Malignant Tumors of Connective Tissue-II

Dept Of Oral Pathology & Microbiology

Purpose Statement

At the end of the lecture student should be able to

- Describe Clinical features & histopathological features of Ewing's Sarcoma
- Describe Clinical features & histopathological features of Chondrosarcoma
- Describe Clinical features & histopathological features of Osteosarcoma

Learning Objectives

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	Enumerate clinical features	Cog nitive	Mus t Know	All	
	Write classification	Cog nitive	Must Know	All	
	Write pathogenesis	Cog nitive	Mus t	All	

Contents

- Ewing's sarcoma/Round cell sarcoma
- Chondrosarcoma
- Osteosarcoma

Ewing's Sarcoma/Round cell sarcoma

- Highly lethal round cell sarcoma
- James Ewing in 1921
- Composed of small undifferentiated round cells of uncertain histogenesis
- Considered to be intraosseous counterpart of PNET
  (Primitive Neuro Ectodermal Tumor)
- 3rd most common osseous neoplasm after osteosarcoma & chondrosarcoma

Etiology

- Genetic mutation
- Reciprocal translocation between chromosomes 11 & 22 [t(11;22) (q24;q12)]

Clinical Features

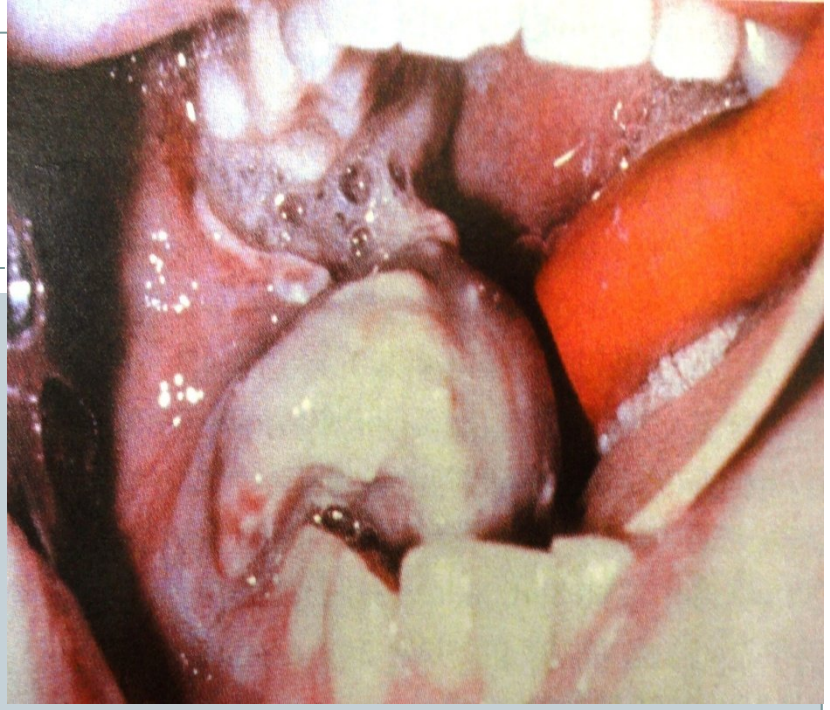
Age: Children & young adults, 5-25 yrs
Sex: M>F
Race: Whites>blacks (never)
Site: long bones, pelvis & ribs
I/o –Mandible>Maxilla

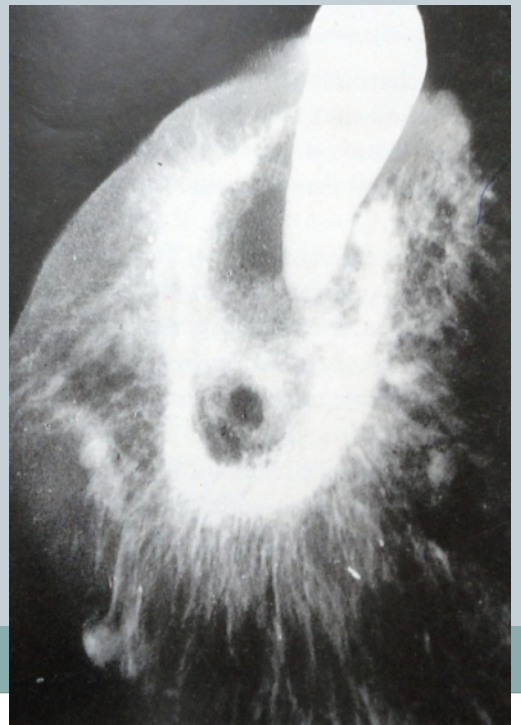
S/S

- Pain (intermittent),dull to severe in nature with swelling
- An episode of trauma often precedes
- Rapid growth of swelling
- Intraoral mass may ulcerate
- Facial neuralgia, lip paresthesia & loosening of teeth (jaw)
- Low grade fever, Leucocytosis, E.S.R. ↑

Radiographic features

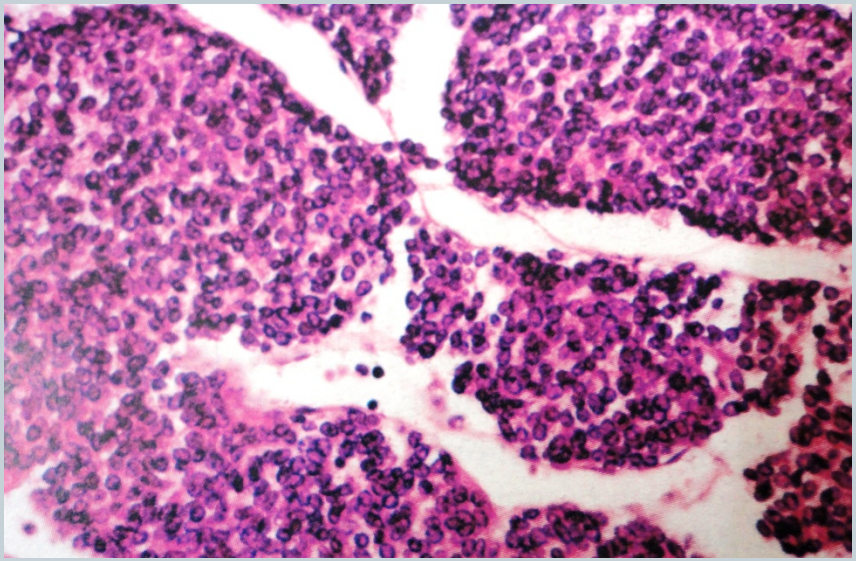
- Irregular diffuse radiolucency with ill-defined borders
- *Onion skin* appearance due to formation of layers of subperiosteal bone - resembling sclerosing osteomyelitis
- *Sun- ray* appearance due to osteophyte formation



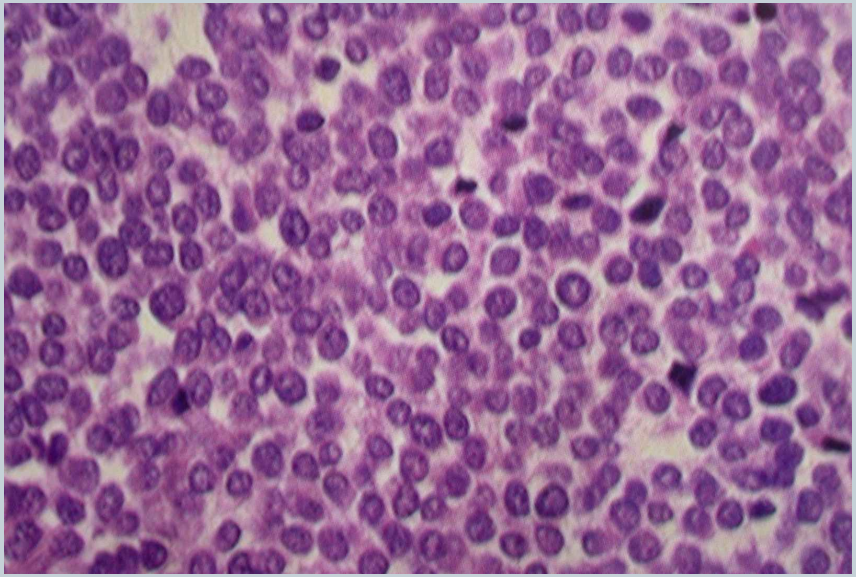


Histopathological Features

- Extremely cellular
- Solid sheets of small round cell with little stroma
- Variably sized nests - separated by fibrovascular septa- lobular pattern



- Small round cells - scanty cytoplasm, ill defined borders, relatively large round or ovoid nuclei, dispersed chromatin and hyperchromasia



Treatment

- Surgery
- Chemotherapy
- Radiotherapy

Chondrosarcoma

- Malignant tumor of cartilaginous tissue, a counterpart of chondroma
- Bones that arise from cartilaginous tissue are more liable to develop chondrosarcoma, therefore a jaw lesion is rather rare
- Jaw lesion has a poorer prognosis than those in other bones

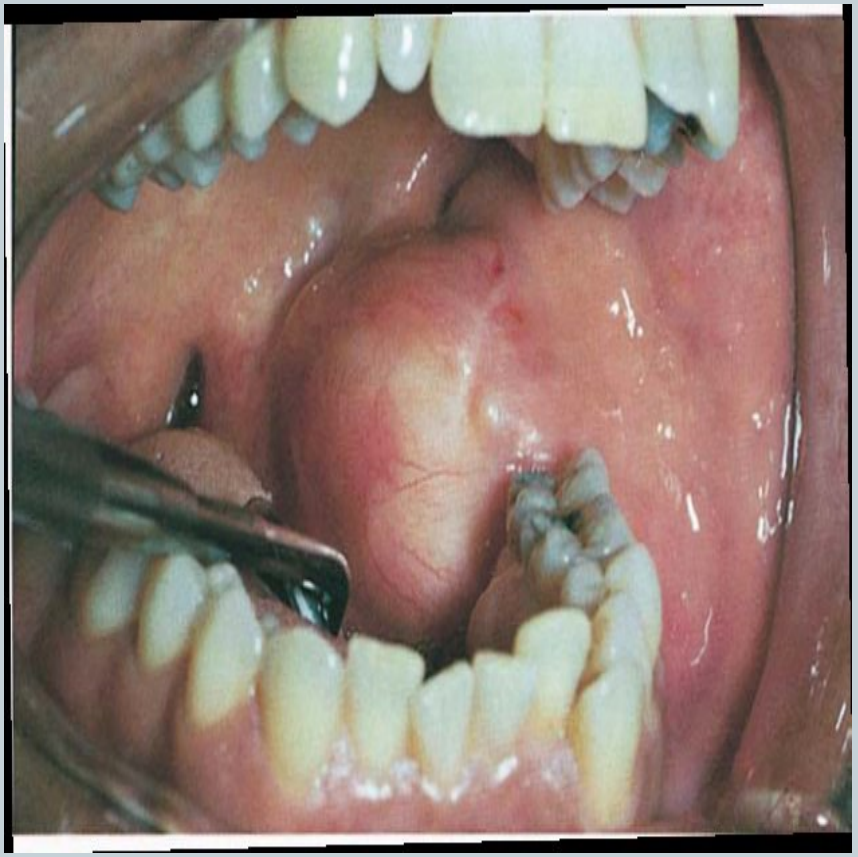
- Primary chondrosarcomas (arising de novo)
- Secondary chondrosarcomas arising most commonly in osteochondroma

Clinical Features

- Any age, 10-80 years, secondary at an earlier age
- M>F
- Metastasis relative rare & occurs late
- No pathognomic signs and symptoms
- Depending on grade-
  High grade-fast growth with excruciating pain
  Low grade- more indolent with pain & swelling

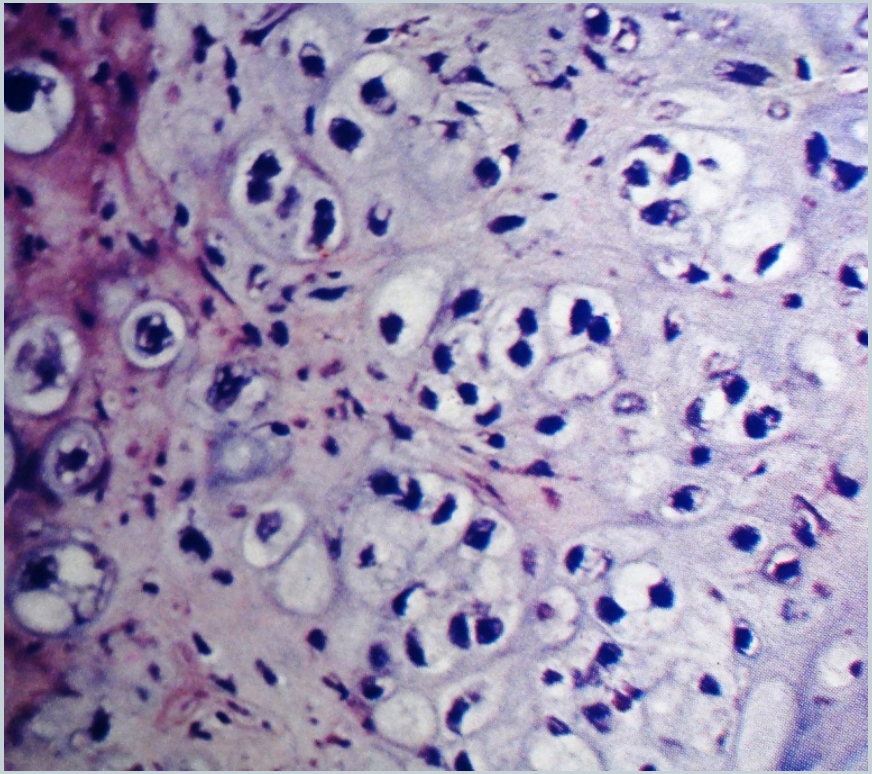
Oral manifestations

- Expanding painless swelling
- Mucosa intact
- Commonly alveolar ridge /near antrum
- Resorption & exfoliation of teeth
- Invasive & destructive lesions & metastasize readily

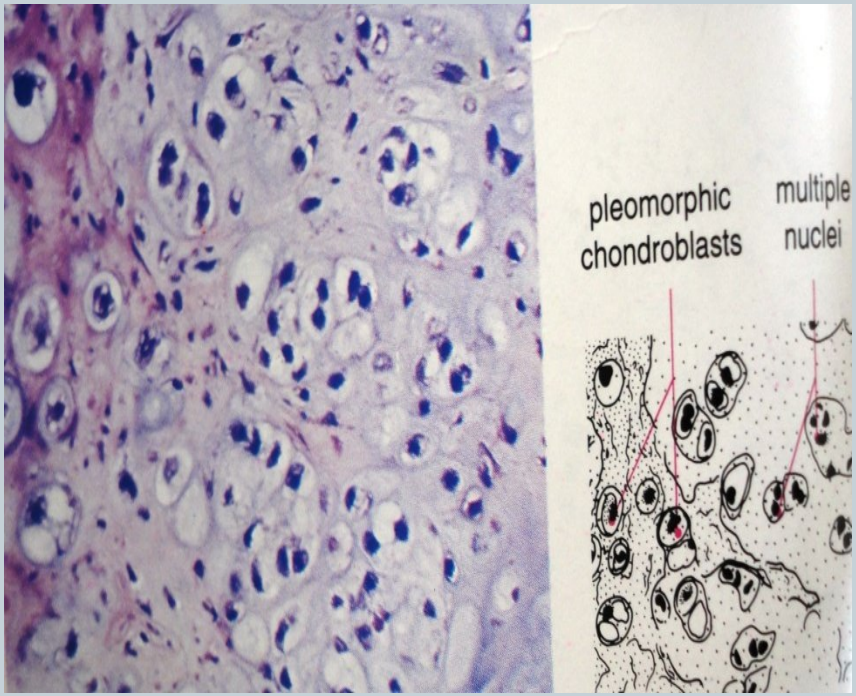


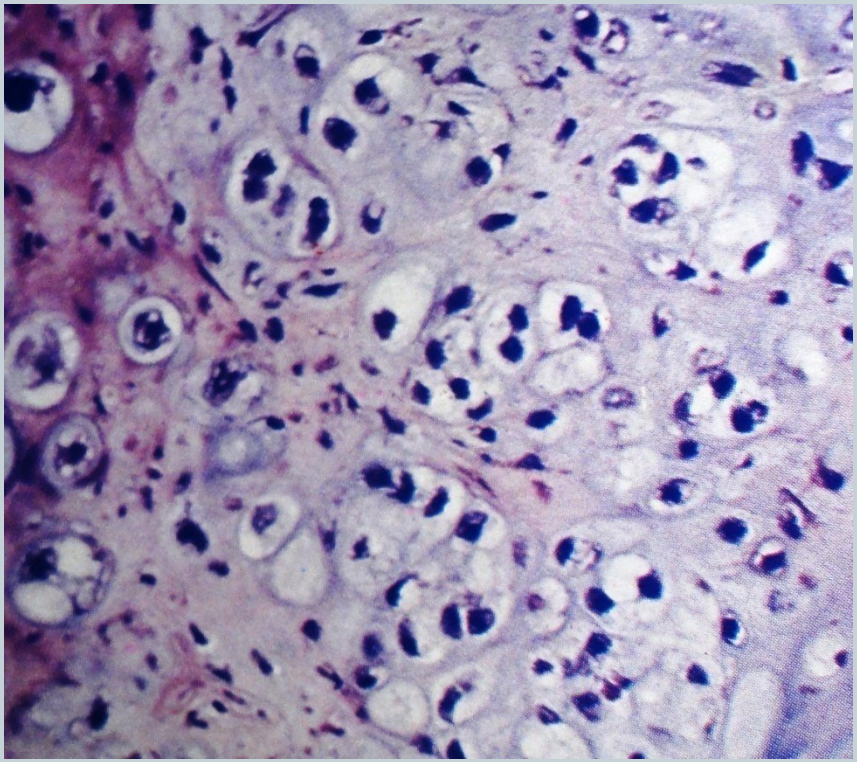
Histopathological Features

- More difficult to diagnose
- Sheets of chondrocytes
- Large, pleomorphic chondroblasts with plump nuclei & binucleated chondrocyte



- Giant cartilage cells with single or multiple nuclei
- Tumor lobules separated by fibrous connective tissue septa






chondrosarcoma
cementum of root
periodontal ligament

- Grading - based on cellularity and cytologic atypia

  - Grade I-closely mimics chondroma
  - Grade II-increased cellularity
  - Grade III-highly cellular, more mitotic figures.

Variants

- Clear cell chondrosarcoma
- Mesenchymal chondrosarcoma
- juxta-cortical chondrosarcoma
- Extra –skeletal chondrosarcoma
- Myxoid chondrosarcoma
- Dedifferentiated chondrosarcoma

T/t: Radical Surgery.

Osteosarcoma/Osteogenic sarcoma

- Third most common cancer in adolescent (lymphoma & brain tumor)
- Arises from primitive mesenchymal cells that have ability to produce osteoid or immature bone
- Except hematopoietic naoplasms,most common malignancy originate within bone

Etiology

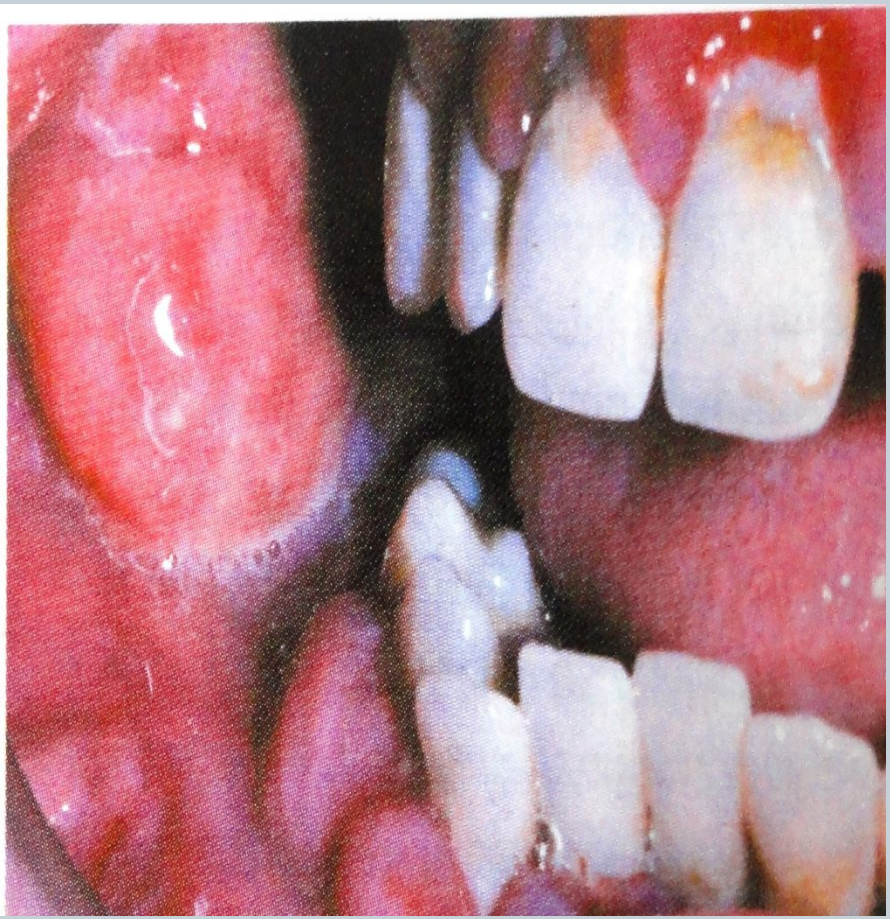
- Etiology –exact cause is unknown
- Predisposing factor -Rapid bone growth, exposure to radiation, trauma
- Secondary to Paget's disease
- Genetic predisposition –familial cases (RB gene), bone dysplasias, Li-Fraumeni syndrome, Rothmund-Thomson syndrome

Clinical features

- Bimodal Age distribution
- 10 to 25 years, young persons(corresponds with growth spurts) & over age of 50
- Initial peak during period of greatest bone growth
- Long bones of extremities near metaphyseal growth plates.
- Site – Femur, tibia, humerus, skull /jaw & pelvis
- Sex- M>F
- Pain & swelling, particularly with activity of involved bone

Oral manifestations

- Maxilla= mandible
- Mandibular –posterior body & horizontal ramus
- Maxillary-inferior portion (alveolar ridge, sinus floor, palate)
- Swelling with Facial deformity
- Pain, Tooth ache & Loosening of teeth, Paresthesia,
- Nasal obstruction

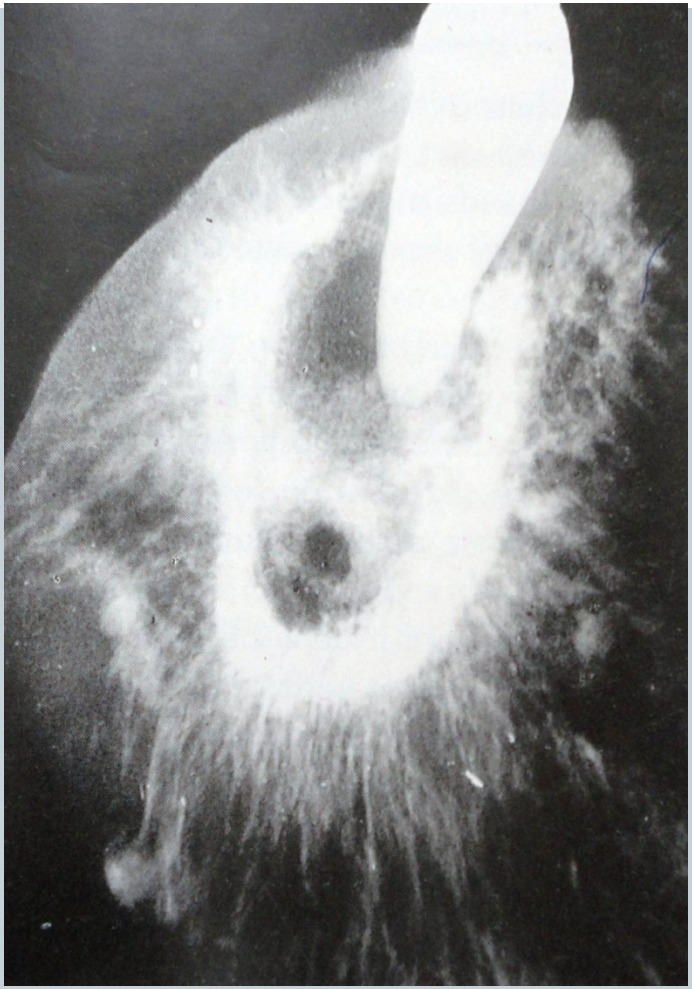


Expansion of mandible in osteosarcoma

Radiographic features

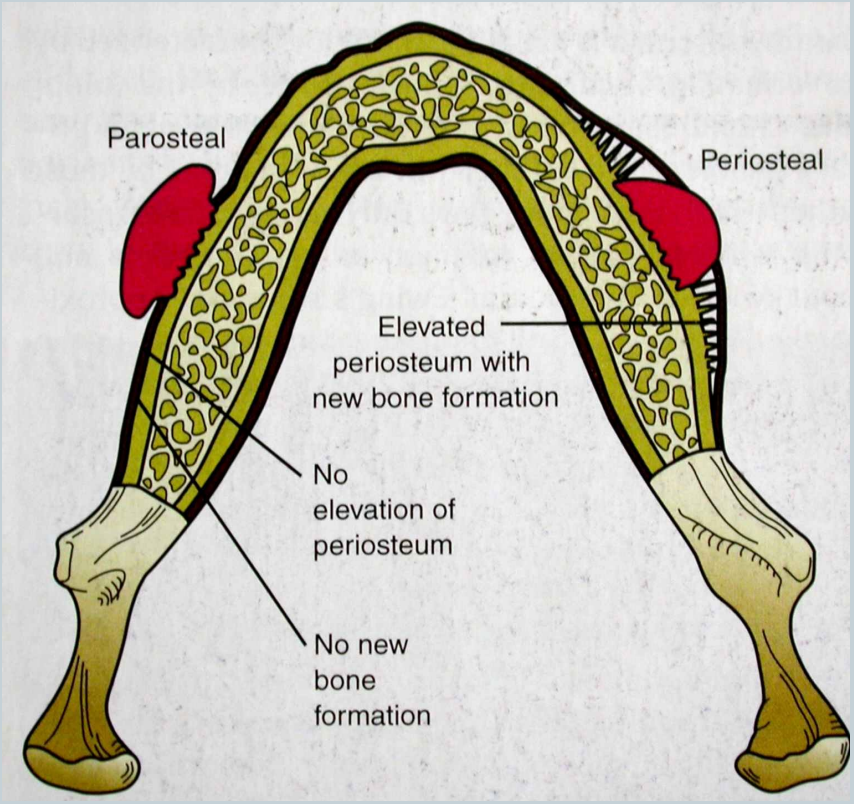
- Variable
- Depends on amount of tumor bone synthesized by malignant osteoblasts
  - Radiolucent
  - Mixed sclerotic & radiolucent lesion
  - Dense sclerotic

- Three features
- Sunray (sunbrust) pattern
- Uniform widening of periodontal ligament space
- Codman's triangle



Types

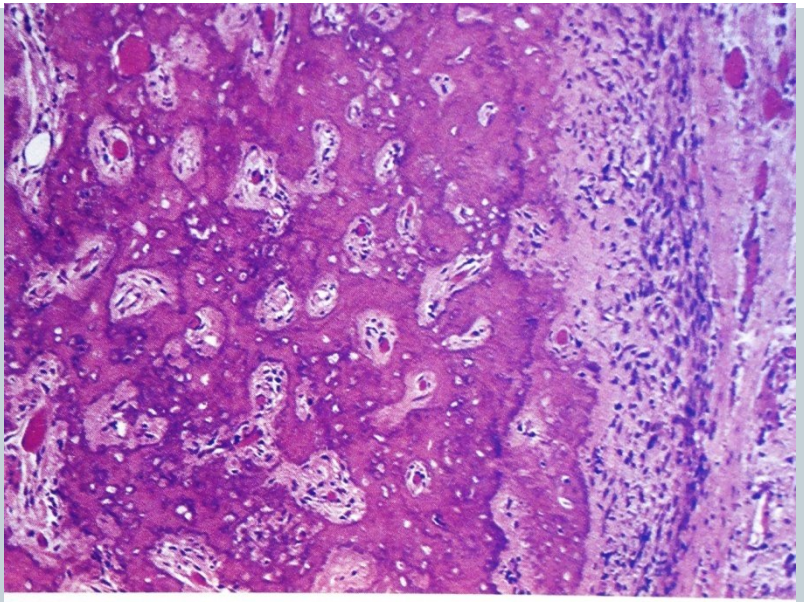
- Parosteal (juxtacortical) osteosarcoma-slow growth & good prognosis
- Periosteal osteosarcoma-more aggressive
- Extraosseous osteosarcoma-soft tissue

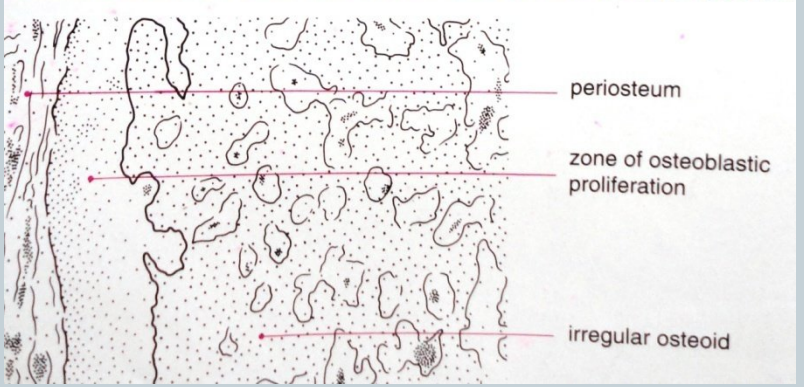


Histopathological features

Histologic Variants:

- Osteoblastic osteosarcoma
- Chondroblastic osteosarcoma
- Fibroblastic osteosarcoma
- Characterized by presence of osteoid formed by malignant osteoblasts
- Stromal cells may be spindle shaped & atypical with irregular shaped nuclei





Summary

Clinical features & histopathological features of Ewing's Sarcoma

Clinical features & histopathological features of Chondrosarcoma

Clinical features & histopathological features of Osteosarcoma

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THANK YOU