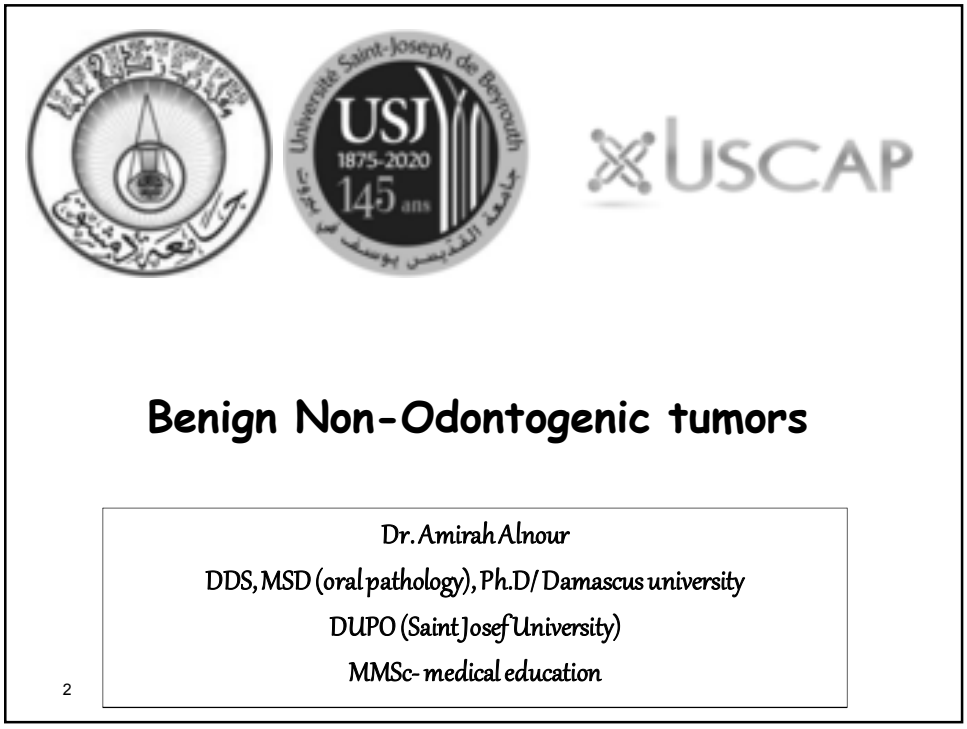

Dr. Amirah Alnour
DDS, MSD (oral pathology), Ph.D/ Damascus university
DUPO (Saint Josef University)
MMSc- medical education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Benign Non-Odontogenic tumors
Dr. Amirah Alnour
DDS, MSD (oral pathology), Ph.D/ Damascus university
DUPO (Saint Josef University)
MMSc- medical education
2

Lecture outlines

1. Ossifying Fibroma
2. Fibrous Dysplasia
3. Cemento-Osseous Dysplasia
4. Osteoblastoma/Osteoid Osteoma
5. Osteoma
6. Desmoplastic Fibroma
7. Chondroma
8. Central Giant Cell Granuloma
9. Giant Cell Tumor
10. Hemangioma of Bone
11. Langerhans Cell Disease
12. Tori and Exostoses

3

Fibro-Osseous Lesions of the Jaws: Entities Most Commonly Included

1. Ossifying fibroma
2. Fibrous dysplasia
3. Cemento-osseous dysplasia:
   - Periapical/focal
   - Florid
4. Chronic osteomyelitis

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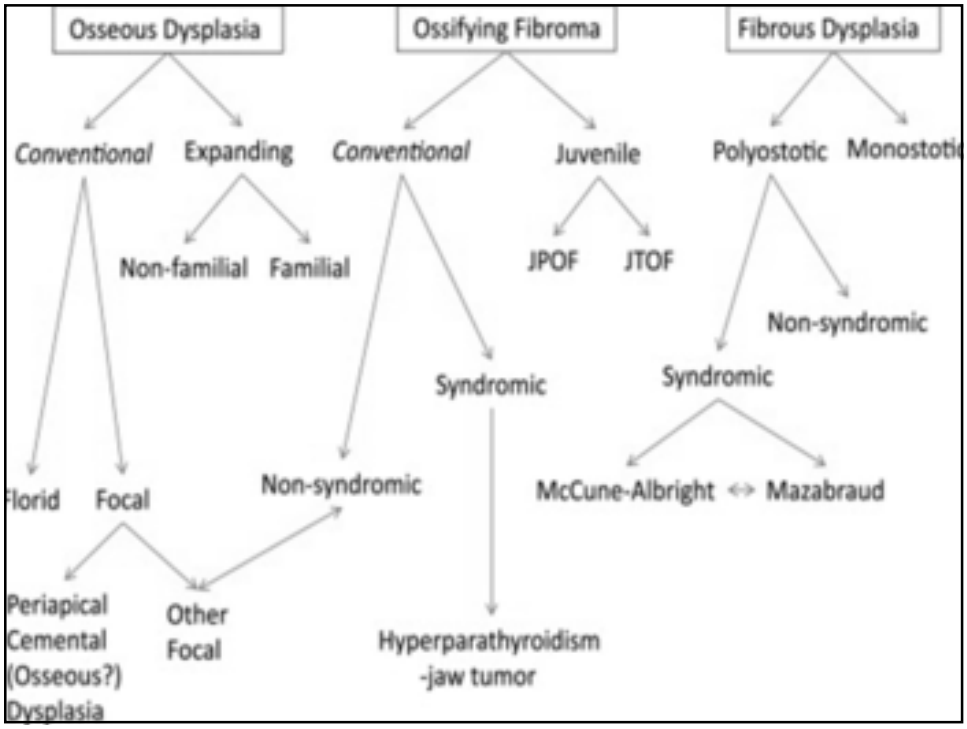

Ossifying Fibroma

Clinical Features

- Third and fourth decades
- Mandible > maxilla
- Well circumscribed
- Lucent or lucent/opaque pattern
- Continuous growth

Histopathology

- Cellular fibrous matrix
- Islands/trabeculae of new bone
- Osteoblasts; no osteoclasts
- Relatively homogeneous pattern
- No inflammatory cells

Treatment

- Curettage/excision

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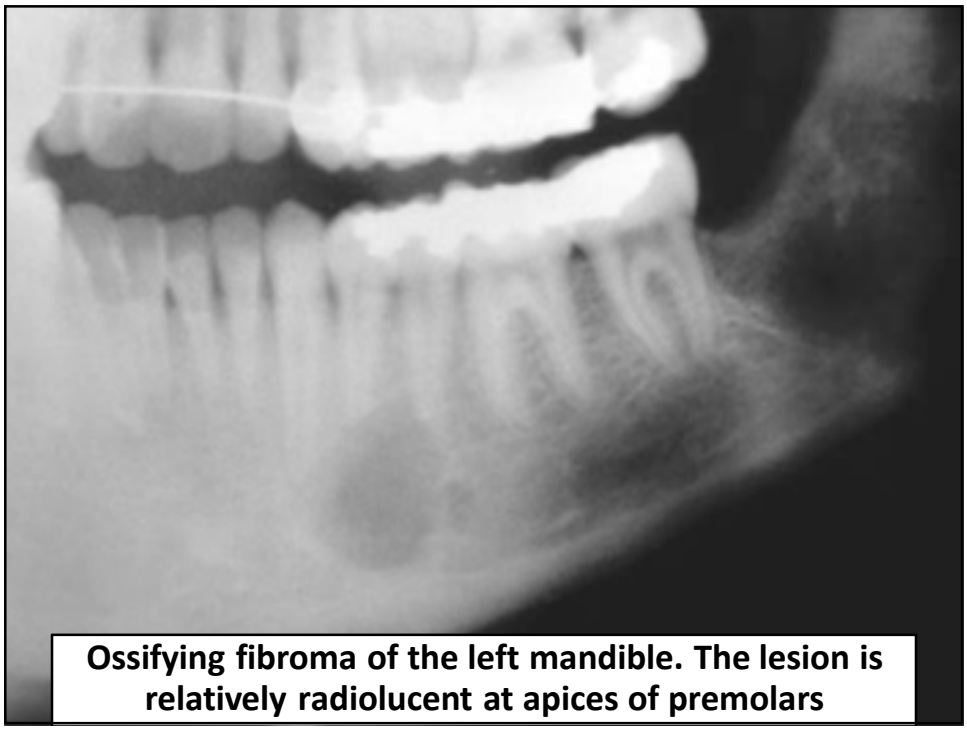
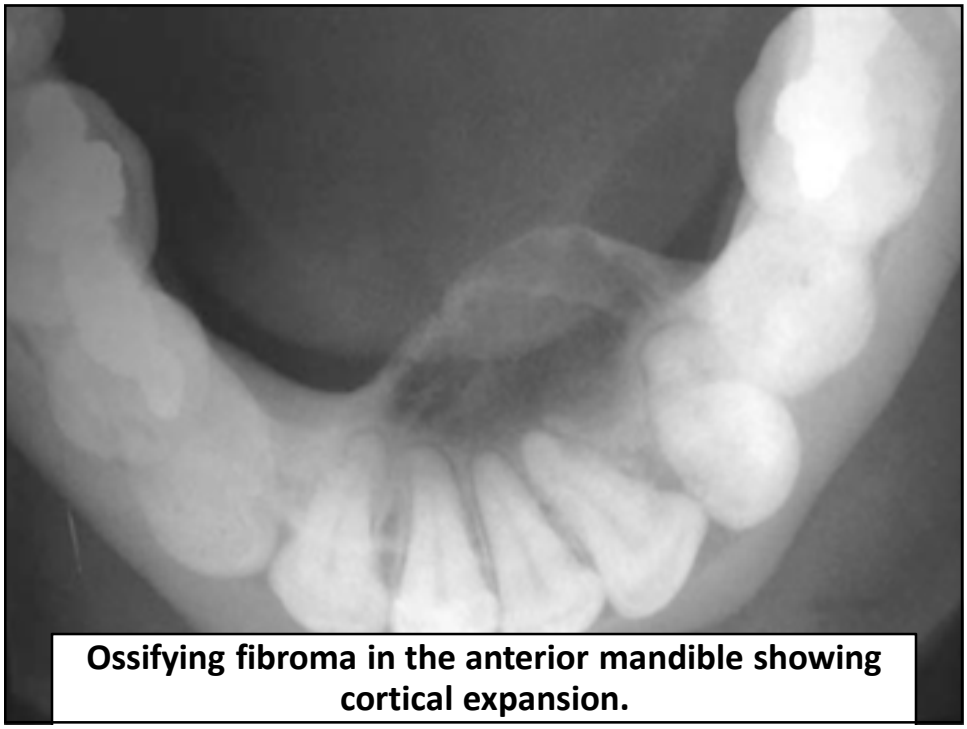
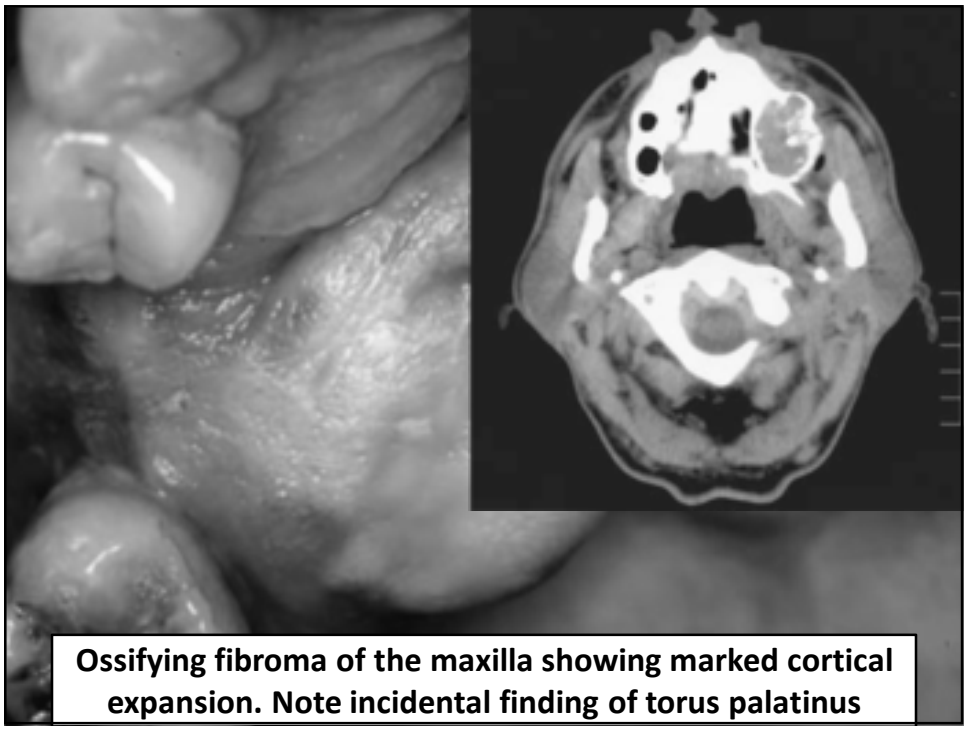


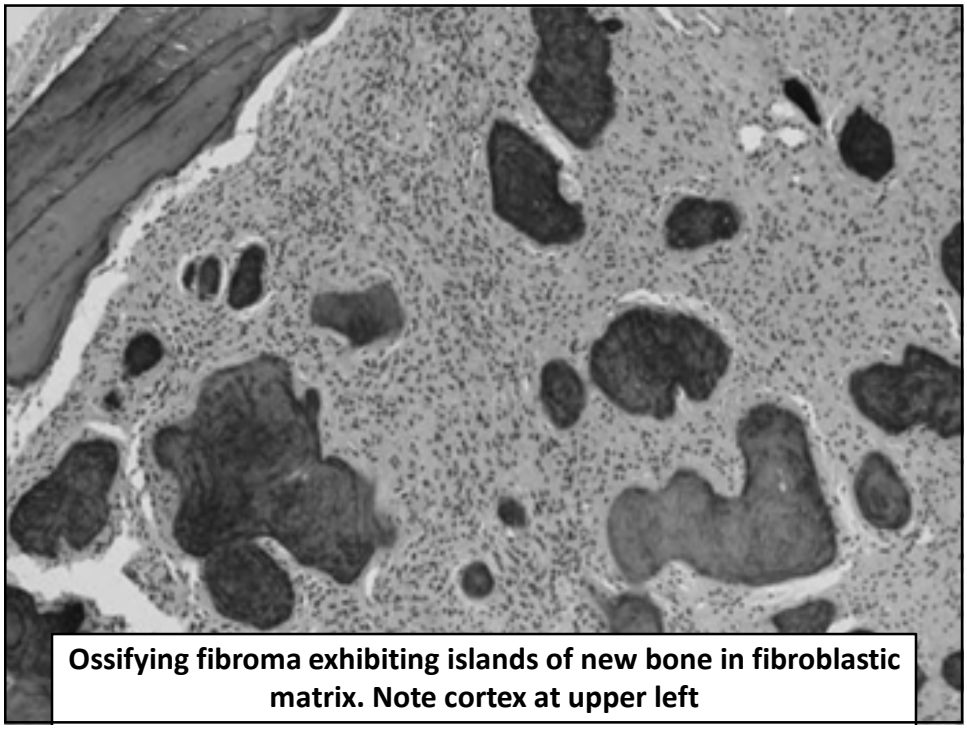
Ossifying fibroma of the left mandible. The lesion is relatively radiolucent at apices of premolars



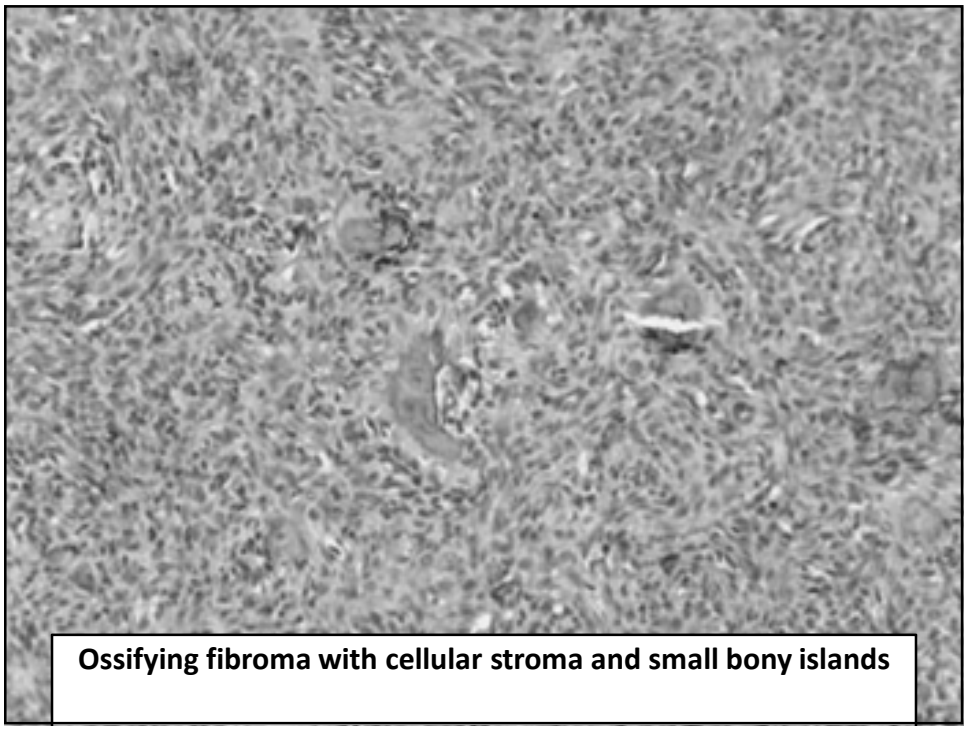
Ossifying fibroma in the anterior mandible showing cortical expansion.



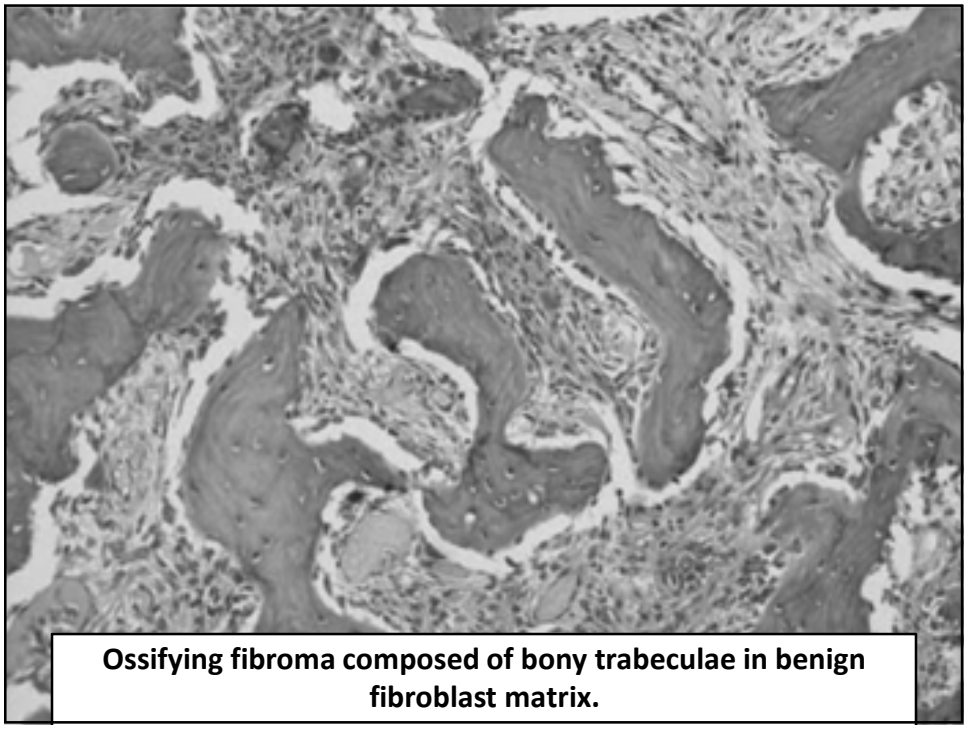
Ossifying fibroma of the maxilla showing marked cortical expansion. Note incidental finding of torus palatinus



Ossifying fibroma exhibiting islands of new bone in fibroblastic matrix. Note cortex at upper left



Ossifying fibroma with cellular stroma and small bony islands



Ossifying fibroma composed of bony trabeculae in benign fibroblast matrix.

Ossifying Fibroma Variants

Juvenile Trabecular Ossifying Fibroma

- Younger patients
- Aggressive clinical course
- Cellular (benign) stroma
- Trabecular or spherical bone

Juvenile Psammomatoid Ossifying Fibroma

- Biologically same as ossifying fibroma
- Spherical islands of bone (cementum)
- Bone and cementum microscopically identical

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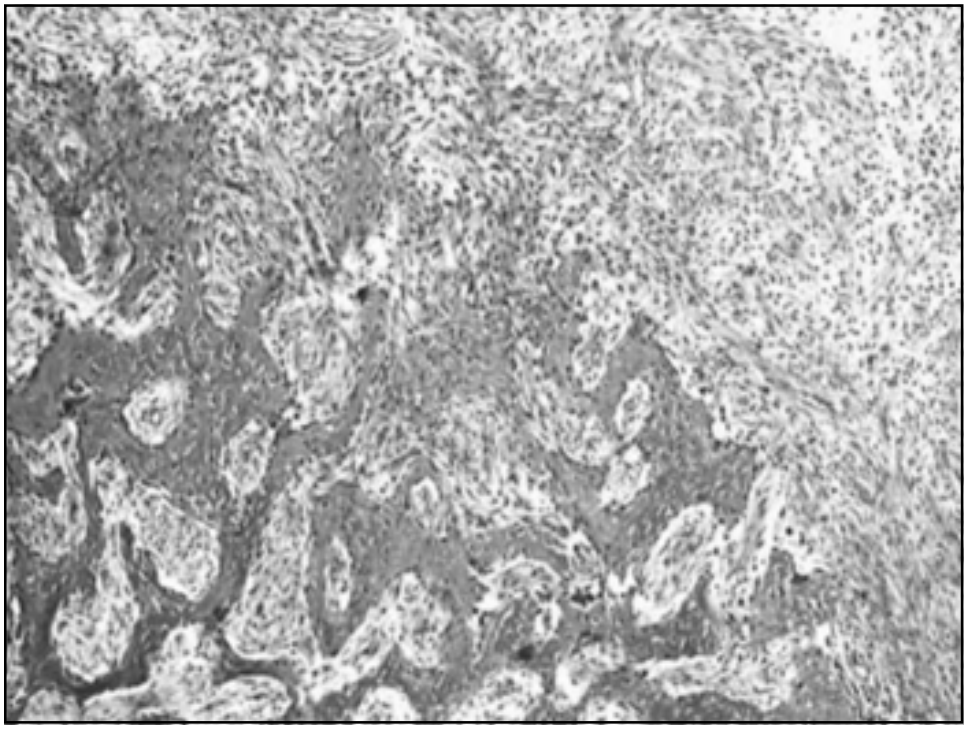

Fibrous Dysplasia

Clinical Features

- First and second decades (stabilizes at puberty and very slow
- growth thereafter)
- Maxilla >mandible (one or more bones)
- Ribs, femur, tibia also affected
- Unilateral diffuse opacity
- Asymptomatic; self-limiting
- Serum laboratory values normal

Histopathology

- New fibrillar bone trabeculae
- Few osteoblasts; no osteoclasts
- Homogeneous pattern
- Vascular matrix
- No inflammatory cells

Treatment

- Surgical recontouring for cosmetics (after growth spurt)
- Regrowth in 25% of treated cases

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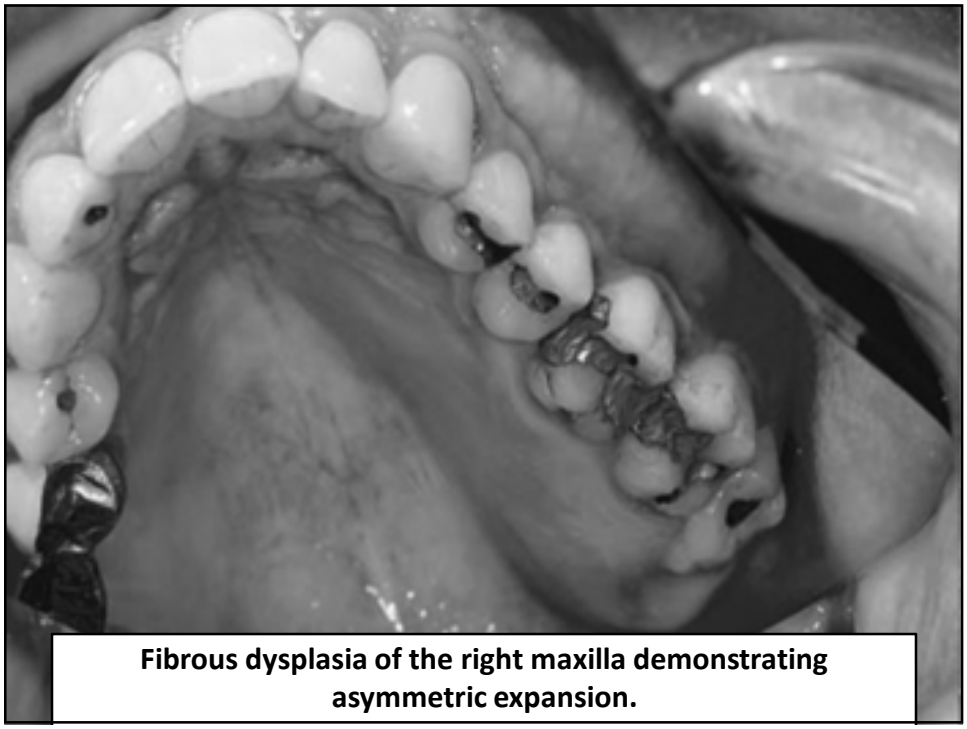
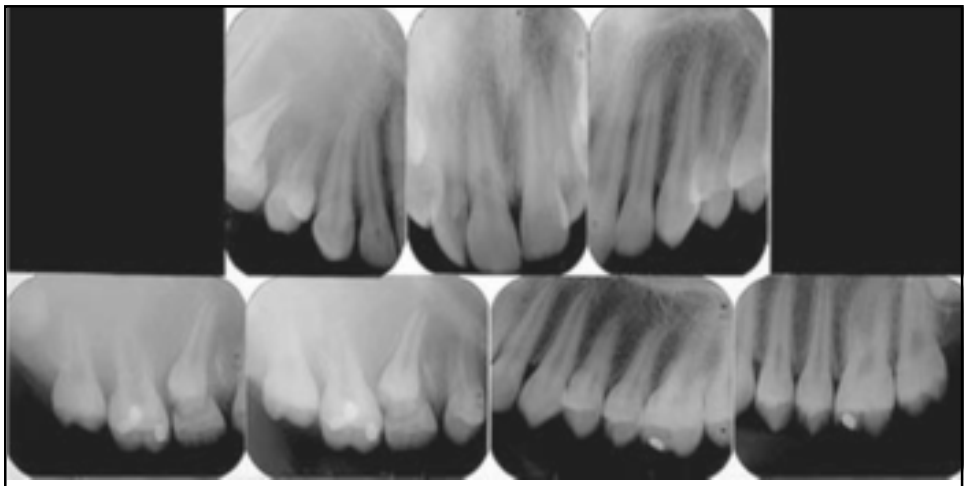


Fibrous dysplasia of the right maxilla demonstrating asymmetric expansion.



Fibrous dysplasia of the right maxilla causing a characteristic diffuse ground-glass effect.

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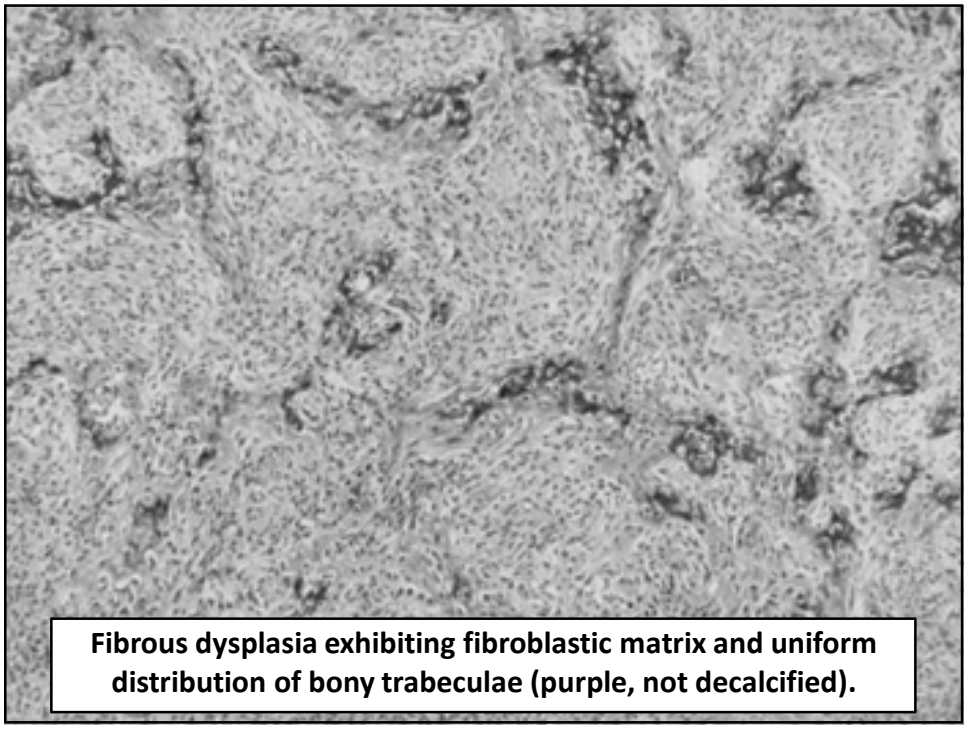
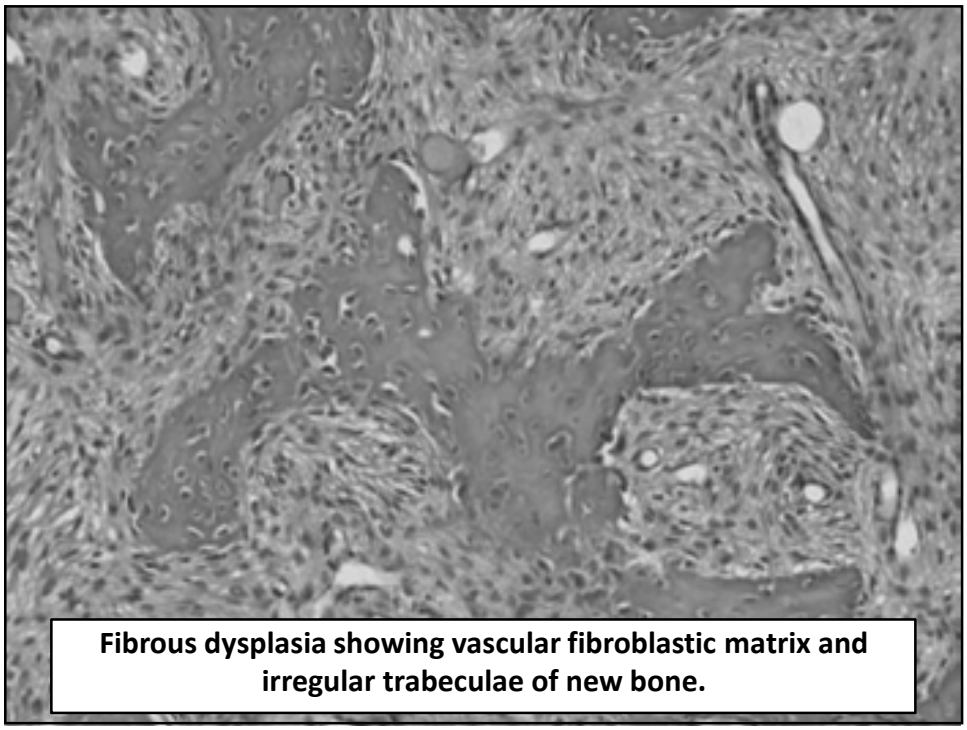


Fibrous dysplasia exhibiting fibroblastic matrix and uniform distribution of bony trabeculae (purple, not decalcified).



Fibrous dysplasia showing vascular fibroblastic matrix and irregular trabeculae of new bone.

Fibrous Dysplasia vs. Ossifying Fibroma

Fibrous Dysplasia	Ossifying Fibroma
1. First and second decades	1. Third and fourth decades
2. Maxilla > mandible	2. Mandible > maxilla
3. Diffuse opacity	3. Circumscribed
4. Self-limited	4. Continuous growth
5. One or more bones	5. One bone
6. Vascular matrix	6. Cellular fibrous matrix
7. Woven bone trabeculae	7. Bony islands and trabeculae
8. Stabilizes at puberty	8. Not hormone related
9. Recontour for cosmetics	9. Excise
10. Majority with mutations in GNAS gene	10. No genetic mutations identified

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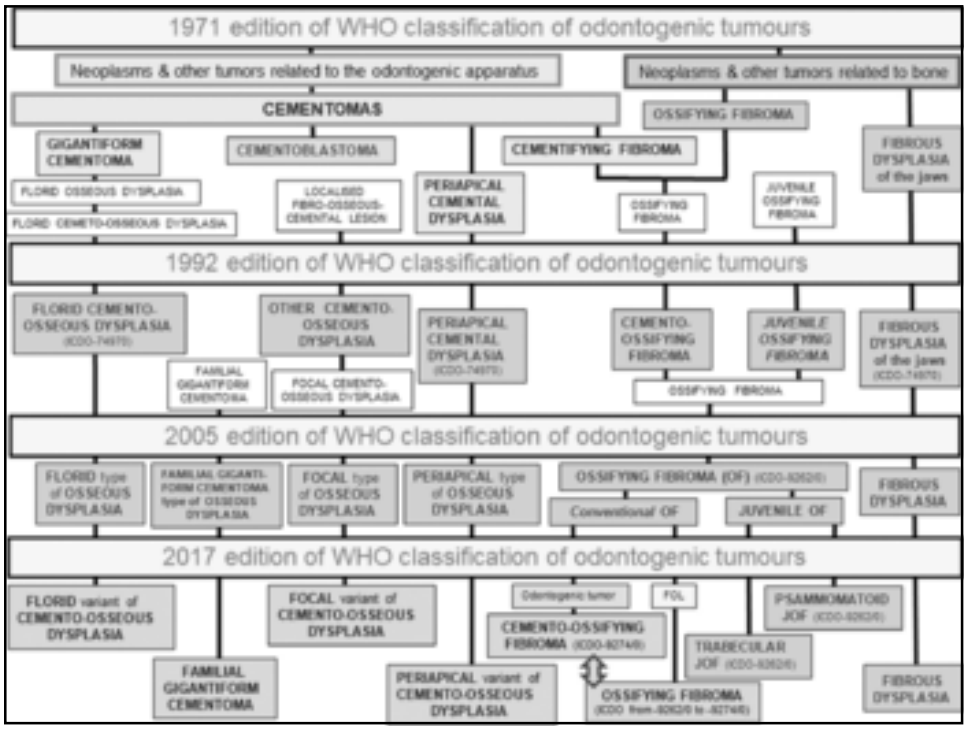

Osteoblastoma/Osteoid Osteoma

- Osteoblastoma: uncommon primary lesion of bone that occasionally arises in the maxilla or the mandible
- Osteoid osteoma: represent a smaller version of the same tumor, although some prefer to separate these lesions into two distinct entities.
- These are benign neoplasms of undetermined cause, although a genetic defect has been suggested. Clinically and histologically, they may be confused with osteosarcoma.

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Osteoblastoma/Osteoid Osteoma

	Osteoid Osteoma	***Osteoblastoma***
Age	10-20 years	10-20 years
Sex	2:1 males	2:1 males
Site	Femoral neck	Spine
Pain	Mod.-severe Worse at night , Aspirin response 90%	dull aching pain Worse at night, Aspirin relief,<50%
Nidus	less than 2.0 cm	2.0 -10.0 cm
Recurrence	No	10%

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Osteoblastoma

- Large counterpart of osteoid osteoma
- Osteoblastoma > 1.5 cm
- Osteoid osteoma < 1.5 cm
- 50% are painful
- Second decade is characteristic age
- Circumscribed
- Benign cellular (osteoblasts) neoplasm with new bone in scant fibrous stroma
- Treatment by excision; few recurrences

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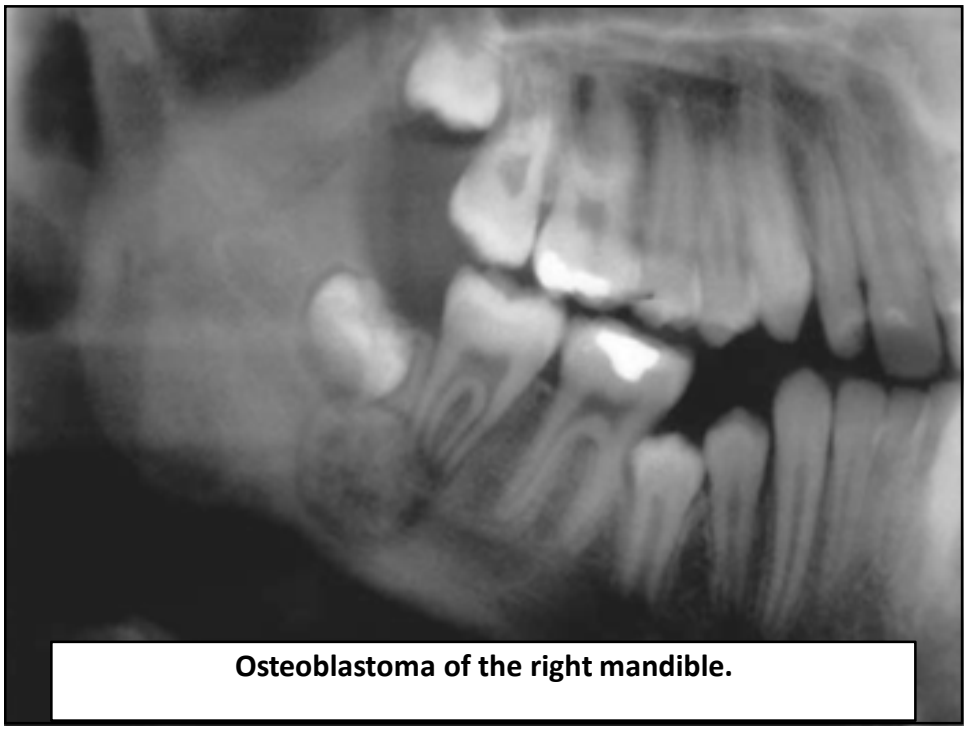
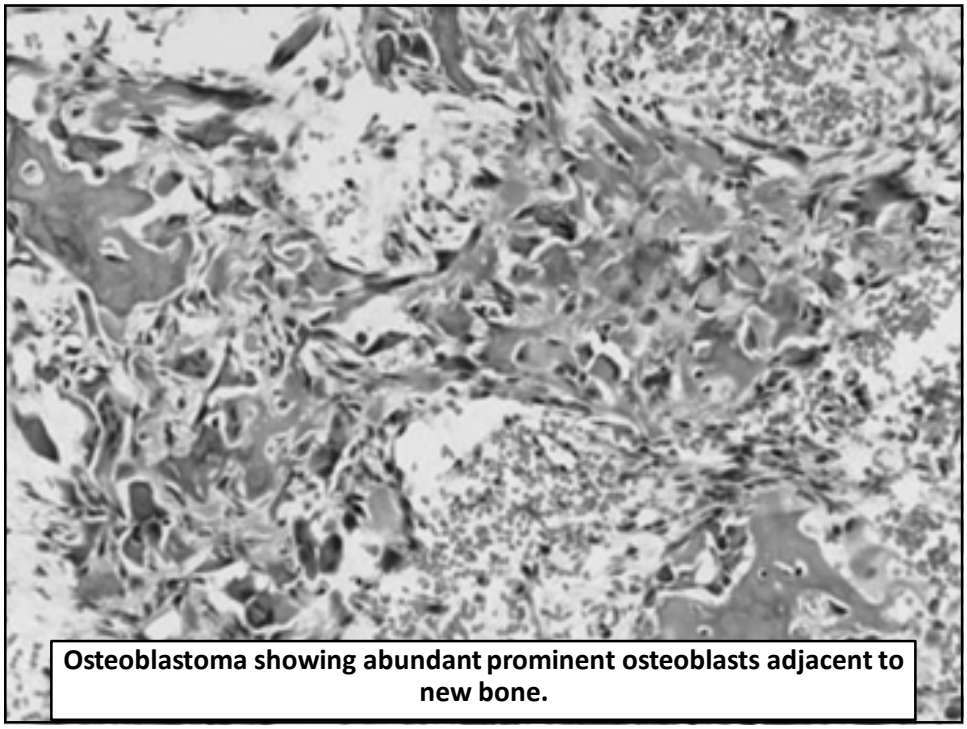


Osteoblastoma of the right mandible.



Osteoblastoma showing abundant prominent osteoblasts adjacent to new bone.

Osteoma

- Benign tumors that consist of mature, compact, or cancellous bone.
- Osteomas are relatively rare in the jaws.
- Unknown cause
- Contributing factors: trauma, infection, genetic/congenital, and developmental abnormalities

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Osteomas called:

- Periosteal osteomas: on surface of bone
- Endosteal or solitary central osteomas: centrally within bone

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Periosteal osteomas:

asymptomatic, slow-growing, bony, hard masses. Asymmetry may be noted when lesions enlarge to sufficient proportion.

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Endosteal osteomas

- discovered during routine radiographic examination as dense, well-circumscribed radiopacities, because extensive growth must take place before cortical expansion is evident.

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- 70% of cases in mandible as well as in facial and skull bones and within paranasal sinuses.
- Symptoms: Headaches, recurrent sinusitis, and ophthalmologic complaints have been noted, depending on the lesion location.

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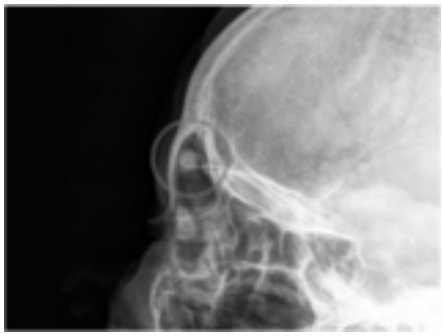
- 2-5 decades of life,
- M>F males
- usually solitary, except in patients with *Gardner syndrome.*



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Gardner syndrome, inherited as an autosomal-dominant disorder, is characterized by:

1. intestinal polyposis,
2. multiple osteomas,
3. fibromas of the skin,
4. epidermal and trichilemmal cysts,
5. impacted permanent and supernumerary teeth,
6. odontomas .

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- The genetic defect is found in a small region on the long arm of chromosome 5 (5q21), where the familial adenomatous polyposis (APC) gene resides.
- Most patients with Gardner's syndrome do not exhibit the complete spectrum of clinical disease expression.
- Osteomas associated with this syndrome may be found in the jaws (especially the mandibular angle) and in facial and long bones.
- Intestinal polyps associated with Gardner syndrome are commonly located in the colon and rectum. Significantly, these polyps, found microscopically to be adenomas, exhibit a very high rate of malignant transformation to invasive colorectal carcinoma.

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Histopathology

Two distinct histologic variants of osteoma have been described.

1. composed of relatively dense, compact bone with sparse marrow tissue.

2. consists of lamellar trabeculae of cancellous bone with abundant fibrofatty marrow.

Osteoblasts may be numerous, but osteoclasts are sparse.

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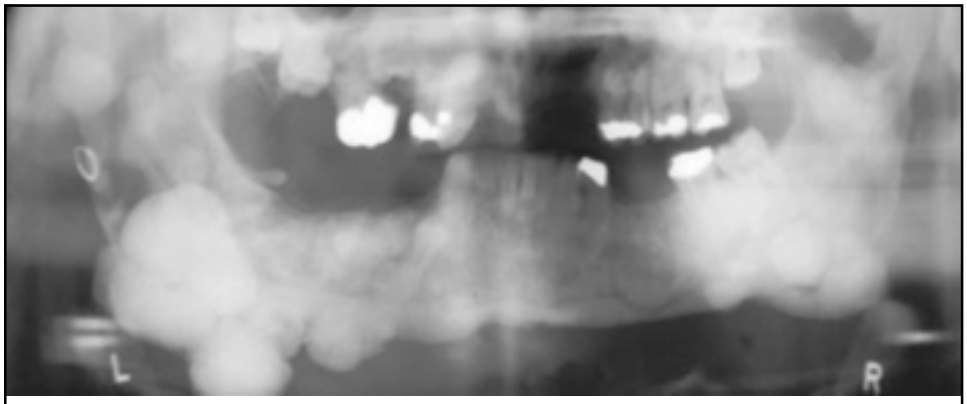

Osteomas of Gardner's syndrome.

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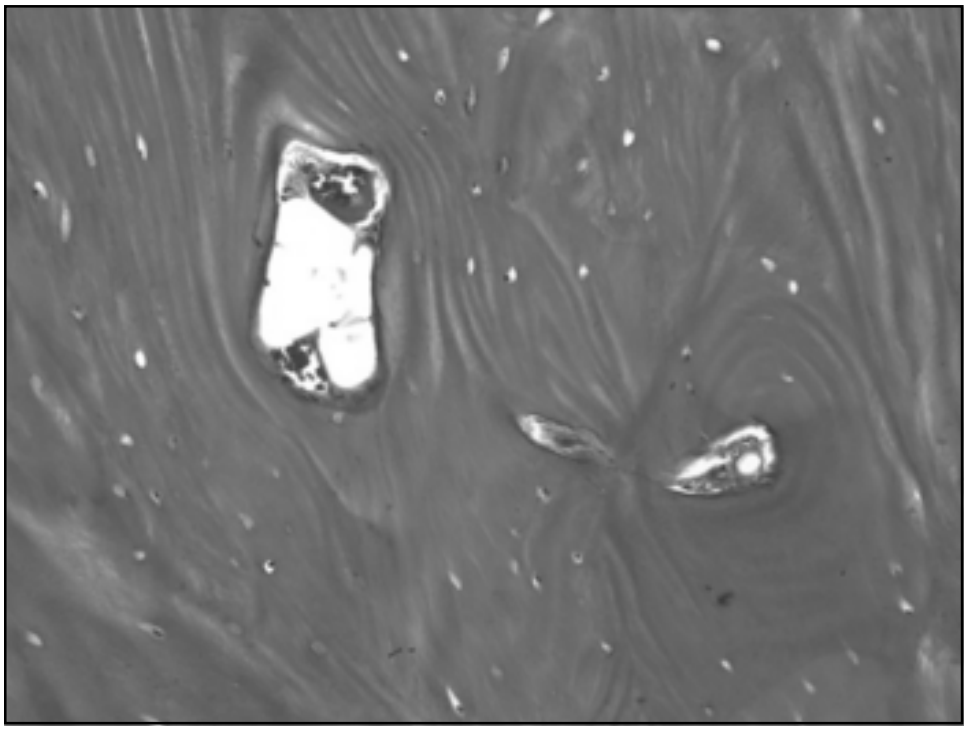

Desmoplastic Fibroma

- Young adults (< 30 years of age)
- Bony counterpart of fibromatosis
- Microscopic DDx:
1. Odontogenic fibroma
2. Odontogenic fibromyxoma
3. Low-grade fibrosarcoma
4. Follicular sac
- Recurrence potential

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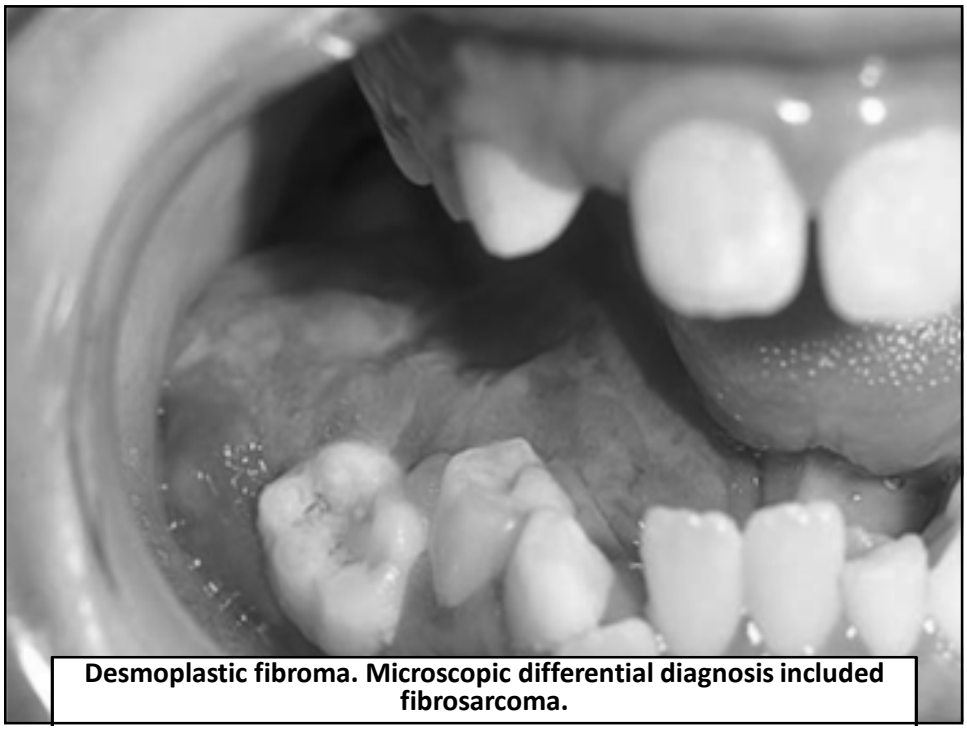
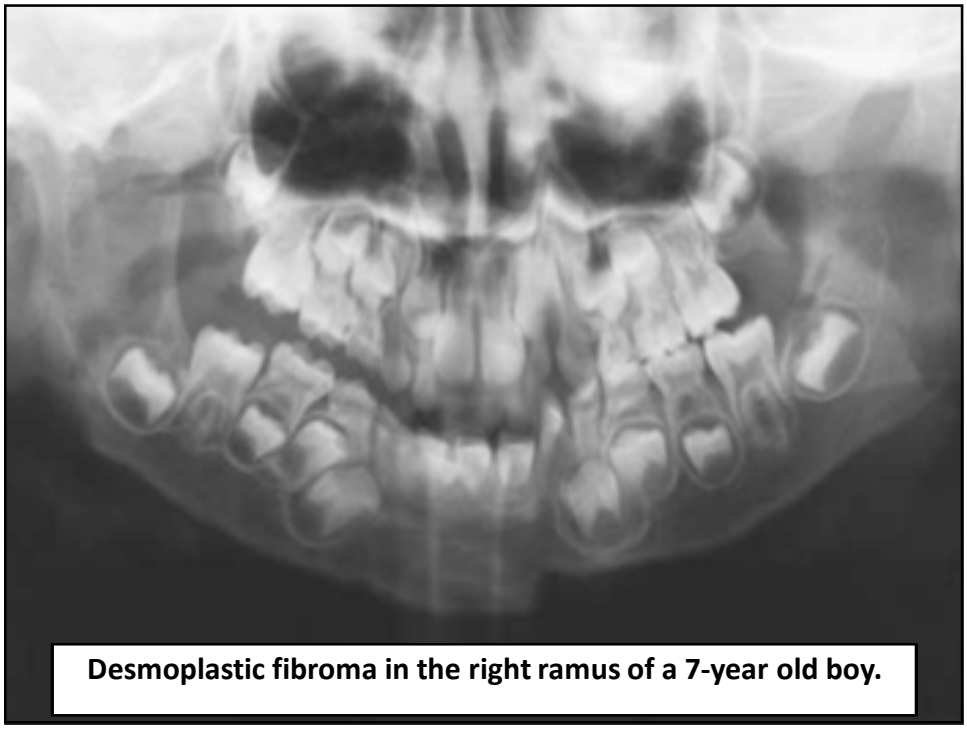
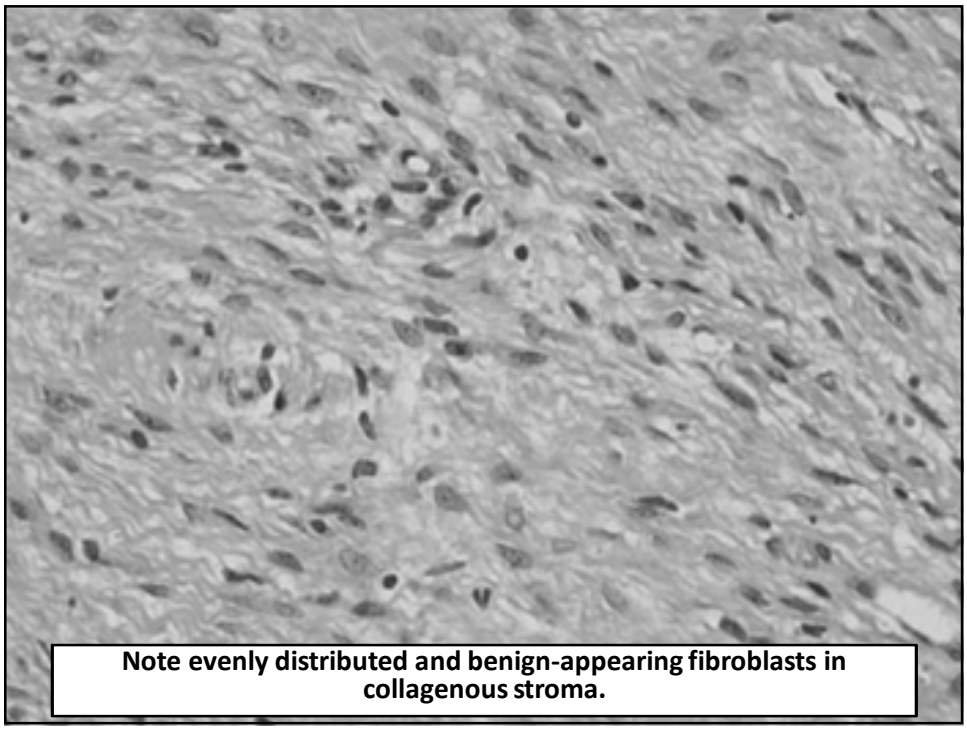


Desmoplastic fibroma. Microscopic differential diagnosis included fibrosarcoma.



Desmoplastic fibroma in the right ramus of a 7-year old boy.



Note evenly distributed and benign-appearing fibroblasts in collagenous stroma.

Chondroma

- Benign cartilaginous tumor of unknown cause.
- very rarely seen in the jaws,
- painless, slowly progressive swelling.
- Most lesions of the craniofacial complex arise in the nasal septum and ethmoid sinuses.
- M=F , with the majority of tumors appearing before 50 years of age.

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In maxilla:

in the anterior region, where cartilaginous remnants of development are located.

In mandibular:

in the body and symphysis areas, as well as in the coronoid process and the condyle.

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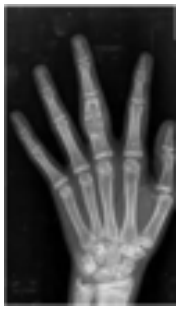

- The radiographic appearance: variable, but it often presents as an irregular radiolucent area. Foci of calcification may be evident within the radiolucent lesion.
- Within the **TMJ**, lesions histologically similar to chondromas are likely to represent pseudotumors including synovial chondromatosis, osteochondroma, and other entities.

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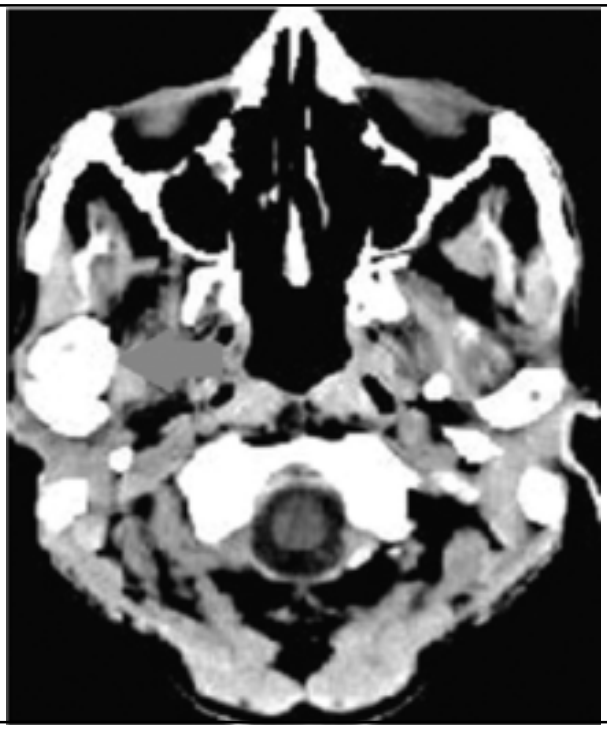

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Histopathology:

- well-defined lobules of mature hyaline cartilage. The chondrocytes are small and contain single, regular nuclei. The degree of cellularity varies
- The principal diagnostic problem rests in microscopically distinguishing chondroma from a well-differentiated chondrosarcoma.
- The latter exhibits a heterogeneous pattern with cytologically atypical and irregularly spaced chondrocytes.

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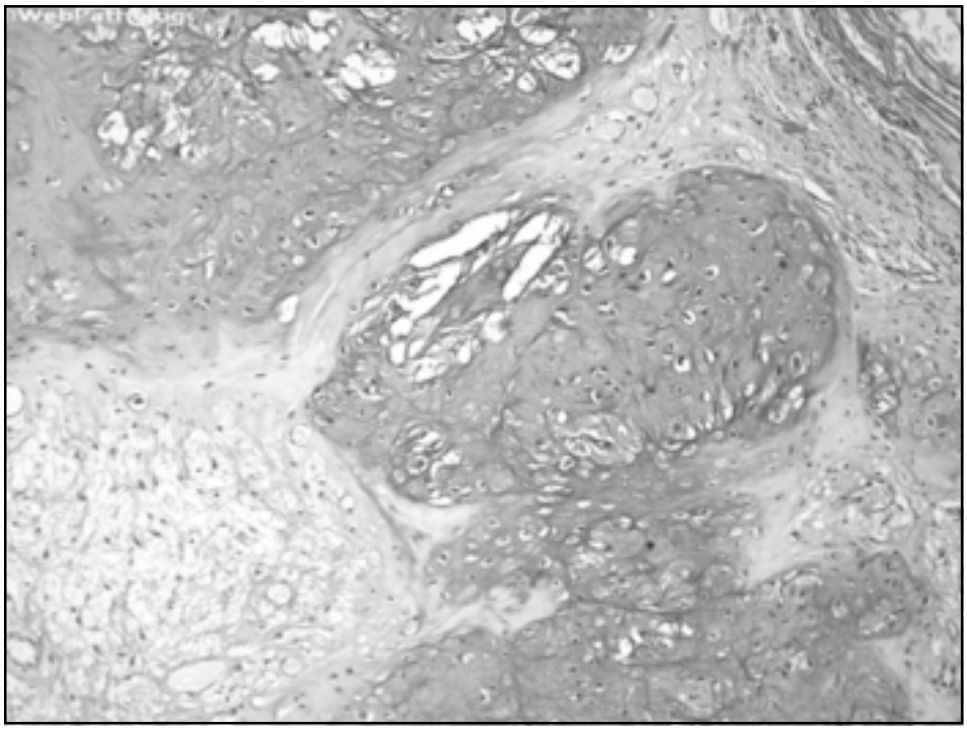

Treatment & prognosis

Chondromas are surgically excised, and recurrence is unusual. Any recurrence should be cause for reconsidering the original diagnosis in favor of the possibility of low-grade malignancy.

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Central Giant Cell Granuloma

- (CGCG), or giant cell lesion, is a benign proliferation of fibroblasts and multinucleated giant cells within a well vascularized stroma that occurs almost exclusively within the **jaws**
- The tumor typically presents as a solitary radiolucent lesion of the mandible or maxilla.

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Clinical Features

- Most patients younger than 30 years of age; females affected more often than males
- Radiolucency; mandible > maxilla; anterior jaw . posterior jaw
- Recurrences unpredictable (10%-50%)

Histopathology

- Benign fibroblast matrix (in cell cycle)
- Giant cells variable (size, number, distribution)
- Few to many mitotic figures
- Cannot separate aggressive from nonaggressive lesions

Treatment

- Traditional excision vs. medical management—calcitonin (osteoclast inhibition)

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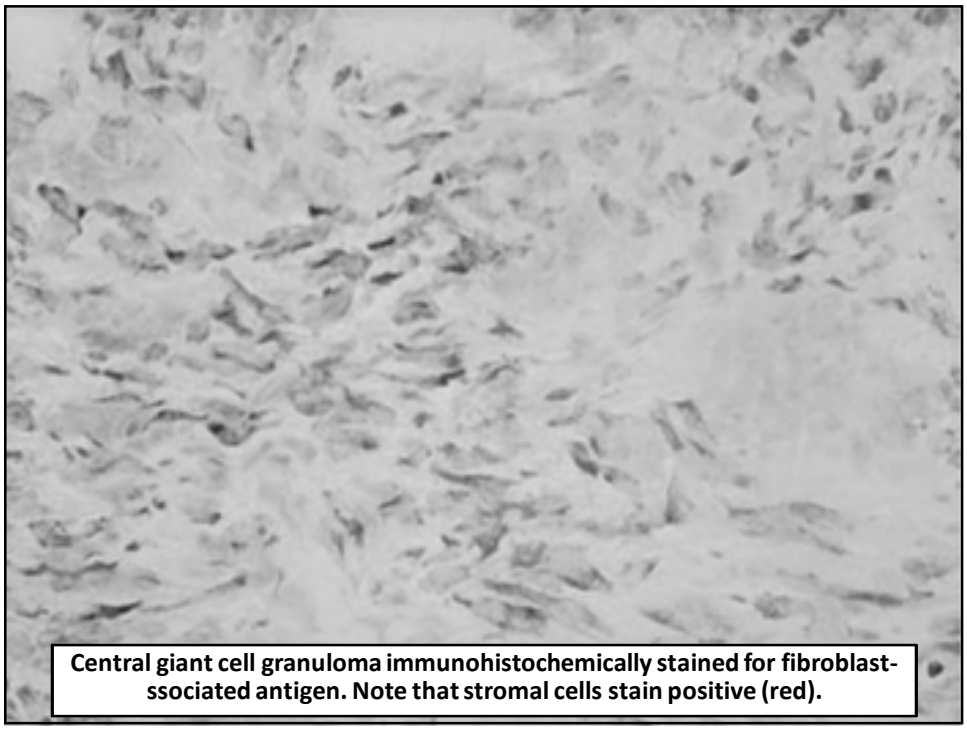
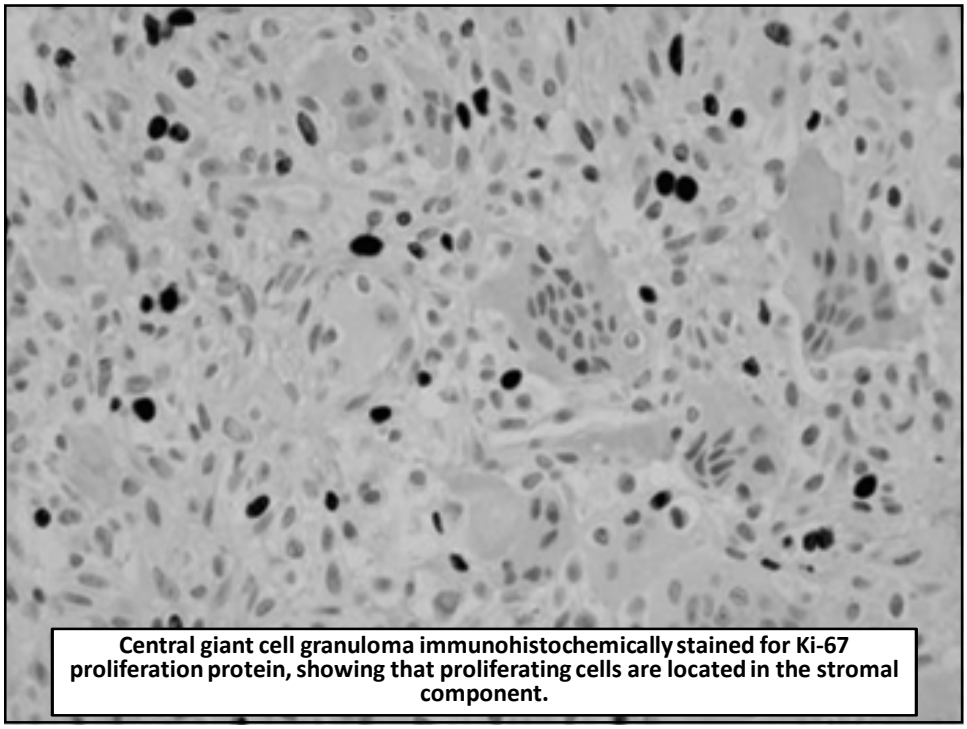
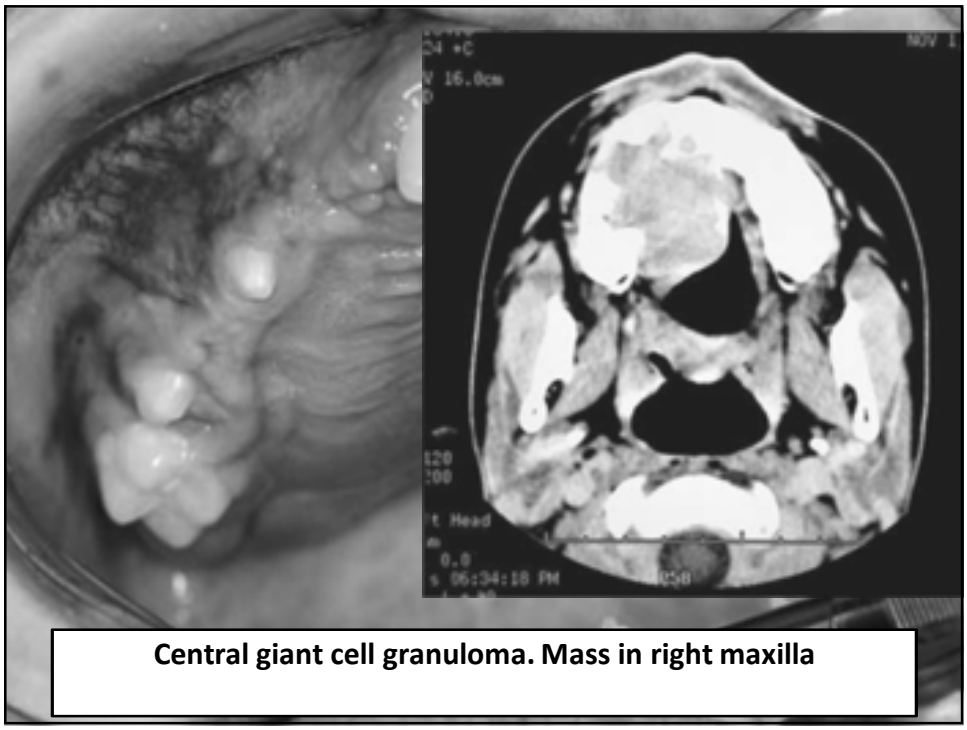


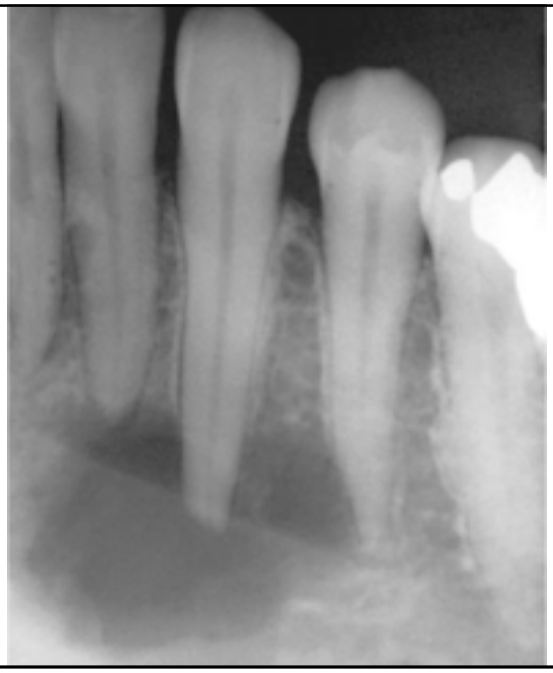
Central giant cell granuloma immunohistochemically stained for fibroblast-ssociated antigen. Note that stromal cells stain positive (red).



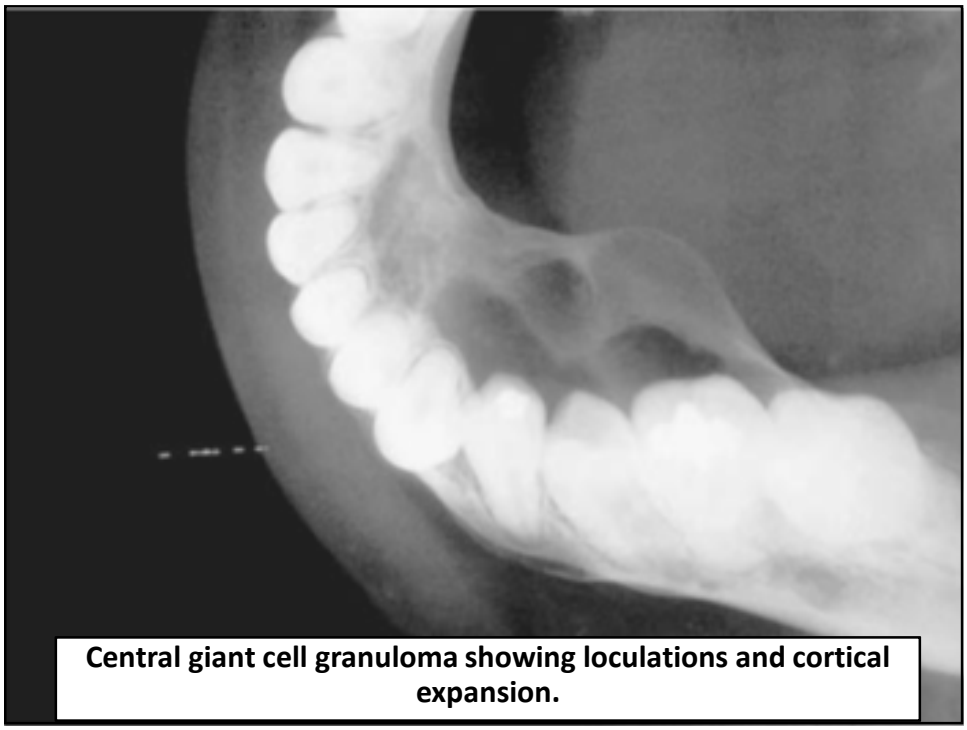
Central giant cell granuloma immunohistochemically stained for Ki-67 proliferation protein, showing that proliferating cells are located in the stromal component.



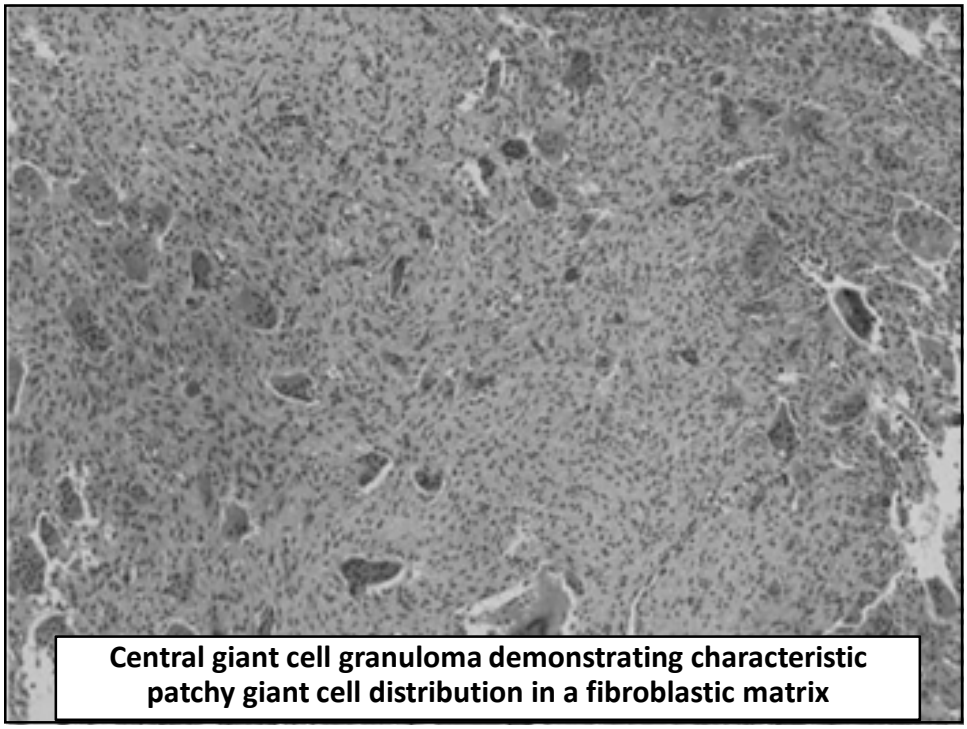
Central giant cell granuloma. Mass in right maxilla



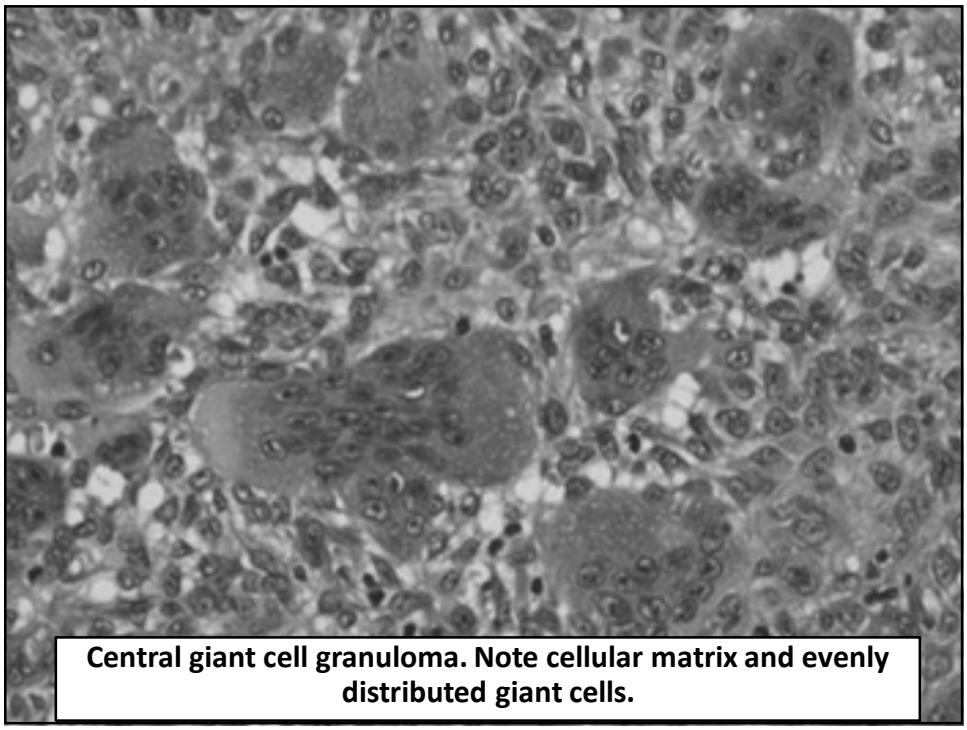
Central giant cell granuloma of the anterior mandible



Central giant cell granuloma showing loculations and cortical expansion.



Central giant cell granuloma demonstrating characteristic patchy giant cell distribution in a fibroblastic matrix



Central giant cell granuloma. Note cellular matrix and evenly distributed giant cells.

Treatment & prognosis:

- Surgical management of these lesions is the treatment of choice. Excision or curettage of the tumor mass followed by removal of the peripheral bony margins results in a good prognosis and a low recurrence rate. A somewhat higher rate of recurrence has been reported in lesions arising in children and young teens. Lesions with aggressive clinical features also exhibit a tendency to recur, often necessitating more extensive surgical approaches, including resection.

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Central Giant Cell Granuloma: Microscopic Differential

Hyperparathyroidism

- Elevated serum parathormone and alkaline phosphatase
- Multiple bone lesions; loss of lamina dura

Aneurysmal Bone Cyst

- Blood-filled sinusoids present

Cherubism

- Symmetric lesions
- Family history
- Perivascular collagen cuffing

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Giant Cell Tumor

- true neoplasms that arise most commonly in long bones, especially in the area of the knee joint.
- These tumors exhibit a wide spectrum of biological behavior from benign to malignant.
- The relationship between this lesion and CGCG is controversial. Most regard the giant cell tumor as distinct from CGCG, acknowledging the very rare occurrence of giant cell tumor within the jaws.

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- Lesions exhibit slow growth and bone expansion, or they produce rapid growth, pain, or paresthesia.
- Radiographically, the giant cell tumor produces a radiolucent image

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- Microscopically, this tumor is characterized by the presence of numerous multinucleated giant cells dispersed evenly among monocyte-macrophages and spindle cells.
- spindle cells: represent the neoplastic cells in this tumor,
- monocyte-macrophages: are reactive, giving rise to giant cells through recruitment and induction factors (e.g., tumor necrosis factor [TNF]-alpha, macrophage colony-stimulating factor) secreted by the tumor spindle cells.
- Stromal cellularity is usually prominent, with minimal collagen production.
- Giant cells in giant cell tumors are usually larger and contain more nuclei than the corresponding cells of CGCG. Significant variation is noted, however, such that any given lesion may present diagnostic difficulty because of considerable histologic overlap. Giant cell tumors may contain inflammatory cells and areas of necrosis while exhibiting a relative absence of hemorrhage and hemosiderin deposition. Osteoid formation is noted less often than in giant cell granulomas.

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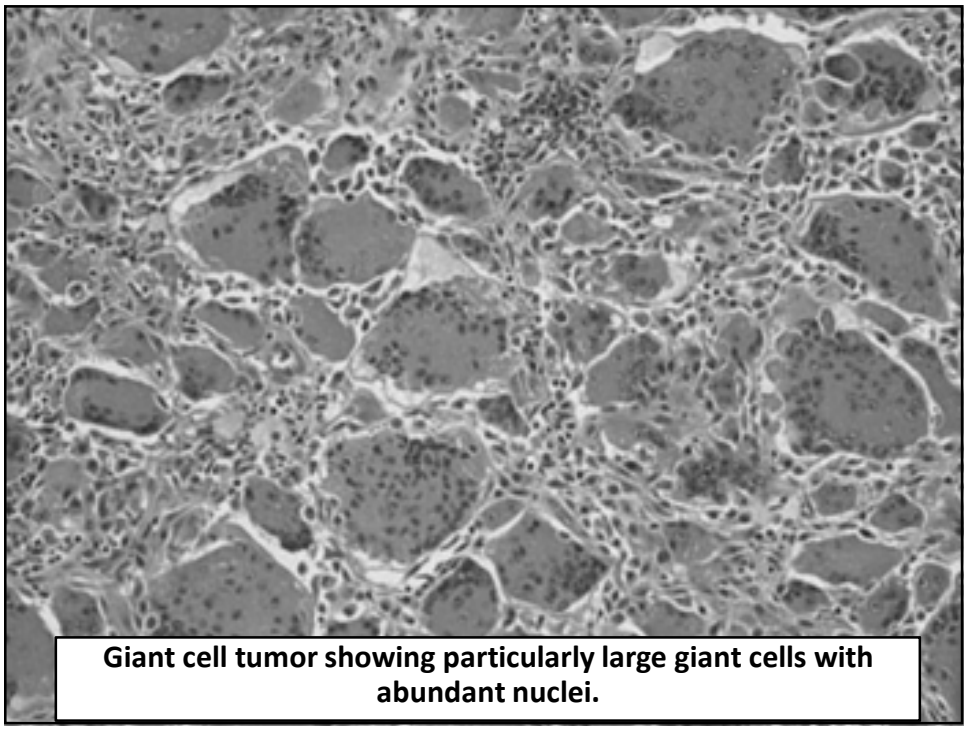

Giant cell tumor showing particularly large giant cells with abundant nuclei.

Treatment & prognosis:

- Surgical excision is the treatment of choice
- Promising clinical results have been associated with the use of anti-osteoclastogenic drugs (bisphosphonates, monoclonal antibody to RANK ligand [denosumab]).
- These lesions exhibit a greater tendency to recur after treatment than do giant cell granulomas. Although too few cases have been reported in the jaws to predict recurrence rates, it is noteworthy that 30% of lesions in long bones recur after curettage.

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Hemangioma of Bone

- Hemangiomas of bone are rare intraosseous vascular malformations that, when seen in the jaws, can mimic both odontogenic and nonodontogenic lesions. Difficult to control hemorrhage is a notable complication of surgical intervention.

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- More than half of central hemangiomas of the jaws occur in the mandible, (posterior regin)
- F= 2M
- second decade of life.
- A firm, slow-growing, asymmetric expansion of the mandible or maxilla
- Spontaneous gingival bleeding around teeth in the area of the hemangioma
- Paresthesia or pain, as well as vertical mobility of involved teeth, is occasionally evident.
- Bruits or pulsation of large lesions may be detected with careful auscultation or palpation of the thinned cortical plates.
- hemangiomas may be present with no signs or symptoms.
- The lesions may produce resorption of the roots of teeth in the area.

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- Radiographically, more than half of jaw hemangiomas occur as multilocular radiolucencies that have a characteristic soap bubble appearance.
- A second form of these lesions consists of a rounded, radiolucent lesion in which bony trabeculae radiate from the center of the lesion, producing angular loculations.
- Less commonly, hemangiomas appear as cystlike radiolucencies.

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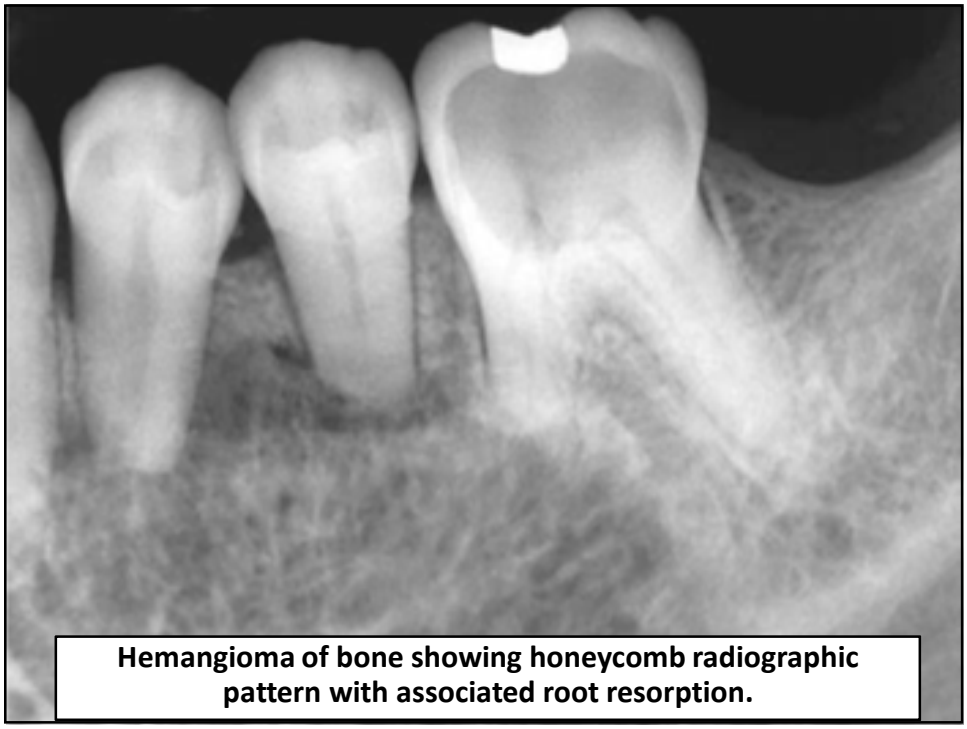

Hemangioma of bone showing honeycomb radiographic pattern with associated root resorption.

Histopathology

- a proliferation of blood vessels. Most intrabony hemangiomas are of the cavernous type (large-caliber vessels), while fewer are of the capillary type (small-caliber vessels).
- However, separation of hemangiomas into one of these two microscopic subtypes is academic, because no differences in biological behavior are noted.

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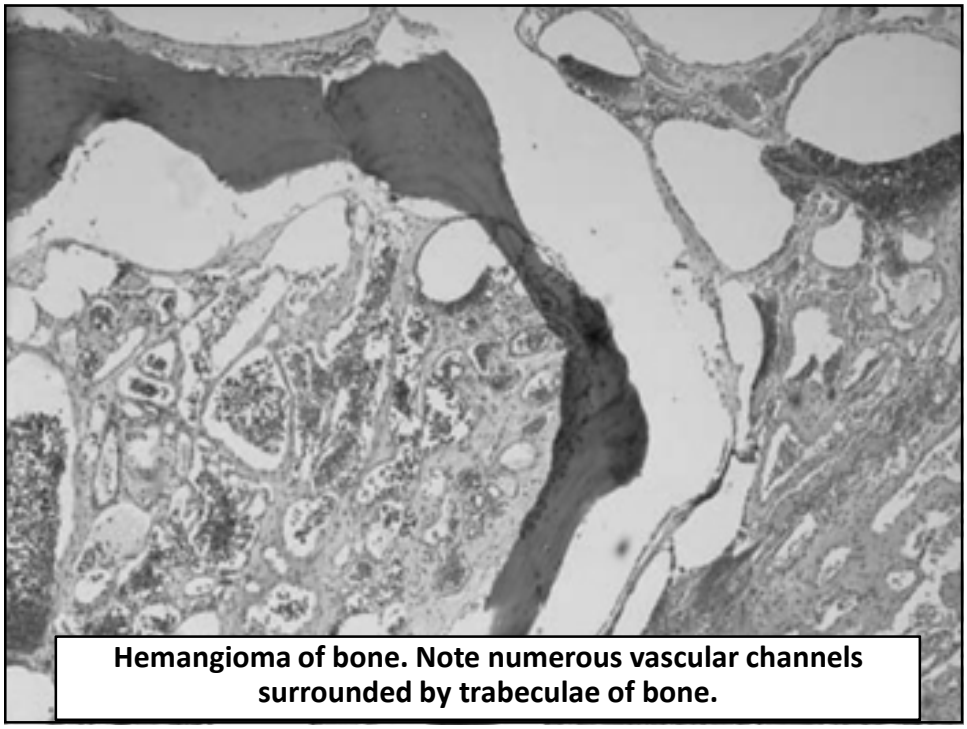

Hemangioma of bone. Note numerous vascular channels surrounded by trabeculae of bone.

Treatment & Prognosis

- may prove life threatening if improperly managed.
- Extraction of teeth in an area involved by a central vascular lesion may result in potentially fatal bleeding. It is imperative to perform needle aspiration of any central lesion that may be of vascular origin before performing a biopsy.
- TRT: surgery, radiation therapy, sclerosing agents, cryotherapy, and presurgical embolization techniques.
- The vascular supply of a given lesion, as well as its size and location, must be evaluated before a given treatment method is selected.

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Langerhans Cell Disease

Langerhans Cell Disease Classification

- Eosinophilic granuloma (chronic localized): solitary or multiple bone lesions
- Hand-Schüller-Christian (chronic disseminated): bone lesions, exophthalmos, diabetes insipidus
- Letterer-Siwe (acute disseminated): bone, skin, internal organs affected

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- Proliferation of dendritic cells with Langerhans cell features
- Cells are CD1a, CD207 and S-100: positive
- Cells contain Birbeck granules (ultrastructure)
- Few macrophages (histiocytes) are present
- Cause unknown
- Any age; three variants
- Radiograph shows punched-out noncorticated lesions or “floating teeth”
- Several treatment options
- Prognosis good to excellent; depends on form

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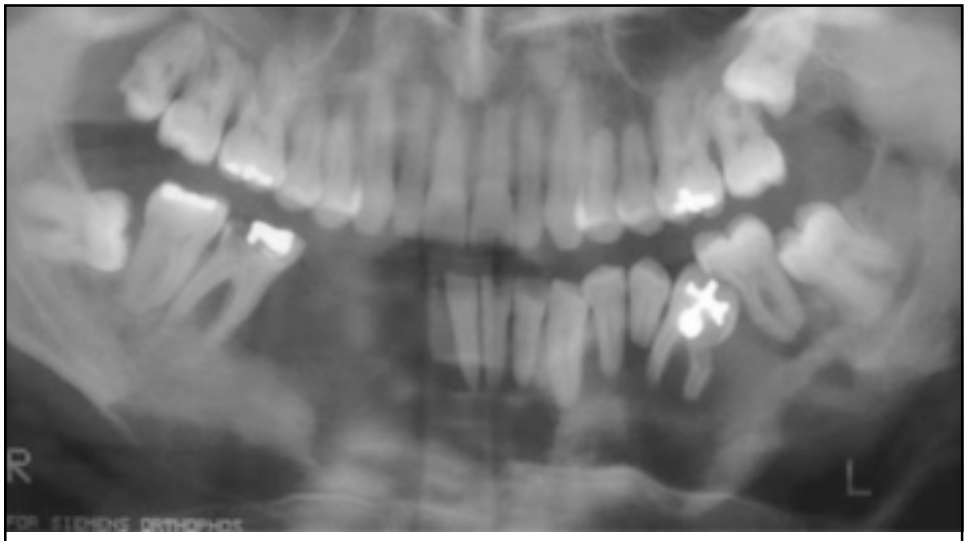

Langerhans cell disease resulting in marked destruction of the mandible

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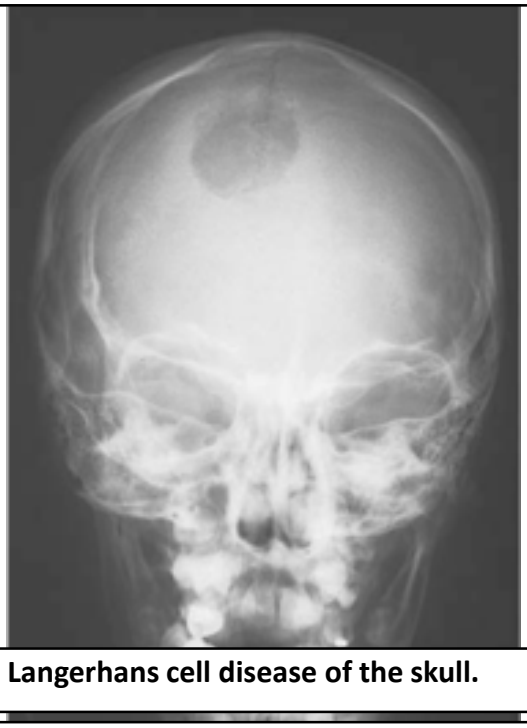
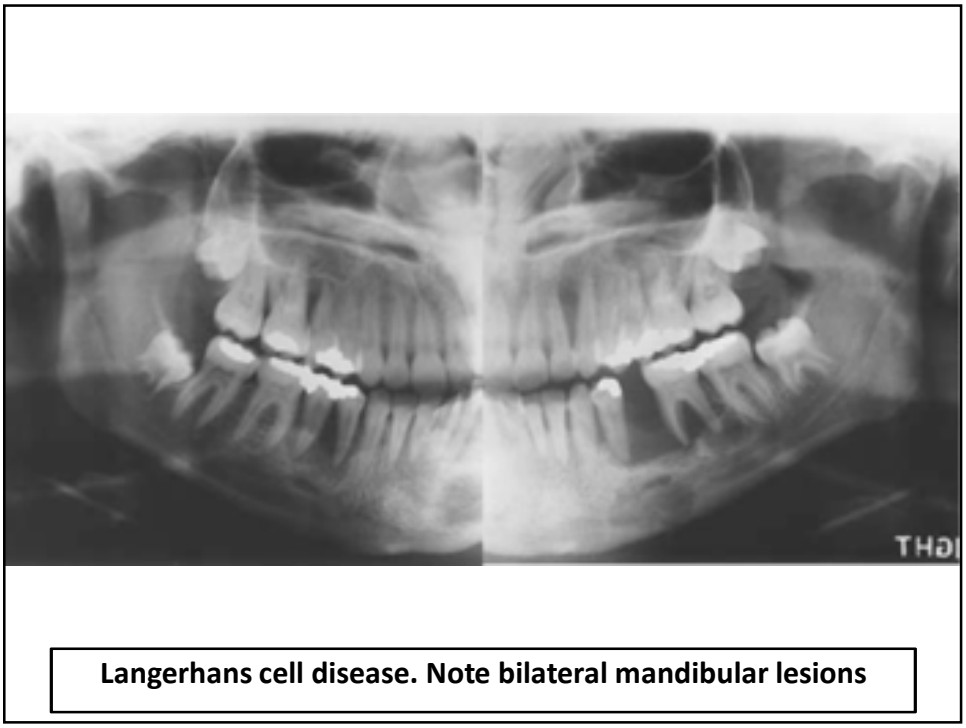
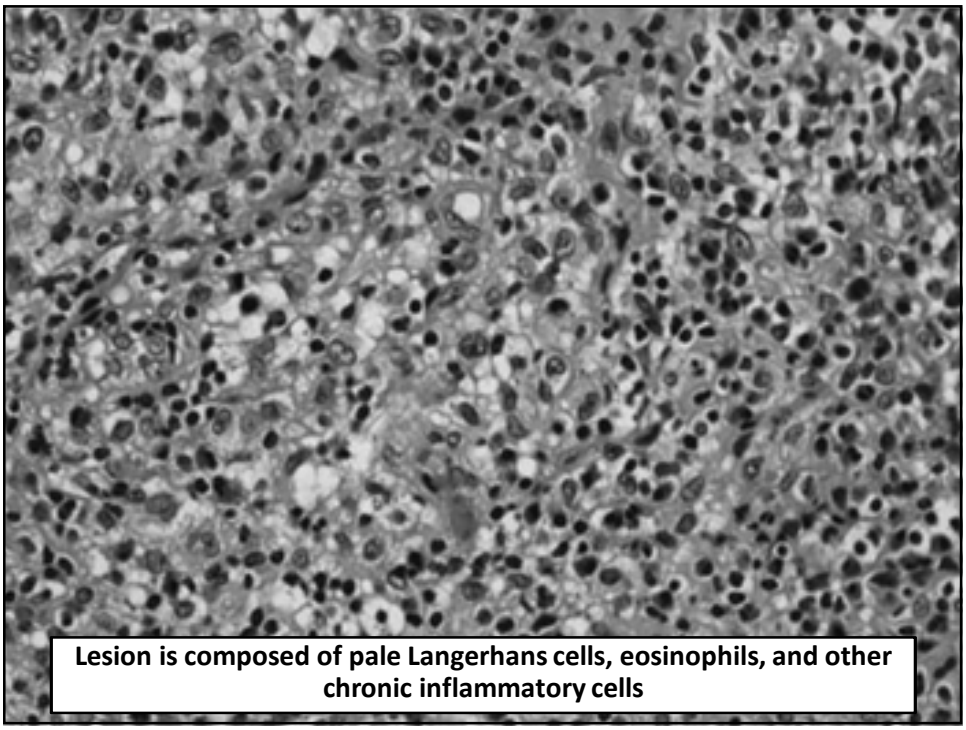


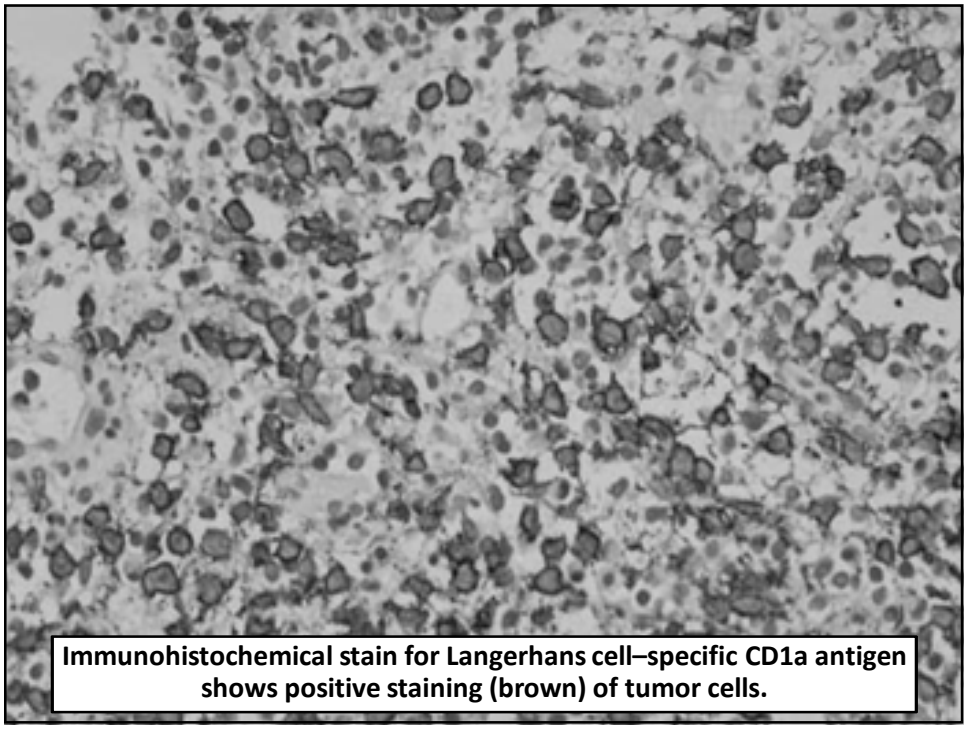
Langerhans cell disease of the skull.



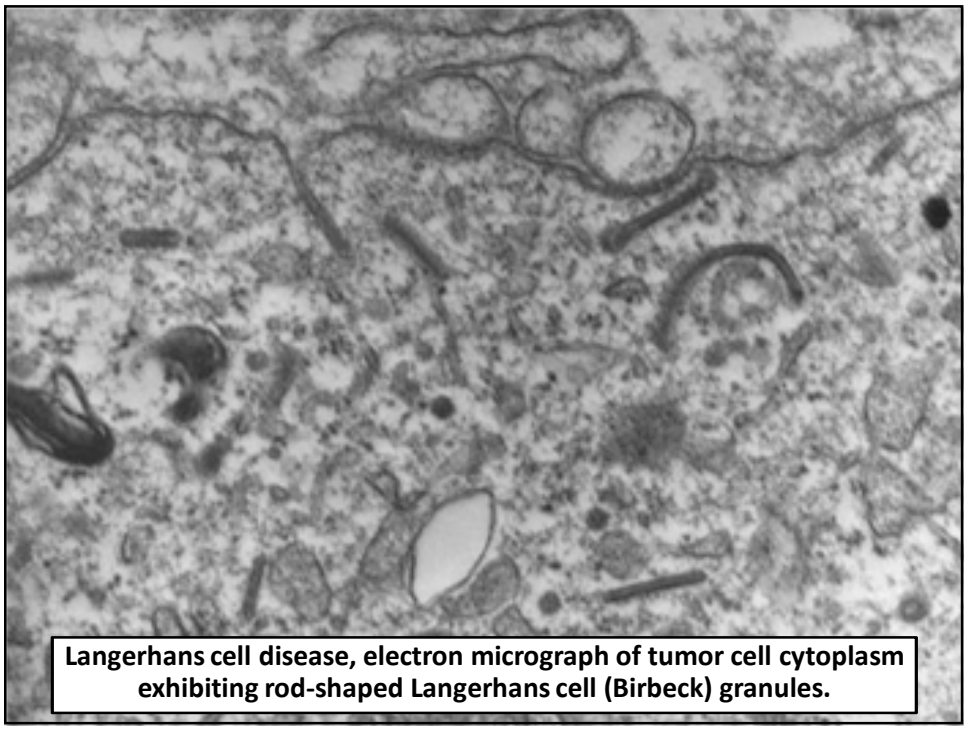
Langerhans cell disease. Note bilateral mandibular lesions



Lesion is composed of pale Langerhans cells, eosinophils, and other chronic inflammatory cells



Immunohistochemical stain for Langerhans cell–specific CD1a antigen shows positive staining (brown) of tumor cells.



Langerhans cell disease, electron micrograph of tumor cell cytoplasm exhibiting rod-shaped Langerhans cell (Birbeck) granules.

Treatment

Localized Disease:

- Curettage
- Radiation, low dose
- Intralesional corticosteroid injection
- Rare spontaneous regression

Disseminated Disease

- Immunosuppressive agents, corticosteroids, cytosine arabinoside

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Tori and Exostoses

- Tori and exostoses are nodular protuberances of mature bone; their precise designation depends on the anatomic location. These lesions are of little clinical significance because they are non-neoplastic and rarely are a source of discomfort. The mucosa surfacing these lesions occasionally may be traumatically ulcerated, producing a slow-healing, painful wound or, less commonly, osteomyelitis. Surgical removal for the

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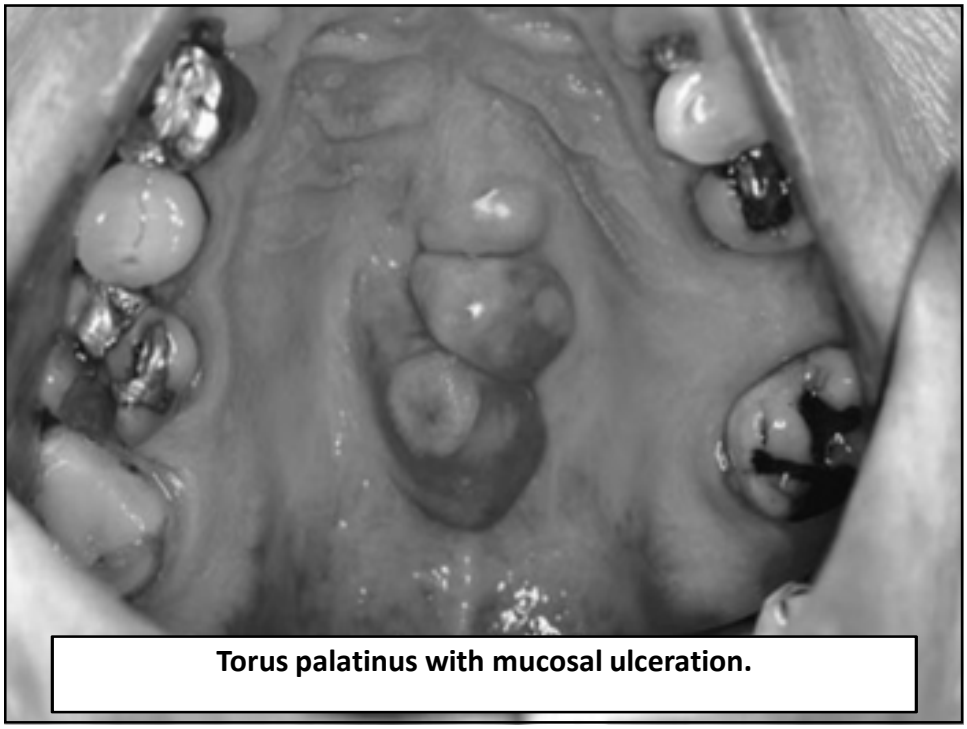
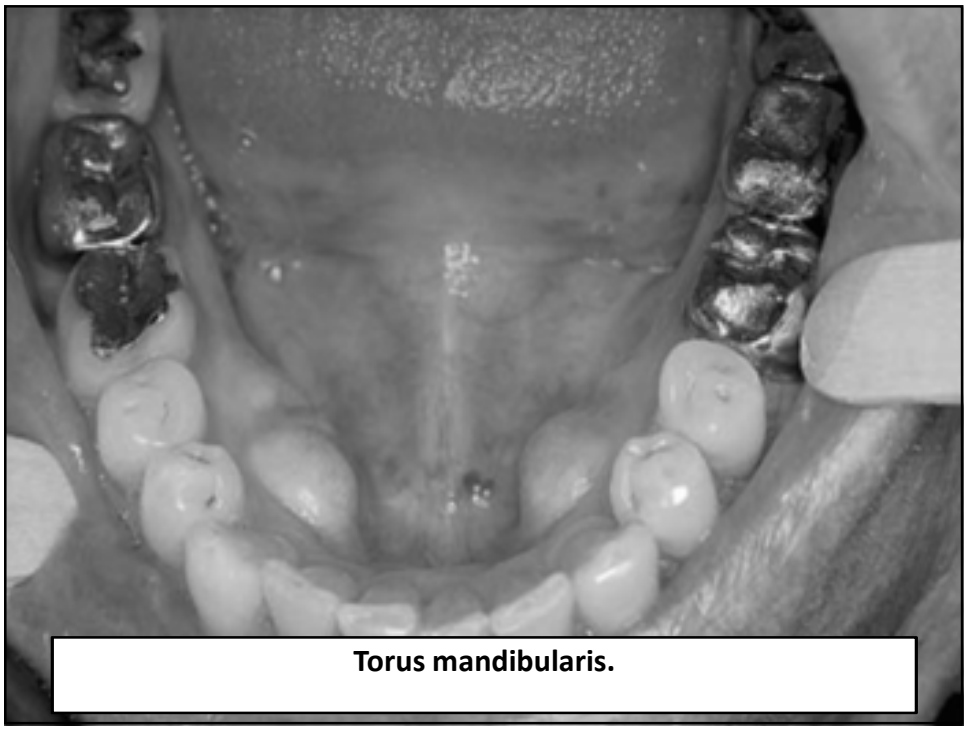
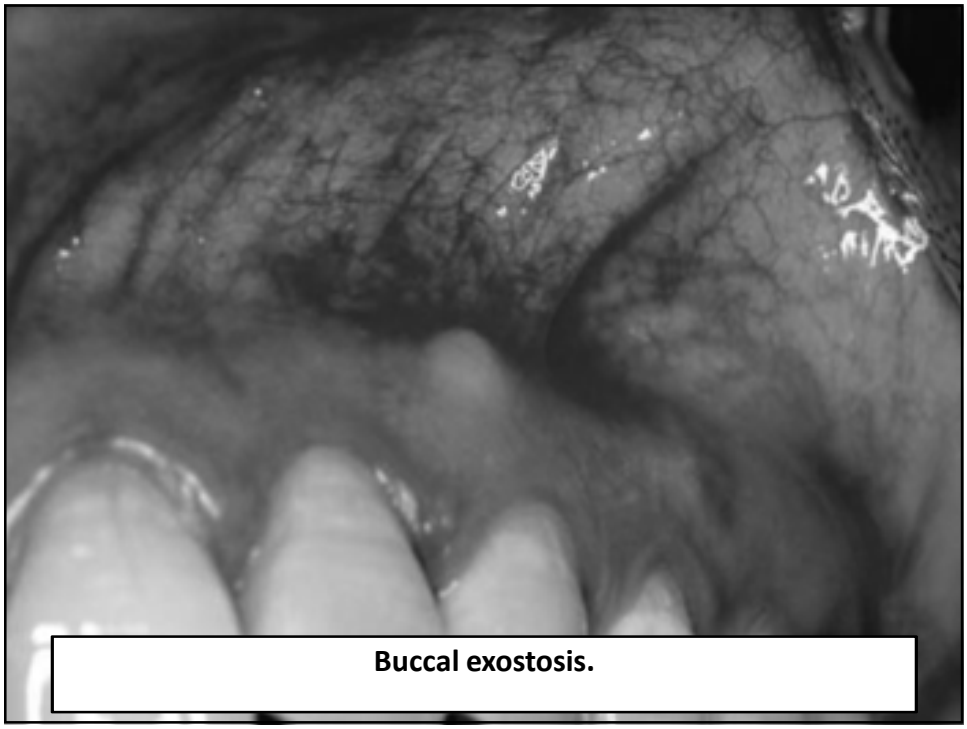


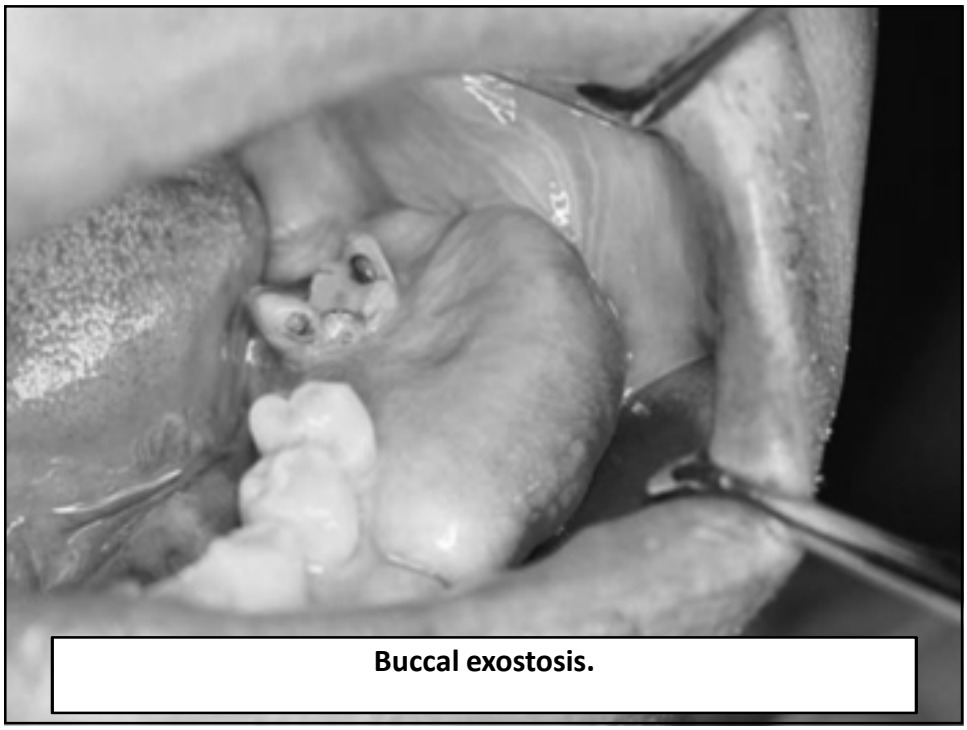
Torus palatinus with mucosal ulceration.



Torus mandibularis.



Buccal exostosis.



Buccal exostosis.


Thank you ...