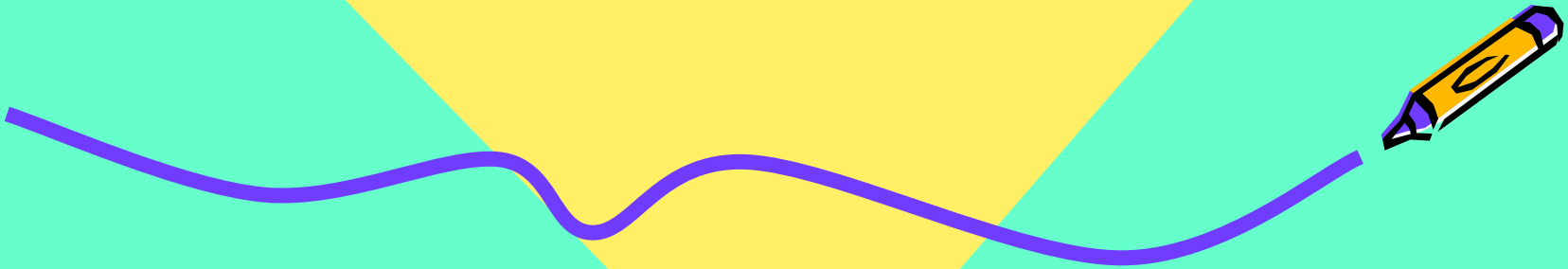
BONE TUMORS



- Primary tumors of bone are uncommon lesions in the jaw.
- They arise from any cells or tissues present in the bone

  (periosteum, N.F, marrow T, B.V, F.T, cartilage)

Osteoma

-Benign tu composed of mature (well-differentiated) compact or cancellous bone.

-Most commonly occur in craniofacial skeleton – rare in other parts of body.

TYPES OF OSTEOMA :

(clinical & radiographical)

Periosteal:-arise on surface of bone

Endosteal:-located in medullary bone

- Extra-skeletal osteomas that occur in buccal mucosa,tongue,nasal cavity, are not true neoplasm &termed **"Choristoma"**

  tumor-like growth of normal tissue in an abnormal location

Palatal and mandibular tori are not considered as osteomas although they are histologically identical.

Hamartoma== Disorganized overgrowth of tissue in their normal location.

An excess of normal tissue in a normal situation

Choristoma== Normal tissue growth in an abnormal location
e.g: gastric tissue located in distal ileum
(Meckles diverticulum)

An excess ot tissue in an abnormal location

CLINICAL FEATURES : -

- Age incidence: Young adults (**25-35 y**)
- Sex incidence: Nil
- Site predilection: Mandible affected more commonly than maxilla.

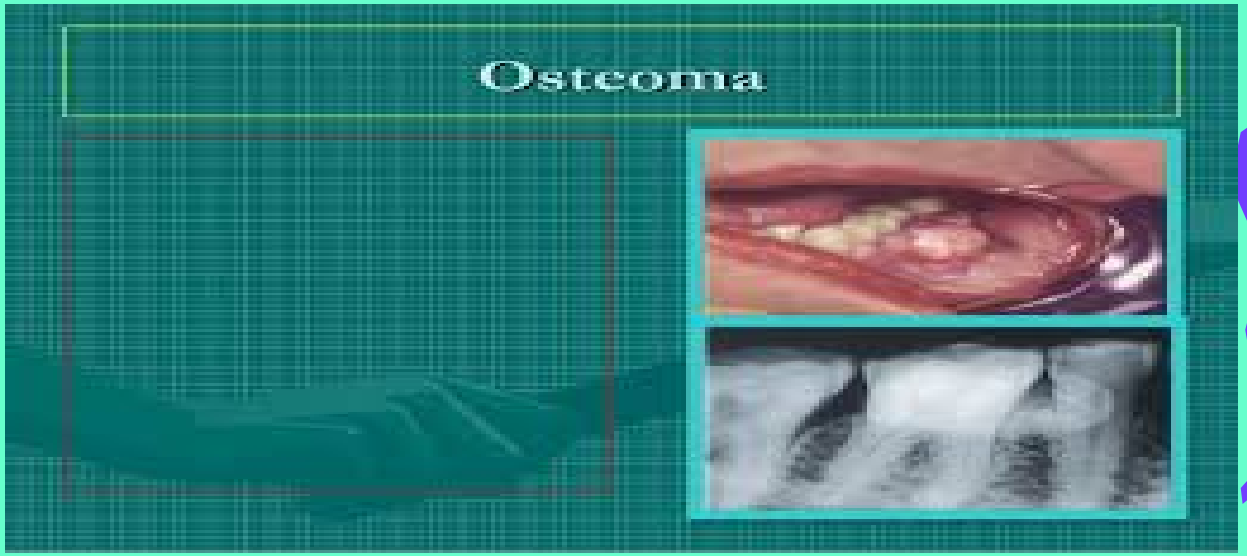
  In mandible – body / condyle.

  Body of mandible – posterior to premolars on lingual surface.

Osteoma involving mandibular condyl may cause a slowly progressing shift in patient's occlusion, with deviation of midline of the chin toward unaffected side.

Other sign & symptomes includes:- Facial swelling, Pain, Limited mouth opening

Symptoms of osteoma in Head & Neck region include:- Chronic sinusitis, Headache, Nasal obstruction, Hearing loss



- **RADIOGRAPHIC FEATURES :**
- Endosteal osteomas appear as radiopaque sclerotic masses.
- Periosteal osteomas may appear as uniform 'opaque mass or sclerotic periphery with central trabecular pattern.

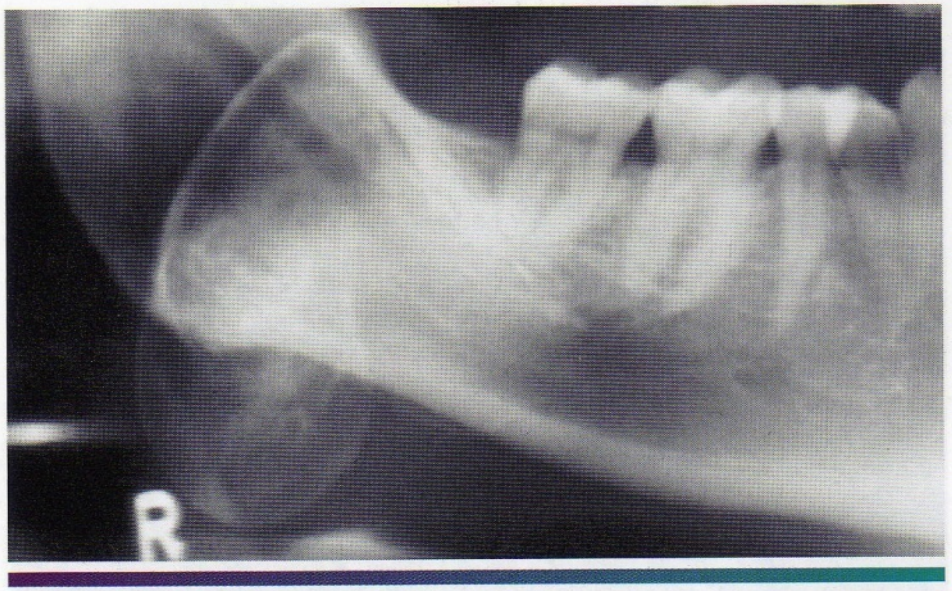


Figure 14-58 • **Osteoma.** The radiograph shows a pedunculated cancellous osteoma arising from the lingual surface of the mandible near the crest of the alveolar ridge.

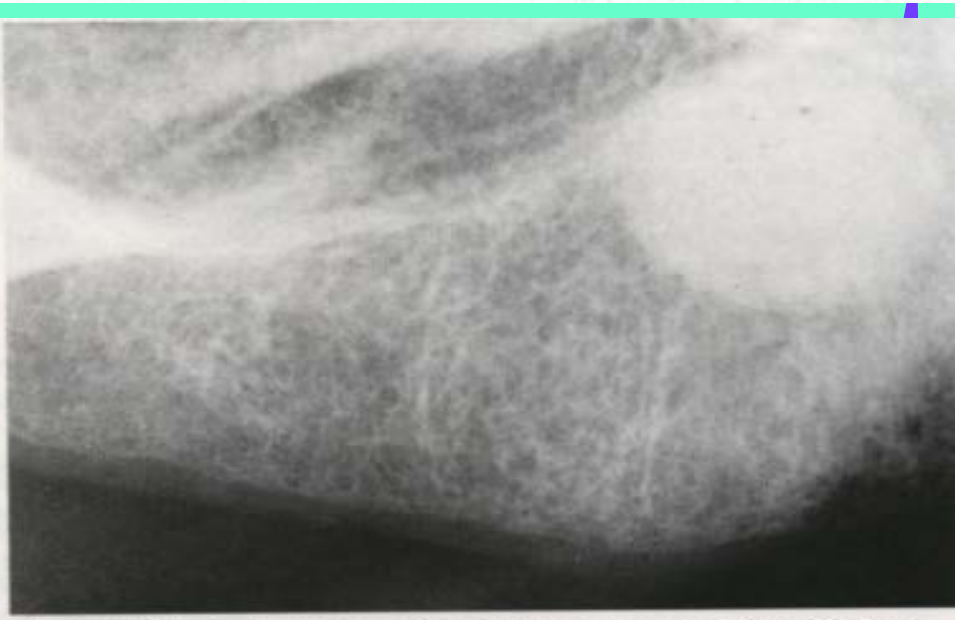


Fig. 7.4 Endosteal osteoma: this compact osteoma was attached to the surrounding bone by a small bony pedicle.

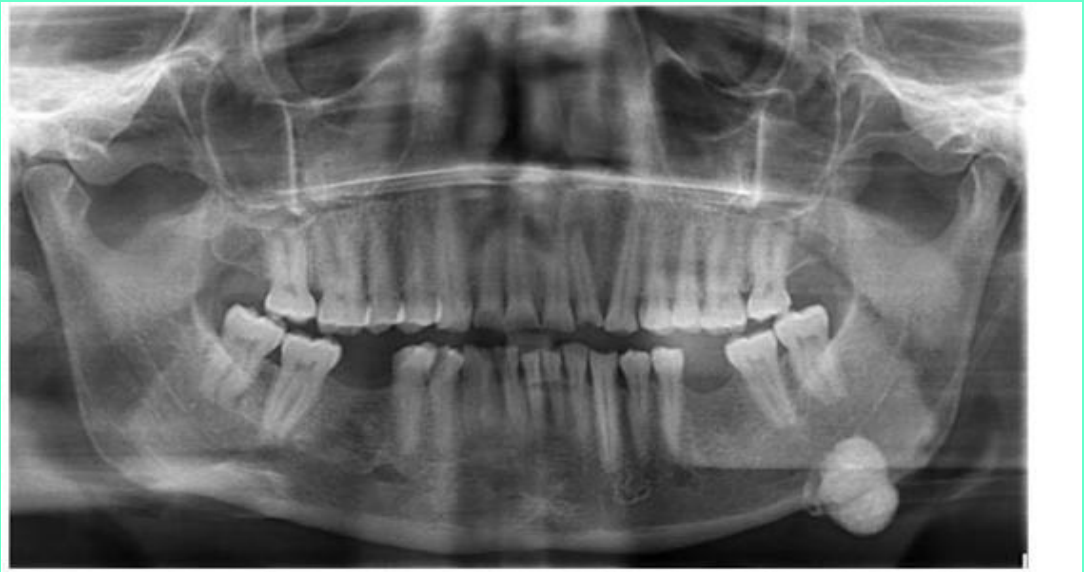
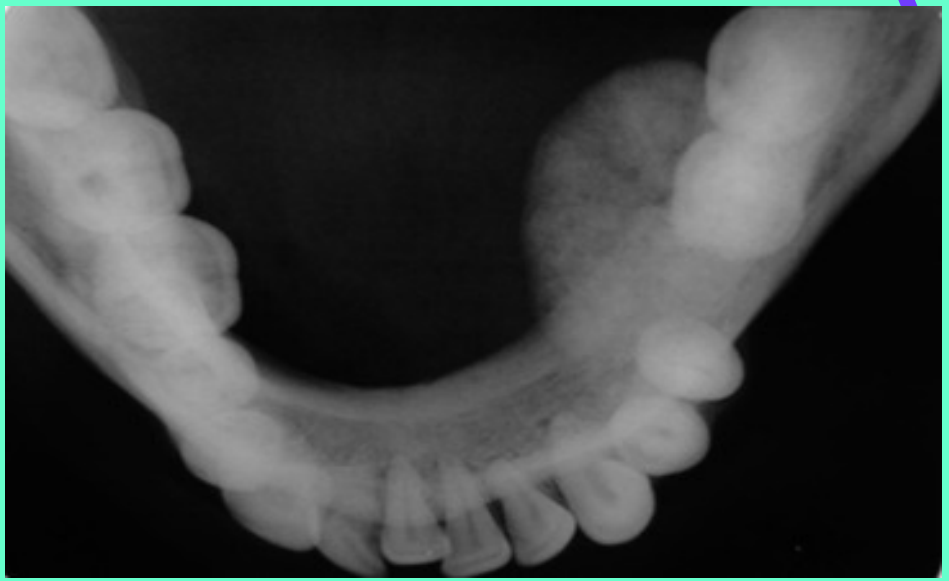
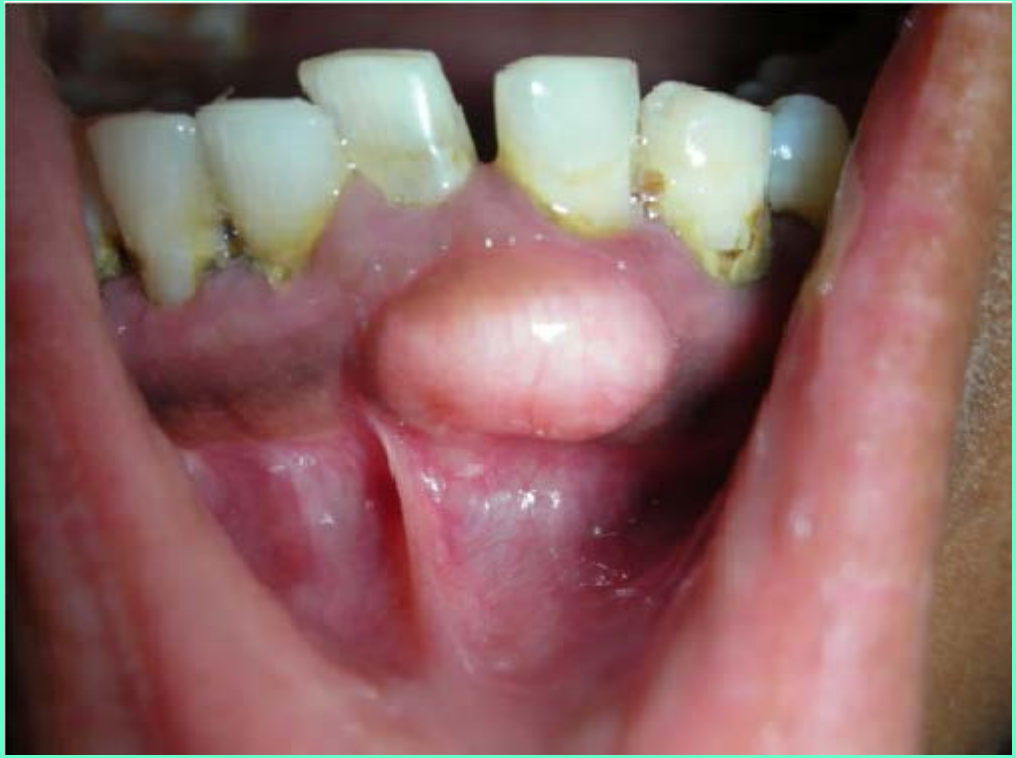
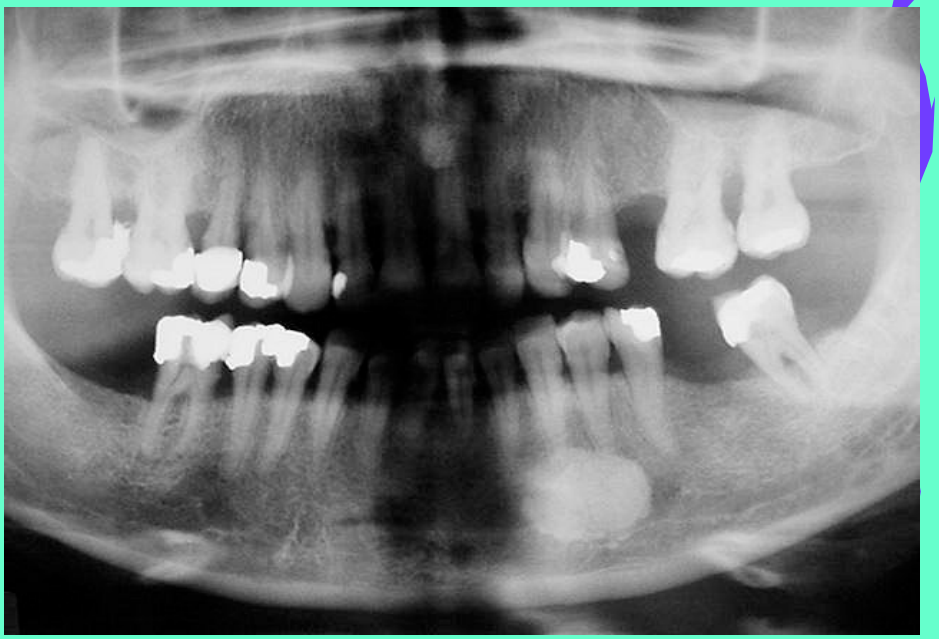


Figure 2: Panoramic radiograph showing a large radiopaque lesion in the left angle of the mandible obscuring the mandibular canal.

Occlusal radiograph







Histopathology

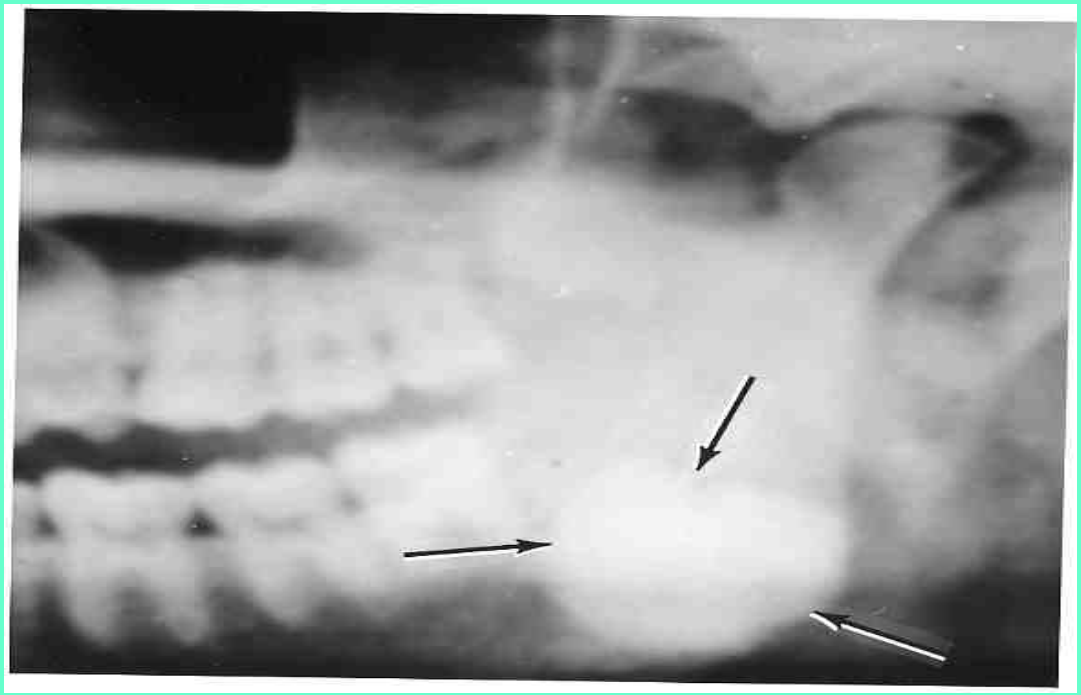
- It can be divided to:

1-Compact (ivory) osteoma

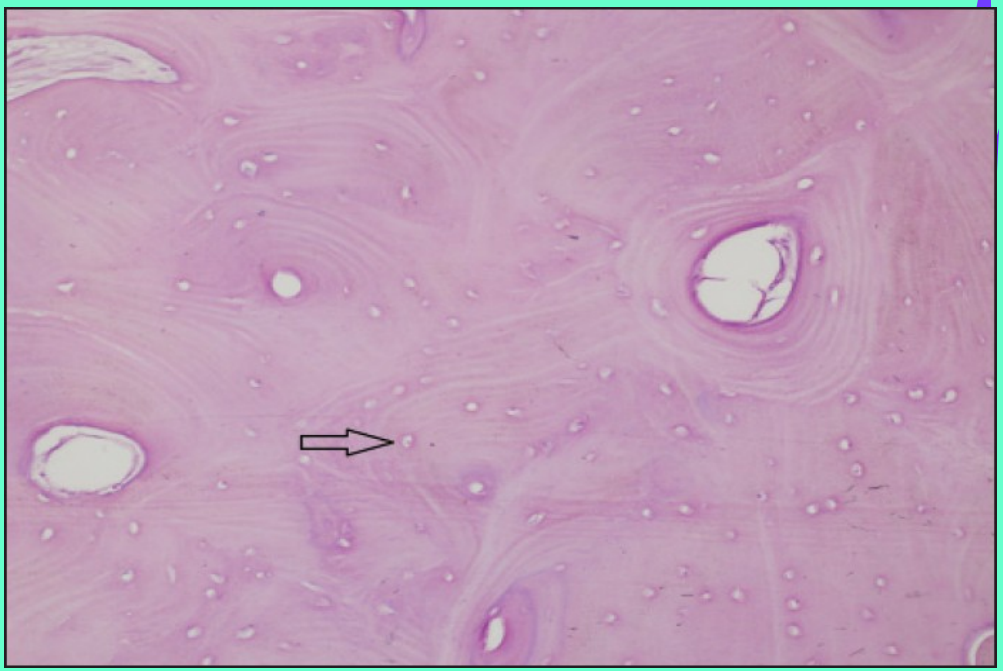
Mass of dense, compact, lamellar bone with a few marrow space(fibrous)

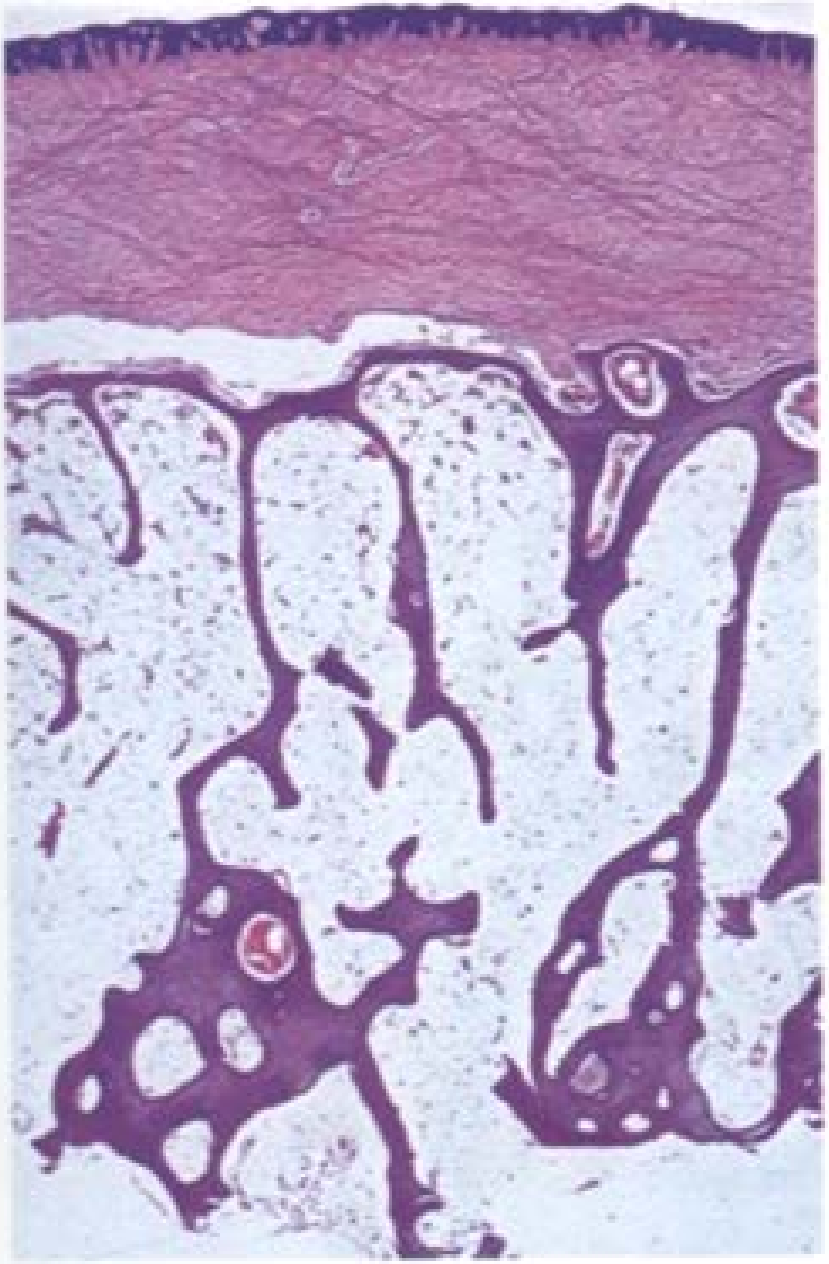
2-Cancellous osteoma

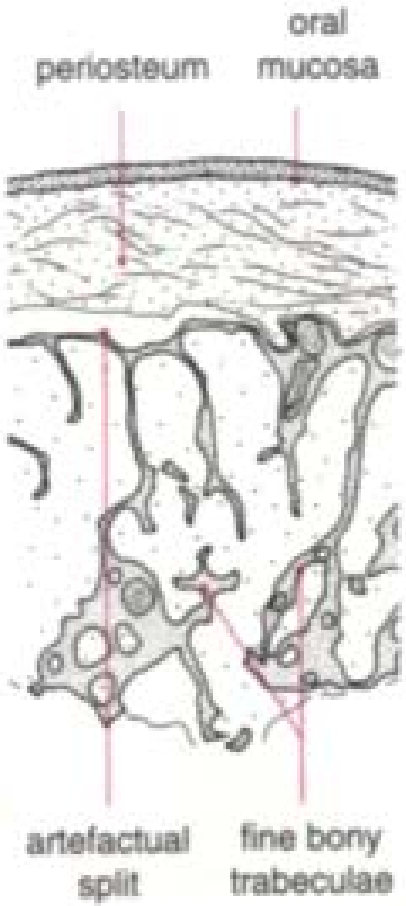
Interconnecting mature trabeculae enclosing fatty or fibrous marrow.



Compact osteoma







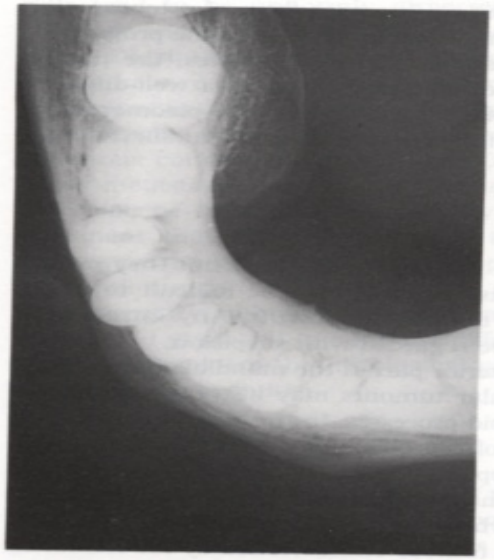


Fig. 17.16

Fig. 17.18

Fig. 7.7 Cancellous osteoma: this section shows thin bony trabeculae covered by periosteum and oral mucosa.

Osteoma

- It is restricted to craniofacial bones, rarely in other bones. Asymptomatic. It is a solitary lesion, multiple osteoma of jaws occur as a features of

Gardner syndrome

Rare familial disorder, transmited as an autosomal dominant trait, consist of:

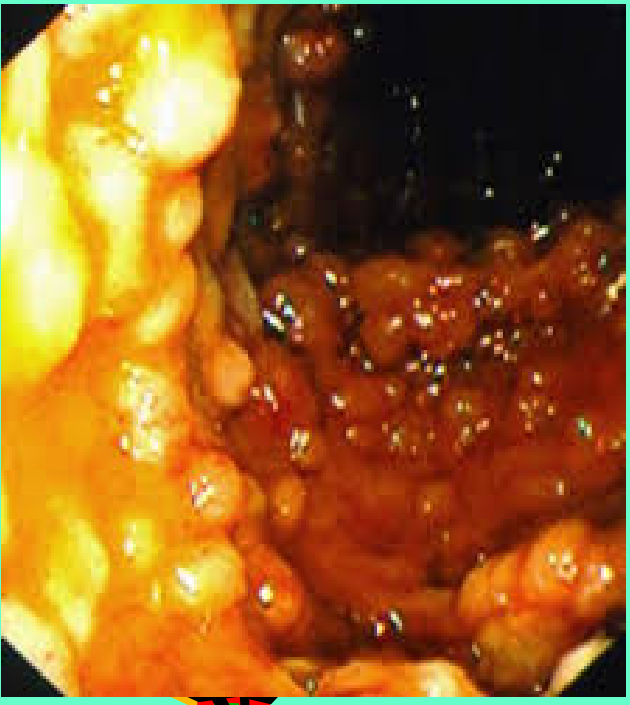
- Multiple intestinal polyp with malignant potential
- Multiple keratinous cysts of skin
- Multiple fibromatosis
- Multiple impacted & supernumerary teeth
- Multiple osteomas of skull & mandible

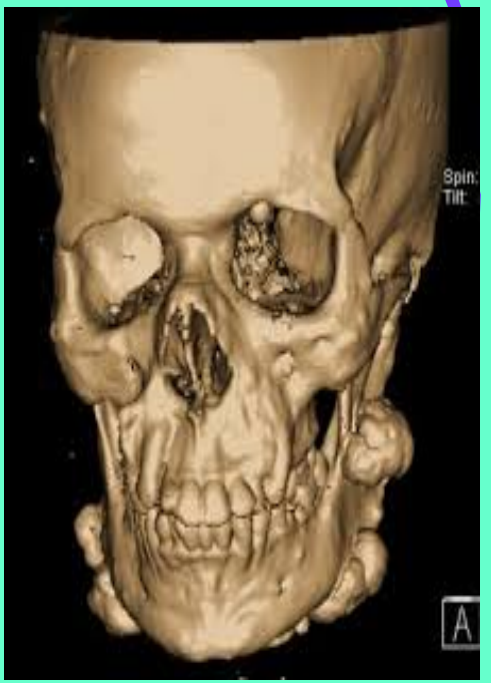
Treatment

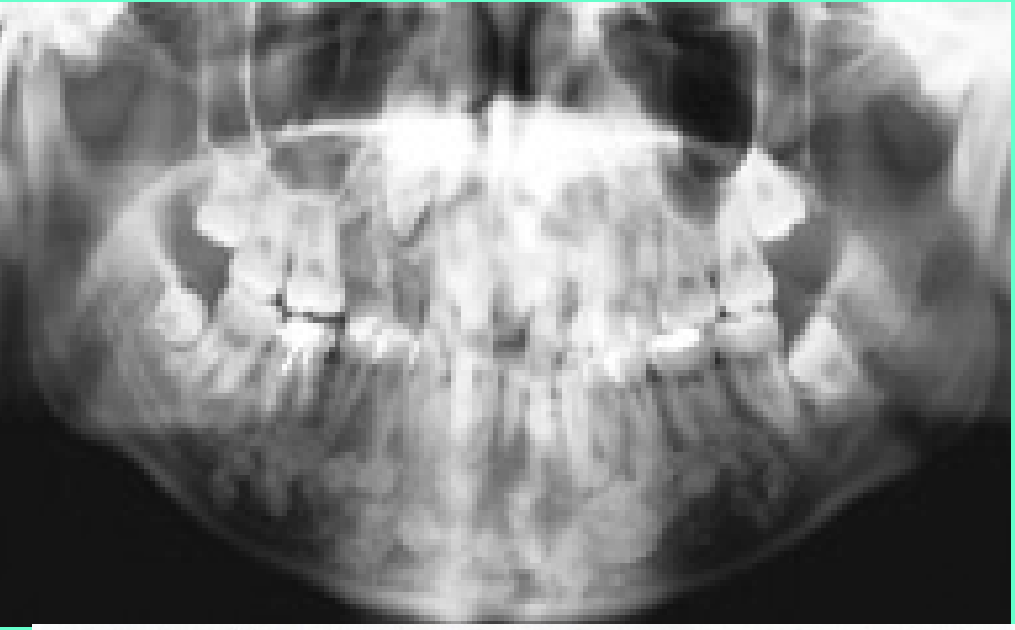
Excision → if it is large enough to cause symptom or difficult denture fitting

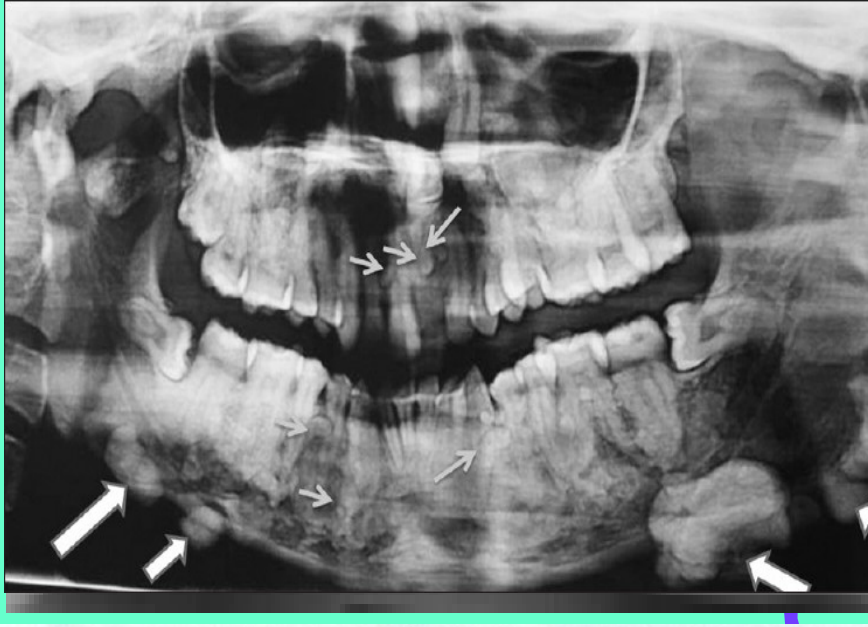
Recurrence is quite unusual

Figura 1. Protuberâncias múltiplas ósseas maxilar









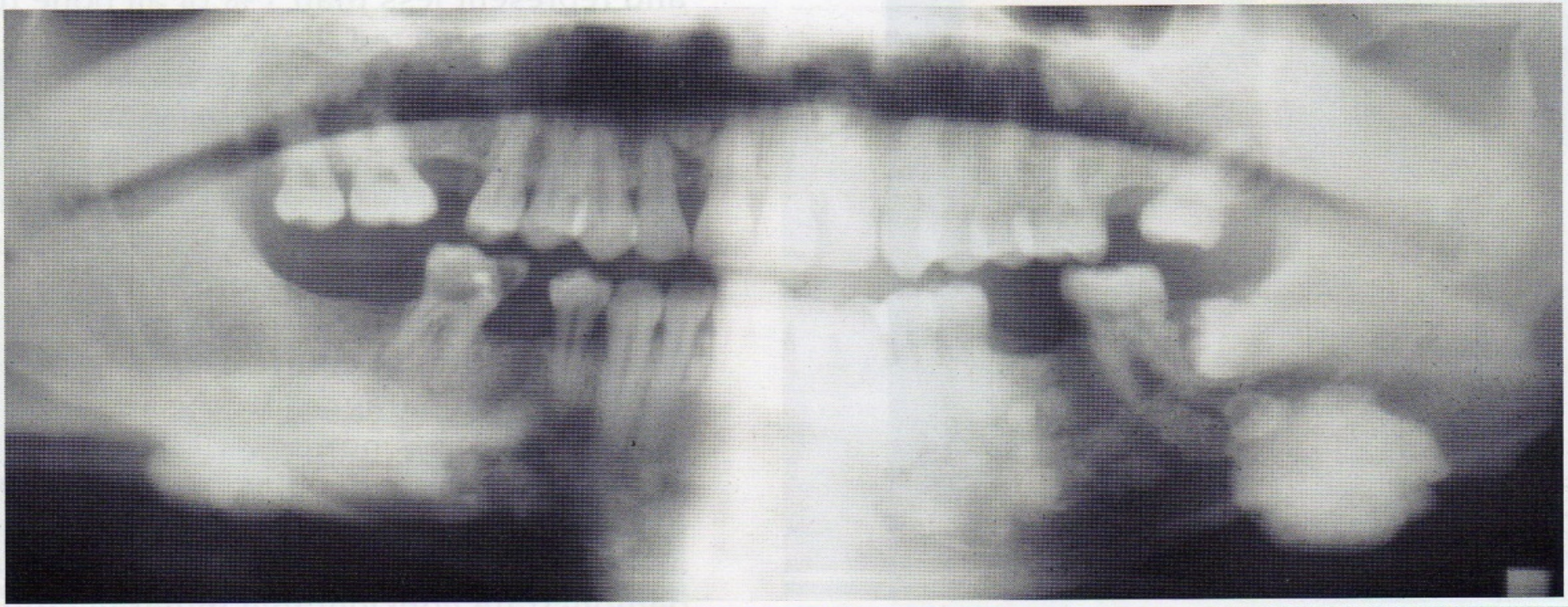


Figure 14-60 ◆ **Gardner syndrome.** Panoramic radiograph showing multiple osteomas of the mandible.

Haemangioma:

The word "hemangioma" comes from the Greek== haema blood";

"

angeio -"vessel"; oma -"tumor

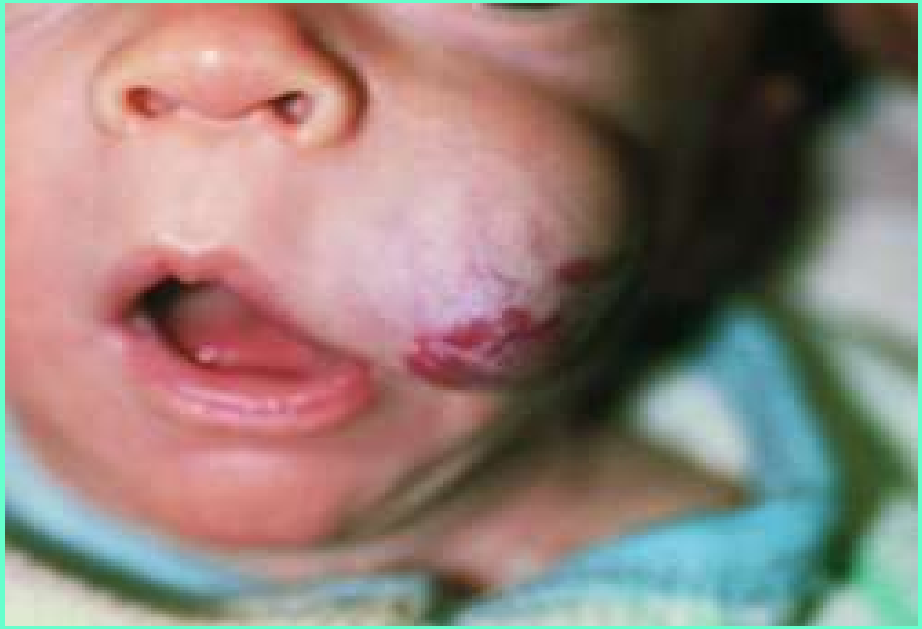
- Rare tu of jaw, more in mandible
- Vascular malformation,, Hamartomas
- Radiographically ⟶ osteolytic defect

⟶ multilocular, honey-comb appearance

-Aspiration ⟶ fresh blood

-Microscopically ⟶ marrow space contain very large blood-filled sinuses lined by endothelial cell,& with thin walls.

Cavernous hemangioma



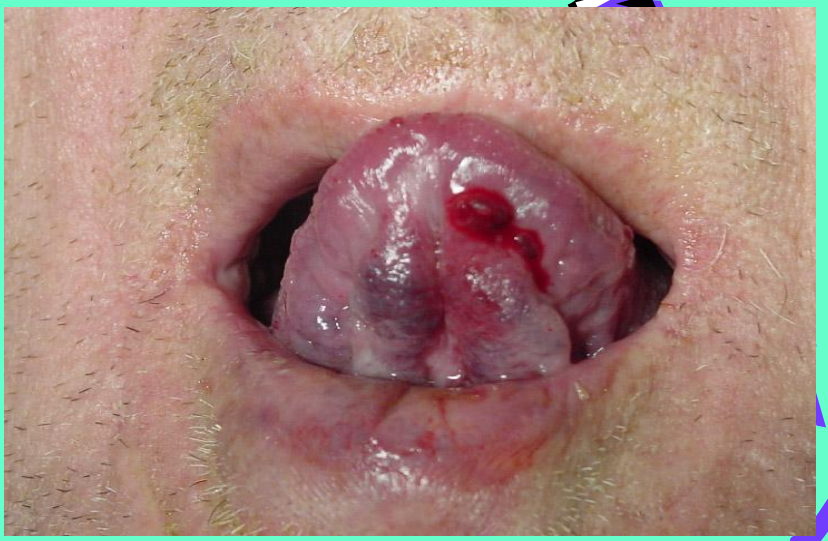
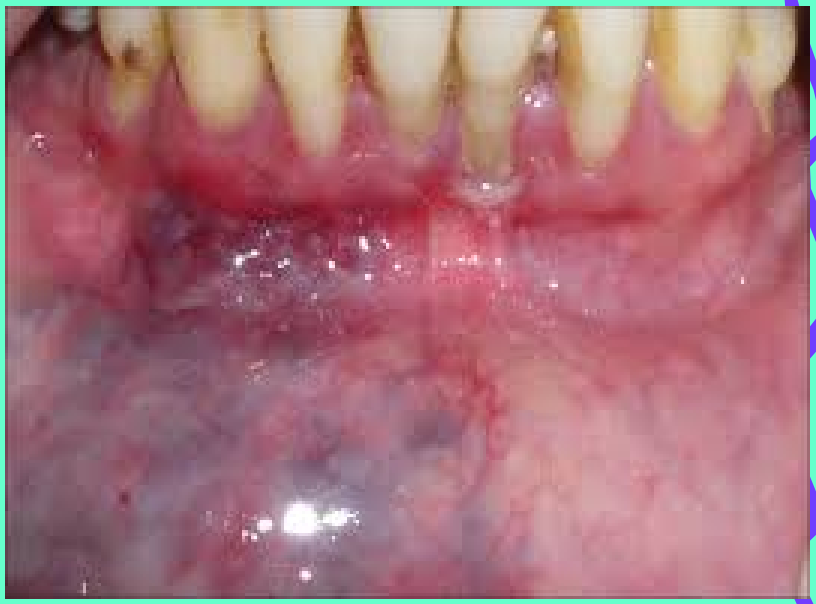
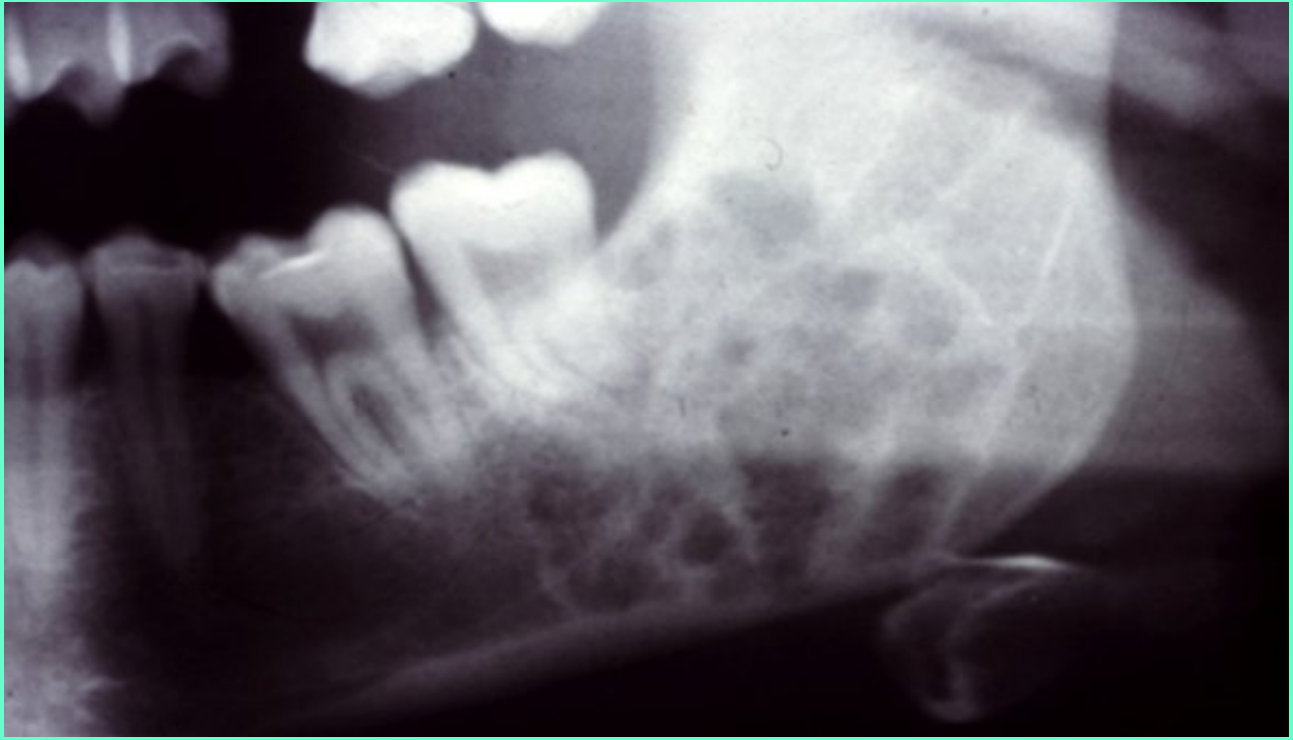
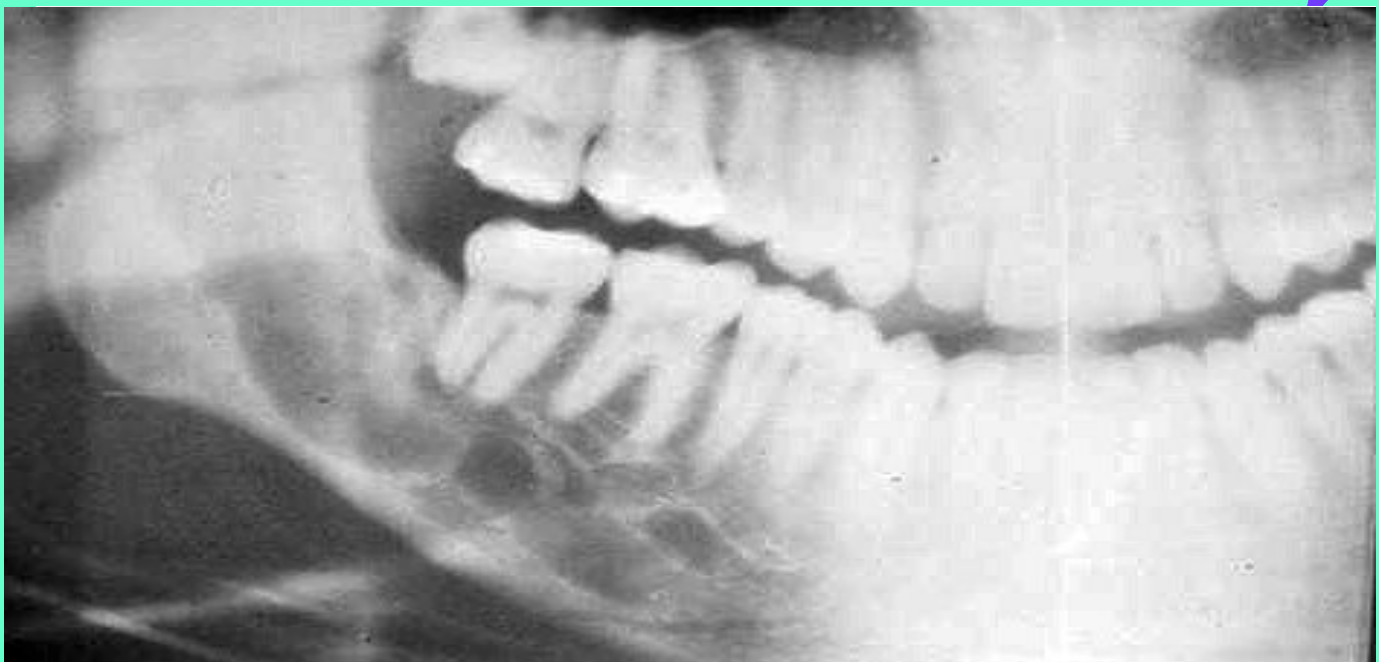


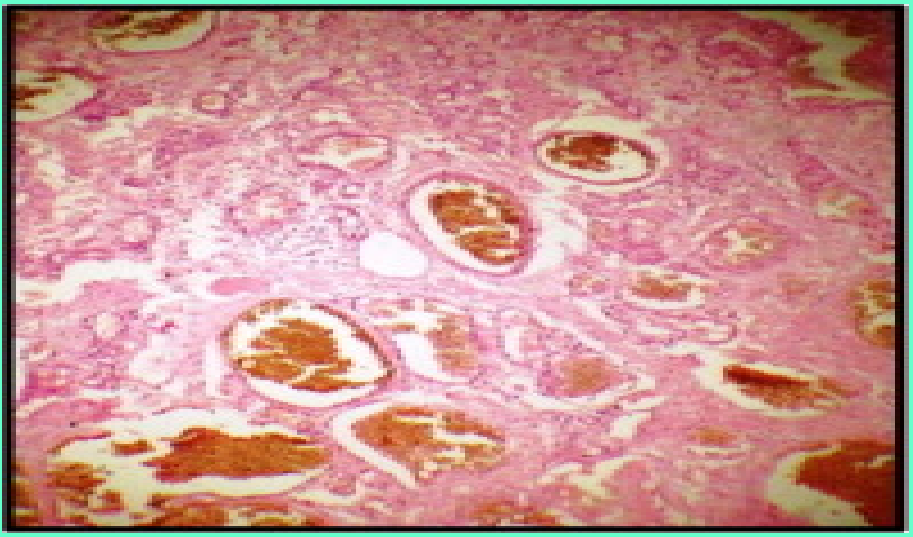
Fig. 1 A capillary hemangioma is an abnormal overgrowth of blood vessels that is sometimes referred to as a "strawberry" birthmark.

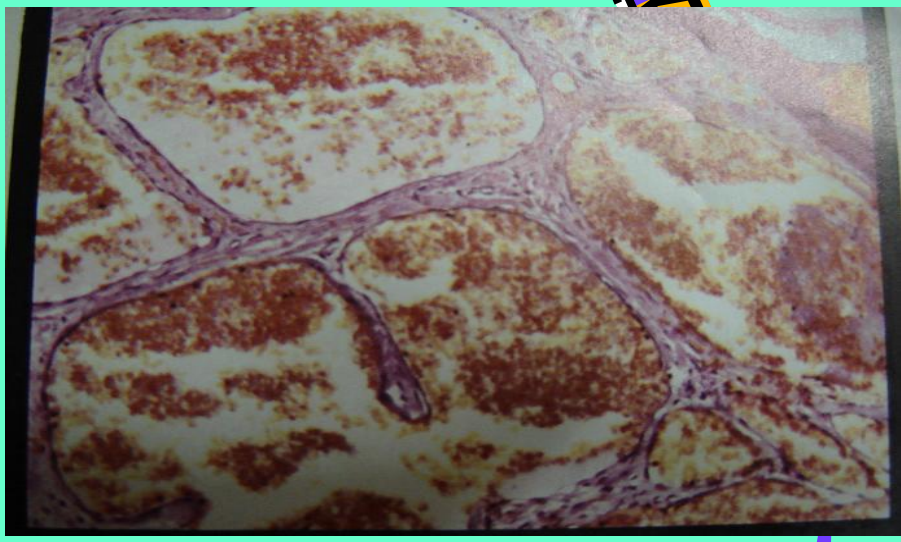


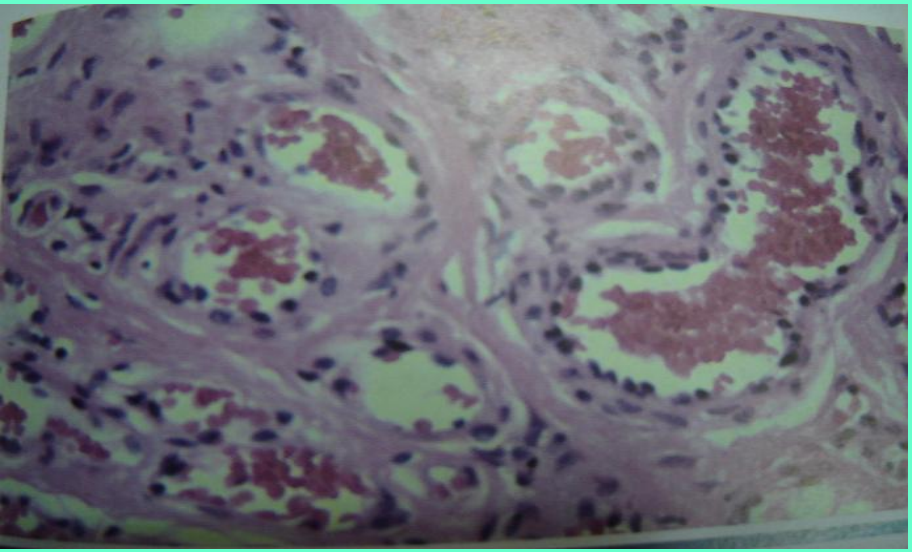
Hemangioma (cavernous)

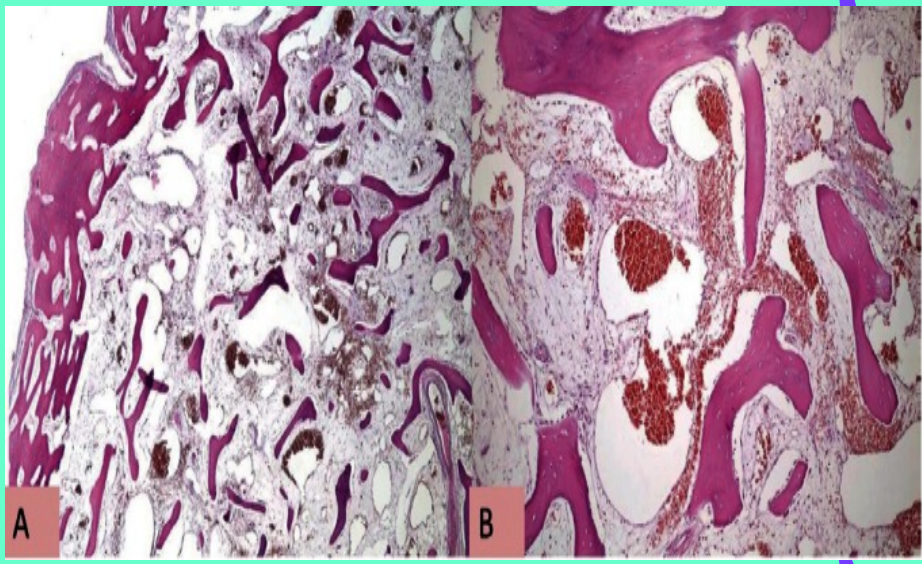












Hemangioma (capillary)

Hemangioma (cavernous)

Treatment:

Life threating lesion
Tooth extraction ------Fatal bleeding

Methods of treatment:

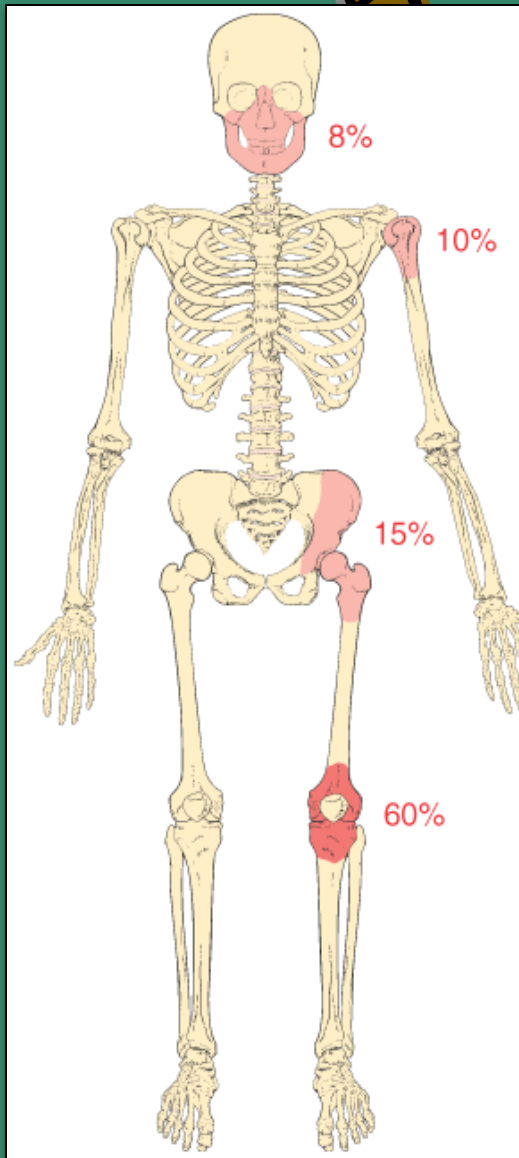
Surgery
Radiation therapy
Sclerotic agents
Cryotherapy

Osteosarcoma

Most common primary neoplasm of bone.
Highly malignant.Rare in jaw
(6-8 %) of all osteosarcomas

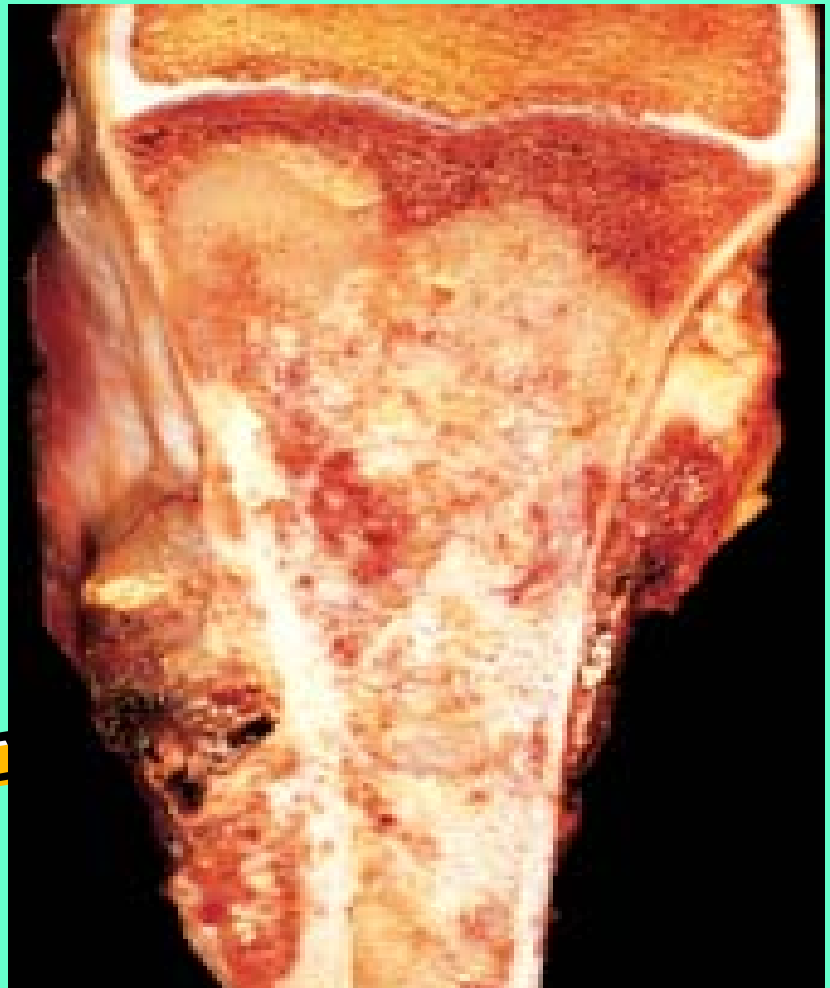
-Pt with jaw tu present around age of 30 yrs (decade later than in tu elsewhere in the skeleton).

-Present in older pt after irradiation, chemotherapy,

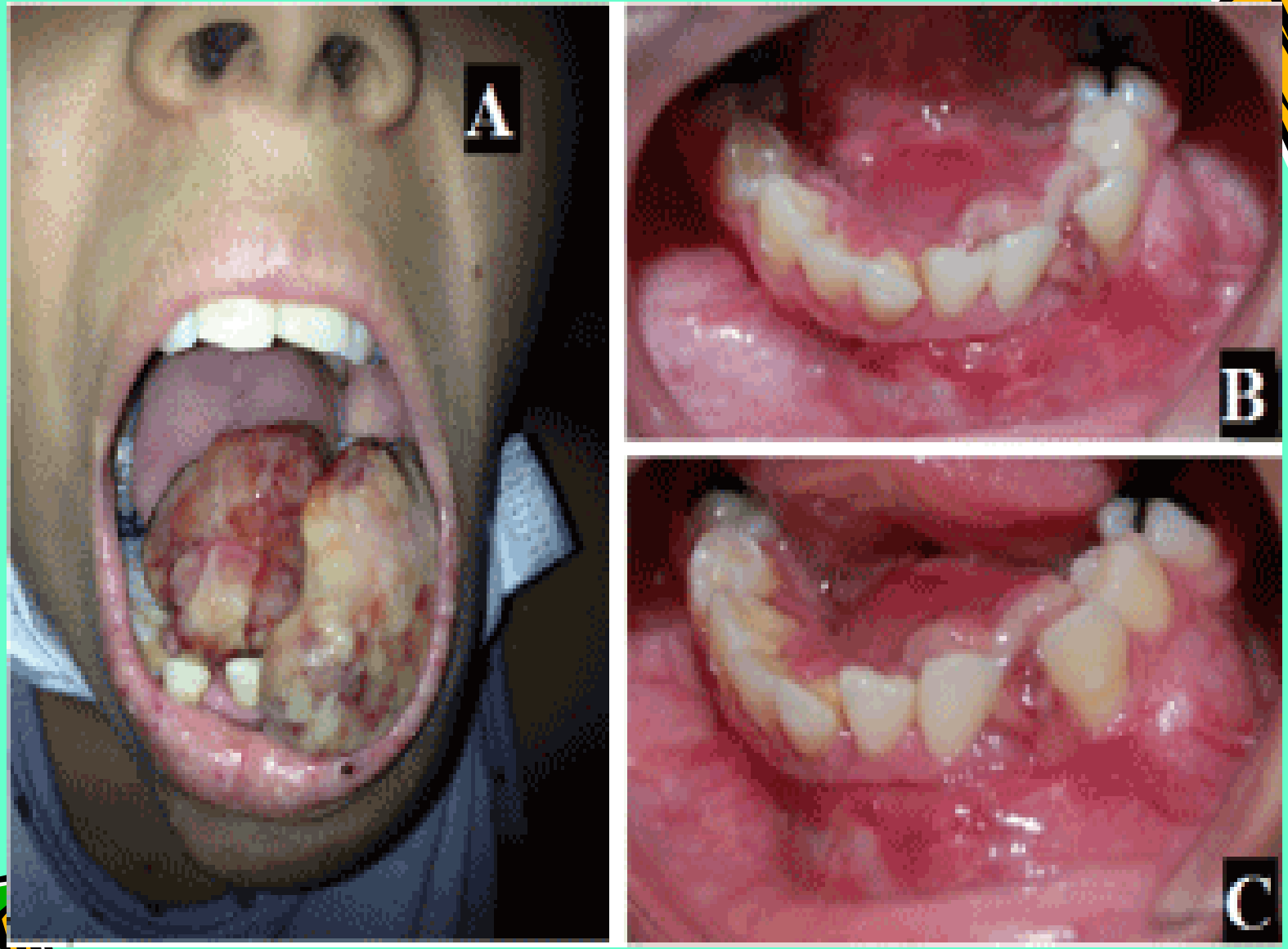


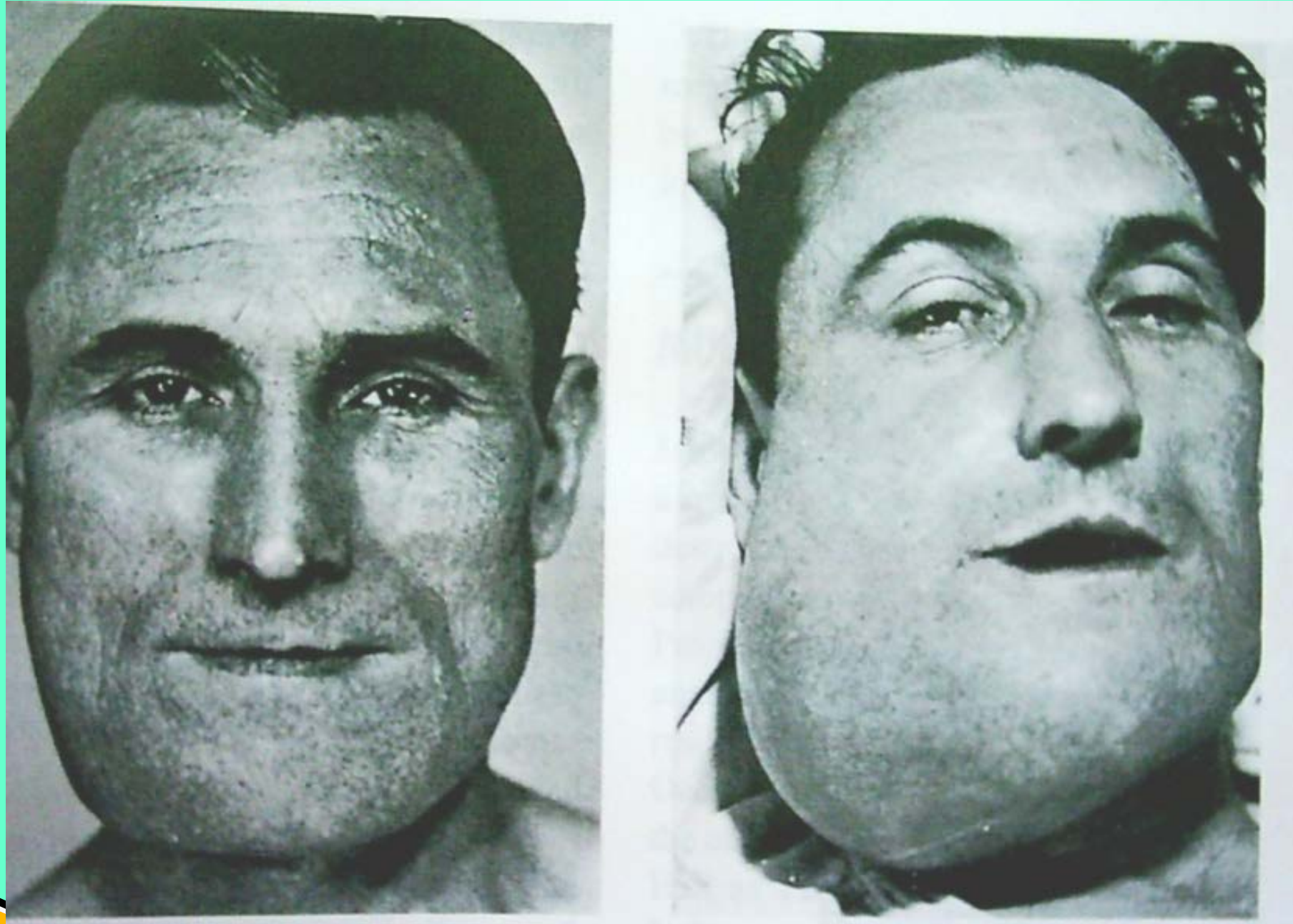
Most osteosarcoma of long bone located in the metaphyseal region.
e.g: The lower end of femur & upper end of humerus.

Jaw tu presents with swelling, pain, parasthesia, loosening of teeth,nasal bstruction.



Osteosarcoma of the upper end of the tibia. The tan-white tumor fills most of the medullary cavity of the metaphysis and proximal diaphysis. It has infiltrated through the cortex, lifted the periosteum, and formed soft tissue masses on both sides of the bone.


A
B
C



. 9.15 Osteosarcoma. The pictures show the progress of the tumour over a period of 3 nths. The patient died with metastases in the lungs, about a month after the second picture

RADIOGRAFICALLY

Variable → osteolytic type → irregular R.L.
→ osteoblastic → irregular R.O.

- If cortical plate perforated by the tu →
  Periosteum raised → tu extend into the
  Surrounding soft T → characteristic radiog
  appearance **Sun-ray, Sun-burst**
  appearance.
  (noticed in 25% of jaw osteosarcoma,
  mostly on occlusal projection)

-Ill-defined border → difficult to
determine the extend of the lesion.

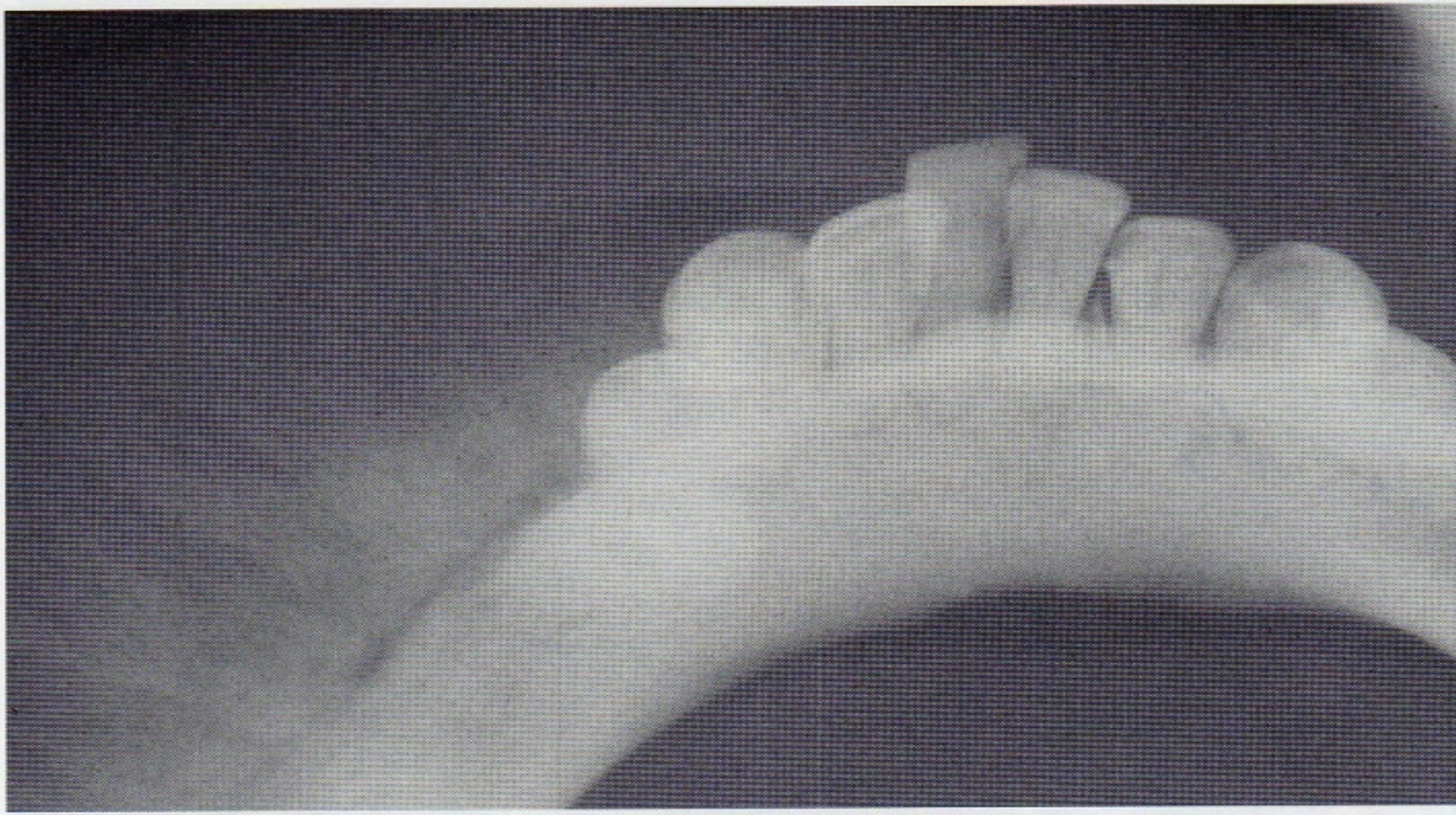


Figure 14-75 ◆ Osteosarcoma. Occlusal radiograph demonstrating prominent exophytic tumor bone production on the buccal surface of the mandible, resulting in the "sunburst" pattern. (Courtesy of Dr. Lewis Gilbert.)

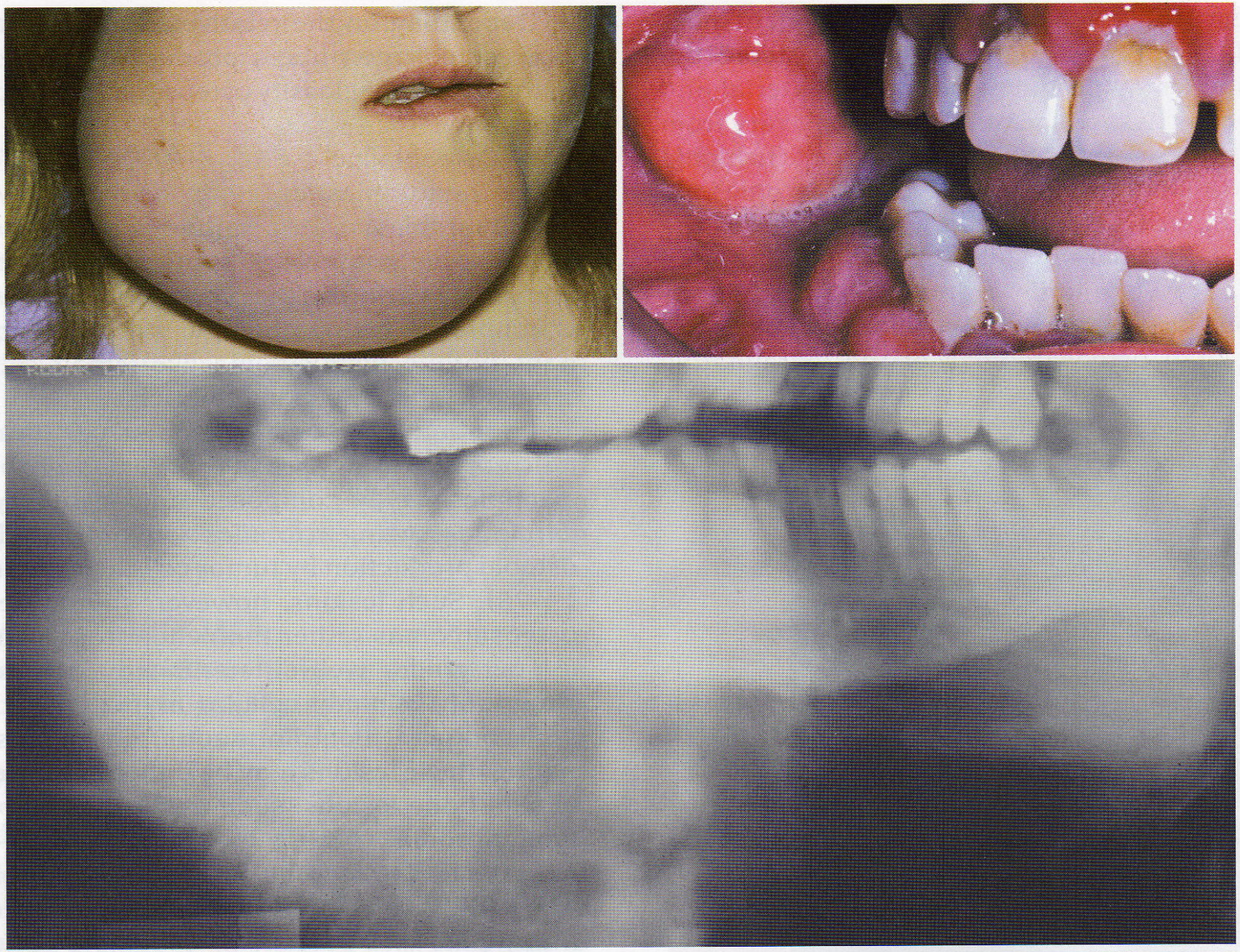
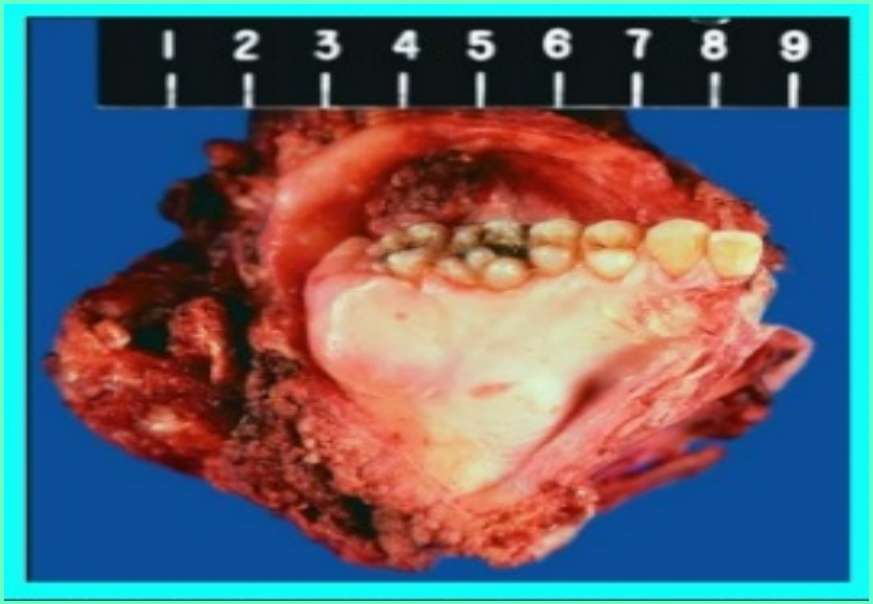
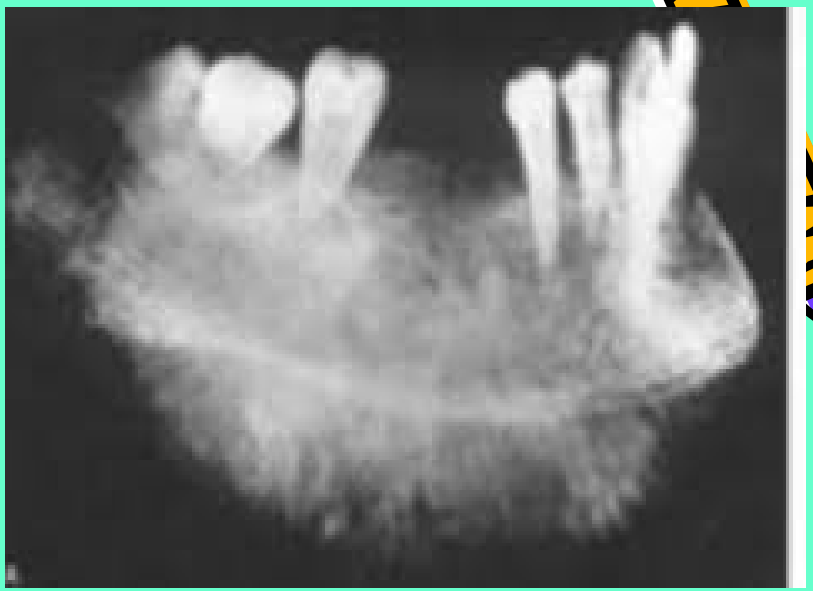
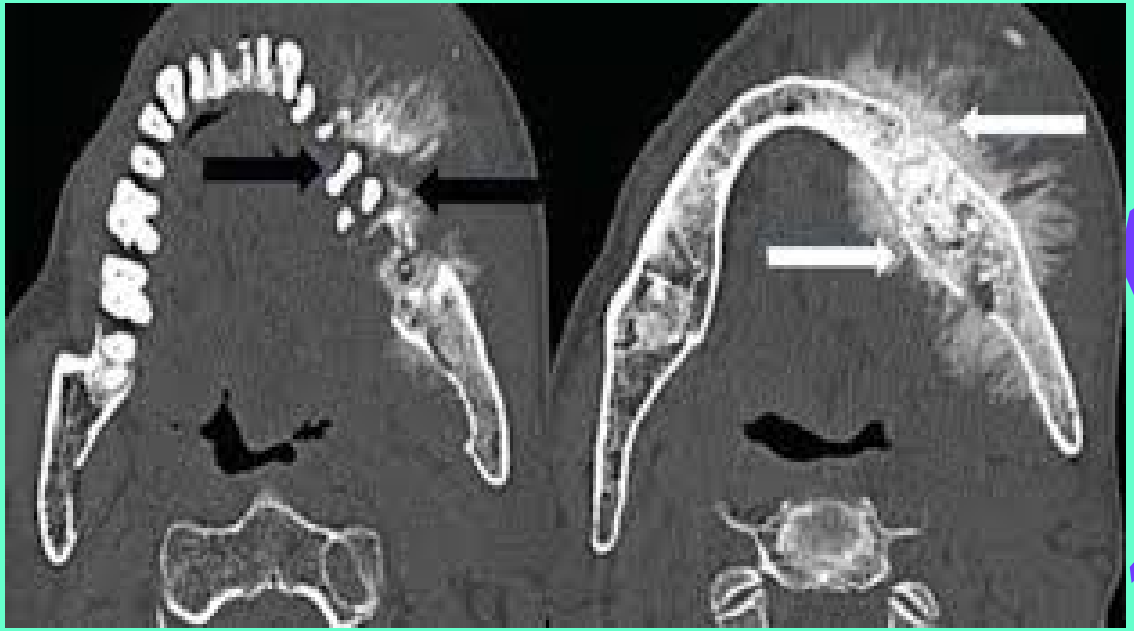


Figure 14-74 ◆ **Osteosarcoma. A,** This massive tumor had been present for many months before the patient sought treatment. **B,** Intraoral photograph of the tumor mass. **C,** The panoramic radiograph shows a "sunburst" pattern of trabeculation within the tumor.





Osteosarcoma



The tumor may spread from following pathways:

1- Spread along the marrow cavity

2- Invade the cortex

3- Elevate or invade the periosteum

4- Extend into the soft tissue

5- Metastasize through the blood stream to distant sites, particularly to the lung

Histologically

- Key feature for this tu diagnosis is the production of abnormal osteoid &/or bone by malignant osteoblast.
  { **Osteoid** → eosinophilic staining,glassy appearance, irregular (coarse) conture, surrounded by rim of osteoblast }

-Tu cell may produce F.T → fibroblastic osteos
or produce cartilage → chondroblastic osteos

Histopathology

-This may resemble fibrosarcoma & chondrosarcoma,,By histochemical staining → ALK . PH activity → +ve in osteosarcoma.

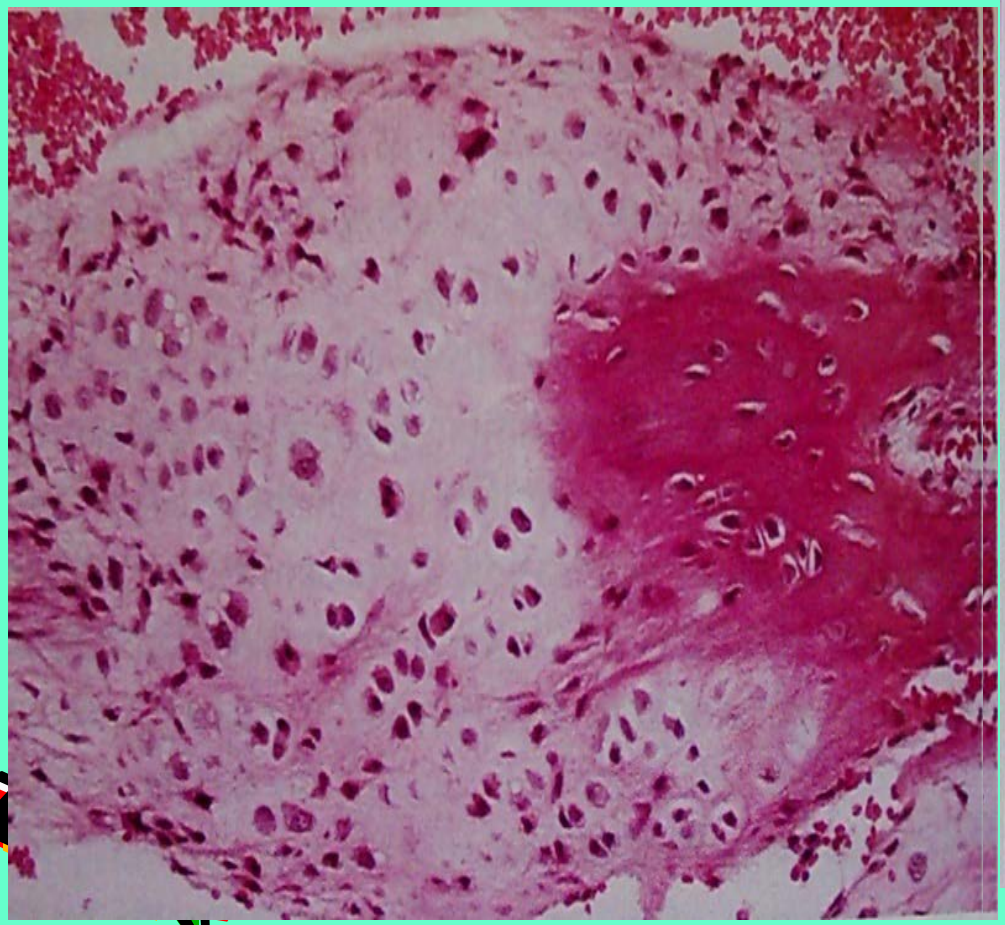
Tu cells may be round, spindle shaped or highly pleomorphic with bizarre nuclear &cytoplasmic shaped,bizarre giant cells are common,common mitosis.

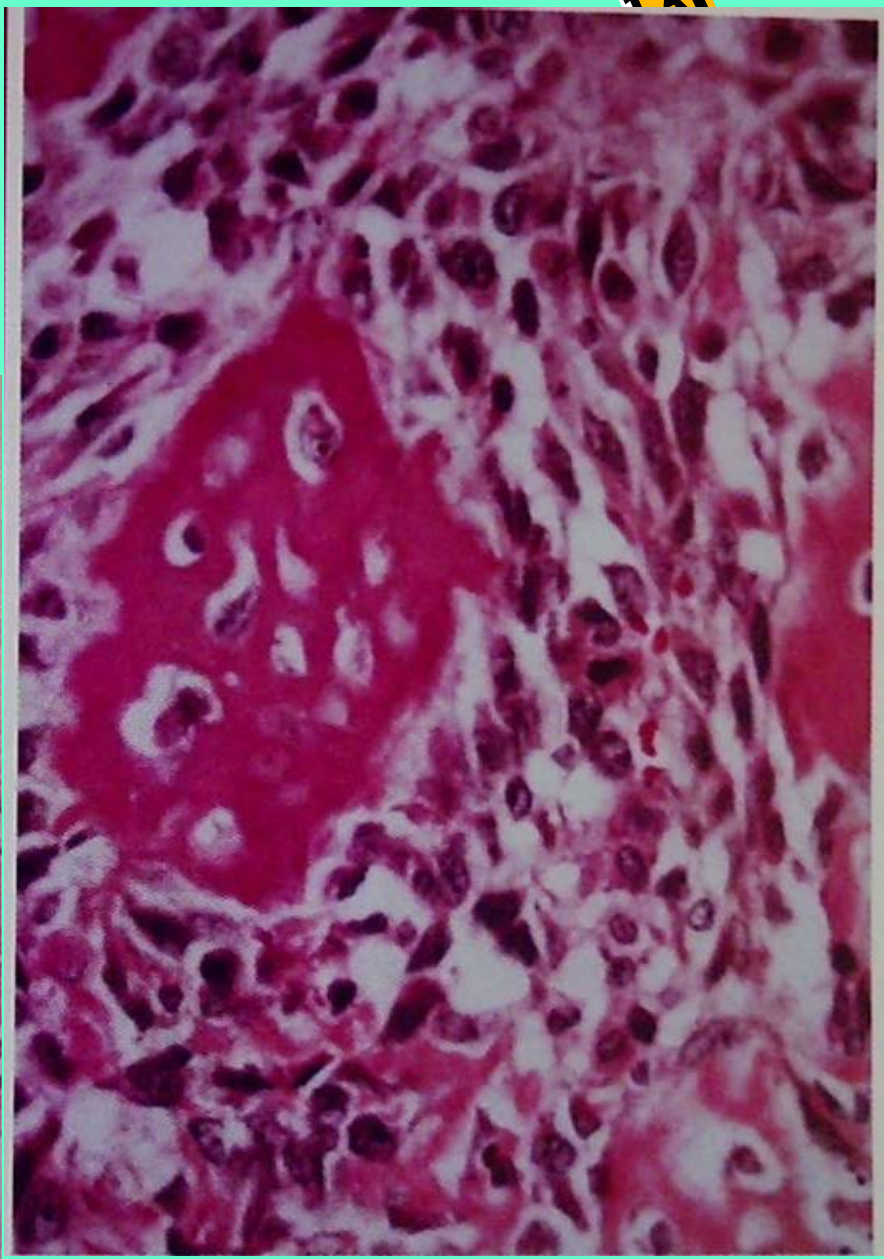
Vascular invasion by tu cells is common.

Necrosis of the tu, is common.

Histological: subtypes

1. Chondroblastic
2. Osteoblastic
3. Fibroblastic





Treatment

- Rapidly invasive tu, metastases early

  → Complete surgical removal

  [Mandiblectomy & Maxillectomy]

- This is followed by chemotherapy.

- Osteosarcoma of jaw is less aggressive than that of the long bone

  Low-grade tu, metastases less frequent

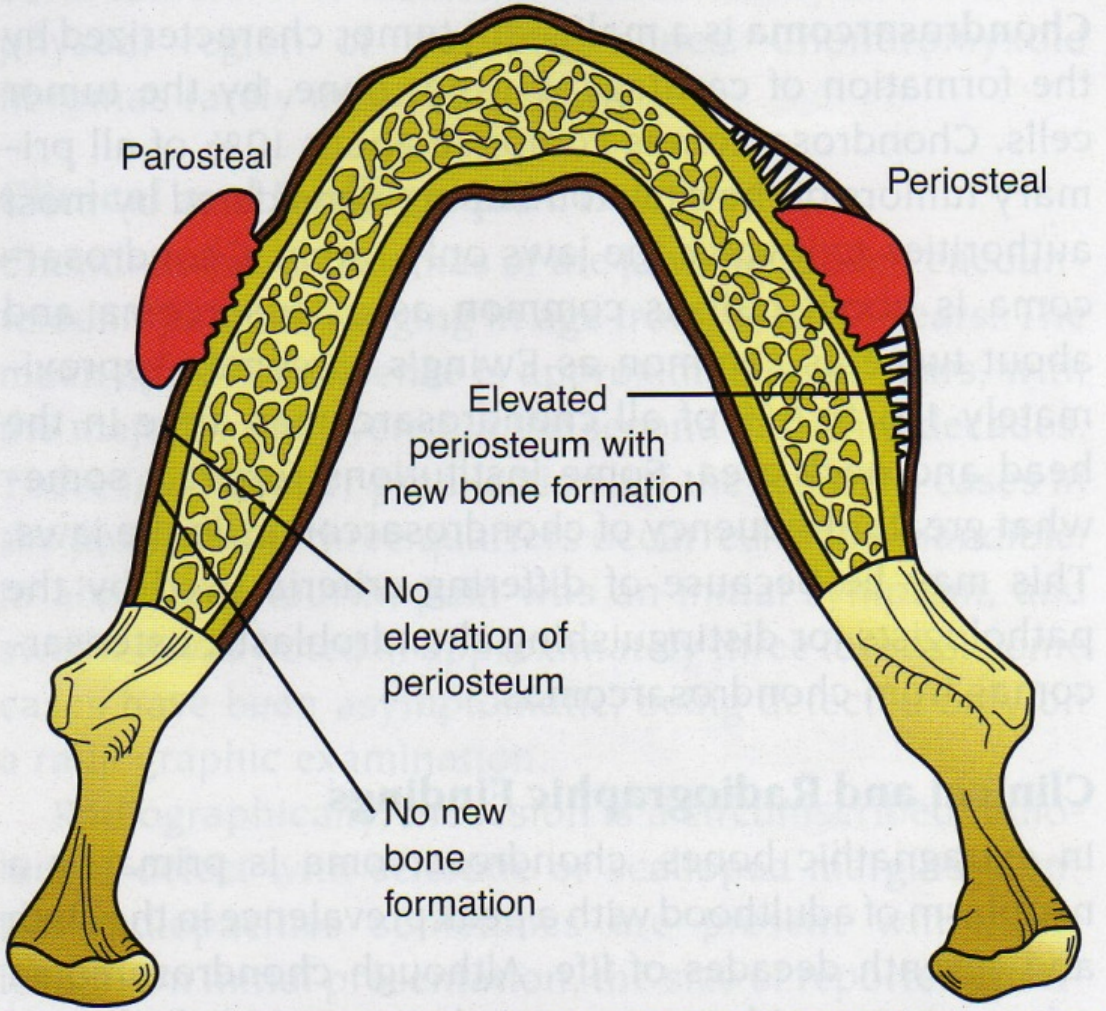


Figure 14-79 • Peripheral (juxtacortical) osteosarcoma. Illustration comparing different types of peripheral osteosarcoma. *Parosteal osteosarcoma* presents as a lobulated nodule without a peripheral periosteal reaction. *Periosteal osteosarcoma* presents as a sessile mass associated with significant periosteal new bone formation.

1. **Intramedullary (central)**
2. **Juxtacortical (peripheral)**

Tu grow outward from surface(not involve medullary bone)

Two types:-

1- Parosteal—nodule attached to cortex

2- Periosteal—lesion within cortex

Chondroma& chondrosarcoma

- Rare in jaw , ant.maxilla & posterior mandible, are the commonest sites.

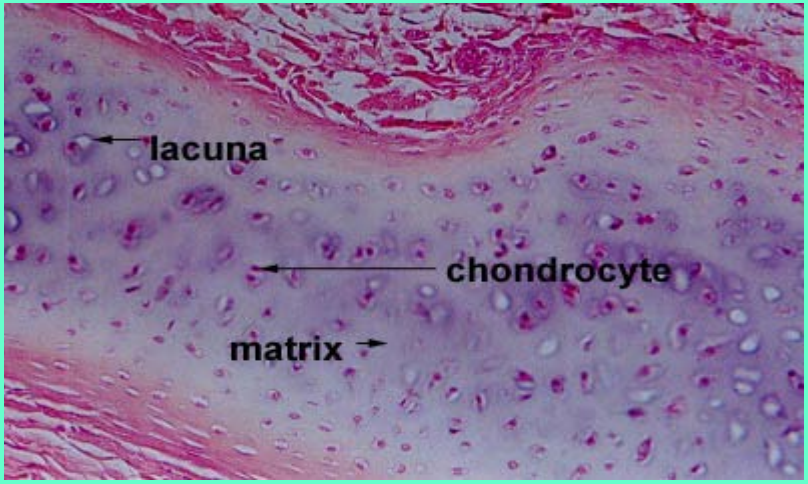
Histology

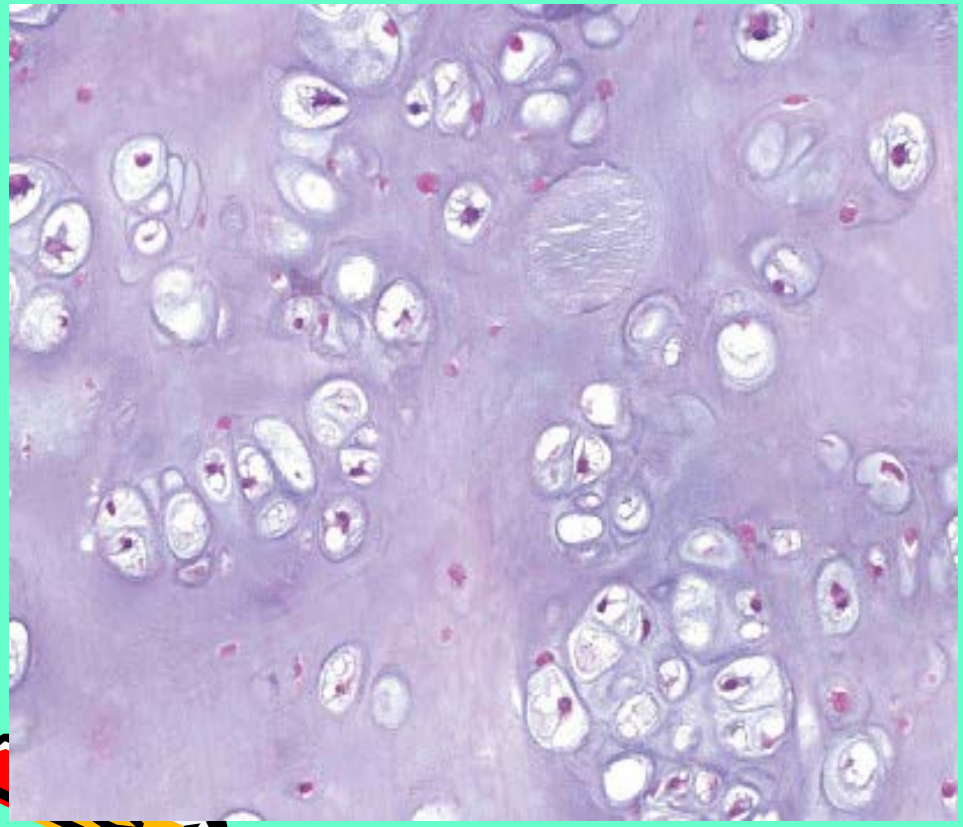
Chondroma:-Benign tu charecterized by circumscribed mass of mature hyaline cartilage with benign chondrocyte.

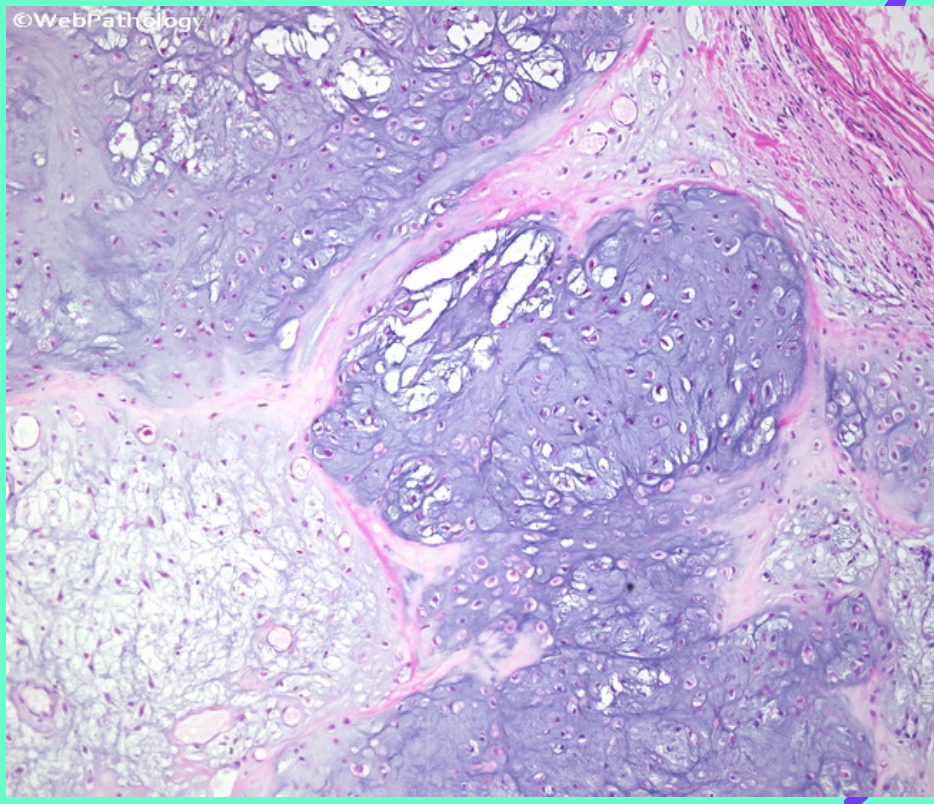
[Well-formed lacunae containing small chondrocyte with pale cytoplasm &small rounded nuclei]

X-ray → R.L with central area of R.O

Normal cartilage







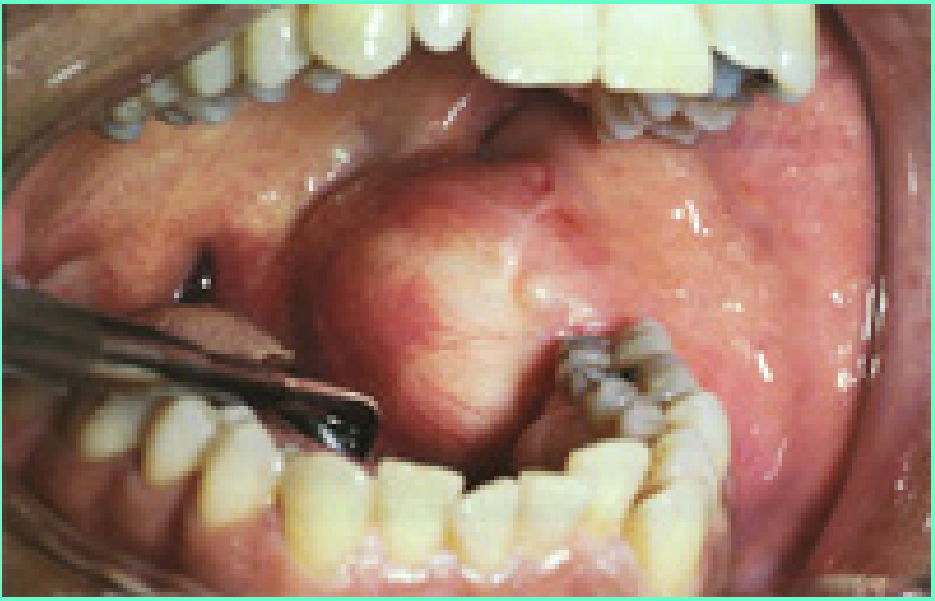
Chondroma

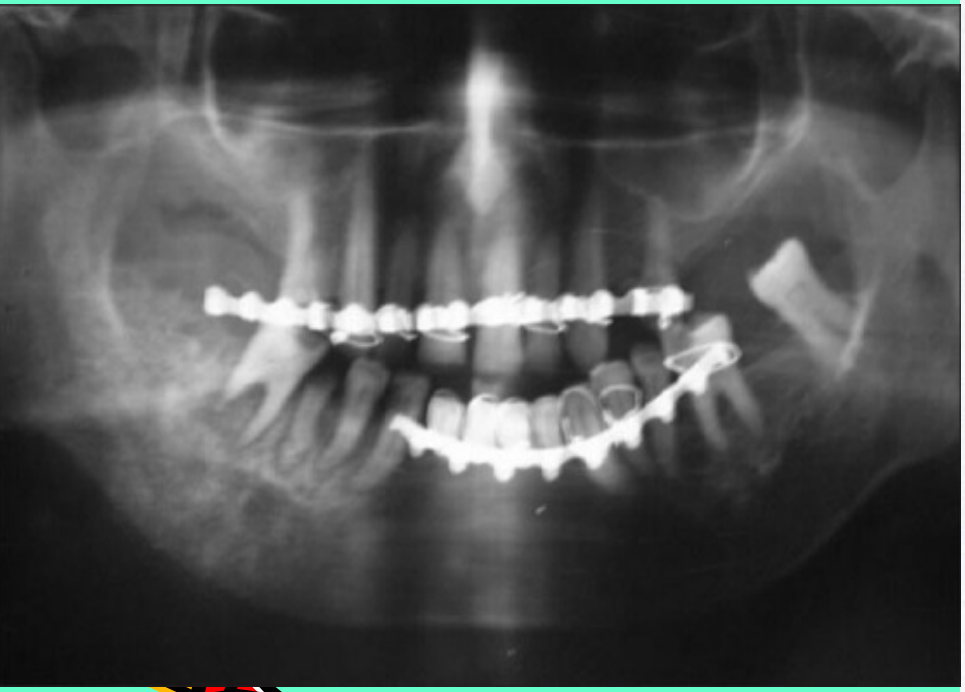
Chondrosarcoma

-Malignant tu producing neoplastic cartilage, showing varying degree of maturation & cellularity.

- Chondrocyte are pleomorphic, binucleated & show mitotic activity.

X-ray → R.L process with ill-defined border. R,L area show variable amount of R.O mass caused by calcification of cartilage matrix.





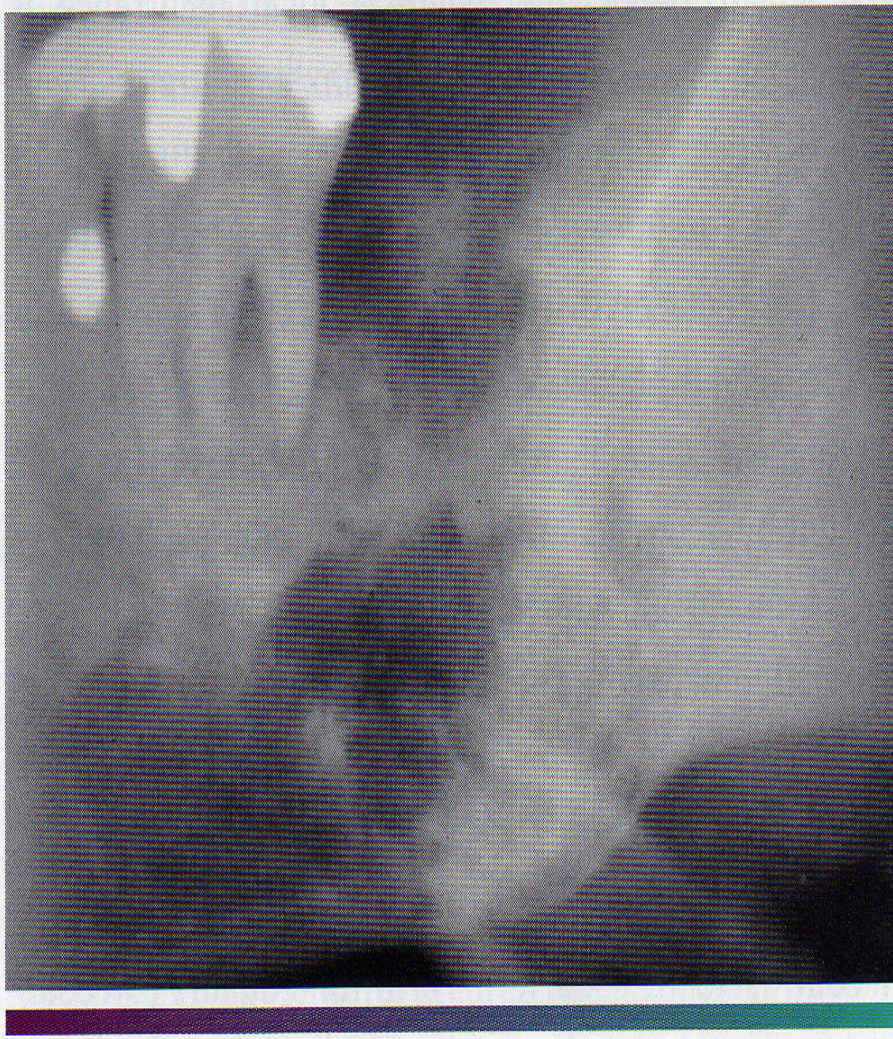


Figure 14-80 ♦ Chondrosarcoma. Ill-defined radiolucent lesion of posterior mandible containing radiopaque foci. (Courtesy of Dr. Ben B. Henry.)

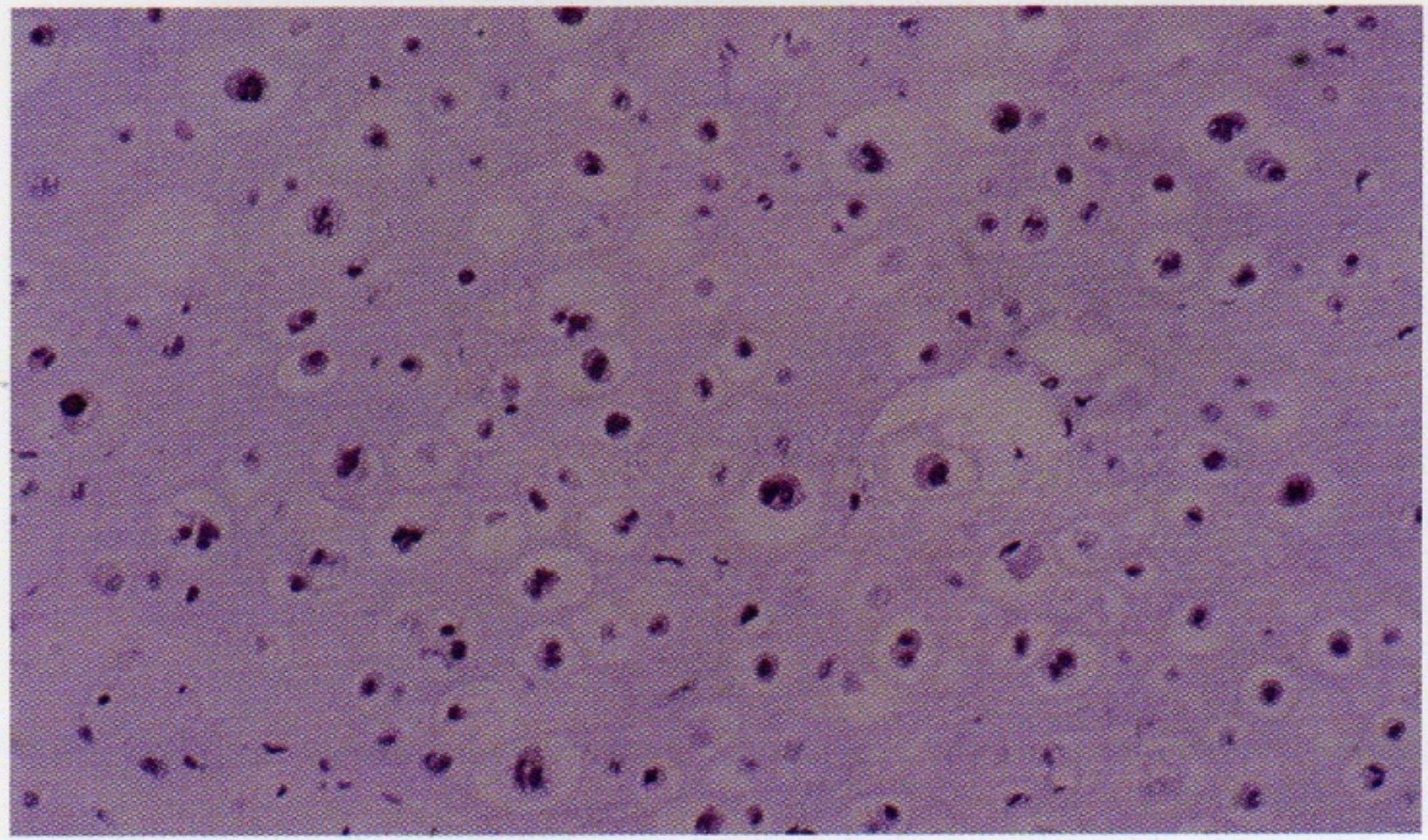
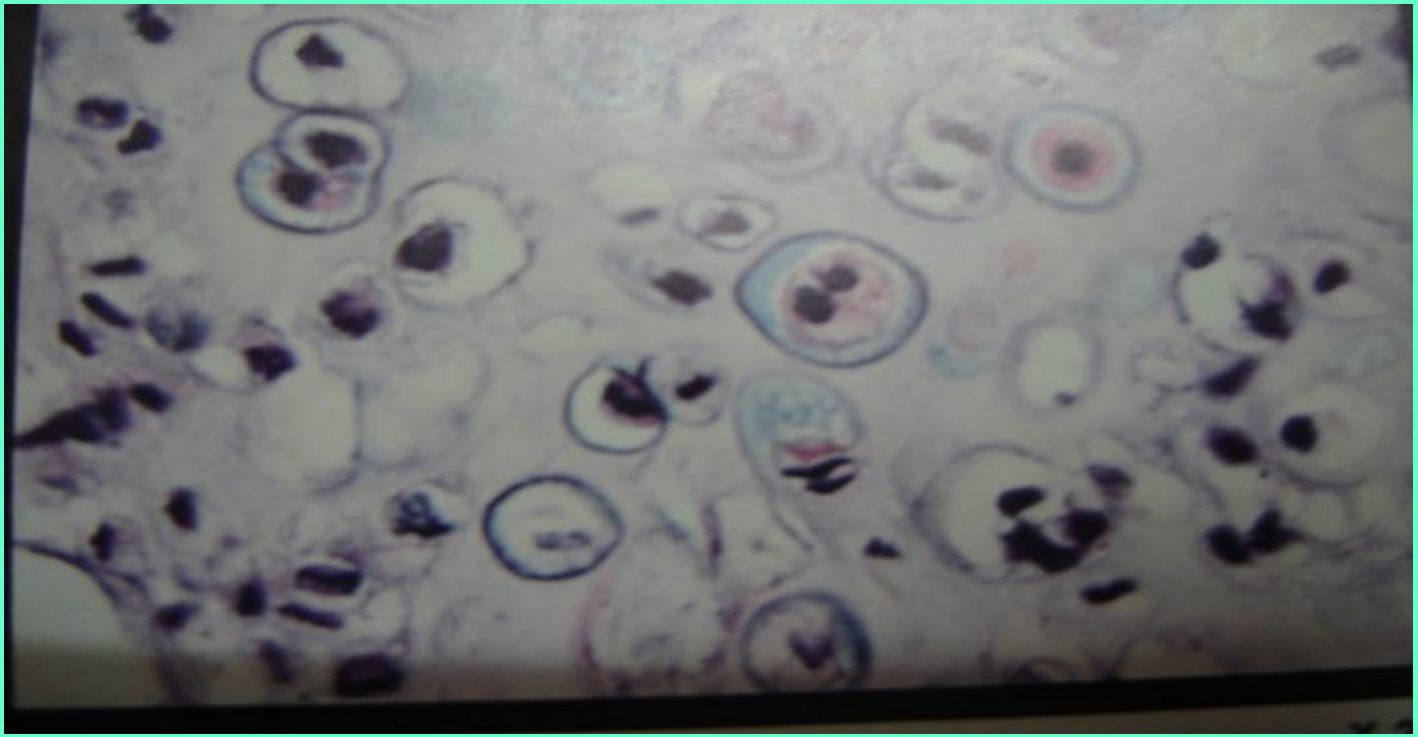
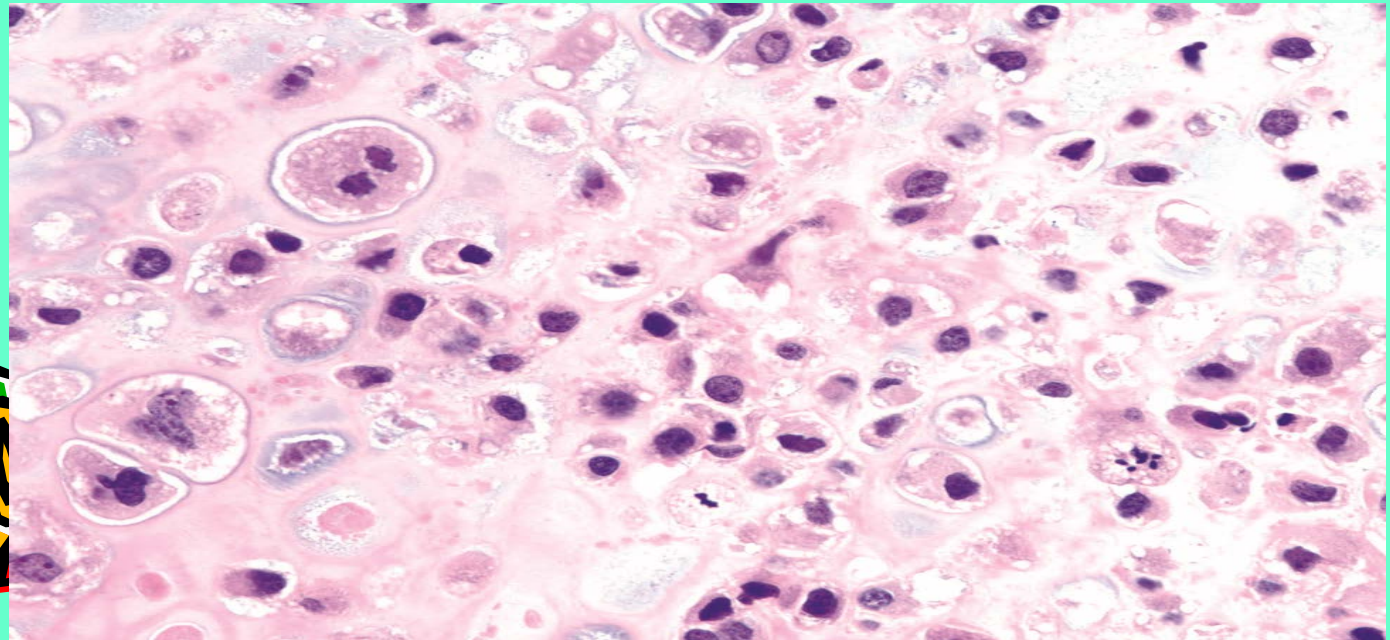


Figure 14-81 • Chondrosarcoma. This grade II chondrosarcoma shows a variation in size of chondrocyte nuclei. Occasional double nuclei are seen in the lacunae.





Treatment

- It is wise to cinsider chondroma of Jaw to represent a potential chondrosarcoma.

Treatment:

Depend on size, location & grade

wide excision with tumor free margins

chemotherapy and radiation are less effective comparing with osteosarcoma

- The prognosis of chondrosarcoma in mandible is better than maxilla.

  (problem of acheving clearance by radical surgery)

Recurrence is common in both Benign & Malignant tu.

Prognosis:

-- not good, five year survival rate is less than 20%

-- high rate of recurrence

-- 10% metastasis usually to lungs or other bones

Ewing's sarcoma

- Adistinctive primary malignant tu of bone composed of small undifferentiated round cells of uncertain histogenesis

  It is linked with primitive neuroectodermal tu "PNET" ---both exhibiting features of a neuroectodermal origin.

(same tu differ only in degree of differentiation)

-Both tu demonstrate a reciprocal translucation bet chromosome **11 & 12**

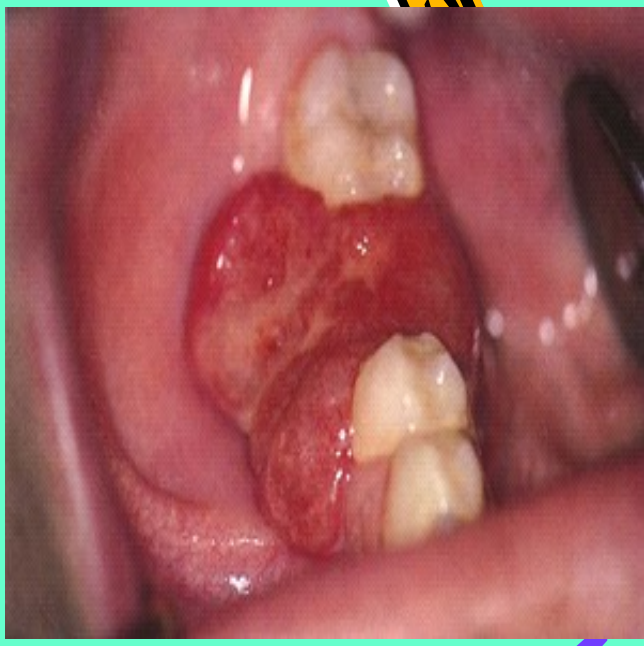
Clinically

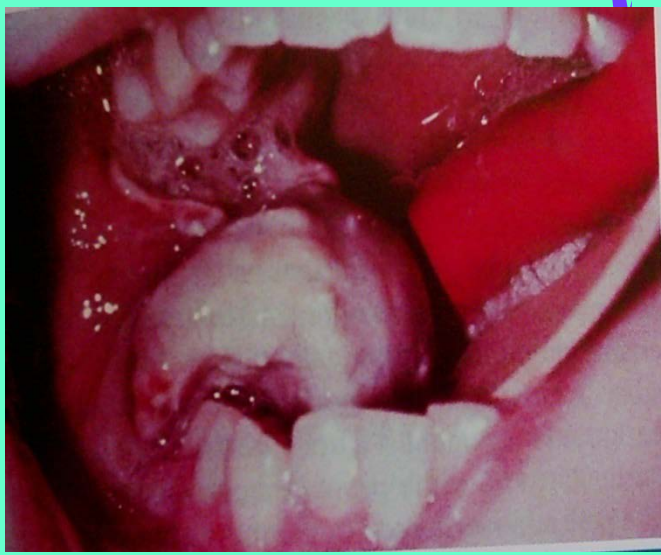
-Children & young adults.

-Tu presents with painful bone swelling,loosening of teeth with mucosal ulceration,systemic sign & symptoms

---fever,leucocytosis,raised ESR,anemia---

-Rare in jaw, **1-2 %** occurs in the gnathic or craniofacial bones.





Histopathology

-Solid sheets of cells divided by fibrous strands into irregular lobules or masses.

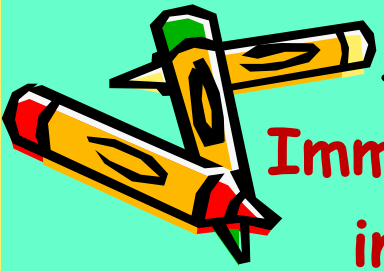
-Cells are uniform & small (like lymphocytes but larger twice in size) & of neuroectodermal origin. Its outline is ill-defind , dark-staind, round nuclei, small nucleoli, with variable mitotic activity.

-Hemorrhage & necrosis.

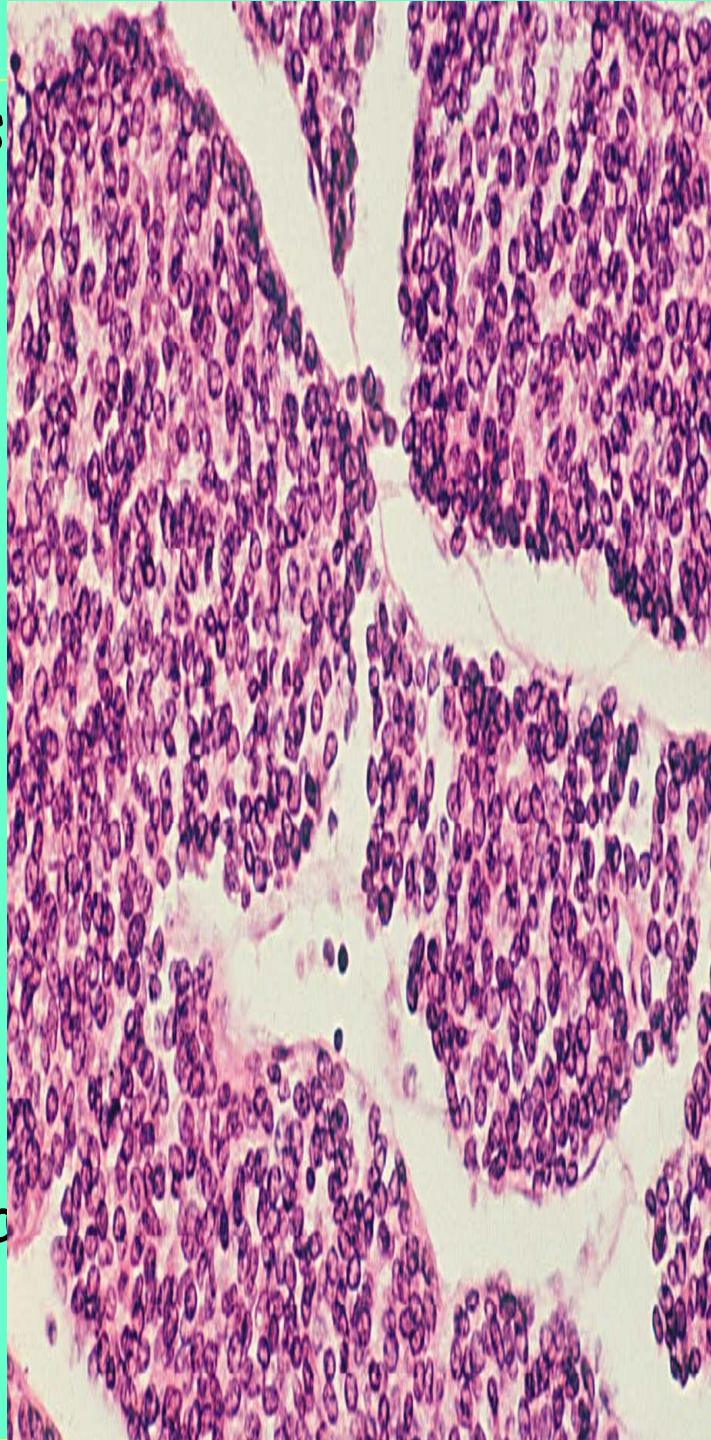
-Cytoplasm with glycogen granules

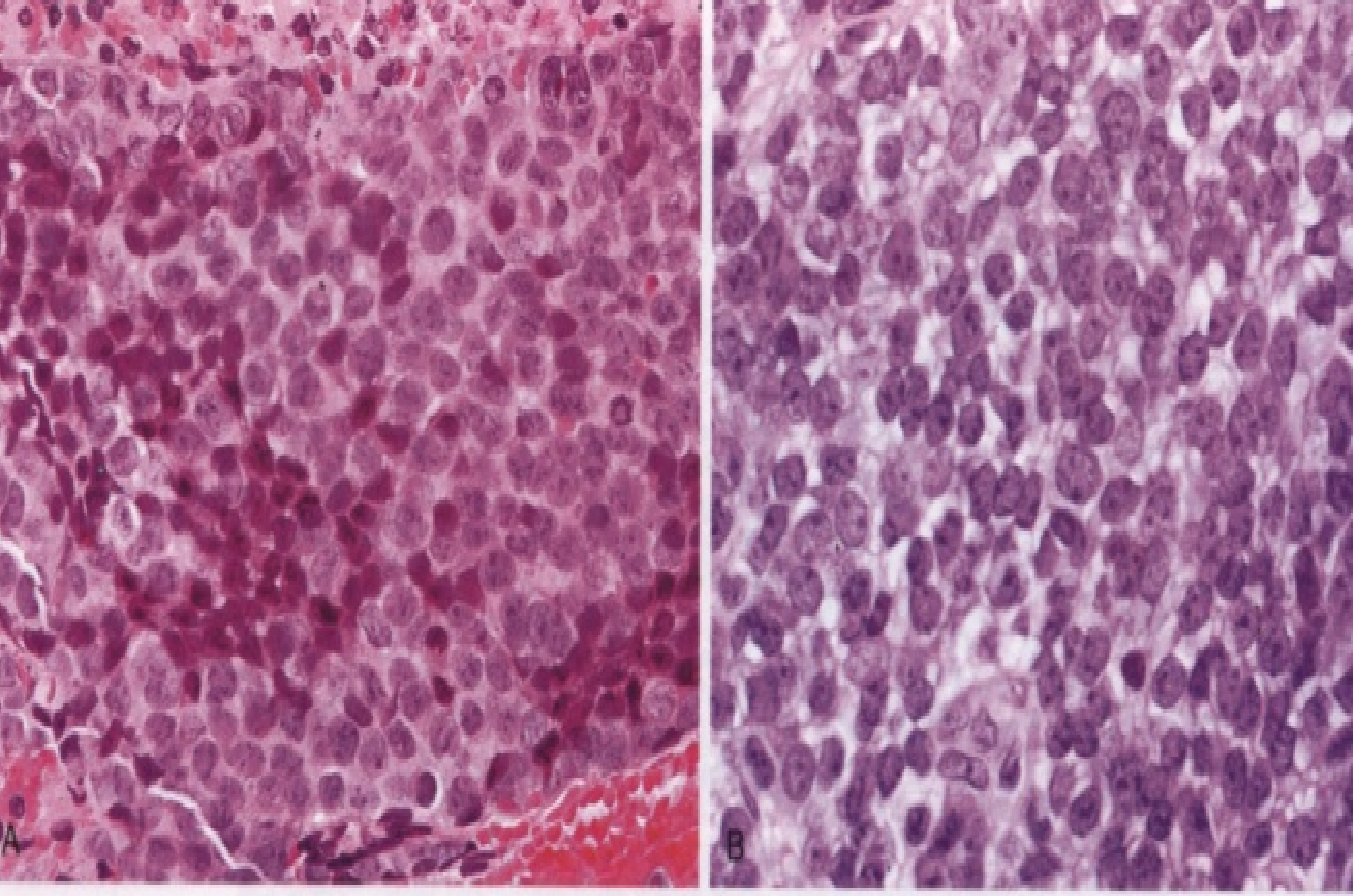
Help in diagnosis

→ & differentiate it from other small roundcell tu,,lymphoma



Immunohistochemistry useful in dx



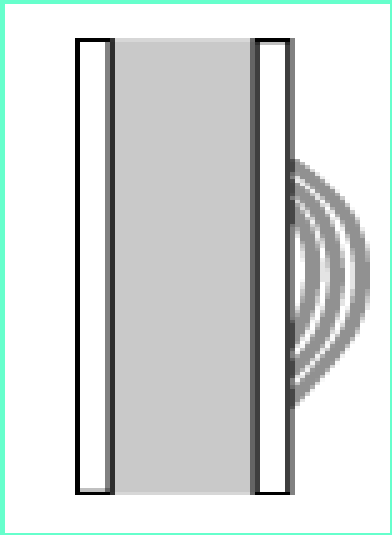


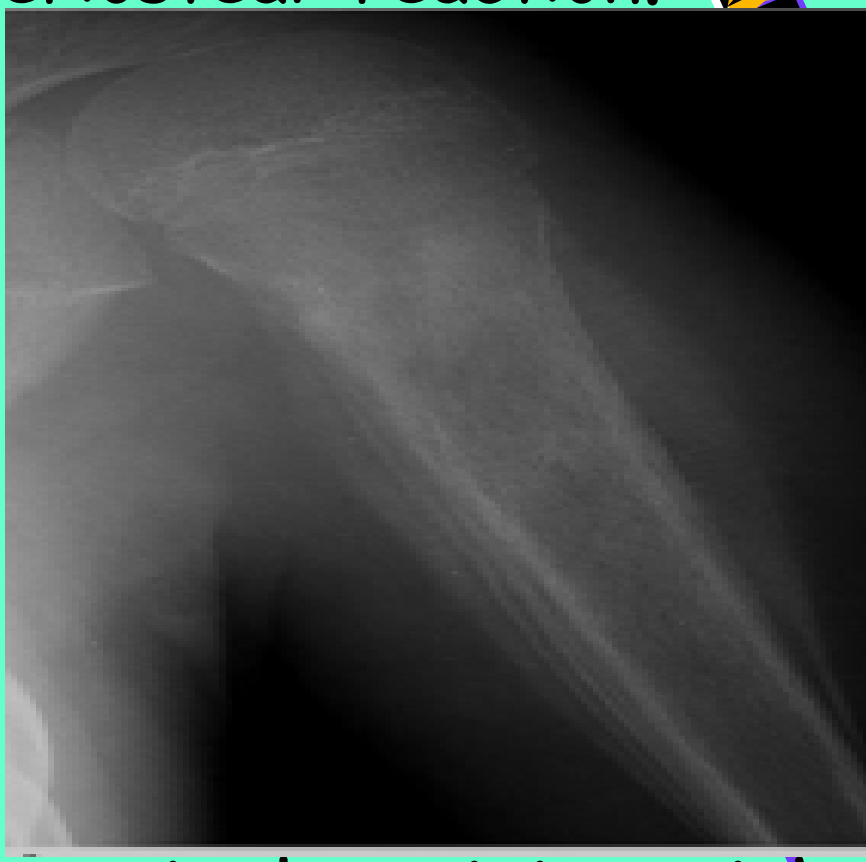
gure 8–15. *A*, Ewing's sarcoma. Uniform small cells with little to no visible cytoplasm. Nuclei are round with uniformly distributed, powdery chroma ed smaller dark cells with hyperchromatic dense nuclei are also present. *B*, Ewing's sarcoma. Cells have somewhat coarser chromatin than the cell part *A* and small nucleoli are present. Some cells have a vacuolated, clear cytoplasm due to the presence of glycogen.

X-ray

Irregular RL bone destruction with ill-defined border

Charecteristic" **onion-skin**" periosteal reaction.





Treatment

Chemotherapy &surgical excision with or without radiation.

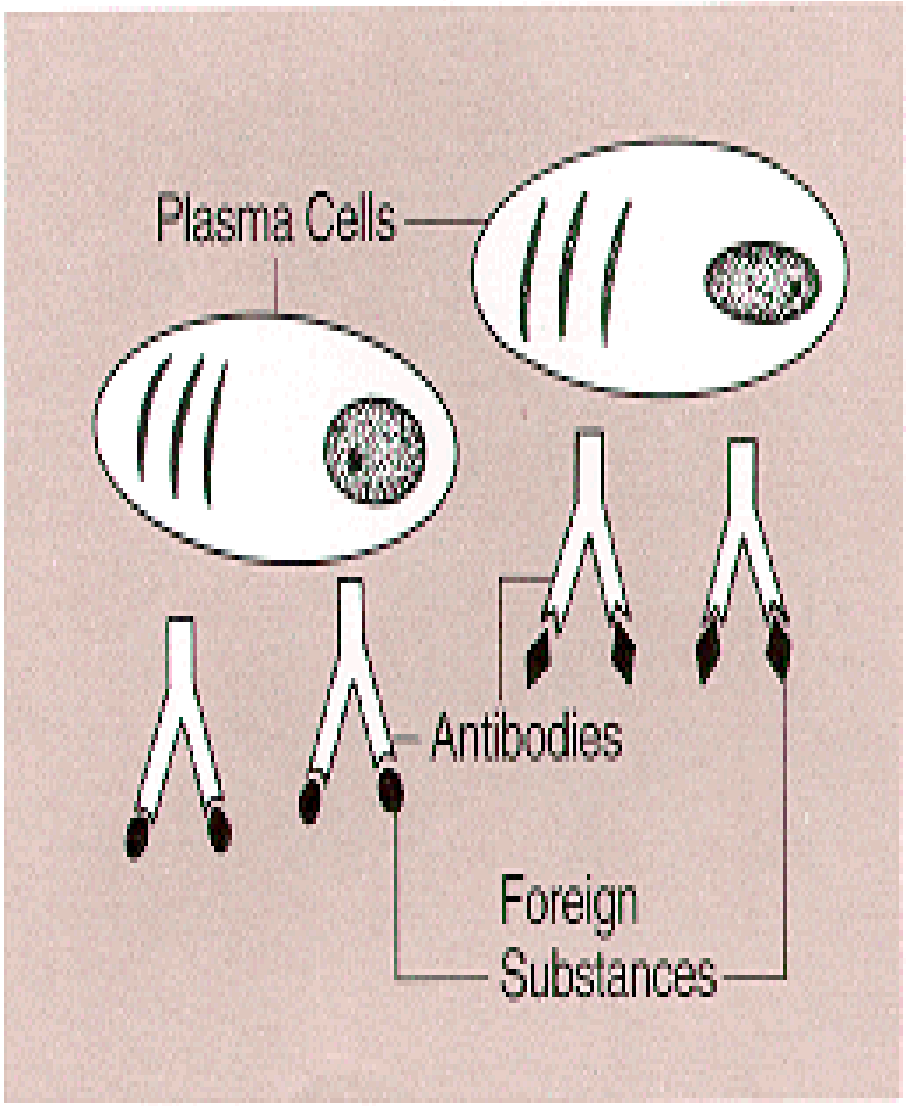
Multiple myeloma

Neoplasm of differentiated B-lymphocyte

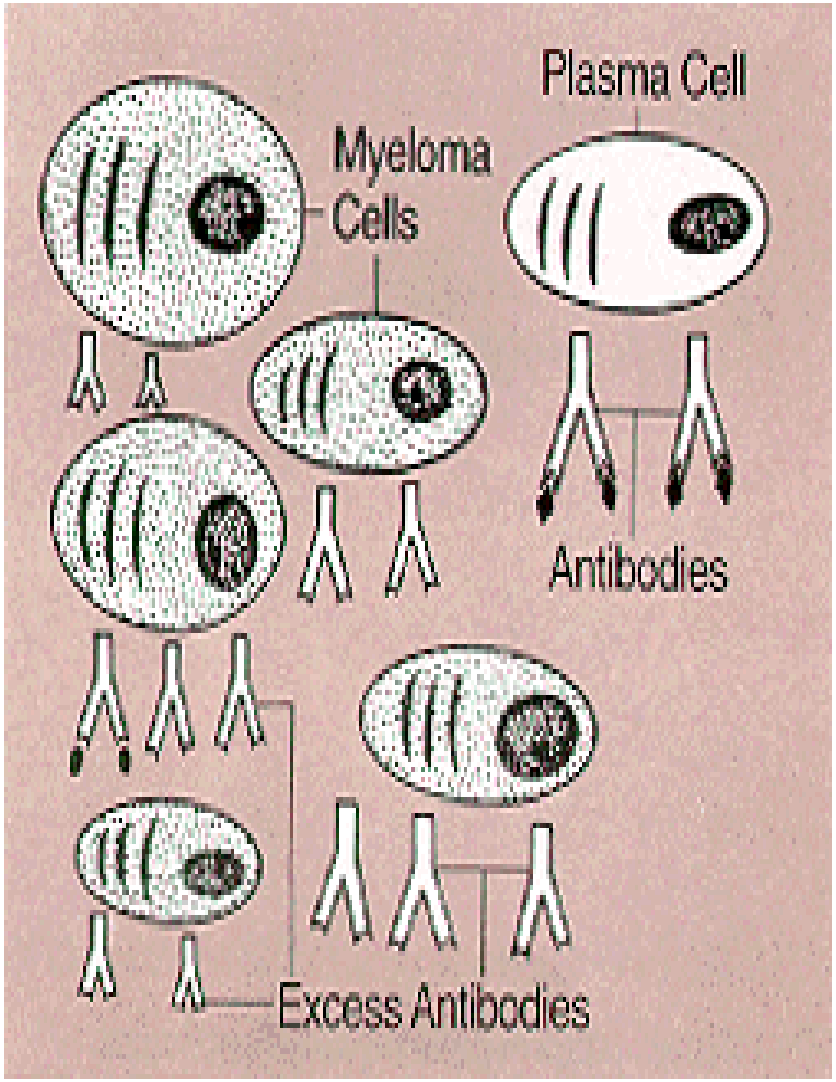
-M.M uncommon malignancy of plasma cell origin ,often appear to have a multicentric origin within bone(multiple bone involvement)

-Account for nearly 50% of all bone tu.

-Malignant plasma cell compose this tu are monoclonal, arise from a single malignant precursors undergone uncontrolled mitotic division & spread through out the body

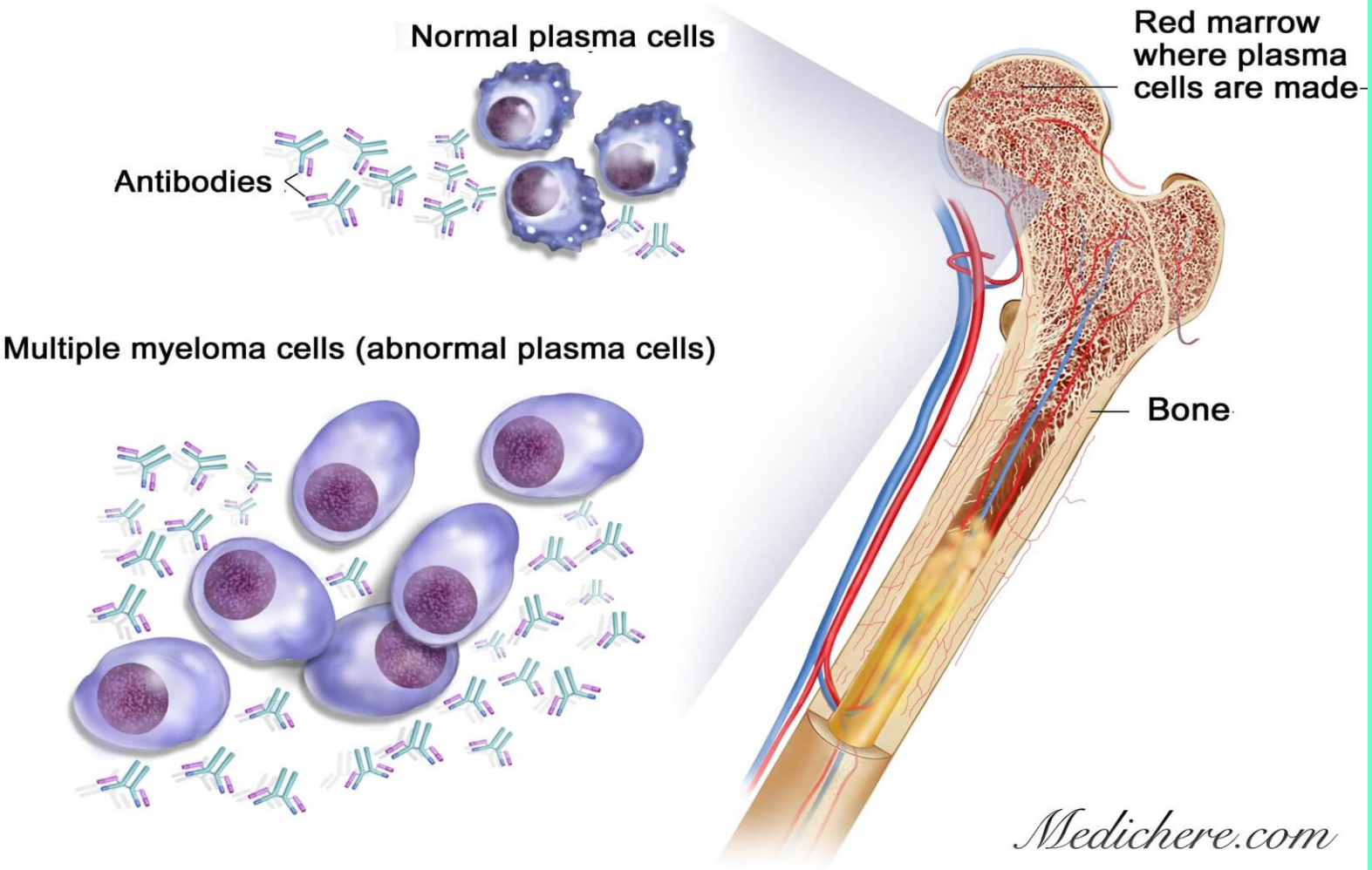


Plasma cells produce proteins called antibodies. Antibodies attach to foreign substances to fight infection and disease.



In multiple myeloma, the body makes too many plasma cells (myeloma cells). These cells produce antibodies that the body does not need.

Multiple Myeloma



-M.M develops from a single cell →

all daughter cells have the same genetic makeup & produce the same protein(immunoglobulin) that the plasma cell normaly produced (**mainly IgG**).

-The effects of tu result due to abnormal cell proliferation &the uncontrolled production of their protein.

Clinically

-It is a disease of adult, median age 60-70 yrs.

-Bone pain is the most characteristic symptoms.

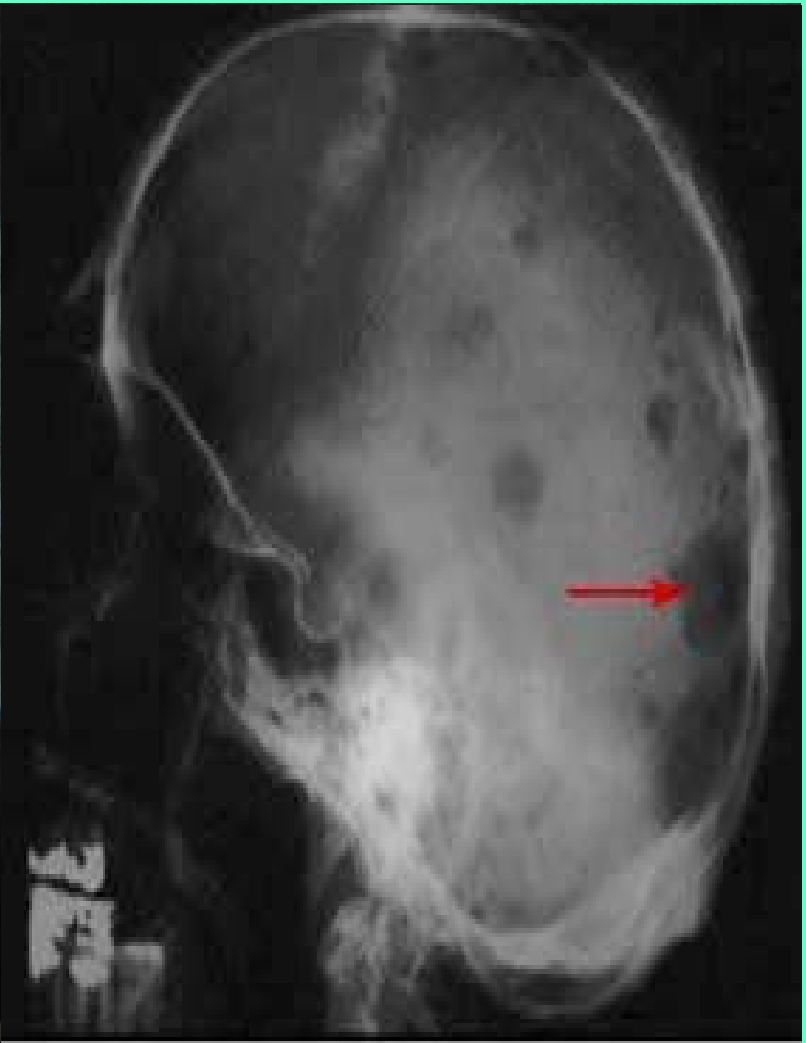
-Patho. fracture,fatique,peticheal hemoorhage of skin,fever, ↑susceptibilityto infection.

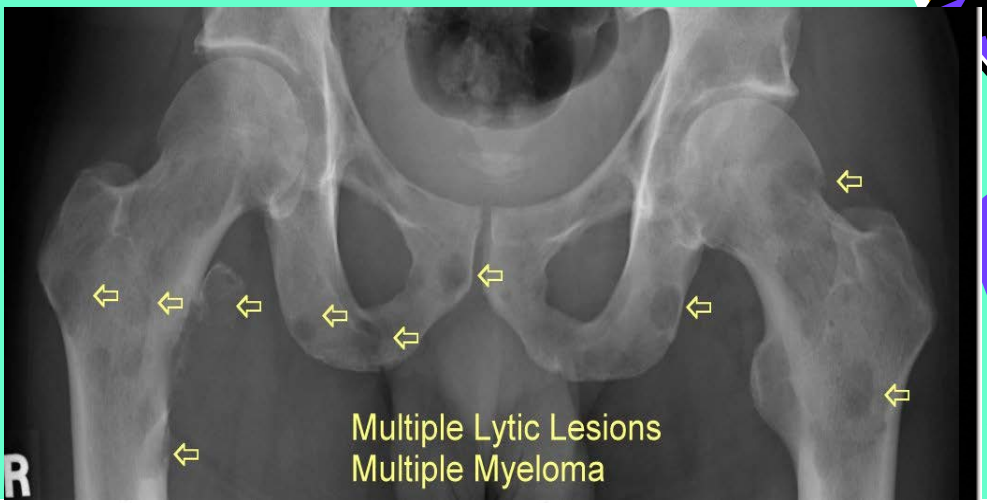
-Jaw lesions associated with pain,numbness, lossening of teeth,patho.fractures &post operative extraction hemorrhage.

-Mandible more affected,in posterior region.

Radiographically

Multiple well-defined punched- out areas of R.L espicially at vault of skull





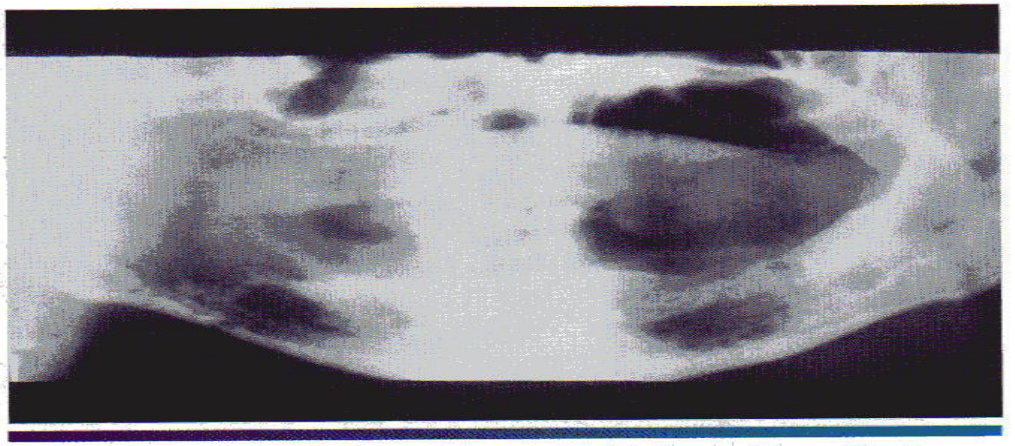


Figure 13-36 ◆ **Multiple myeloma.** Multiple myeloma affecting the mandible. The disease produced several radiolucencies with ragged, ill-defined margins. (Courtesy of Dr. Joseph Finelli.)

Microscopically

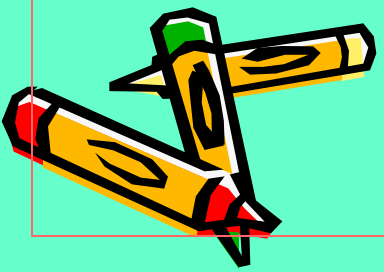
-Dense cellular tu, ↑ no. of myeloma cells (plasma cells).

(Cells with prominent nucleoli,cytoplasm with inclusion containing Ig)

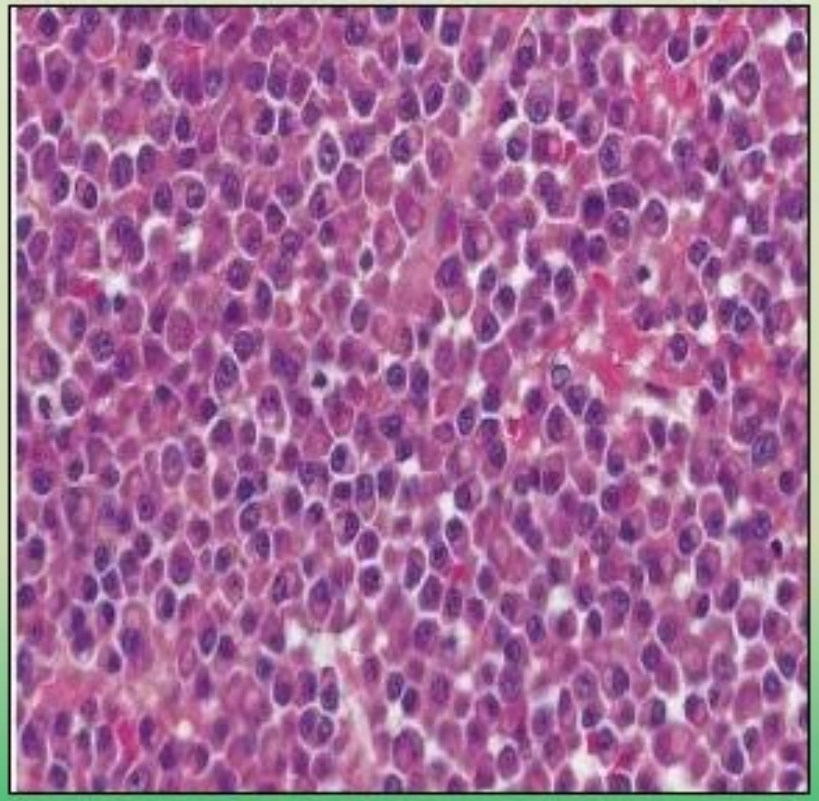
(cart-whell appearance)

- Frequent mitosis

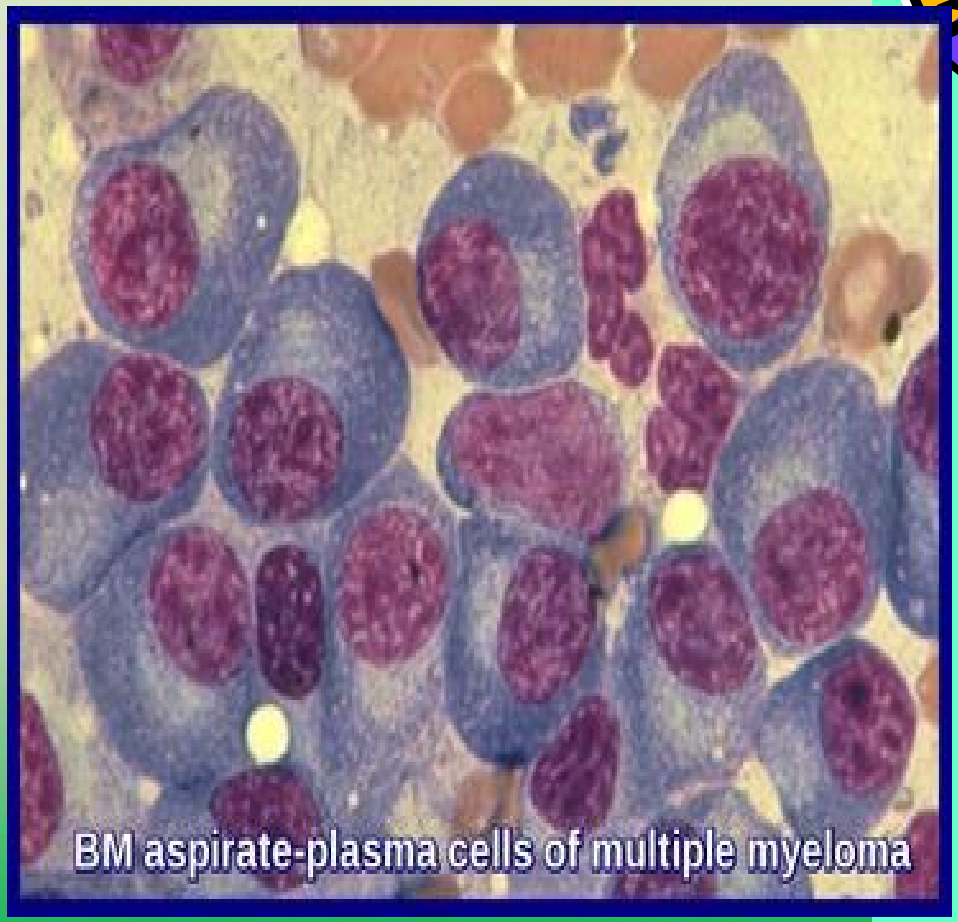
-Little or no supporting stroma.



MICROSCOPY



WELL DIFFERNTIATED MYELOMA



- **Diagnosis ofM.M depends on:**

-Bone marrow biopsy → atypical plasma cells

-X-ray → R.L areas.

-Serum protein electrophoresis → massive production ofabnormal **IgG** by neoplastic clone of cell.

- Bence-Jones protein find in **30-50%** of pt.

Treatment:

Chemotherapy → poor prognosis

5 yrs survival rate **is** **25%** only

Plasmacytoma is a solitary form of myeloma

Langerhans cells histiocytosis

- Old name is **Histiocytosis X**
- A spectrum of disease(similar in histology but differ in behaviour & severity) charecterized by proliferation of histiocyte-like cell, accompanied by eosinophel,lymphocytes, plasma cells & multinucleated giant cells.
- The neoplastic cells are **Langerhans cells** which are dendritic mononuclear cells, normaly found in epidermis, mucosa ,that derived from bone marrow.

Langerhans cells are bone marrow-derived antigen processing cells and represent the most peripheral extension of the immune system. The pathologic proliferation of these cells is referred to as Langerhans cell histiocytosis.

Clinically : Present in one of 3 main ways:

1-Chronic localized histiocytosis:

Monostotic or polyostotic eosinophilic granuloma of bone without visceral involvement. Represents the most common form of this disease and presents typically as a localized, unifocal osteolytic lesion.

2-Chronic dissemenated histiocytosis:

Involving bone, skin,viscera. Pt presents with classical trait of **Hand-Schuller-Christian** syndrome.

(skull defect(osteolytic lesions),exophthalmus, diabetes insipidus)

3-Acute disseminated histiocytosis:

Involving cutaneous, visceral, bone marrow involvement.

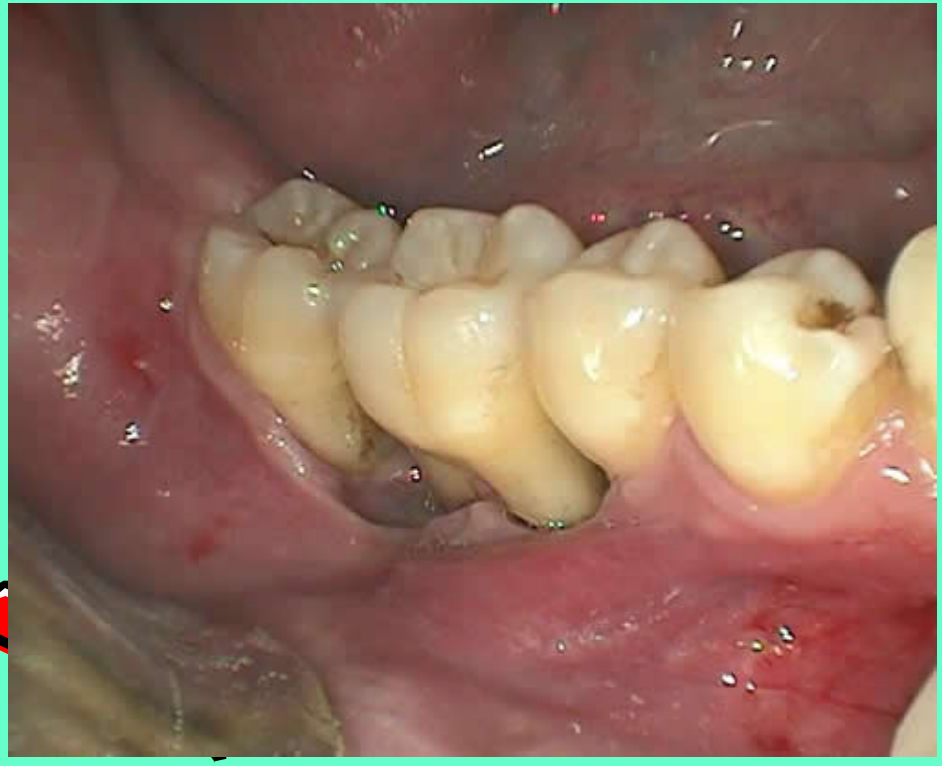
Mainly in infant & young children under age 2 yrs.

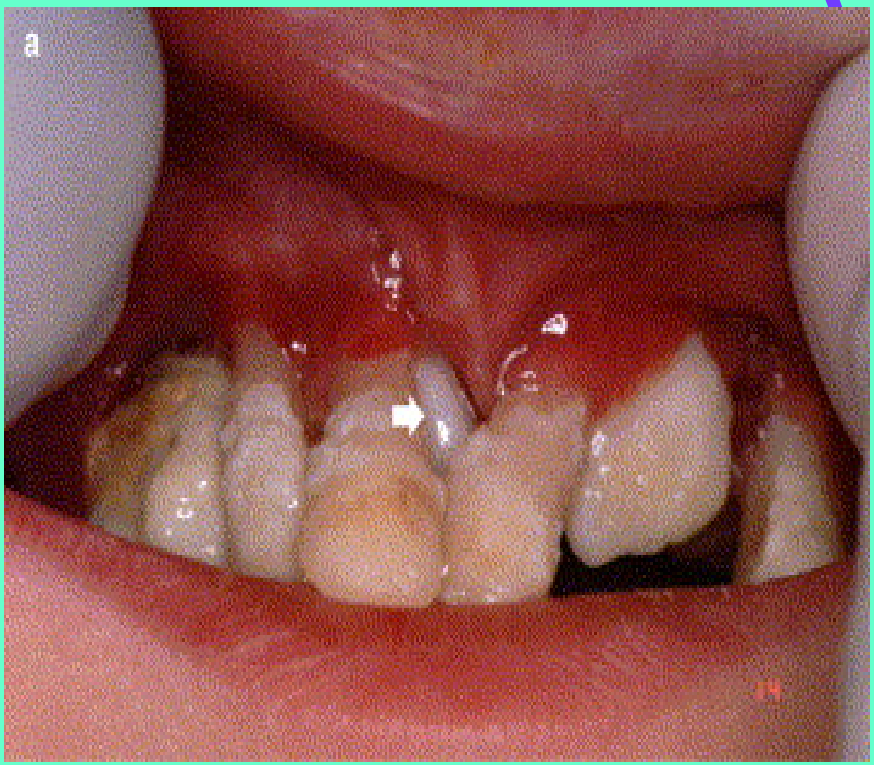
It runs a rapid and fatal course. High mortality rate.

Letterer-Siwe disease

The most common presenting symptom is local bone pain. Other features encountered in the head and neck area include a painful scalp mass, scalp rash, cervical lymphadenopathy, otitis media, and orbital proptosis.

Oral and perioral involvement includes mucosal ulcerations, loosening of teeth, enlarged salivary glands, gingival edema, gingival ulceration and aggressive periodontitis-like lesions.



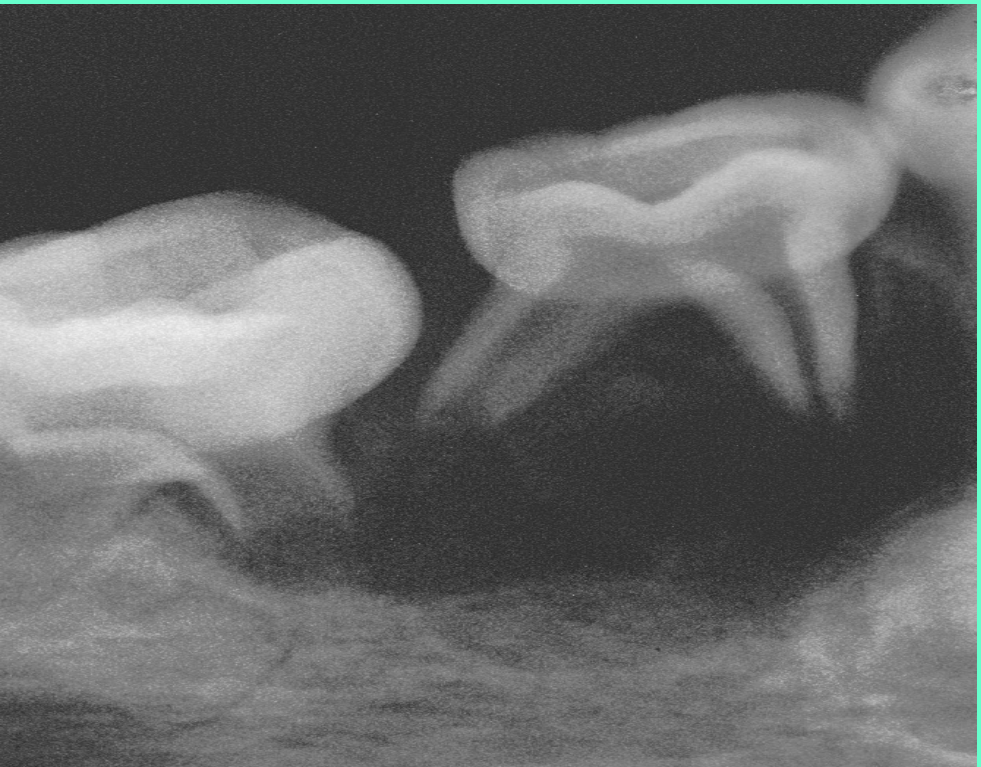


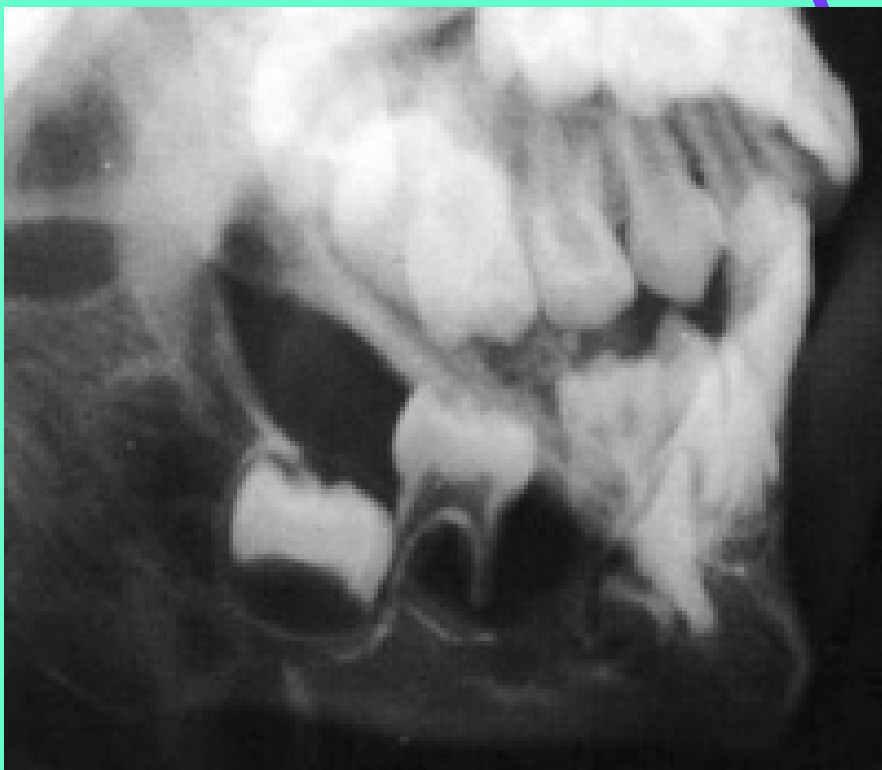
X-ray

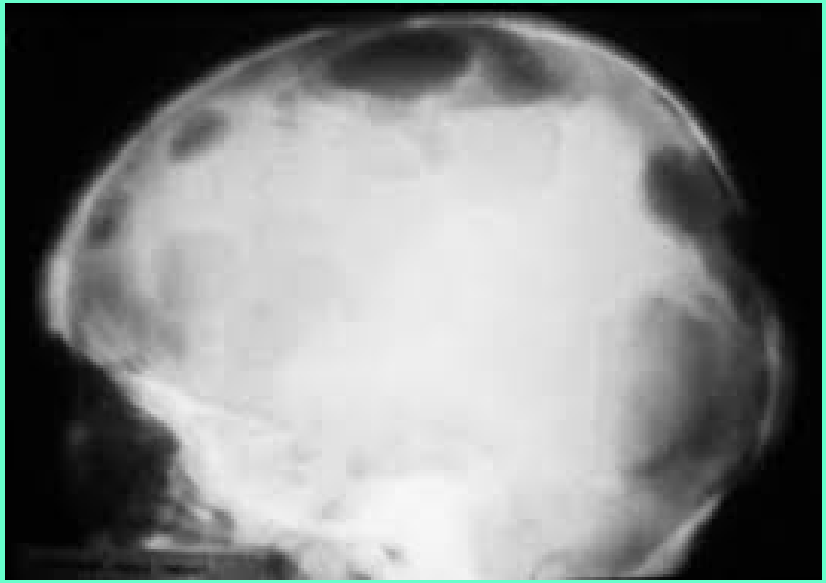
Posterior mandible, alveolar bone destruction,

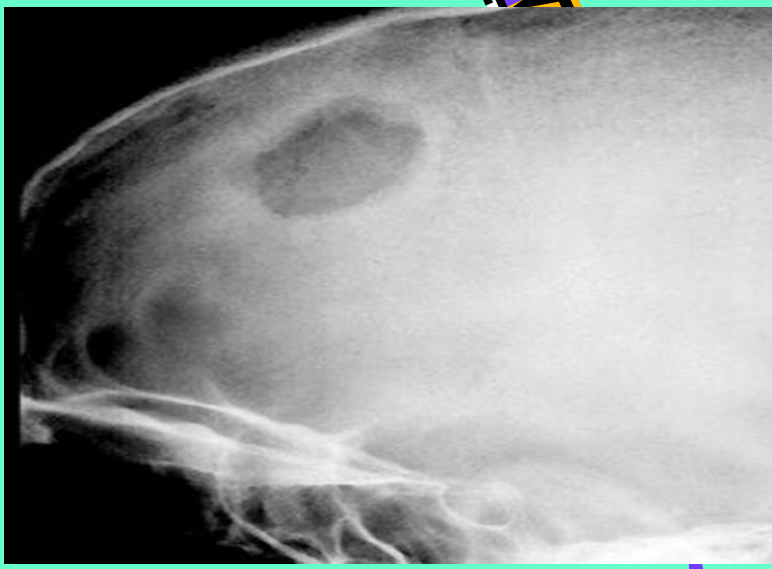
RL areas **Scooped appearance**

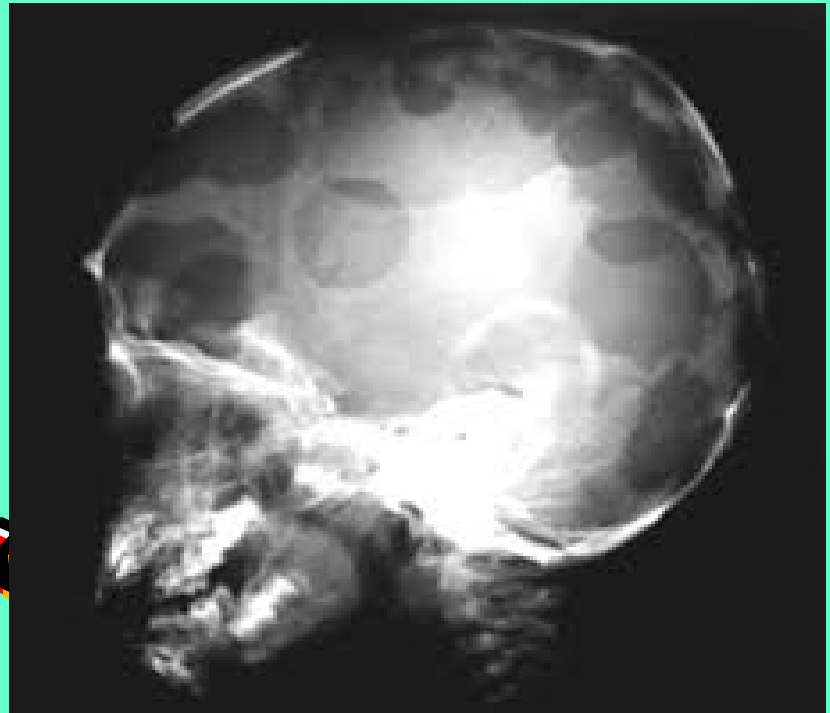
Extensive bone destruction & loosening of teeth "**Floating in air appearance**"

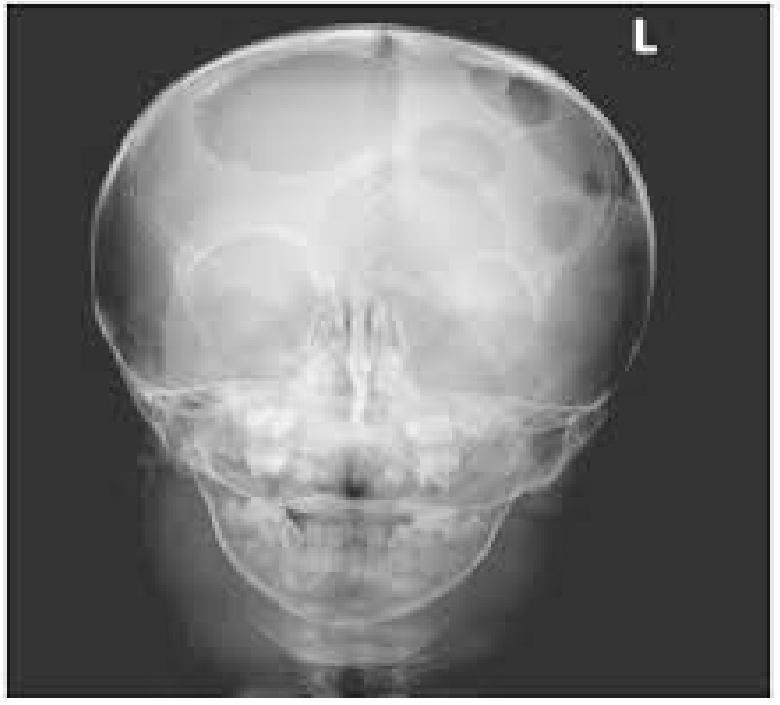








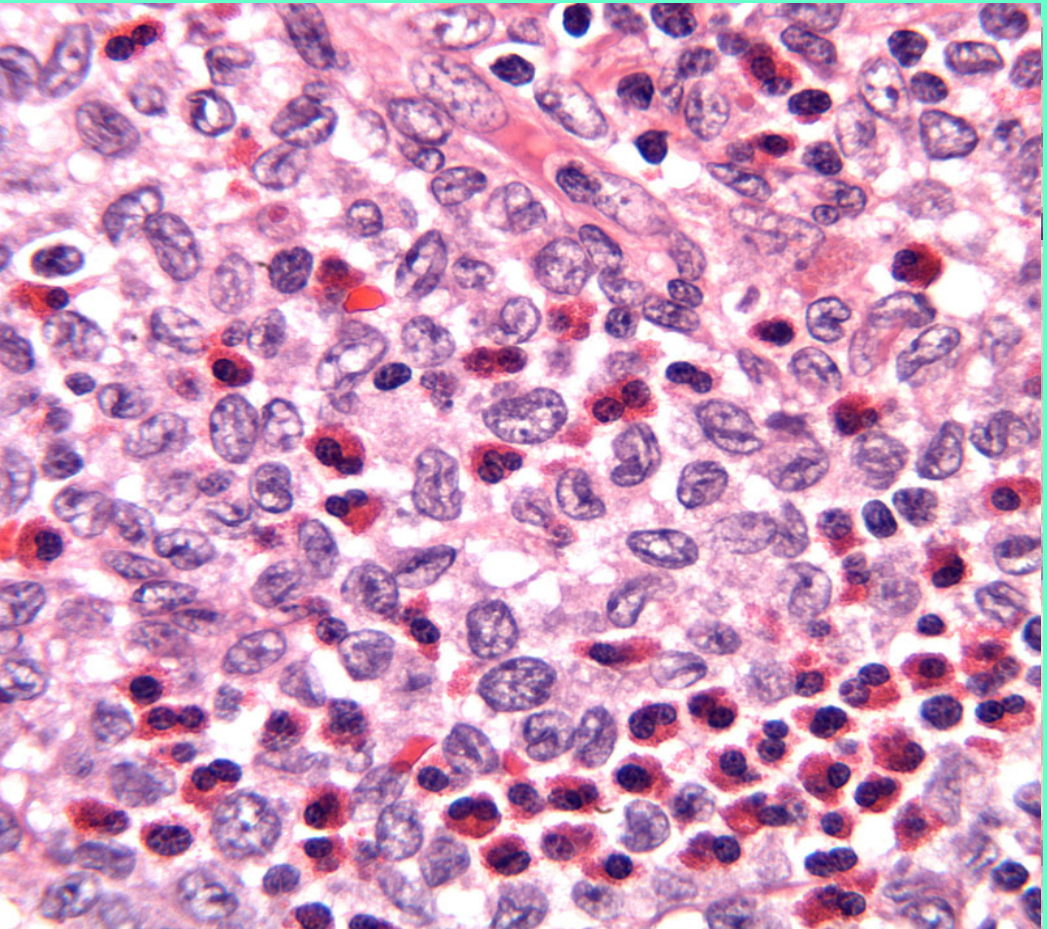


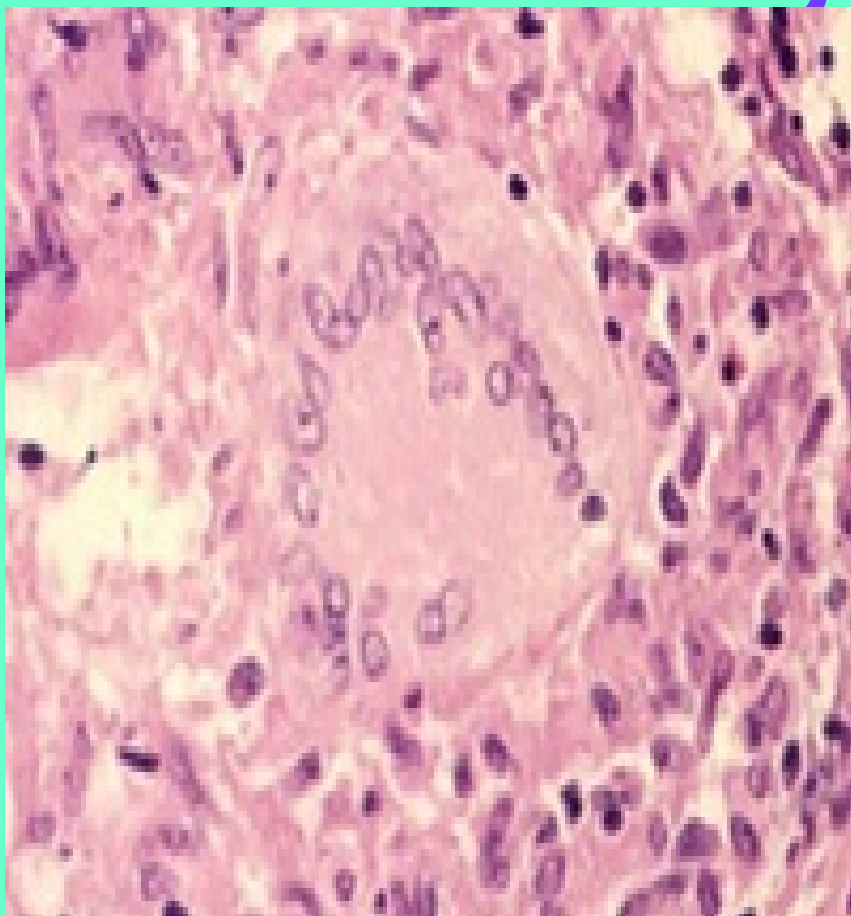

L

Histopathology

Diffuse infiltration of large,pail-staining mononuclear cells that resemble histiocyte with indistinct cytoplasmic border, round vesicular nuclei.

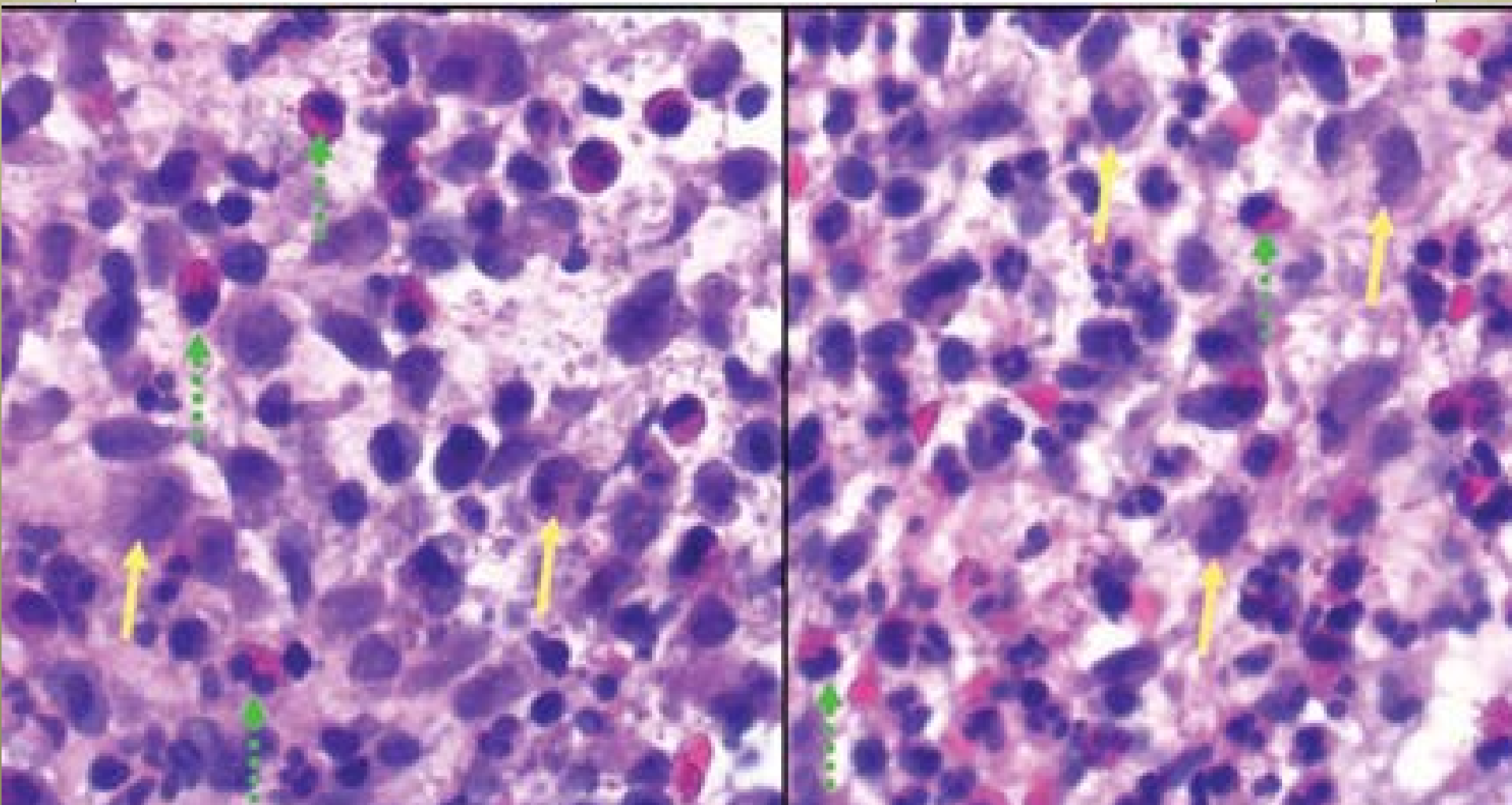
Eosinophel,plasma cell,lymphocyte & multinucleated giant cells are present.





Intermixed of cells : *Eosinophils ,,(interrupted green arrows)*

Histiocytes (large cells) ,, (solid yellow arrows).



Malignant lymphoma

Lymphoma is cancer that begins in infection-fighting cells of the immune system, called lymphocytes. These cells are in the lymph nodes, spleen, thymus, bone marrow, and other parts of the body. When you have lymphoma, lymphocytes change and grow out of control.

There are two main types of lymphoma:

Non-Hodgkin: Most people with lymphoma have this type.
HodgkinLymphoma

Lymphoma starts in infection-fighting lymphocytes.
Leukemia starts in blood-forming cells inside bone marrow.

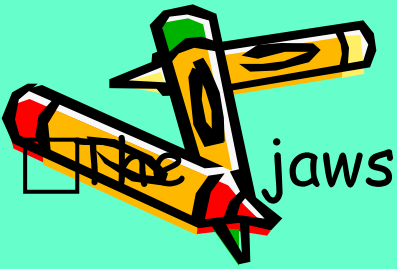
BURKITT'S LYMPHOMA

- High-grade malignant B-lymphocyte lymphoma.It is a form of non-Hodjkin's lymphoma .

- Epstein–Barr virus is closely associated, the virus that causes infectious mononucleosis.,

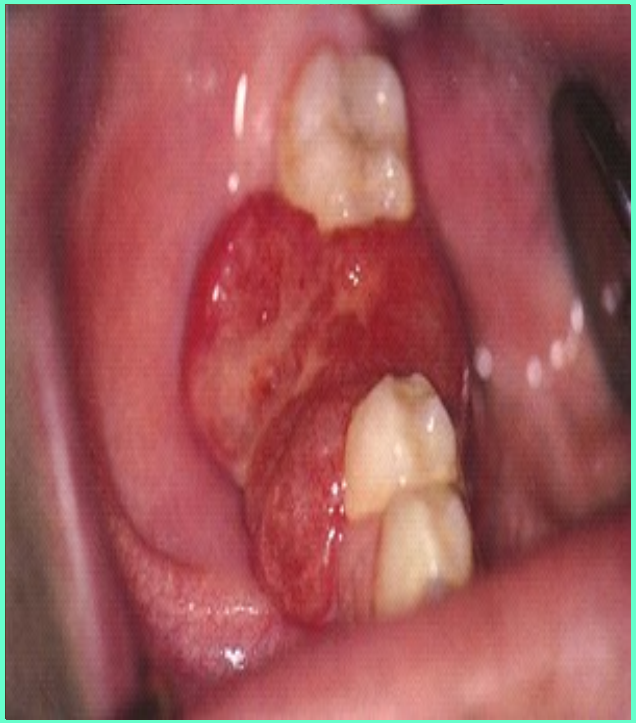
- The malignancy is prevalent in central Africa
  - **(the endemic form)**, and usually affects children 2–12 years of age.

- Cases have also been observed in other countries
  - **(the nonendemic form),**and recently in patients with
  - **AIDS.**



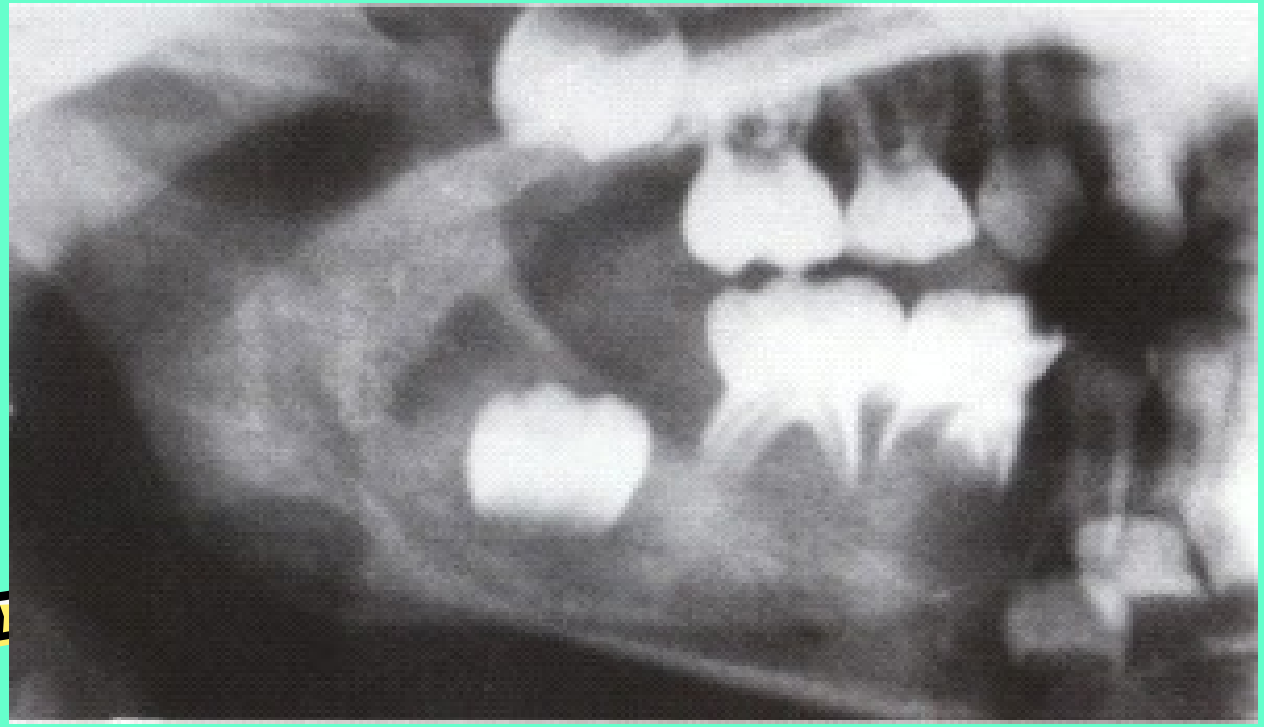
- The jaws are the most common site of lymphoma (50–70%)

Clinically: It presents as a rapidly growing hard swelling that causes bone destruction, tooth loss, and facial deformity, proptosis, tenderness Pain, paresthesia and large ulcerating or nonulcerating masses may also be seen.



Radiographic Features:

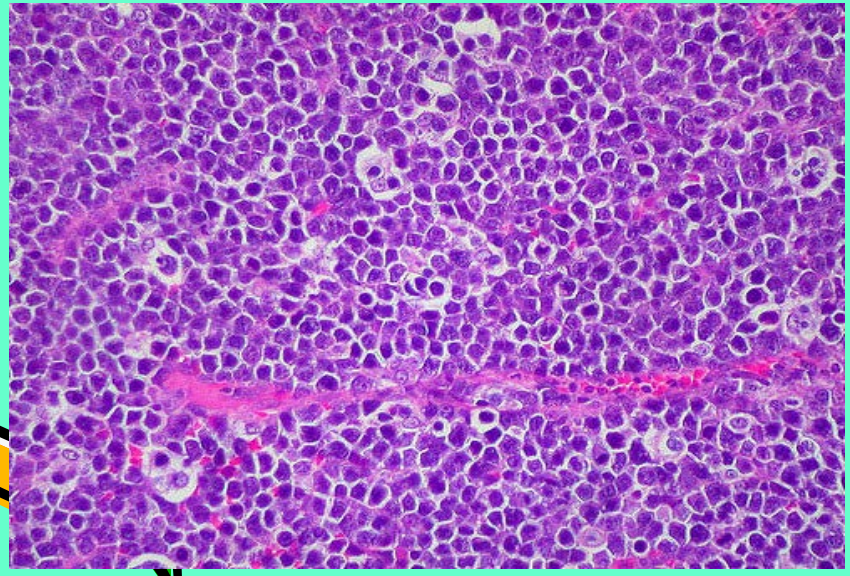
Endemic type: show lesions undergoing osseous resorption with irregular margins and reactive bone.

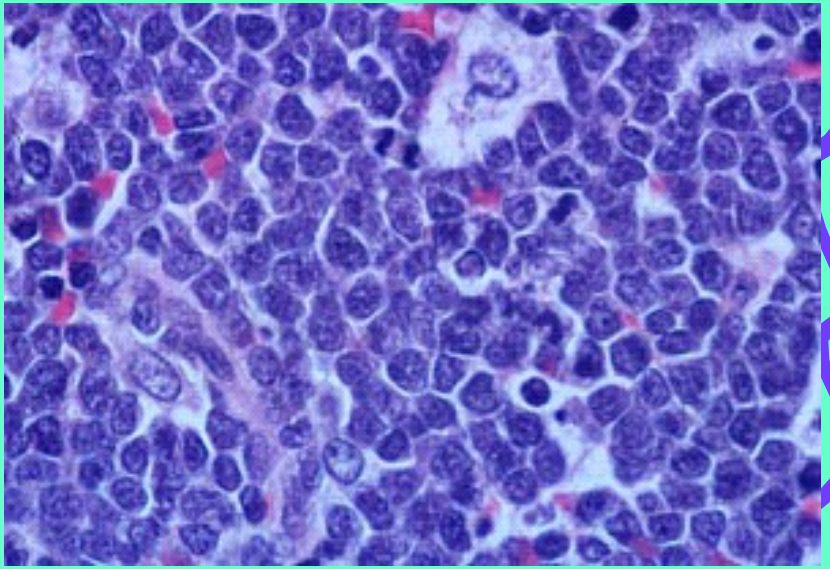


Histopathology:

The lesion is composed of sheets of tumor cells which is undifferentiated small, non-cleaved B-lymphocytes that exhibit round nuclei with minimal cytoplasm, prominent nucleoli, & prominent mitosis.

The clasic **starry sky pattern** associated with the lesion is caused by the presence of histeocytes within the tumor tissue.





Treatmrnt:

intensive chemotherapy can achieve long-term survival in more than half the people with Burkitt lymphoma.

The 5 year survival rate is less thatn 30%

Metastatic tu to jaws

- Account about 1% of malignant tu in oral cavity.
- Hematogenous spread to jaw.
- Metastasis to jaw bone is common than to the oral soft tissue.
- Most common primary tu are Ca breast, bronchus, kidney,thyroid, prostate.
- Elderly individual, Mandible more affected, uncommon in maxilla