

CLASSIFICATION OF BONE TUMOURS AND GENERAL PRINCIPLES OF MANAGEMENT

-Dr Alfven Vieira

Introduction :

- **Neoplasia** : It is defined as a mass of tissue formed as result of abnormal, excessive, uncoordinated, autonomous and purposeless proliferation of cells.
- Term Neoplasia includes both Benign and Malignant.

Contrasting Differences between Benign and Malignant Tumours

FEATURES	BENIGN (DIFFERENTIATED)	MALIGNANT (UNDIFFERENTIATED)
	MACROSCOPIC FEATURES	
Boundaries	Encapsulated or well circumscribed	Poorly circumscribed and irregular
Surrounding tissue	Often compressed	Usually invaded
Secondary Changes	Occur less often	Occur more often
	MICROSCOPIC FEATURES	
Pattern	Usually resemble tissue of origin	No resemblance
Nucleo – Cytoplasmic Ratio	Normal	Increased
Pleomorphism	Absent usually	Often present
Anisonucleosis	Absent	Generally present
Hyperchromatism	Absent	Often present
Growth Rate	Usually Slow	Rapid
Metastasis	Absent	Frequently Present

Routes of Metastasis :

- **Haematogenous Spread** : Most common spread in musculoskeletal tumours
- **Lymphatic Spread** : rare in musculoskeletal tumours.
 - Seen in Rhabdomyosarcoma, Synovial sarcoma, Malignant fibrous histiocytoma .
- **Direct Implantation** :
 - Through surgeons scalpel, needles, sutures, FNAC, diagnostic or excision biopsy
- **Spread through CSF** :
 - malignant tumours of ependyma and leptomeninges rarely spread through CSF to vertebrae.

Classification of Tumours

1. WHO Classification :

widely accepted and is based on histogenesis and histological criteria.

2. Classification based on origin of tumours

3. Classification based on site of lesions.

Modified WHO Classification of BONE TUMOURS

- **1. Bone forming tumours :**

Benign :

- Osteoma
- Osteoid Osteoma
- Osteoblastoma

Intermediate

- Aggressive Osteoblastoma

Malignant

- Osteosarcoma
- Conventional osteosarcoma
- Telangiectatic osteosarcoma
- Juxta cortical or Parosteal Osteosarcoma
- Periosteal Osteosarcoma

2. Cartilage forming Tumours

- **BENIGN :**

- **Chondroma**
- Enchondroma
- **Osteochondroma**
- Chondroblastoma
- **Chondromyxoid fibroma**

- **MALIGNANT :**

- **Chondrosarcoma**
- Juxtacortical
chondrosarcoma
- Mesenchymal
chondrosarcoma
- Clear cell
chondrosarcoma
- De differentiated
chondrosarcoma
- **Malignant
chondroblastoma**

3. Giant Cell Tumours :

- **Osteoclastoma**
- Malignant giant cell tumour
- Giant cell tumour in Pagets disease
- GCT occurring in non epithelial region
- **Giant cell variants** : tumors which histologically show the osteoclastic giant cells
 - 1 - aneurysmal bone cyst
 - 2 osteoclastoma
 - 3 chondroblastoma
 - 4 unicameral bone cyst
 - 5 chondromyxoid fibroma
 - 6 non osteogenic fibroma 7- fibrous dysplasia
 - 8- brown tumor of hyperparathyroidism .

4. Marrow tumours

- **Ewing sarcoma**
- Reticulosarcoma
- Lymphosarcoma
- Myeloma

5. Vascular Tumours

— Benign :

- » Haemangioma
- » Lymphangioma
- » Glomus Tumour

— Intermediate or indeterminate variant :

- » Haemangio endothelioma
- » Hemangio pericytoma

— Malignant :

- » Angiosarcoma
- » Malignant haemangio pericytoma

6. Other Connective tissue tumours

- **Benign :**

- » Benign fibrous histiocyoma

- » **Lipoma**

- **Intermediate :**

- » Desmoplastic fibroma

- **Malignant :**

- » **Fibrosarcoma**

- » **Liposarcoma**

- » Malignant mesenchymoma

- » Malignant fibrous histiocyoma

- » Leiomyosarcoma

- » Undifferentiated sarcoma

7. Other Tumours

- **Chordoma**
- Adamantinoma
- Neurilemmoma
- **Neurofibroma**

8. Unclassified Tumours

9. Tumour Like Lesions

- **Solitary Bone cysts**
- **Aneurysmal bone cyst**
- Juxta articular bone cyst (Intra osseous ganglion)
- Metaphilic fibrous defect (non ossifying fibroma)
- Eosinophilic granuloma
- **Fibrous dysplasia**
- **Myositis ossificans**
- **Brown Tumour or hyperparathyroidism**
- Intraosseous epidermoid cyst
- Giant cell granuloma

Classification based on origin of tumours

1. **Primary Bone tumours** : Derived from bone

2. **Metastatic bone Tumours** : Due to Mets from :

–Breast	Lytic + Blastic
-Kidney	lesions
–Prostate	Lytic Blastic Lytic
- Adrenal	Lytic Lytic Lytic
–Thyroid	
- Intenstine	
- Lung, Liver	
- Urinary Bladder, Uterine Cervix	Lytic lesions

3. **Tumour Like Lesions** : Non neoplastic Conditions that resemble tumours.

Eg : Solitary Bone cyst, Aneurysmal Bone cyst, Fibrous Dysplasia, Brown`s tumour.

Classification based on site of Origin

1. Epiphyseal

Osteoclastoma, Chondroblastoma

2. Metaphyseal

**Osteioid osteoma, Osteochondroma,
Osteoblastoma, Bone cysts, Osteogenic Sarcoma**

3. Diaphyseal

Ewing`s sarcoma, Multiple myeloma

General Concepts in Tumour Terminology

- **True Capsule :**

- Surrounds a benign lesion and is composed of compressed normal cells and mature fibrous tissue.

- **Pseudocapsule :**

- Compressed tumour cells.
 - Fibrovascular zone of reactive tissue with an inflammatory component that interdigitates with normal tissue and contains satellite lesions.

- **Compartment :**

It refers to bone or muscle of origin;

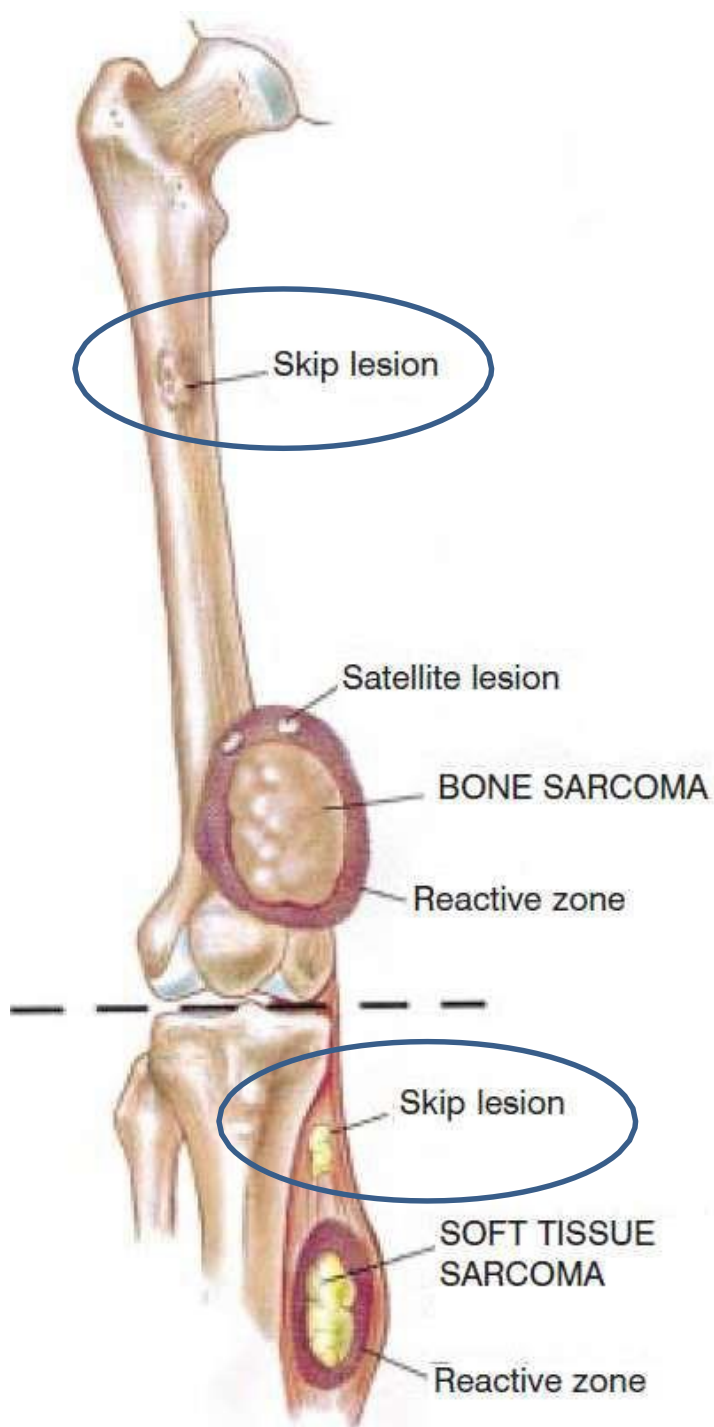
—For Muscle, compartment is that within its Fascia.

—For Bone :

- **Intracompartmental** implies Bone tumour within the cortex
- **Extracompartmental** implies a bone tumour that destroys the cortex and spreads in to the surrounding tissue.

- **Skip Metastasis :**

- A skip metastasis, is defined as a tumor nodule that is located within the same bone as the main tumor or on the opposing side of joint but not in continuity with it.
 - High grade sarcomas have the ability to break through the pseudo capsule and metastasize within the same compartment.
- MRI Scan better identifies them.



Intra Osseous Skip Mets :

Embolization of tumour cells within the marrow sinusoids.

Transarticular Skip Mets :

Occur via periarticular venous anastomosis –
Very poor prognosis

Satellite lesion

Tumour nodule within
reactive zone.

GRADING and STAGING of TUMOURS

- To determine prognosis and choice of treatment.

GRADING :

- It is defined as macroscopic and microscopic degree of differentiation of tumour

- **BORDER`s GRADING :**

- GRADE I : Well differentiated; <25% Anaplastic cells
- GRADE II : Moderately Differentiated; 25-50% Anaplastic cells
- GRADE III : Moderately differentiated; 50-75% Anaplastic cells
- GRADE IV: Poorly differentiated; >75% Anaplastic cells

Enneking`s Grading of Tumours

- **G0 Histologically benign**

(well differentiated and low cell to matrix ratio)

- **G1 Low grade malignant**

– (few mitoses, moderate differentiation and local spread only); Have low risk of metastases

- **G2 High grade malignancy**

– (frequent mitoses, poorly differentiated); High risk of metastases

STAGING OF TUMOURS

- **STAGING** is defined as extent of spread of tumour.
 - It is determined by clinical examination, Investigations and pathological studies.
 - Common staging systems are

1. ENNEKING `S STAGING SYSTEM

2. AJCC SYSTEM

3. TNM STAGING (Union International Cancer centre Geneva Staging System)

ENNEKING`s STAGING OF BENIGN TUMOURS

- 1. Latent**—low biological activity; well marginated;
remains static or heals spontaneously;
often incidental findings (i.e., nonossifying
fibroma)
- 2.Active**—symptomatic; limited bone destruction;
progressive growth but limited by natural barriers;
may present with pathological fracture (i.e., aneurysmal bone cyst)
- 3.Aggressive**—aggressive; bone destruction/soft tissue
extension; do not respect natural barriers (i.e., giant cell tumor)

Enneking`s staging of malignant tumours

- **GTM** classification described by Enneking is adopted by **Musculoskeletal Tumour society** and is based on surgical grading (G), location (T), lymphnode involvement & metastasis (M)

STAGE	GRADE	SITE
I A	Low – G1	Intracompartmental T1
I B	Low – G1	Extracompartmental T2
II A	High G2	Intracompartmental T1
II B	High G2	Extracompartmental T2
III A	Any grade with regional or distal Metastases	Intracompartmental T1
III B	Any grade with regional or distal Metastases	Extracompartmental T2

American joint committee on cancer system bone sarcoma classification (AJCC Classification)

The AJCC system for bone sarcomas is based on tumor grade, size, and presence and location of metastases.

Stage	Grade	Size	Metastases
I-A	Low	≤ 8 cm	None
I-B	Low	> 8 cm	None
II-A	High	≤ 8 cm	None
II-B	High	> 8 cm	None
III	Any	Any	Skip metastasis
IV-A	Any	Any	Pulmonary metastases
IV-B	Any	Any	Nonpulmonary metastases

American Joint Committee on Cancer System for Staging Soft Tissue Sarcomas

STAGE	GRADE	SIZE	DEPTH	METASTASES
I	Low	Any	Any	None
II	High	5 cm	Any	None
	High	>5 cm	Superficial	None
III	High	>5 cm	Deep	None
IV	Any	Any	Any	Regional or distant

TNM STAGING (Union International Cancer centre Geneva Staging System)

- **T – Primary Tumour – T_0 to T_4**

- In Situ lesion T_0 to largest and most extensive T_4 primary tumour

- **N – Nodal involvement – N_0 to N_3**

- No lymph nodes involvement N_0 to wide spread nodal involvement N_3

- **M – Metastasis – M_0 to M_2**

- No Metastasis M_0 to distant metastasis M_2

CLINICAL PRESENTATION

- **Pain :**

- Initially may be activity related, but in case of malignancy there could be progressive pain at rest and at night.
- In benign tumours, pain may be activity related when it is large enough to compress surrounding soft tissue or when it weakens bone.
- A benign Osteoid osteoma may cause night pain initially that classically gets relieved with Aspirin.

- In case of soft tissue sarcomas patients may come with mass rather than pain but in some exceptions like nerve sheath tumours, they have pain and neurological conditions.
- **Age** : It is the most important denominator because most musculoskeletal tumours occur within specific age ranges.
- **Sex** : Very few tumours show sex predilection.
 - Eg GCT is commoner in females.
- **Family History** : may be present in tumours like exostosis/ von recklenghausen`s disease.

INVESTIGATIONS

Serological investigations :

•1. Complete Blood Picture :

- **Haemoglobin** : to rule out anaemia that may be due to replacement of bone marrow by neoplastic process.
- **ESR** : raised in mets, Ewing`s sarcoma, lymphoma, leukemias

•2. Increased ***Prostate Specific Antigens (PSA)*** with ***Prostatic Acid Phosphatase levels*** in a case of blastic lesions of x ray is the diganostic of Mets secondary to Prostate Carcinoma

■ **3. Serum alkaline phosphatase (ALP)**

- Biological marker of tumour activity.
- Increases significantly when tumour and metastasis are highly osteogenic.
- ALP levels decline after Surgical removal of primary tumour and elevates if metastasis aggravates.
- Good prognostic tool.

- **Increased in following conditions: -**

- Osteoblastic bone tumors (metastatic or osteogenic sarcoma)
- 5-Nucleotidase and GGT (Gamma glutamyl Trasferase) are elevated in liver pathology along with Alkaline phosphatase, where as in bone pathologies only ALP is increased.

- **4. Antisarcoma Antibodies :**
 - Monoclonal antibodies can be detected by immunohistochemical assays.
 - Antibodies binding to sarcoma cell surface antigens have specificity.

- **5. Osteocalcin – A :** Helpful in diagnosing heavily bone producing types of tumours.

- **6. Serum Calcium :** Hypercalcemia is often due to Mets, Myeloma, Hyperparathyroidism.

- **7. Abnormal Serum protein electrophoresis** along with ***bence jones proteins in urine*** is classical of Multiple myeloma

Flow Cytometry

- A sample of cells are treated with special antibodies that stick to the cells only if certain substances are present on their surfaces.
- The cells are then passed in front of a laser beam. If the cells now have antibodies attached to them, the laser will cause them to give off light, which can be measured and analyzed by a computer.
- Flow cytometry can help determine type of those abnormal cells and help in diagnosing a tumour early.

INVESTIGATIONS: RADIOGRAPHS

- **Phemister's Law** = the most common site of infection & tumours is the fastest growing site of the long bone
 - To see a lucent lesion in bone, an estimated **30 to 50 %** of the bone must first be lost
- [[Harris & Heaney, *N Engl J Med* 1969](#)]

Radiographic Evaluation

- **Five important parameters in evaluating a tumour on a X RAY are**
 1. Anatomic site
 2. Borders
 3. Bone destruction
 4. New Matrix (Bone)
 5. formation Periosteal reaction

A. Anatomic Sites – X ray

- **Anatomic site** Specific anatomic sites of the bone give rise to specific groups of lesions.

CORTICAL FIBROUS DYSPLASIA
ADAMANTINOMA

EWING SARCOMA
RETICULUM CELL SARCOMA
MYELOMA

Diaphysis

Metaphysis

Epiphysis

OSTEOID OSTEOMA

FIBROUS DYSPLASIA

CHONDROMYXOID FIBROMA

FIBROSARCOMA

FIBROXANTHOMA
(FIBROUS CORTICAL DEFECT;
NON-OSSIFYING FIBROMA).

OSTEOSARCOMA

{ BONE CYST
OSTEOBLASTOMA

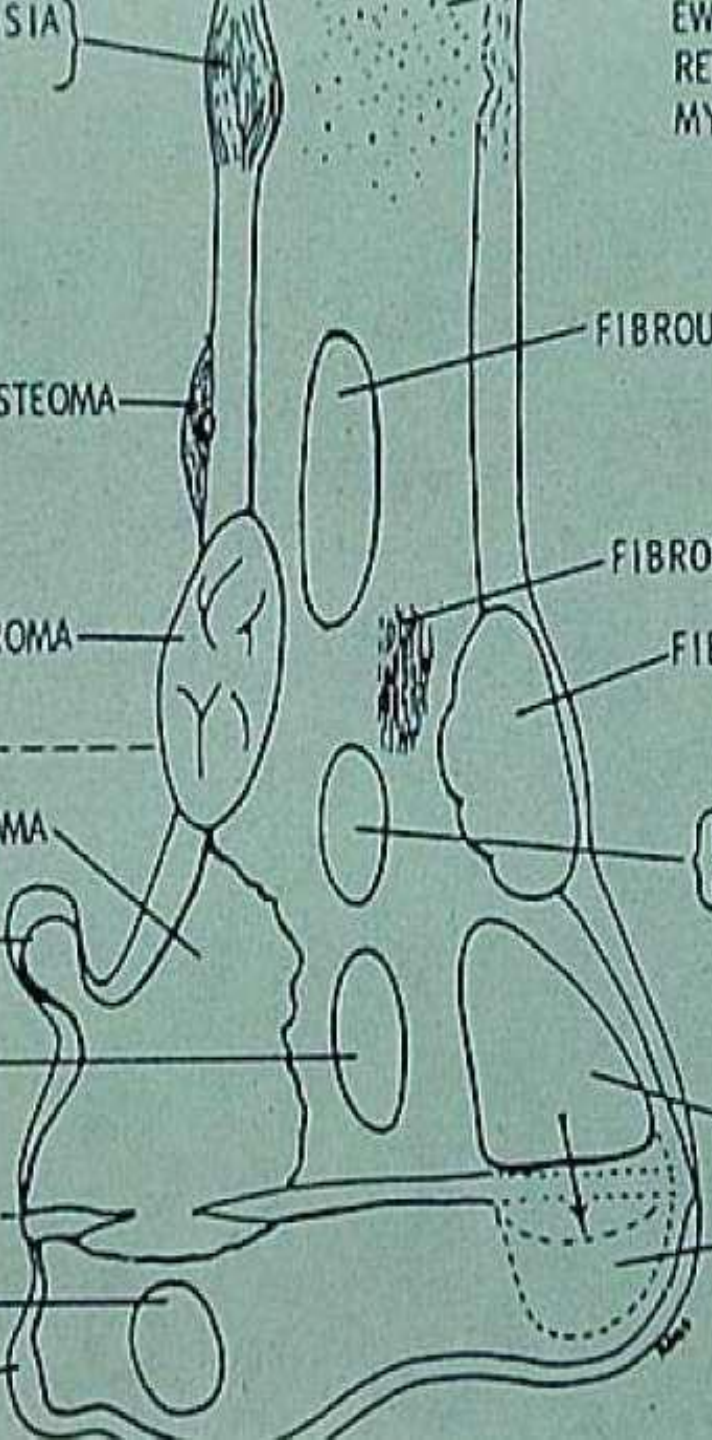
OSTEOCHONDROMA

ENCHONDROMA
CHONDROSARCOMA

GIANT CELL TUMOR
CHILD: METAPHYSEAL
ADULT: "END OF BONE"

CHONDROBLASTOMA

ARTICULAR OSTEOCHONDROMA
(DYSPLASIA EPIPHYSALIS)



Characteristic Locations

- **Simple bone cyst**

- Proximal humerus**

- Chondroblastoma
a Epiphyses
 - Giant Cell tumor
Epiphyses
 - Adamantinoma
Tibia
 - Chordoma

Sacrum, clivus

- Osteoblastoma
Spine, posterior



Characteristic Locations

- Simple bone cyst Proximal humerus

- **Chondroblastom**

a

Epiphyses

- Giant Cell tumor
Epiphyses
- Adamantinoma
Tibia
- Chordoma
Sacrum, clivus
- Osteoblastoma
Spine, posterior



A



B

Characteristic Locations

- Simple bone cyst Proximal humerus
- Chondroblastoma Epiphyses

- **Giant Cell
tumor**

Epiphyses

- Adamantinoma

Tibia

- Chordoma Sacrum, clivus
- Osteoblastoma Spine, posterior



Characteristic Locations

- Simple bone cyst Proximal humerus
- Chondroblastoma Epiphyses
- Giant Cell tumor Epiphyses
- **Adamantinoma**
a

Tibia

- Chordoma Sacrum, clivus
- Osteoblastoma Spine, posterior



Characteristic Locations

- Simple bone cyst
Proximal humerus
- Chondroblastoma
Epiphyses
- Giant Cell tumor
Epiphyses
- Adamantinoma Tibia
- **Chordoma**
Sacrum, Pelvis
- Osteoblastoma Spine,
posterior



Characteristic Locations

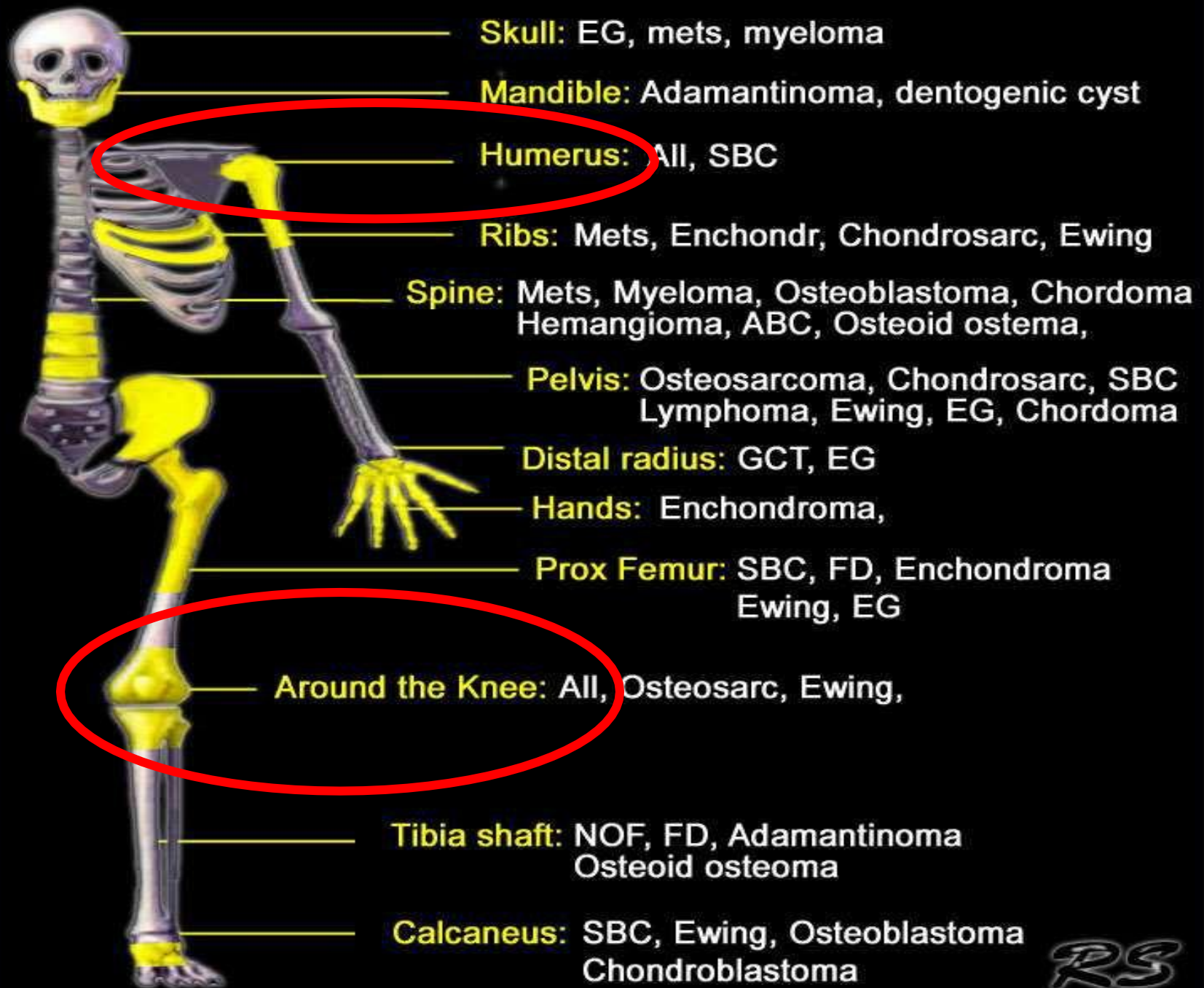
- Simple bone cyst Proximal humerus
- Chondroblastoma Epiphyses
- Giant Cell tumor Epiphyses
- Adamantinoma Tibia
- Chordoma

Sacrum, clivus

- **Osteoblastoma**
a

**Spine - posterior
elements**





B. Borders

- The border reflects the growth rate and the response of the adjacent normal bone to the tumor.
- Most tumors have a characteristic border
- **Benign lesions** (e.g., nonossifying fibromas and unicameral bone cysts) have ***well-defined borders and a narrow transition area*** that is often associated with a reactive sclerosis.
- **Aggressive or benign tumors** (e.g., chondroblastoma and GCTs) ***tend to have faint borders and wide zones of transition with very little sclerosis***, reflecting a faster-growing lesion.
- ***Poorly delineated or absent margins*** indicate an aggressive or malignant lesion

C. Bone destruction

- Bone destruction is the hallmark of a bone tumor.
- Three patterns of bone destruction are described
 1. **Geographic,**
 2. **Moth-eaten,**
 3. **Permeative.**

Geographic Bone Destruction

Complete destruction of bone from boundary to normal bone

- Non-ossifying fibroma
- Chondromyxoid fibroma
- Eosinophilic granuloma



Non-ossifying fibroma

Moth-eaten Bone Destruction

- Areas of destruction with ragged borders
- Implies more rapid growth
 - Probably a malignancy

Examples:

- Myeloma
- Metastases
- Lymphoma
- Ewing's sarcoma



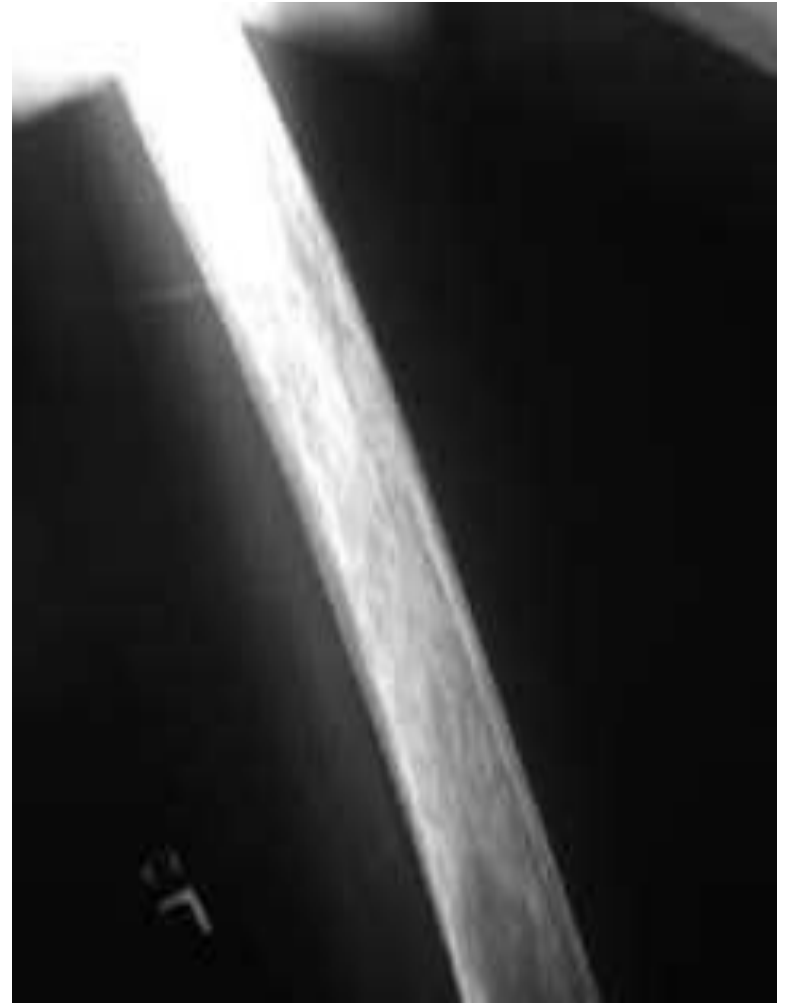
Multiple Myeloma

Permeative Bone Destruction

- Ill-defined lesion with multiple “worm-holes”
- Spreads through marrow space
- Wide transition zone
- Implies an aggressive malignancy
 - Round-cell lesions

Examples:

- Lymphoma, leukemia
- Ewing's Sarcoma
- Myeloma
- Osteomyelitis
- Neuroblastoma



Leukemia

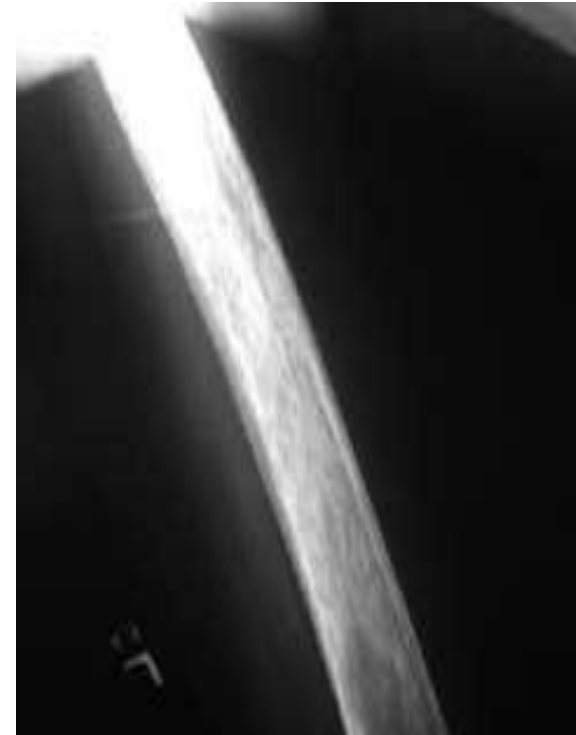
Patterns of Bone Destruction



Geographic



Moth-eaten



Permeative



Less malignant

More malignant

D. Matrix formation

- Calcification of the matrix, or new bone formation, may produce an area of increased density within the lesion.
- Calcification typically appears as flocculent or stippled rings or clusters.
- The appearance of the new bone varies from dense sclerosis that obliterates all evidence of normal trabeculae to small, irregular, circumscribed masses described as "wool" or "clouds."
- Calcification and ossification may appear in the same lesion.
- Neither type of matrix formation is diagnostic of malignancy.

Tumor Matrix

- **Osteoblastic**
 - Fluffy, cotton-like or cloud-like densities
 - Osteosarcoma



Tumor Matrix

Cartilaginous :

– Comma-shaped, punctate, annular, popcorn-like as Enchondroma, chondrosarcoma, chondromyxoid fibroma

Chondrosarcoma



Expansile Lesions of Bone

Multiple myeloma

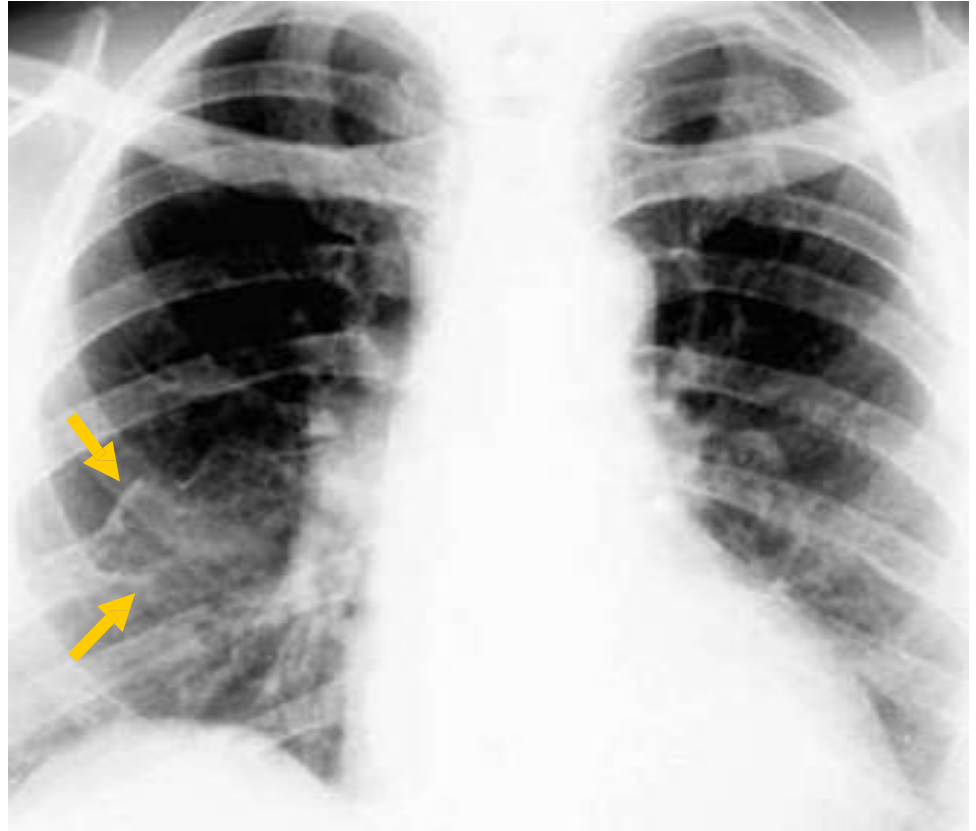
Mets

Brown tumor

Enchondroma

Aneurysmal bone cyst

Fibrous dysplasia



Expansile Lesions of Bone

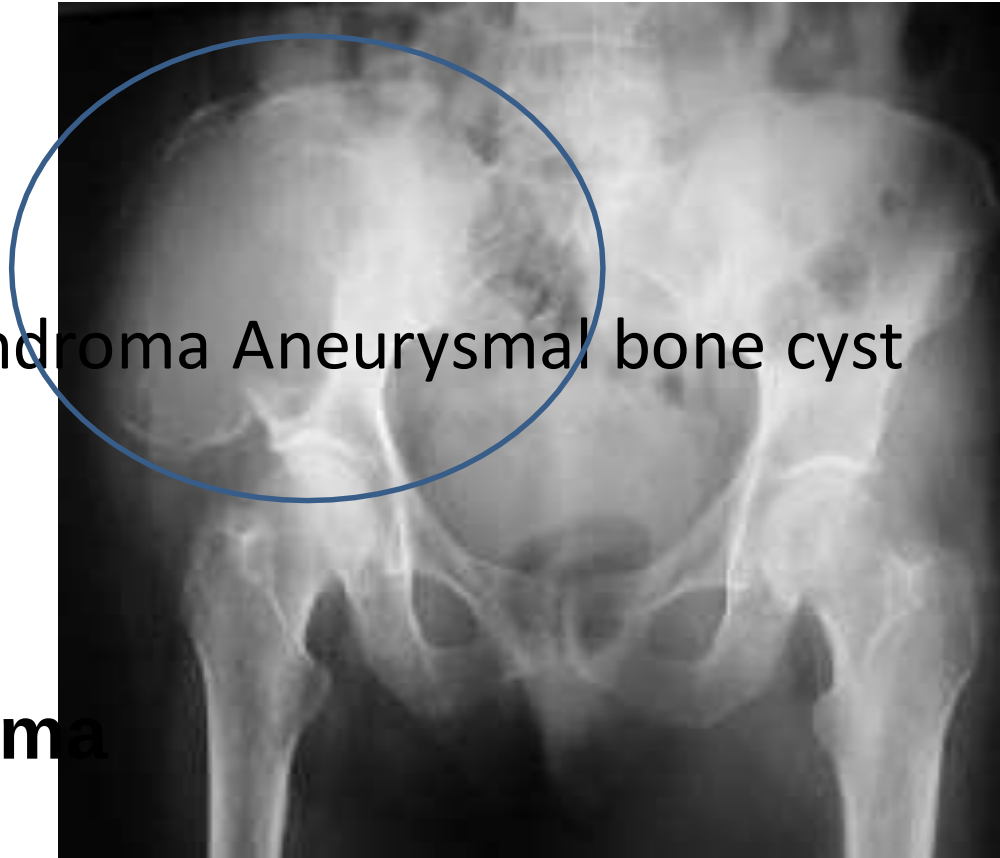
Multiple myeloma

Mets

Brown tumor Enchondroma Aneurysmal bone cyst

Fibrous dysplasia

Renal cell carcinoma



Expansile Lesions of Bone

Multiple myeloma

Mets

Brown tumor

Enchondroma

Aneurysmal bone

cyst Fibrous dysplasia



Expansile Lesions of Bone

Multiple myeloma

Mets

Brown tumor

Enchondroma

Aneurysmal bone cyst

Fibrous dysplasia



E. Periosteal reaction

- Periosteal reaction is indicative of malignancy but not pathognomonic of a particular tumor.
- Any widening or irregularity of bone contour may be regarded as periosteal activity.

Solid Periosteal Reactions

- Single solid layer or multiple closely apposed and fused layers of new bone attached to the outer surface of cortex resulting in cortical thickening.
- It is uninterrupted or continuous.



Chronic osteomyelitis

Types of Solid Periosteal Reaction

- **1. *SOLID BUTTRESS***

- Seen in Aneurysmal bone cyst, chondromyxoid fibroma

- **2. *Solid Smooth or Elleptical layer***

- Seen in Osteoid osteoma and osteoblastoma

- **3. *Undulating type :***

- Seen in long standing varicosities, periosteitis, chronic lymphoedema.

- **4. *Single Lamellar reaction :***

- Seen in Osteomyelitis, Stress Fractures, Langerhans cell histiocytosis.

Interrupted Periosteal Reactions

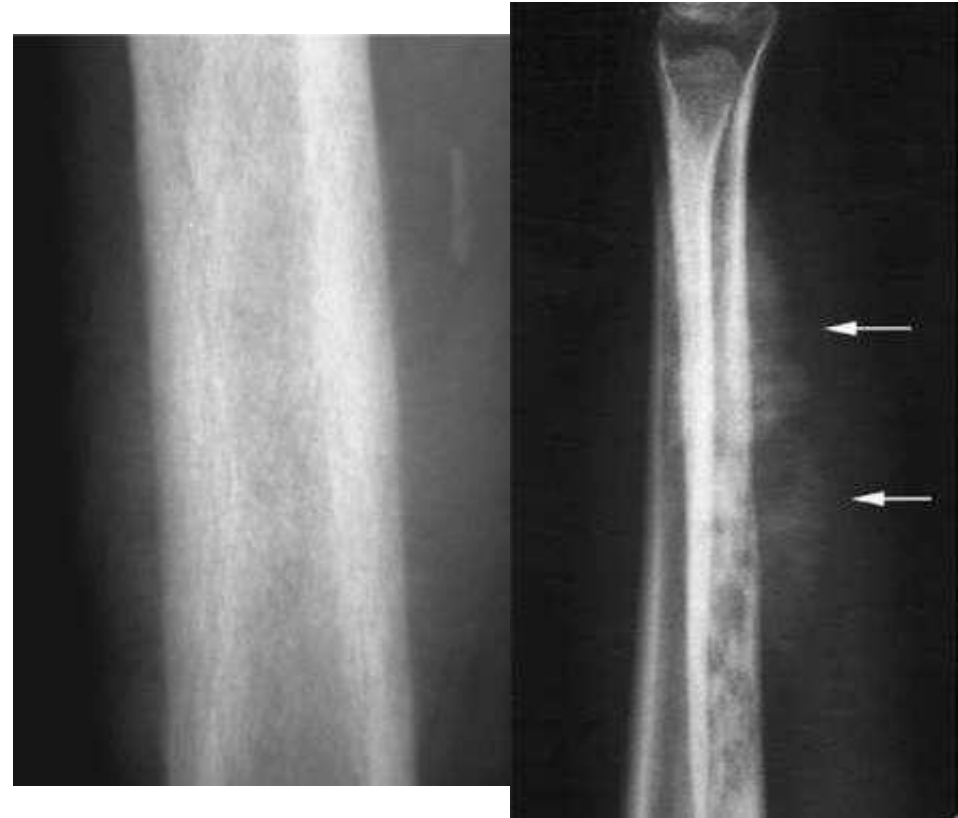
- Commonly seen in Aggressive/malignant tumours.
 - **Onion-Peel (lamellated)**
 - **Ewings sarcoma**
 - **Gouchers disease**
 - Sunburst
 - Codman's triangle



Ewing sarcoma

Sunburst type of periosteal reaction

- ❖ Fine lines of increased density representing newly formed specules of bone radiate laterally from and at right angles to the surface of the shaft giving a typical sun ray appearance.

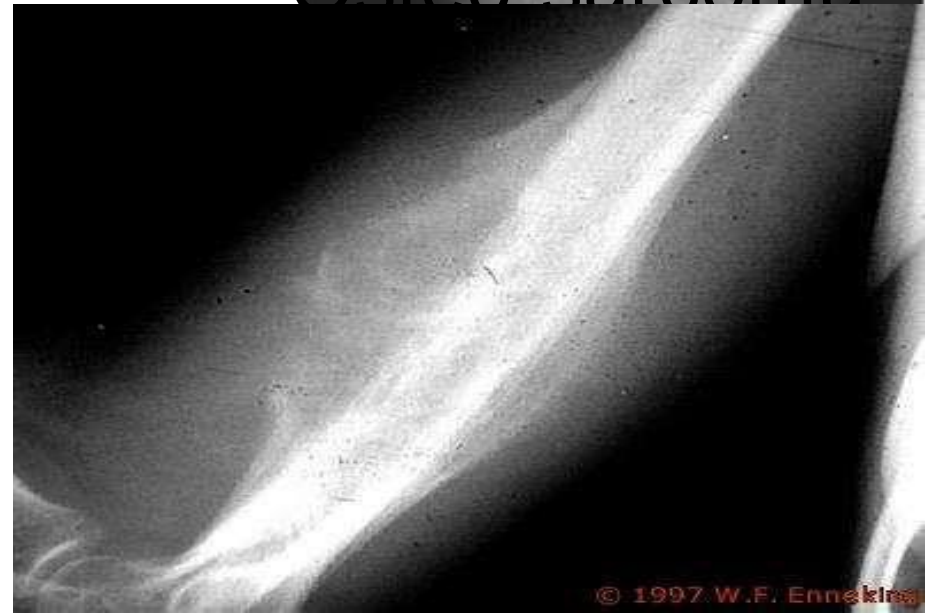


Osteo-sarcoma

Codman's triangle

- When the tumour breaks through the cortex and destroys the newly formed lamellated bone, the remnants of the latter on both ends of the break through area may remain as a triangular structure known as Codman triangle.

**Also seen in
Osteosarcoma, Ewing's
sarcoma,
Chronic Osteomyelitis**



Periosteal Reactions



Solid



Sunburst



onion-peel



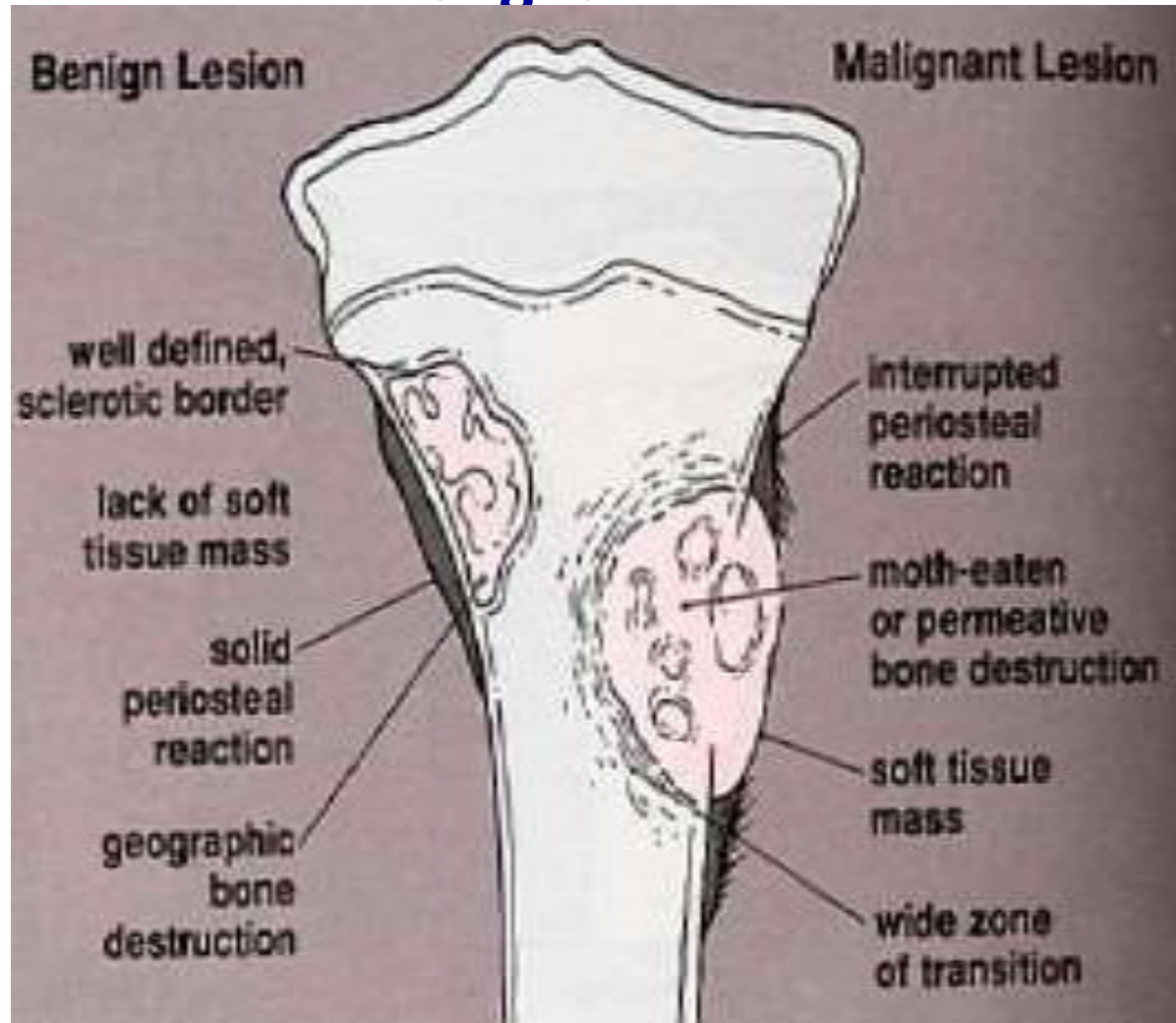
**Codman's
triangle**



Less malignant

More malignant

Radiographic Features in a Benign vs. Malignant



Mnemonic for LUSCENT bone lesions

=

Fibrous Dysplasia **O**steoblastoma **G**iant Cell Tumour

FOGIMACHINES

Metastasis/ Myeloma

Aneurysmal Bone Cyst

Chondroblastoma/ Chondromyxoid

Fibroma **H**yperparathyroidism (brown tumour)/ **H**aemangioma

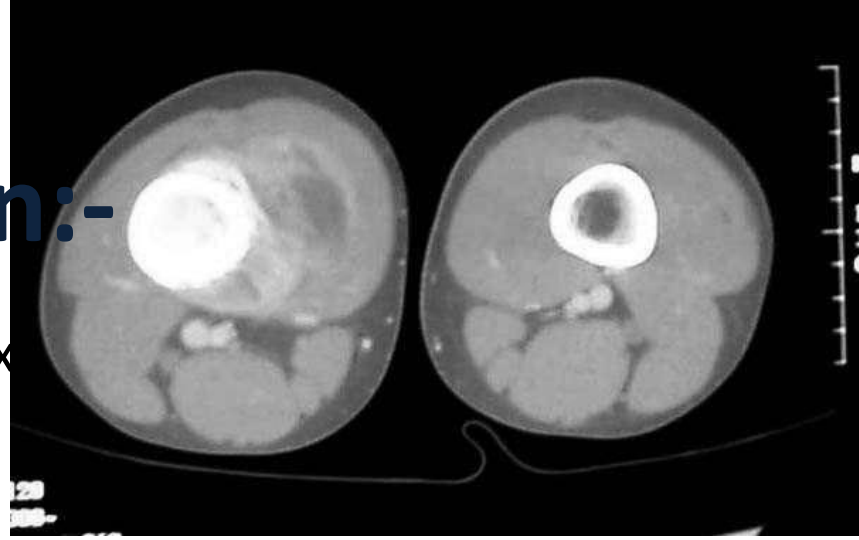
Infection

Non-ossifying Fibroma

Eosinophilic Granuloma/ Enchondroma

Simple Bone Cyst

CT scan:-



- It delineates intra and extra osseous extension.
- It can reliably distinguish between infection and tumor.
- CT scan identifies accurately area of cortical break through, soft tissue extension, medullary spread and proximity of the tumour to neurovascular bundle and evaluating integrity of cortex
- To differentiate solid and cystic lesions.
- Most sensitive investigation to detect Pulmonary mets.

- Best imaging
 - **to localise the nidus** of an osteiod osteoma,
 - to detect a thin rim of reactive bone around an aneurysmal bone cyst,
 - **to evaluate calcification** in a suspected cartilagenous lesion and
 - **to evaluate endosteal cortical erosion** in a suspected chondrosarcoma.
- **To differentiate between the neoplastic mass and inflammatory condition** : Neoplastic masses displace soft tissue fat planes where as they are obliterated in inflammatory conditions.
- It cannot differentiate benign from malignant tumours accurately.
- Except in detecting pulmonary mets, Contrast CT is better than plain CT.

MRI:-

- It has better contrast discrimination than any other modality.
- **Helps in detecting skip lesions**
- Assesses the tumor relationship with adjacent soft tissue, joints and blood vessels.
- It can visualize bone marrow content and demonstrate intramedullary extension of neoplasm.



- It is the investigation of choice in local staging of musculoskeletal tumours.
- On MRI, it is not possible to accurately differentiate benign from malignant tumours. But if the following criteria are present, lesion can be considered as a malignant one :
 - 1. Mass with irregular Border
 - 2. Non homogenous signal intensity with extra compartmental extension
 - 3. Peri tumoral edematous reaction.
 - 4. Soft tissue mass situated deep to fascia and measuring more than 5 cm in greatest diameter is likely to be a sarcoma.

PET- Positron Emission Tomography

- It uses radioactive glucose to locate cancer by observing high glycolysis rates in a malignant tissue metabolism.
- It has low specificity as the FDG (Fluoro labelled deoxy glucose) can also accumulate in benign aggressive and inflammatory lesions.
- **Also helpful in evaluating the tumour after chemotherapy.**
- **Micromets are better visulaised.**

Angiography

Most reliable means of determining vascular anatomy.

- Reactive zone is best seen on early arterial phase, while the intrinsic vascularity is best seen on late venous phase as a tumour blush.
- Transcatheter embolisation is done as a definitive treatment in some benign vascular tumours.

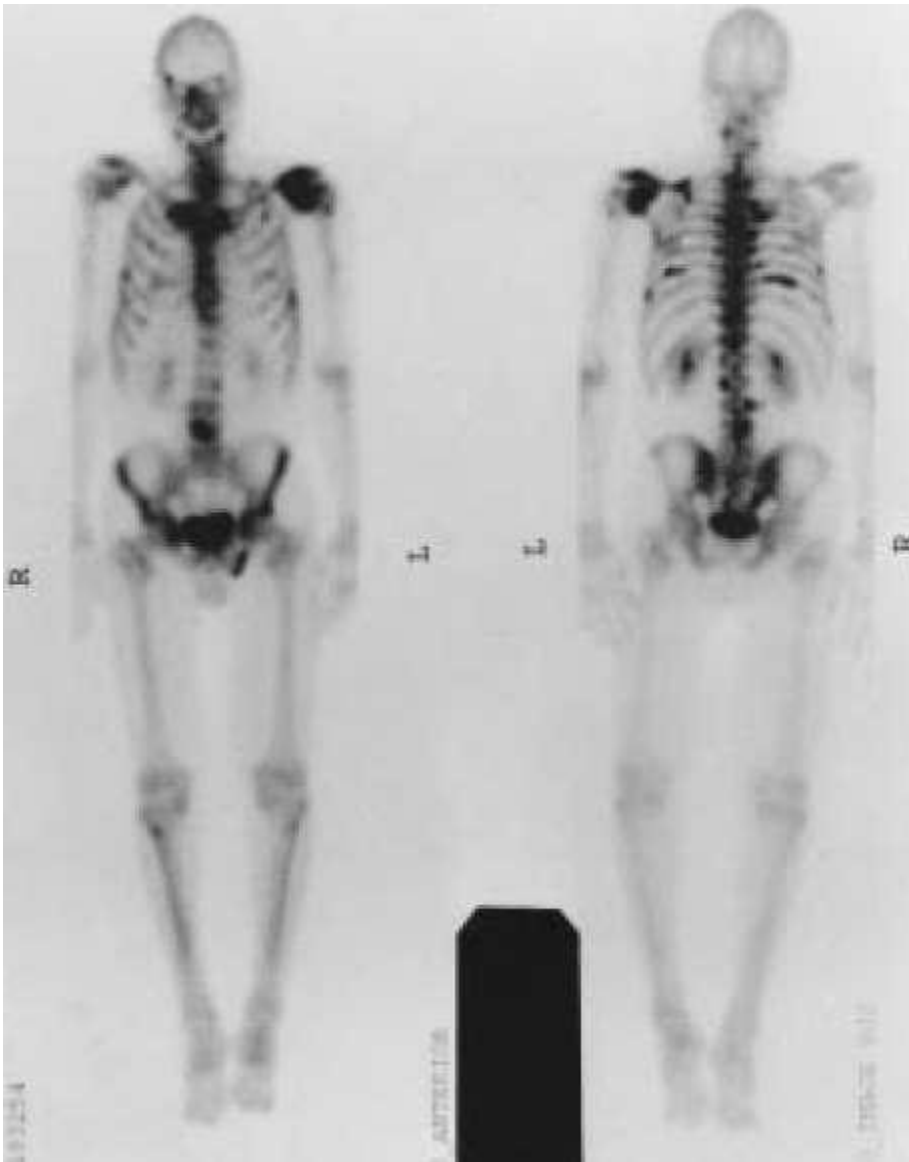


Angiography demonstrating vascularity of a tumour

Embolization of a vascular lesion, performed at least 6 hours prior to surgery, is expected to significantly reduce intraoperative blood loss.

Nuclear Imaging -Bone scan Scinitigraphy

- Technetium (^{99m}Tc) bone scans are used.
- It is an indicator for mineral turnover.
- Whenever there is altered local metabolism in remodeling bone, increased vascularity or mineralization, the isotope uptake is enhanced mainly in reactive zone surrounding the lesions.
- Confirms epiphyseal spread of tumour.
- ***Helps in detecting multiple lesions like multiple osteochondroma, enchondroma.***
- ***Where as a MRI helps in detecting skip lesions***



Bone scan showing
HOT SPOTS over
proximal humerus and
ribs

It detects the presence
of skeletal metastasis
and delineates it from
primary else where in
the body.

- Bone scintigraphy tends to show larger area of extension of medullary involvement of tumour as the radio active agent also localises the area of hyperemia and edema adjacent to tumour.
- ***Nuclear imaging is advantageous only to identifying whether skeletal involvement is solitary or multiple.***

Ultrasound

- Not routinely used in diagnosis of sarcoma; as it better differentiates only bony cystic lesions.
- However Ultrasound is used in guided percutaneous biopsy.
- In patients treated with prosthetic implants, USG is the modality that depicts early recurrence as MRI produces blurred and artifact images due to metallic implants.

Biopsy

- ▶ Used for definitive diagnosis.

- **Principles of biopsy:**

- ▶ Opted only after all other investigations are performed.

- ▶ A biopsy can be done by FNAC, core needle biopsy, or an open incisional procedure.

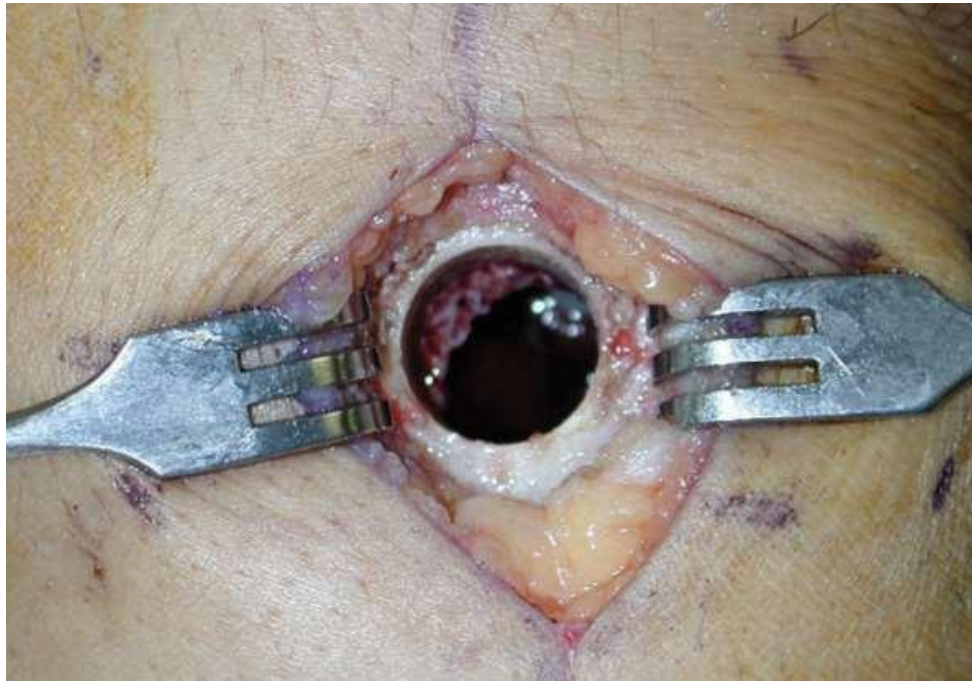
- ▶ **FNAC** may be 90% accurate at determining malignancy; however, its accuracy at determining specific tumor type is much lower.

- **Trephine or core biopsy** is recommended and often yields an adequate sample for diagnosis.

- Complications are greater with **incisional biopsy**; but least likely to be associated with a sampling error, and provides the sample for additional diagnostic studies, such as cytogenetics and flow cytometry.

- Core biopsy is preferred if limb spraying is an option as it entails less contamination than open biopsy.
- **A small incisional biopsy** can be performed if core biopsy specimen is inadequate.
- Performed under tourniquet (possibly) - the limb may be elevated before inflation but should not be exsanguinated by compression bandage.
- ▶ Longitudinal incisions preferred as transverse excision are extremely difficult or impossible to excise with the specimen.
- ▶ NV bundle not exposed. Dissection through muscle (not between) to prevent contamination of tumour.

- ▶ Approach for open biopsy is made through region of definitive surgical excision. If a drain is used, it should exit in line with the incision so that the drain track also can be easily excised en bloc with the tumor. Wound is closed tightly in layers.
- ▶ Meticulous haemostasis is arrested by use of bone wax/ Poly Methyl Metha Acrylate(PMMA) to plug the cortical window.
- ▶ Always sample the tissues from periphery of lesion which contains most viable tissue.
- ▶ Never biopsy a periosteal reaction / codmans triangle as it contains a new reactive bone and could be false negative.



- If hole must be made in bone during biopsy, **defect should be round or oval to minimize stress concentration**, which otherwise could lead to pathological fracture.
- Torsional strength is not affected by length of defect. Always attempt to keep defects less than 10% of bone diameter.
- When biopsy size is greater than 20% of bone diameter, torsional strength decreases to 50%.

Examples of poorly performed biopsies



Biopsy resulted in irregular defect in bone, which led to pathological fracture

Examples of poorly performed



**Transverse incisions
should not be used**



**Needle biopsy track
contaminated patellar
tendon**



**Multiple needle
tracks contaminate
quadriceps tendon**



Drain site was not placed in line with incision



Needle track placed posteriorly, location that would be extremely difficult to resect en bloc with tumor if it had proved to be sarcoma

- Biopsy should be done only after clinical, laboratory, and radiographic examinations are complete.
- Completion of the evaluation before biopsy aids in planning the placement of the biopsy incision, helps provide more information leading to a more accurate pathological diagnosis, and avoids artifacts on imaging studies.
- If the results of the evaluation suggest that a primary malignancy is in the differential diagnosis, Biopsy is not done unless it is possible to operate the case in the centre.

SURGICAL OPTIONS

- Curettage resection and restoration of function by limb salvage procedures or amputation is primary form of surgical correction.
- Based on surgical plane of dissection in relation to tumour, Enneking formulated following types of resection.
 - 1) Intralesional Resection**
 - 2) Marginal resection**
 - 3) Wide (Intracompartmental) resection**
 - 4) Radical (Extracompartmental) resection**

1) Intralesional Resection :

- An intralesional margin is one in which the plane of surgical dissection is within the tumor.
- This type of procedure is often described as debulking because it leaves behind gross residual tumor.
- This procedure may be appropriate for symptomatic benign lesions when the only surgical alternative would be to sacrifice important anatomical structures.
- This also may be appropriate as a palliative procedure in the setting of metastatic disease.
- ***Intralesional resection*** is through the pseudocapsule of the tumor directly into the lesion. Macroscopic tumour is left behind. Curettage is intralesional procedure.

CURETTAGE

- Cortical window with rounded margins is made
- When possible, the window is sized larger than the tumour so that the entire tumour is readily seen.
- The rounded margins reduce the risk of subsequent fracture.
- Large curetts should be used to remove the lesional tissue.
- Tumour margin should be treated with cryotherapy, PMMA cementage or phenol – alcohol cauterisation, argon beam coagulation in case of aggressive tumours.
- If curettage weakens the bone, graft using allograft or autograft with or without internal fixation is indicated.

2) Marginal Resection

- A marginal margin is achieved when the closest plane of dissection passes through the pseudocapsule.
- This type of margin usually is adequate to treat most benign lesions and some low-grade malignancies.
- In high-grade malignancy, however, the pseudocapsule often contains microscopic foci of disease, or satellite lesions.
- A marginal resection often leaves behind microscopic disease that may lead to local recurrence if the remaining tumor cells do not respond to adjuvant chemotherapy or radiation therapy.

3) Wide Resection

- Intracompartmental
- Wide margins are achieved when the plane of dissection is in normal tissue.
- Although no specific distance is defined, the **Resection includes removal of entire tumour, + Reactive zone & cuff of normal tissue.**
- If the plane of dissection touches the pseudocapsule at any point, the margin should be defined as being marginal and not wide.
- Although sometimes impossible to achieve, ***wide margins are the goal of most procedures for high-grade malignancies.***

4] Radical

- Radical margins are achieved when all the compartments that contain entire tumor and structure or origin of lesion are removed en bloc.
- The plane of dissection is beyond the limiting fascial & bone borders.
- For deep soft-tissue tumors, this involves removing the entire compartment (or multiple compartments) of any involved muscles.
- However, with improvements in imaging studies, radical procedures now are rarely performed because equivalent oncological results usually can be obtained with wide margins

ENNEKING`s Classification of Surgical Procedures for Bone Tumors

Margin	Local (Mode of resection) LIMB SALVAGE OPTIONS	Amputation(Mode of amputation)
Intralesional	Curettage or debulking	Debulking amputation Marginal
Marginal	Marginal excision	amputation
Wide	Wide local excision	Wide through bone, amputation
Radical	Radical local resection	Radical disarticulation

Radical resection

Wide resection

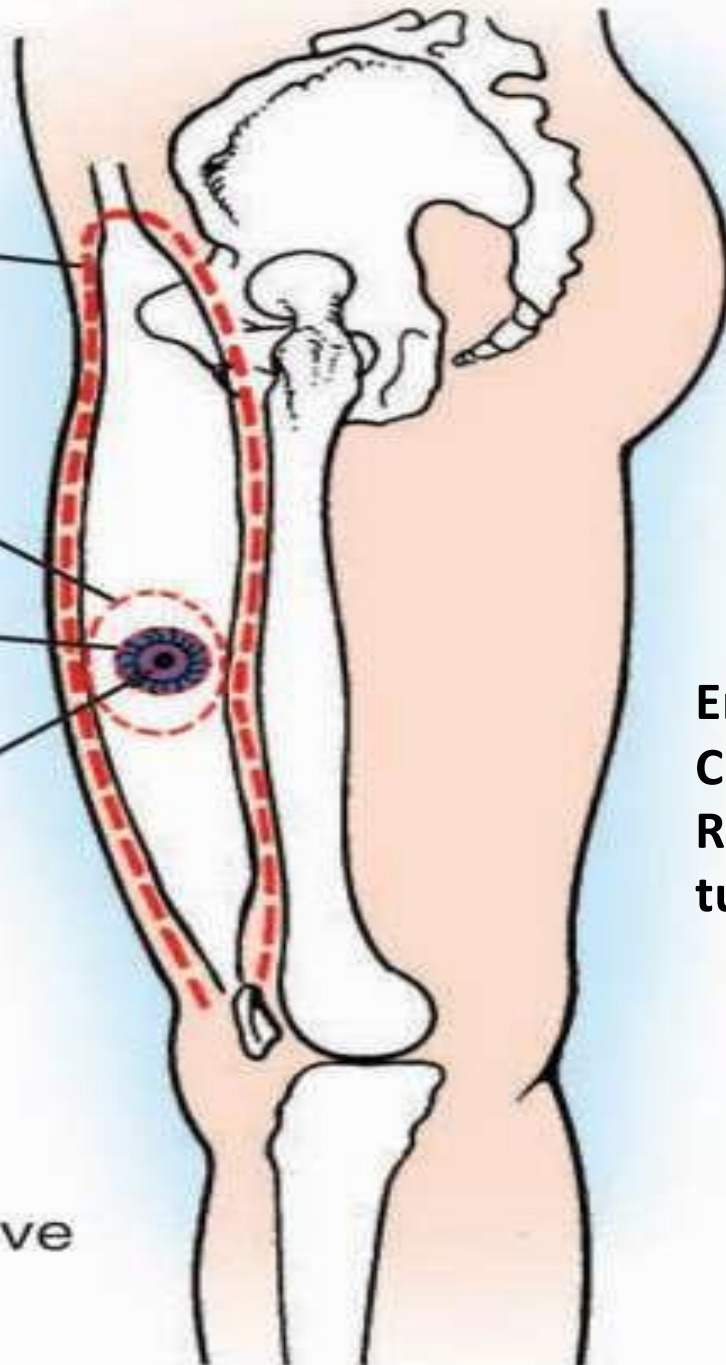
Marginal resection

Intralesional resection (debulking)

● = Tumor

● = Reactive zone

**Enneking's
Classification of
Resection of
tumours.**



SURGICAL OPTIONS

1] Limb Salvage Procedures

2] Amputations

LIMB SALVAGE PROCEDURES

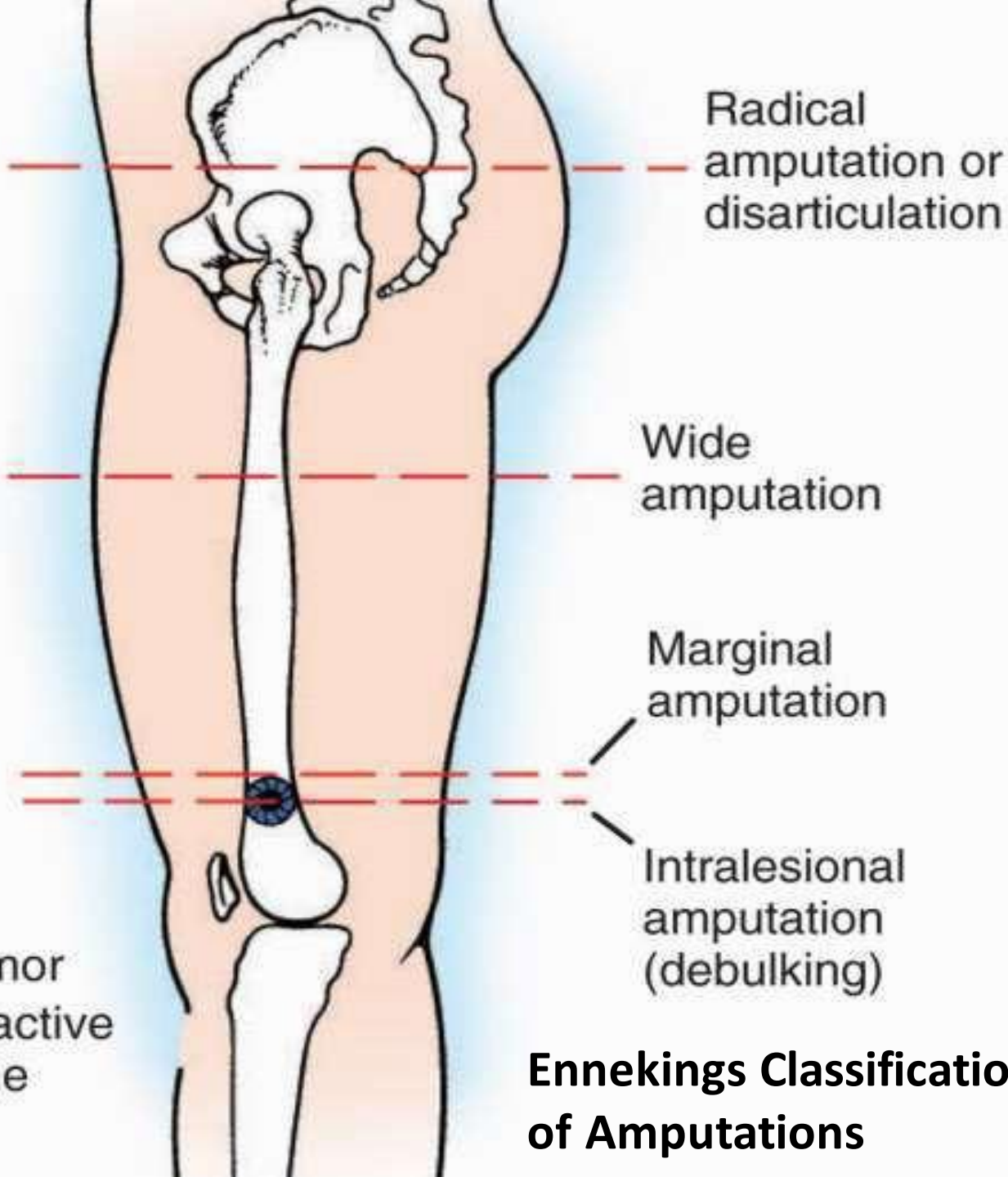
- It is designed to accomplish removal of a malignant tumour & reconstruction of the limb with an acceptable oncologic, functional & cosmetic result.
- It is sub amputative wide resection with preservation of the limb & its function.
- **Indications :**
 - Stage IA Stage IIA & Stage IIIA (All intracompartmental tumours) with good response to pre-operative chemotherapy
 - Skin should be uninvolved and free
 - There should be feasibility of keeping a cuff of normal tissue surrounding the tumour
 - Upper extremity lesions are more suitable for limb sparing surgery
- ▶ Tumours with good pre-operative chemotherapy response

Surgical reconstructive options :

- Reconstruction of bone defect may be done by
 - 1] Osteoarticular allograft reconstruction
 - 2] Allograft-prosthesis composite reconstruction
 - 3] Endoprosthetic reconstruction.
 - 4] Allograft arthrodesis
 - 5} Rotationplasty
 - 6} Turnoplasty

AMPUTATIONS

- Amputation provides definitive surgical treatment when limb sparing is not a prudent one.
- Common amputations in malignant tumours:
 - Proximal humerus : fore quarter amputation
 - Distal femur : hip disarticulation
 - Proximal tibia : mid thigh amputation



**Enneking's Classification
of Amputations**

Chemotherapy

- **Adjuvant chemotherapy:**
 - To treat presumed micrometastasis
- **Neo adjuvant[induction]**
 - Before surgical resection of the primary tumour

Advantages:

- It controls micro metastasis and improves survival rate.
- Chemotherapy makes limb salvage surgery easier.
- Decreases tumor size and vascularity.
- The response to Chemotherapy can be evaluated after surgery.

RADIOTHERAPY

- With Megavoltage radiotherapy tumor cell can be destroyed.
- High voltages are administered in short sessions.
- Radiation therapy should be started immediately after diagnosis before surgery to prevent metastasis .
- Chemotherapy increases the susceptibility of tissues to irradiation.
- Protect all normal tissue biopsy scars to prevent radiation necrosis.
- Distribute the dose in accordance with distribution of tumor.