

Biopsy and Staging of Musculoskeletal Neoplasms

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Definitions

- Bone / Soft tissue tumors (Primary)
 - Mesenchymally derived tumors (Mesodermal)
 - Benign or Malignant (Sarcoma)
 - Sarcoma=fleshy (Greek), fish flesh
 - Sarcoma—ability to metastasize systemically and invade locally

Classification

- Derived from primitive pluripotential mesenchymal cell
- Pluripotential mesenchymal cell can form
 - Bone
 - Cartilage
 - Fibrous Tissue
 - Lipogenic
 - Blood Vessels
 - Nervous tissue
 - Small Round Blue Cells

Classification

- Bone and soft tissue tumors are classified according to the predominant type of tissue (Pattern of Differentiation)
- Important to think in terms of these categories when evaluating
- Unique findings on imaging studies and pathology
- Specific types of tumors in each age group and anatomic site

<i>Histologic Type*</i>	<i>Benign</i>	<i>Malignant</i>
Hematopoietic (41.4%)		Myeloma Reticulum cell sarcoma
Chondrogenic (20.9%)	Osteochondroma Chondroma Chondroblastoma Chondromyxoid fibroma	Primary chondrosarcoma Secondary chondrosarcoma Dedifferentiated chondrosarcoma Mesenchymal chondrosarcoma
Osteogenic (19.3%)	Osteoid osteoma Benign osteoblastoma	Osteosarcoma Parosteal osteogenic sarcoma
Unknown origin (9.8%)	Giant cell tumor	Ewing's tumor Malignant giant cell tumor Adamantinoma
Fibrogenic (3.8%)	(Fibrous) histiocytoma Fibroma Desmoplastic fibroma	(Fibrous) histiocytoma Fibrosarcoma
Notochordal (3.1%)		Chordoma
Vascular (1.6%)	Hemangioma	Hemangioendothelioma Hemangiopericytoma
Lipogenic (<0.5%)	Lipoma	
Neurogenic (<0.5%)	Neurilemmoma	

Natural History

- **Benign**
 - Latent
 - Active
 - Aggressive
- **Malignant**
 - Low Grade
 - Intermediate
 - High Grade

Growth and Behavior

- Sarcomas grow locally in a centrifugal manner
- Form “Ball –Like” masses
- Periphery is least mature
- Benign aggressive and malignant tumors compress adjacent tissue into a pseudocapsular layer
- Pseudocapsular layer-microscopic extension of main tumor mass (satellite nodules)

Growth and Behavior

- Pseudocapsule: 2 zones
 - Compressed tumor cells
 - Fibrovascular zone of reactive tissue with an inflammatory component that interdigitates with normal tissue—contains satellite lesions
- True capsule—surrounds a benign lesion; composed of compressed normal cells and mature fibrous tissue

Growth and Behavior

- Surgical resection must include the pseudocapsule to ensure removal of the entire lesion
- Skip metastasis: High grade sarcomas have the ability to break through the pseudocapsule and metastasize within the same compartment

Growth and Behavior

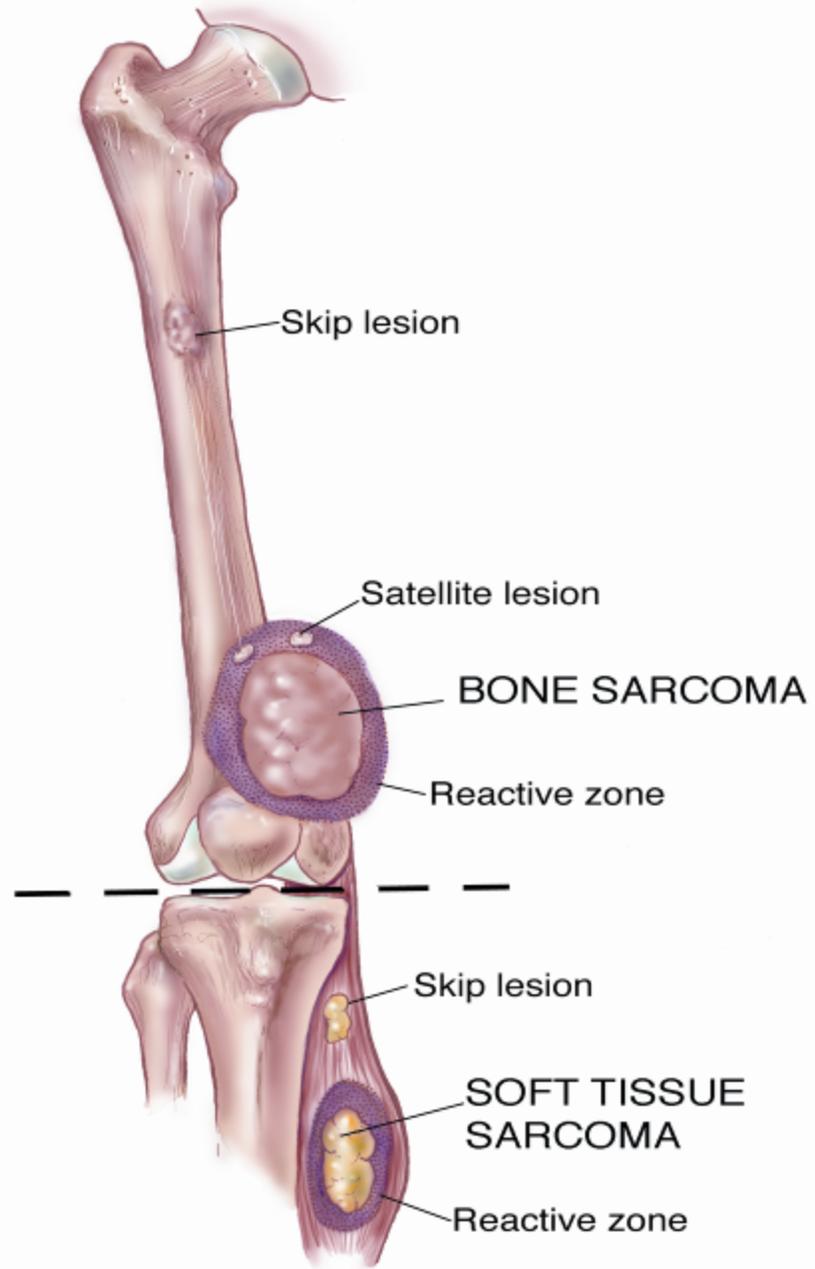
- High Grade Bone Sarcomas
 - Intraosseous Skip Mets---embolization of tumor cells within the marrow sinusoids
 - Transarticular Skip Mets---occur via periarticular venous anastomoses
- Clinical Incidence of Skip Mets < 1%
- Very poor prognosis (0% cure)

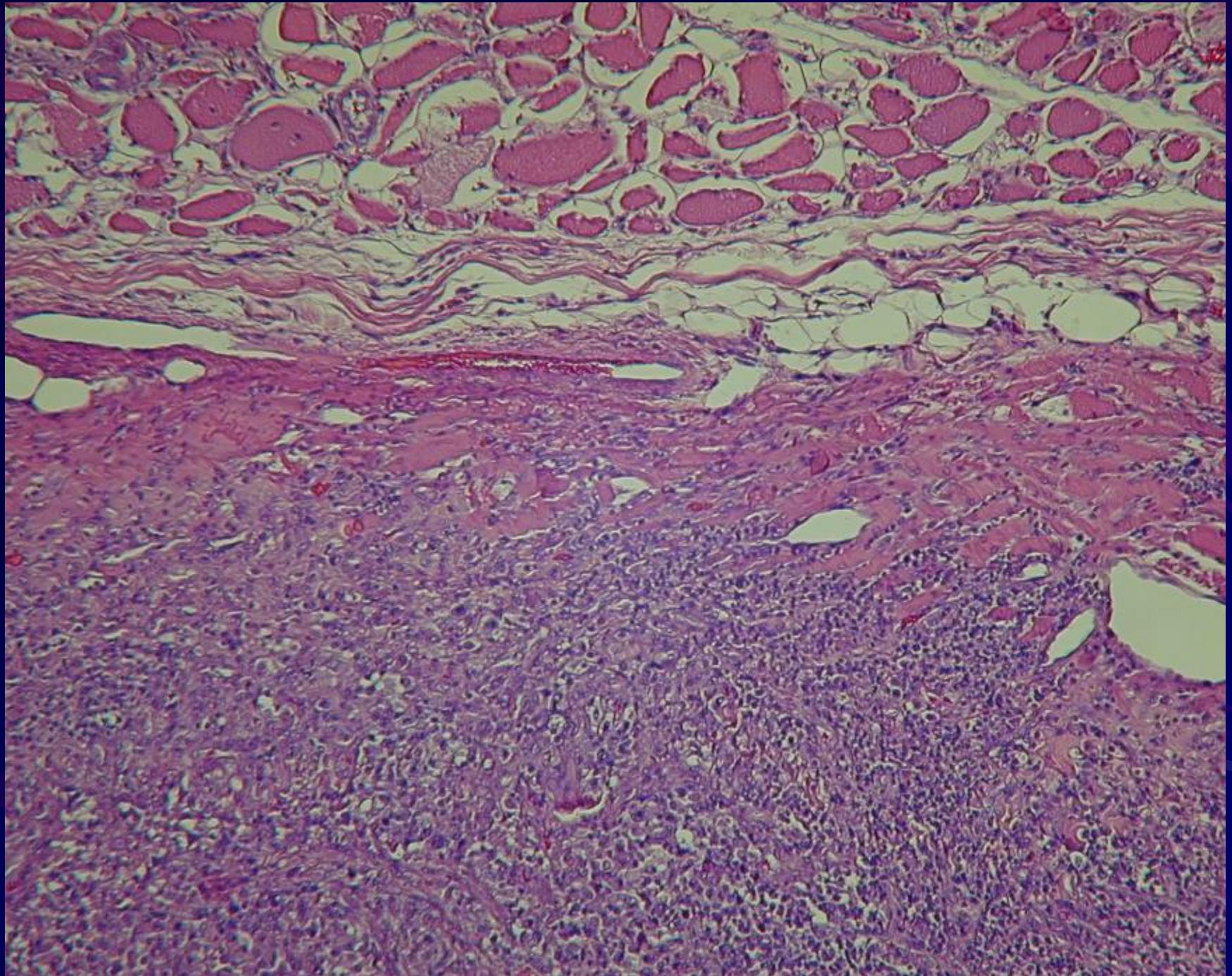
Growth and Behavior

- Sarcomas have the ability to metastasize systemically—hematogenously (contradistinction to carcinomas—lymphatic spread primarily)
- Most common sites:
 - Lungs
 - Bones
 - Liver (primarily retroperitoneal soft tissue sarc)

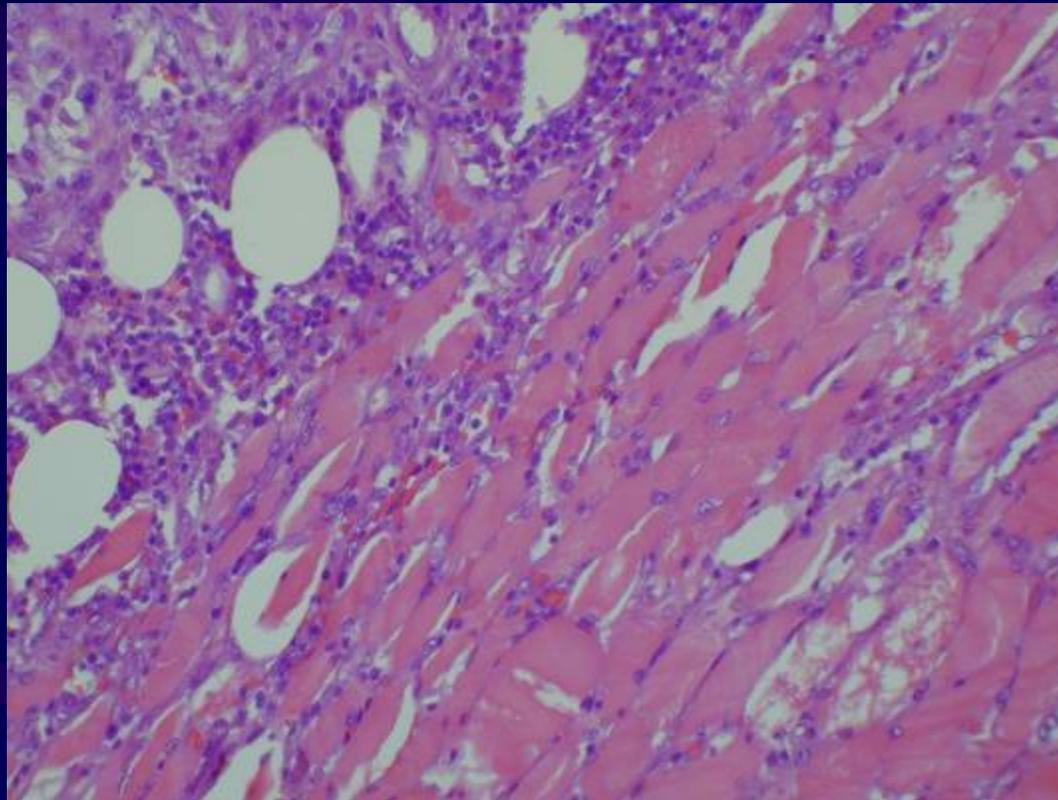
Growth and Behavior

- Low Grade tumors—mets < 5-10%
- High grade lesions—mets 60%-100%
- Some benign aggressive lesions can metastasize to the lungs, other bones (rare event)
 - Giant Cell Tumor
 - Chondroblastoma
- Multicentricity: Multiple bony sites at presentation (??synchronous mets)
 - GCT, Osteosarcoma, Ewings

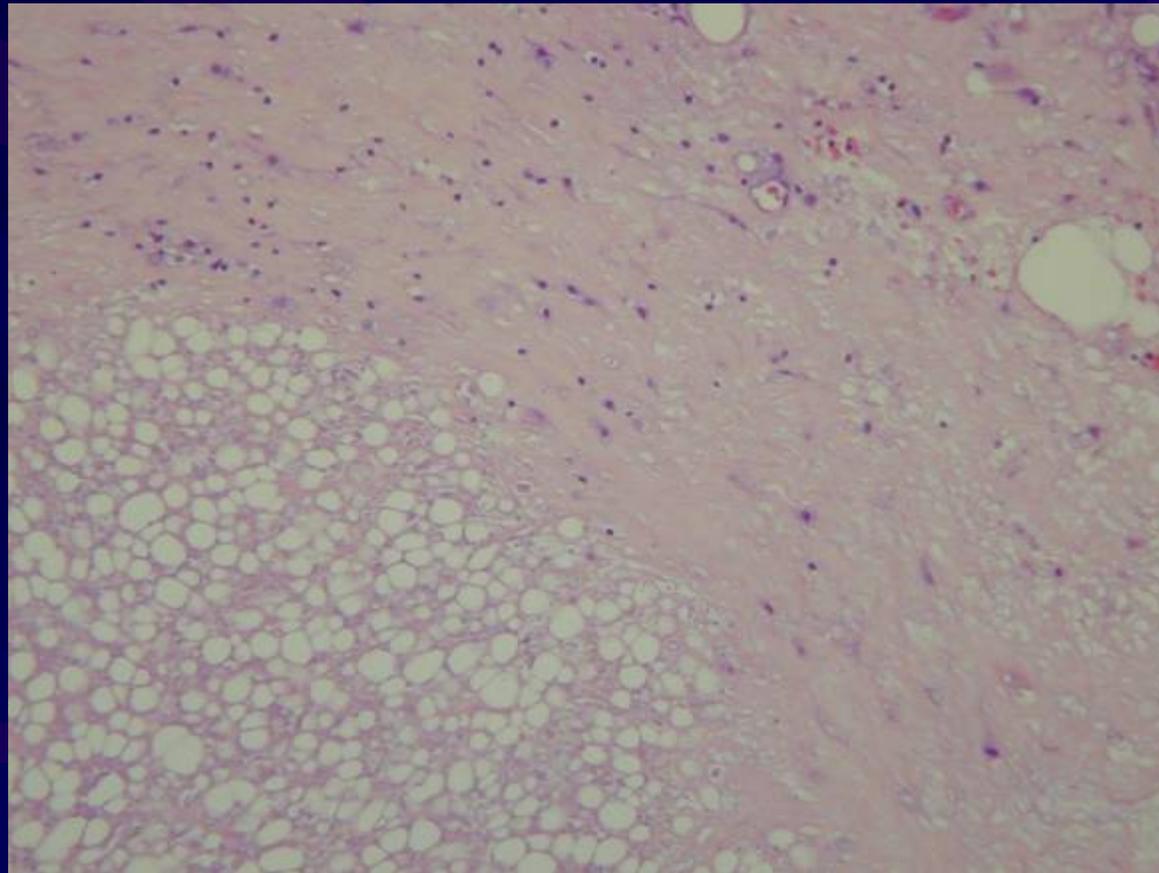


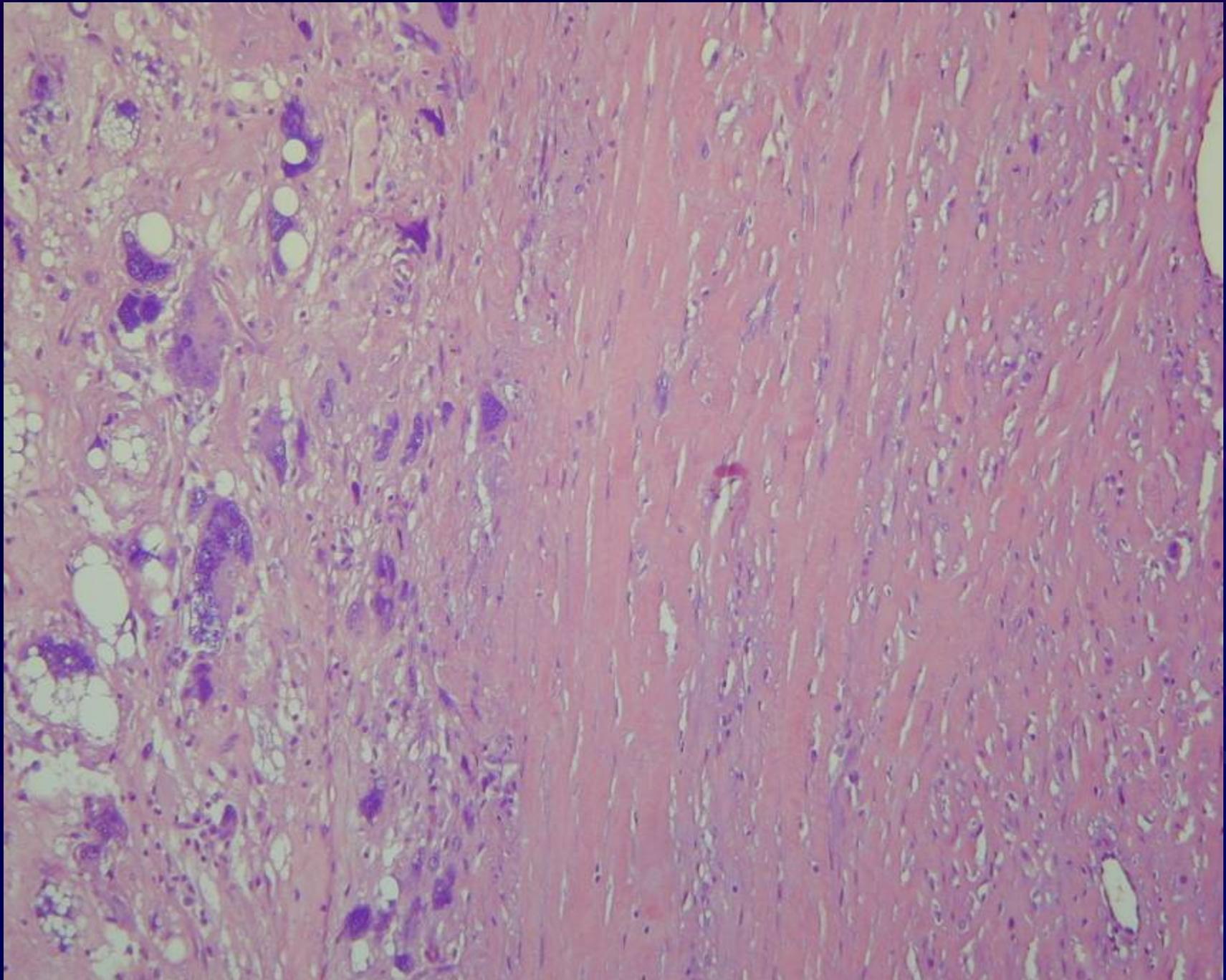


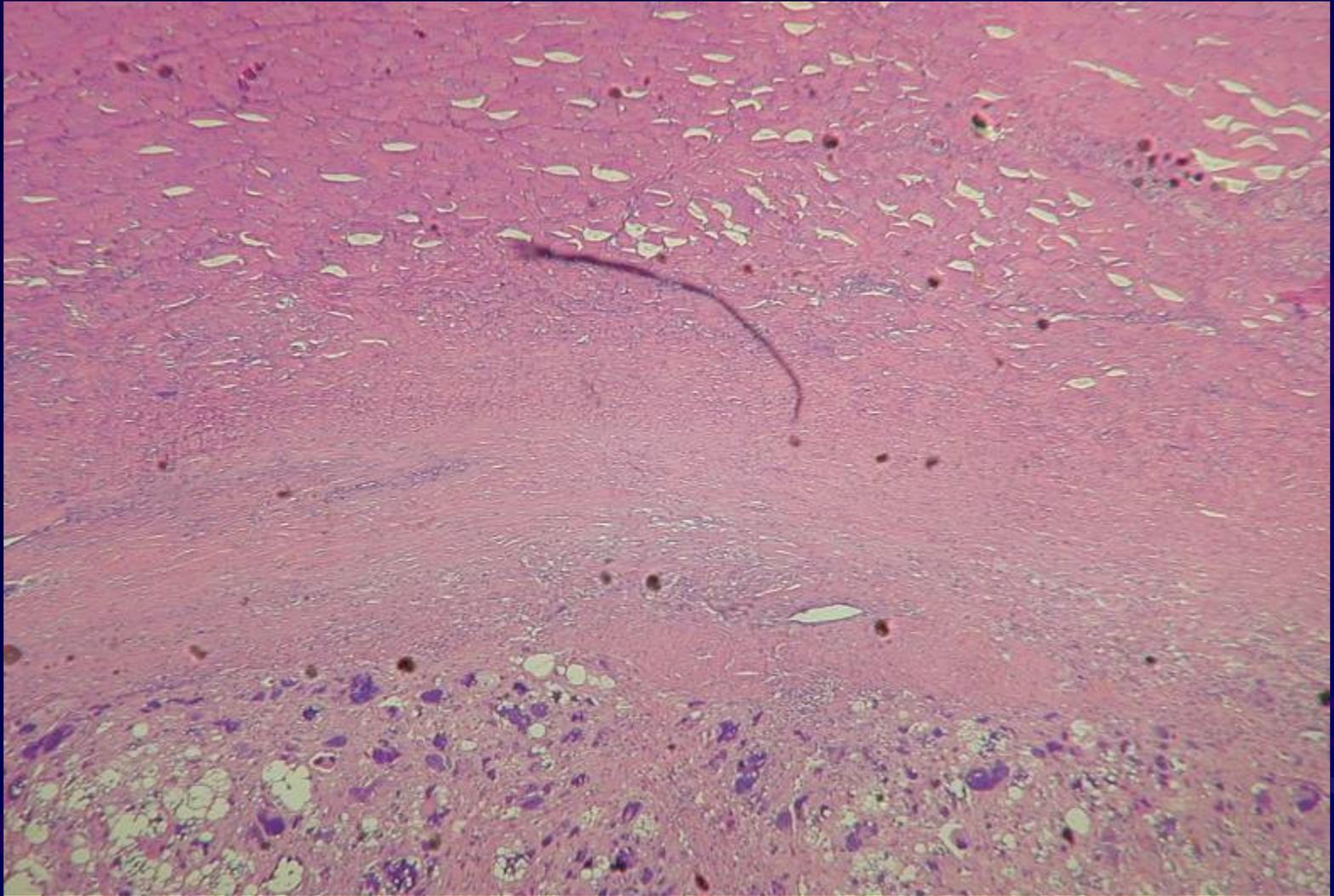
Reactive Zone or Pseudocapsule
Tumor Compressing Muscle and Infiltrating between
Muscle Fibers



High Grade Sarcoma After Good Response to Chemotherapy: the Pseudocapsule is Converted to a True Fibrous Capsule







Local Growth of Sarcomas

- Local growth obeys fascial borders/compartmental borders
- Fascial borders resist tumor penetration
- Compartment refers to the bone or muscle of origin; muscle compartment surrounded by fascia (investing fascia on all sides—resists tumor penetration)
- When a bone tumor destroys the cortex and spreads into the surrounding soft tissue---extracompartmental

Growth and Behavior

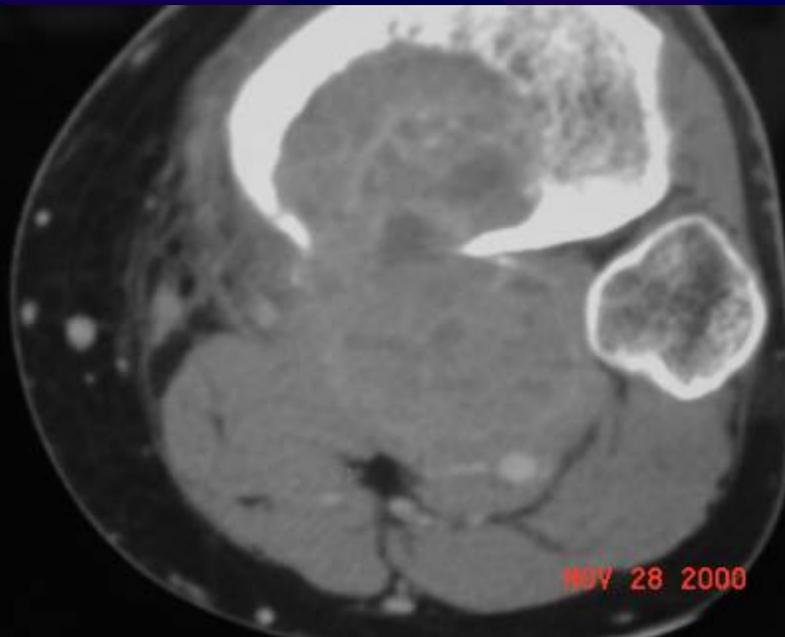
- Bone Tumors that extend extracompartmental compress the surrounding muscles into a pseudocapsule
(the fascia of the surrounding muscle usually contains the tumor and protects other muscles and structures)
 - Distal Femur: Vastus Intermedius
 - Proximal Tibia: Popliteus
 - Proximal Humerus: Subscapularis
 - Scapula: Rotator Cuff

Growth and Behavior

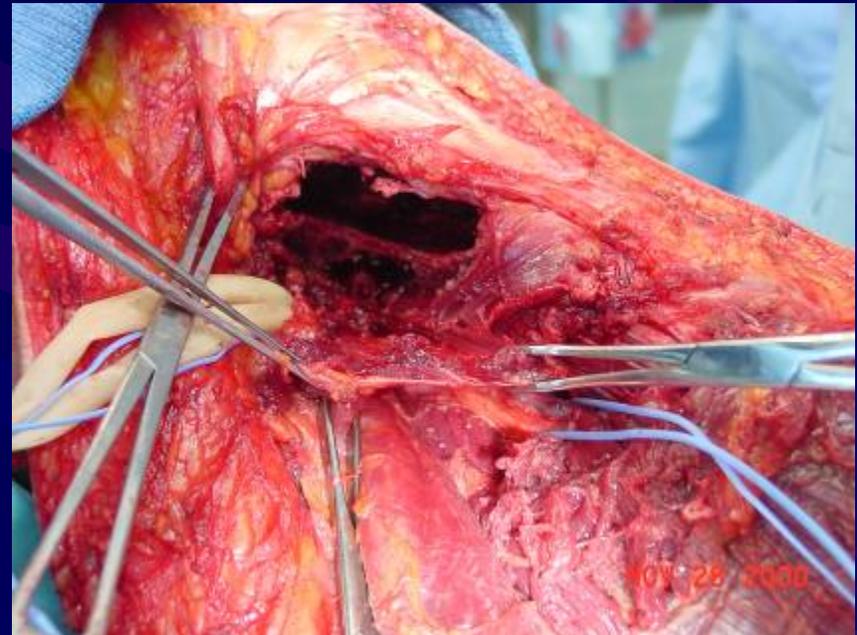
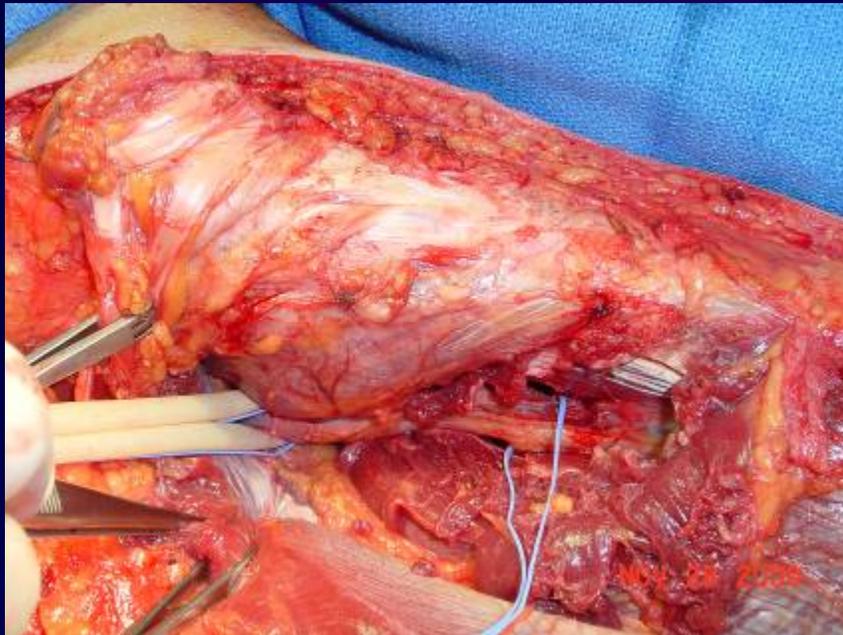
- Soft Tissue Sarcomas
 - Intramuscular---if extends beyond fascia—extracompartmental
 - Intermuscular—extracompartmental

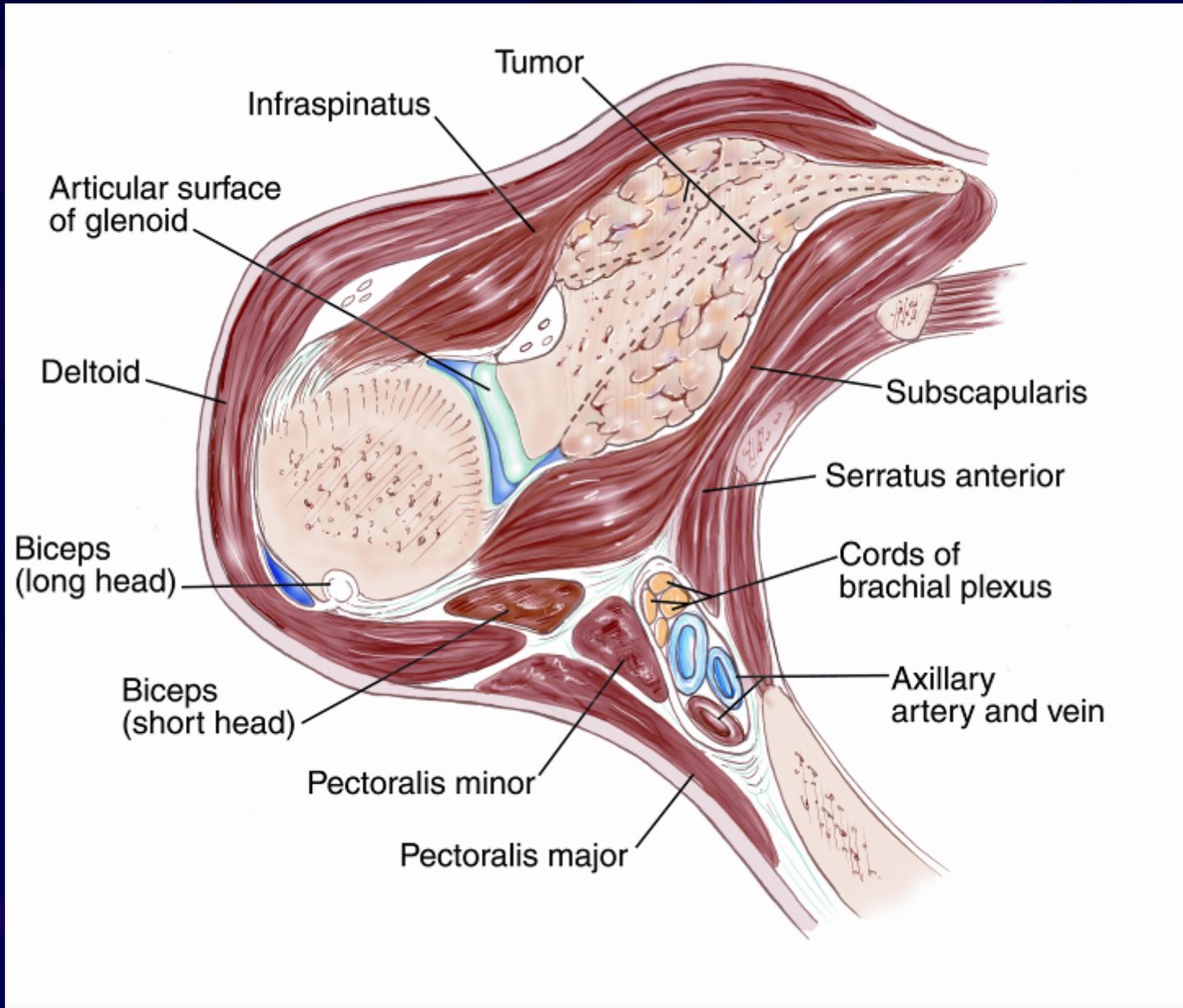


GCT Proximal Tibia

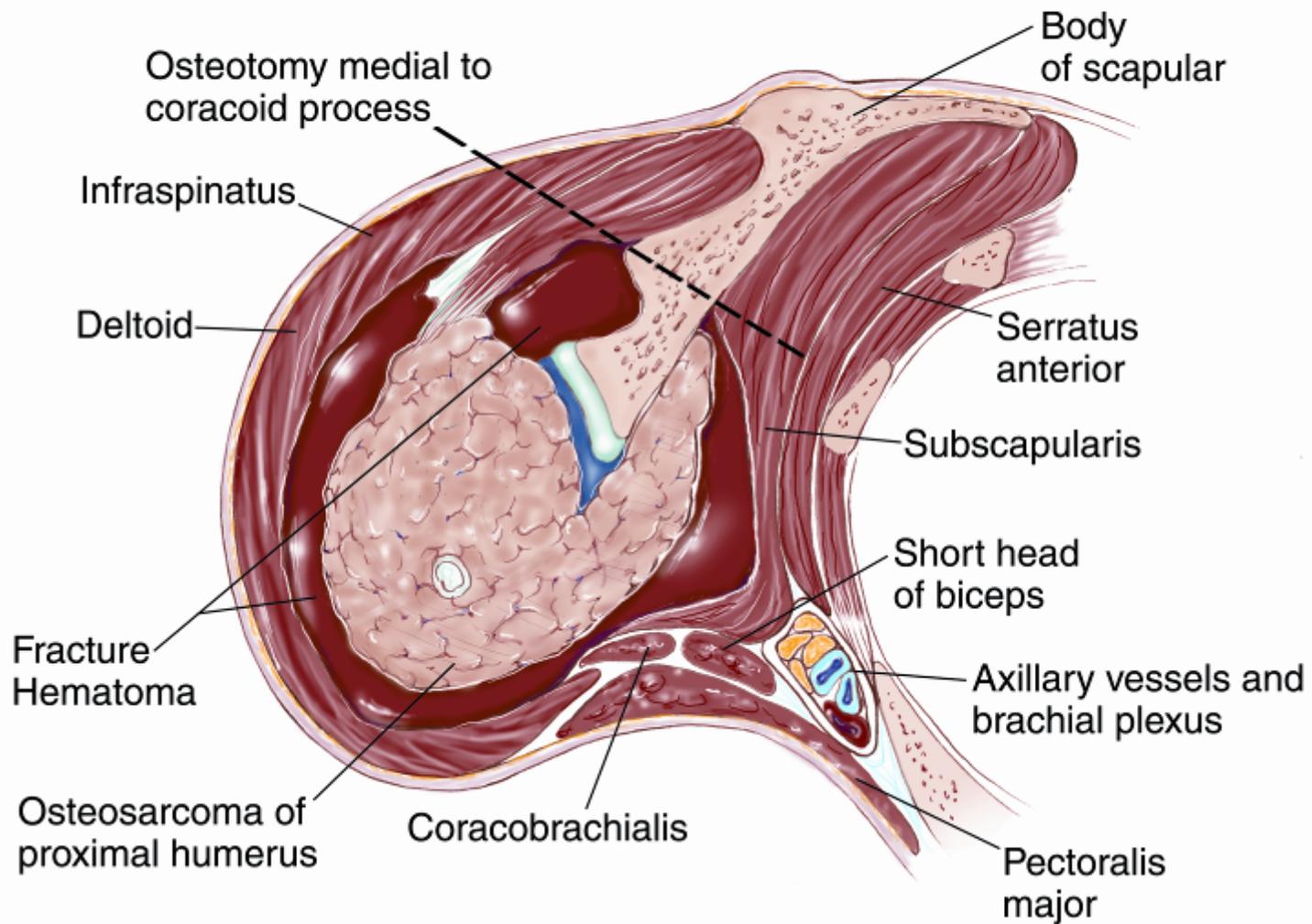


Popliteus (Pseudocapsule)





LOCAL SPREAD OF HEMATOMA SECONDARY TO PATHOLOGICAL FRACTURE



Staging

- Purpose
 - Determine tumor type
 - Determine prognosis
 - Guide treatment
 - Compare results between study groups
 - Delineate extent of local and distant disease

Staging Studies

- Plain Radiograph
- MRI
- CT scan
- Chest CT
- Bone Scan

Plain Radiographs

- Rate of tumor growth
- Tumor interaction with surrounding non-neoplastic tissue
- Internal composition of tumor

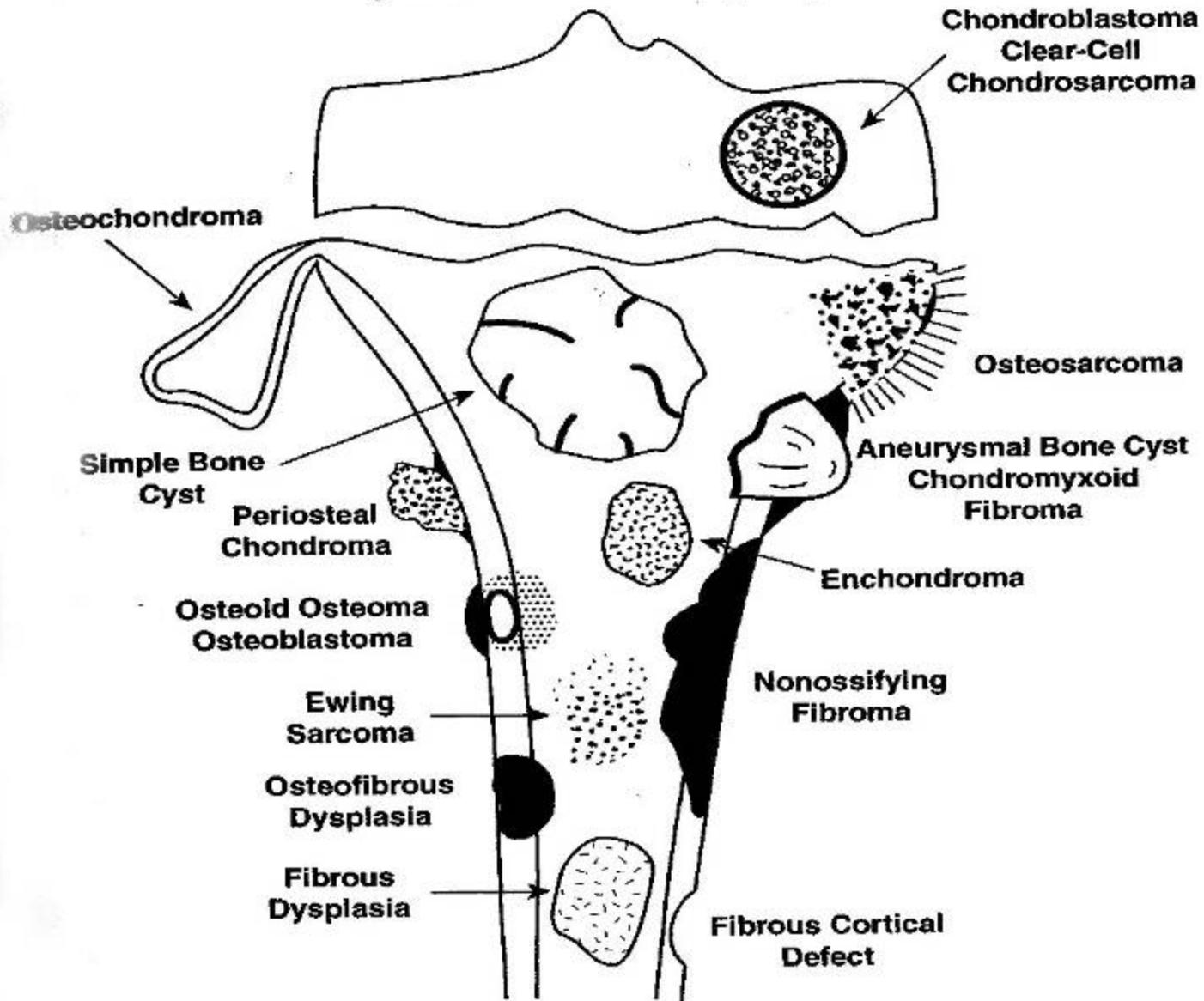
Plain Radiographs

- Bone involved
- Is involved bone normal?
- What part of the bone?
- Open or closed growth plate
- Epicenter of lesion (cortex or medullary canal)
- Tumor contour and zone of transition between tumor and host bone

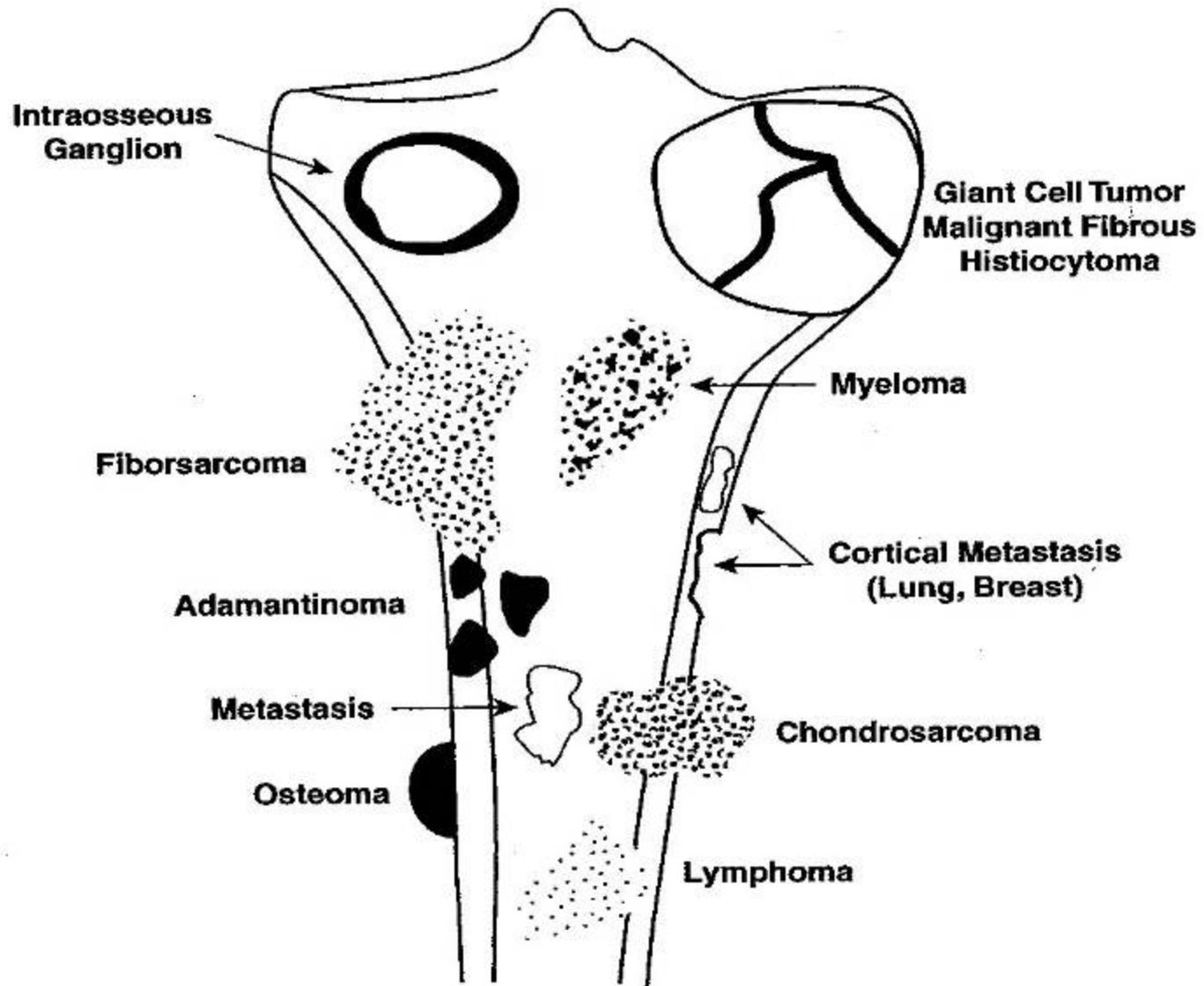
Plain Radiographs

- Mineralized matrix?
- Cortical destruction?
- Periosteal reaction? What type
- Involvement of joint space?
- Tumor multifocal?
- Is tumor of uniform appearance or does it have several different components?

Immature Skeleton (Growth Plate Open)



Mature Skeleton (Growth Plate Closed)



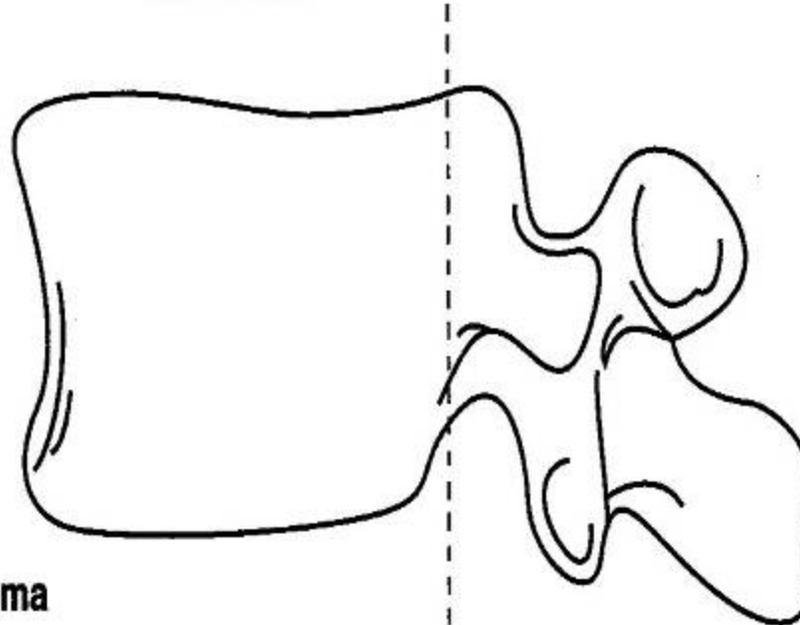
Lymphoma
Hodgkin
Myeloma
Ewing
Osteosarcoma
Chondrosarcoma
Metastasis

Exceptions:

Hemangioma
Langerhans-cell Granuloma
Fibrous Dysplasia

MALIGNANT

BENIGN

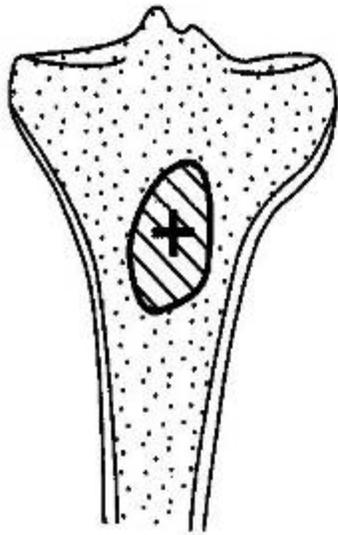


Osteblastoma
Osteoid Osteoma
Aneurysmal Bone Cyst
Osteochondroma
Chondromyxoid Fibroma

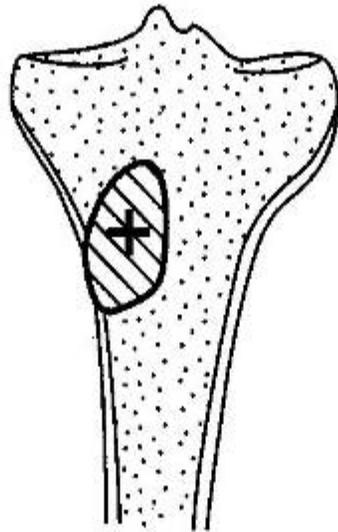
Anterior

Posterior

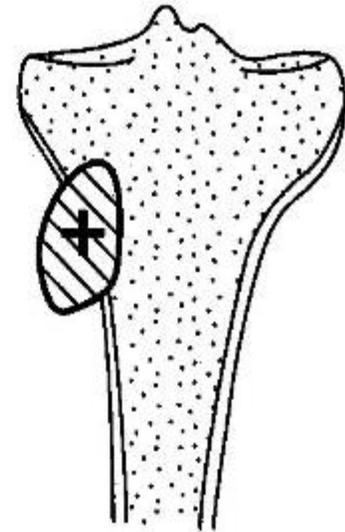
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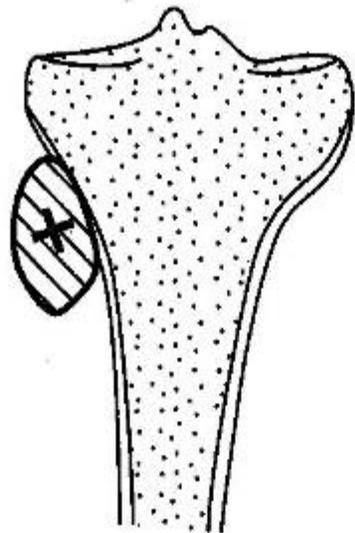
Central



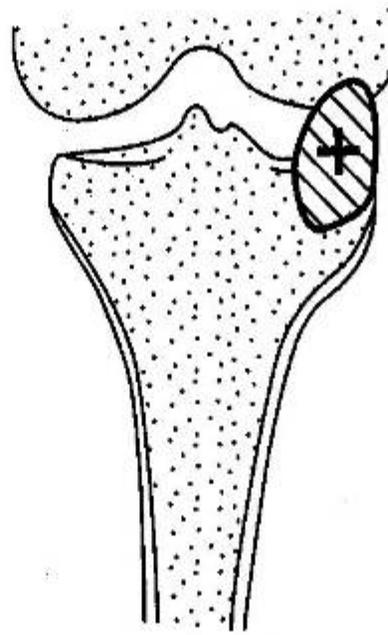
Eccentric



Cortical

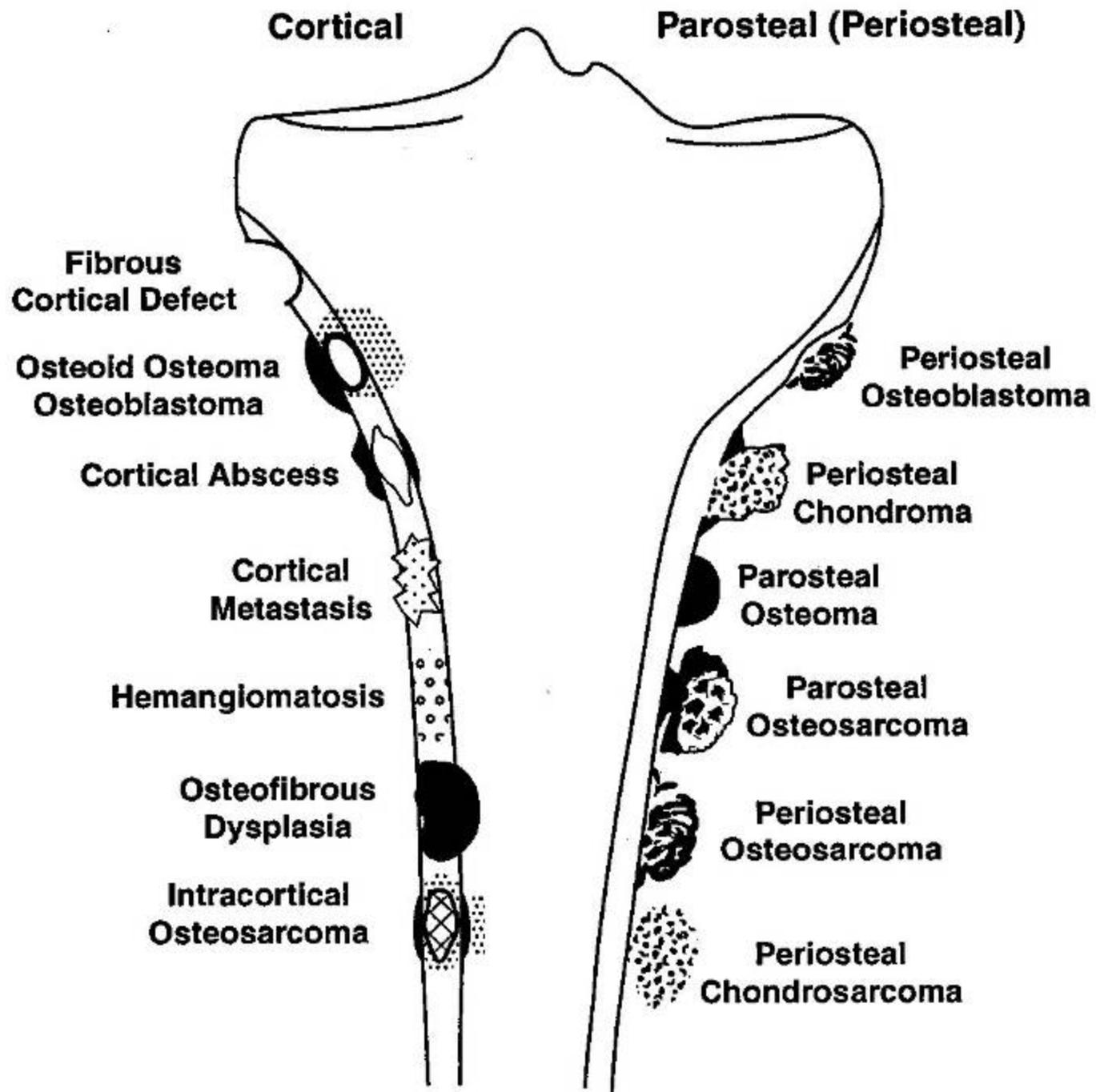


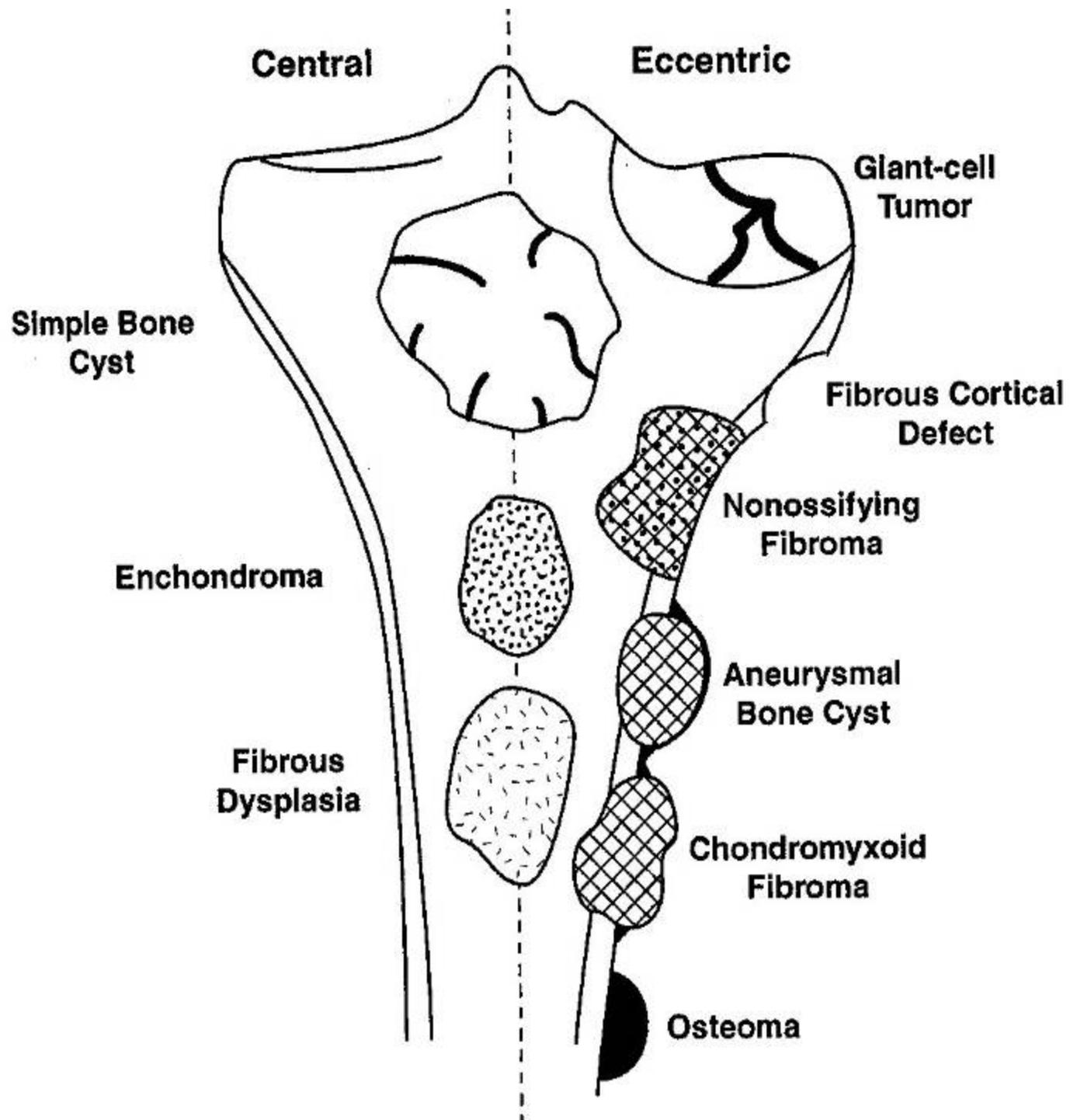
Parosteal (periosteal)

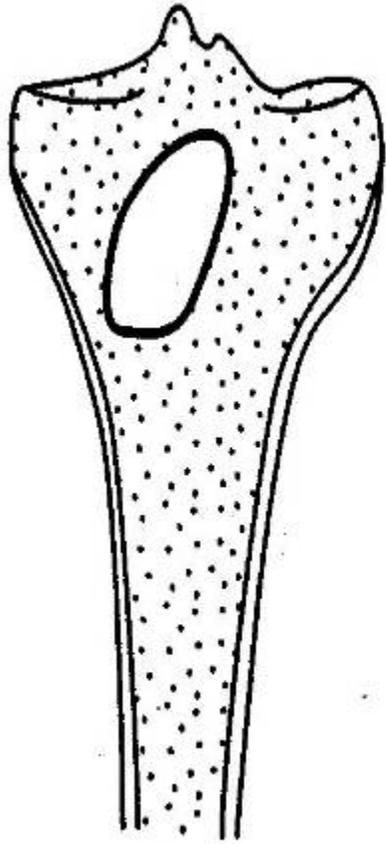


Intra-articular

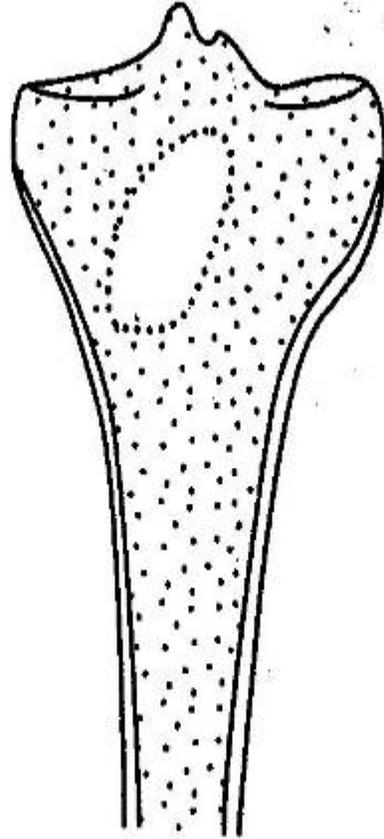
A



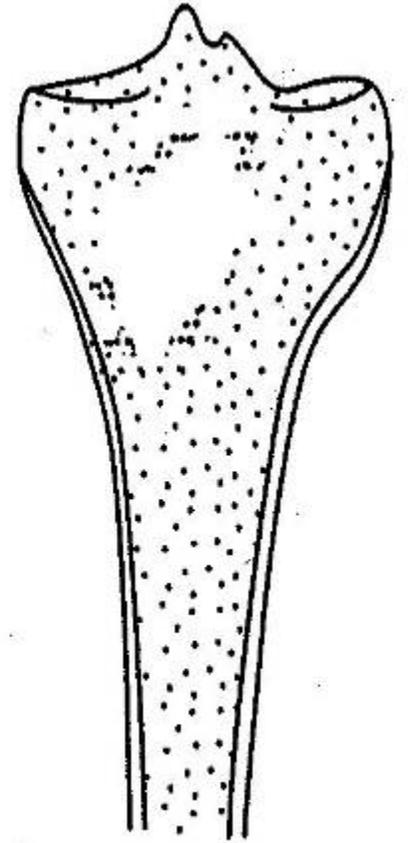




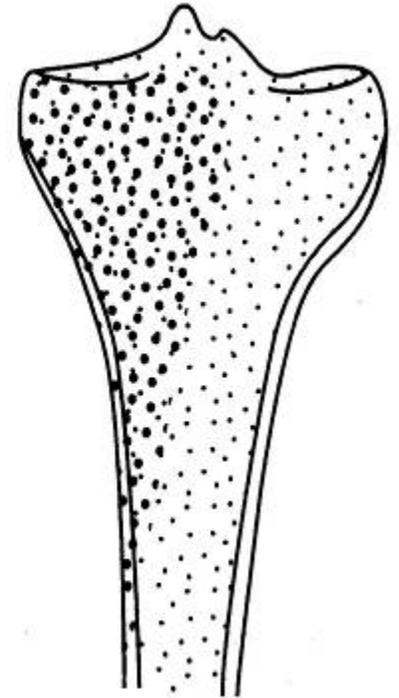
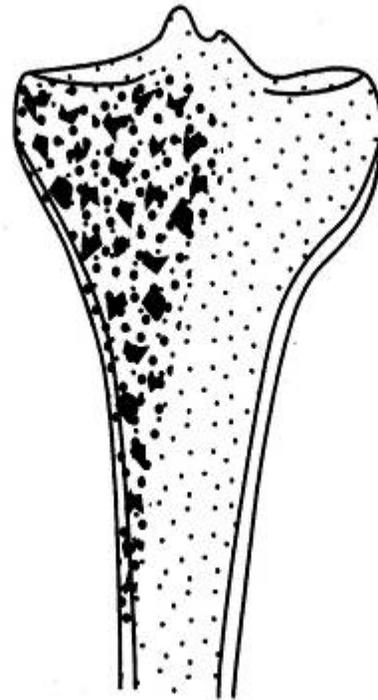
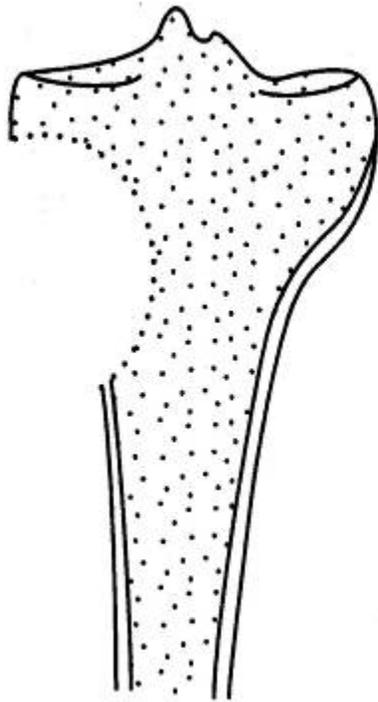
1A



1B



1C



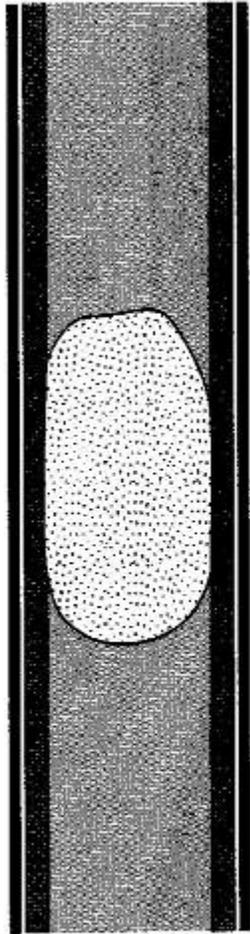
A Geographic

Motheaten

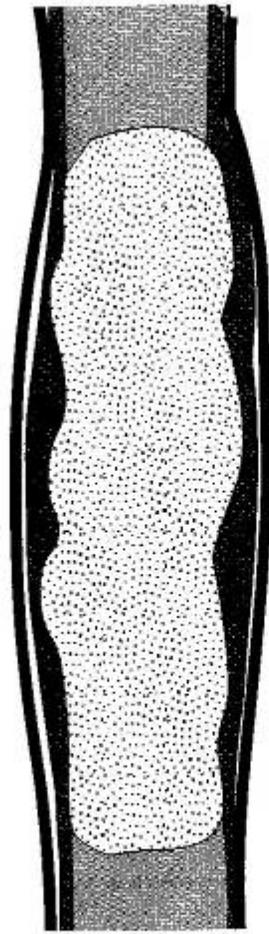
Permeative

SIMPLIFIED RADIOLOGIC GRADING OF BONE TUMORS*

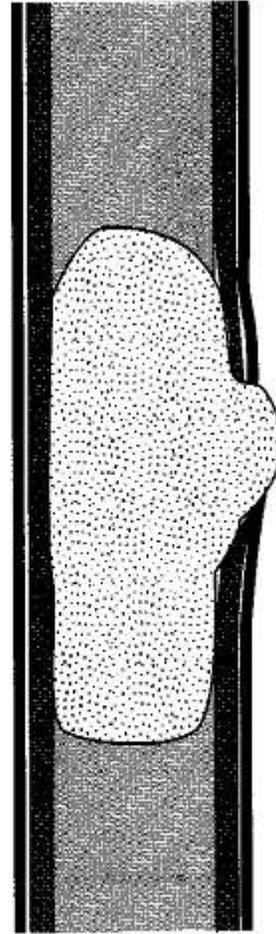
Grade	Radiologic Features
Low grade, nonaggressive	Geographic destruction with sclerotic rim
Medium grade, moderately aggressive	Geographic destruction, no sclerotic rim, and/or cortex "expanded" more than 1 cm or completely penetrated
High grade, very aggressive	Moth-eaten and/or permeative destruction only



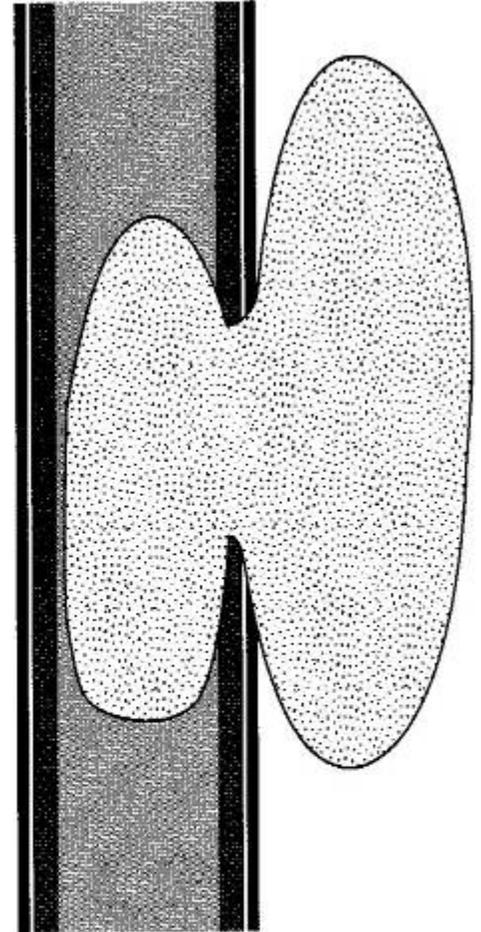
Intact cortex



Scalloped cortex

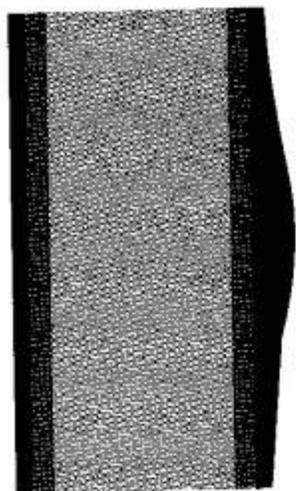


Complete cortical
disruption

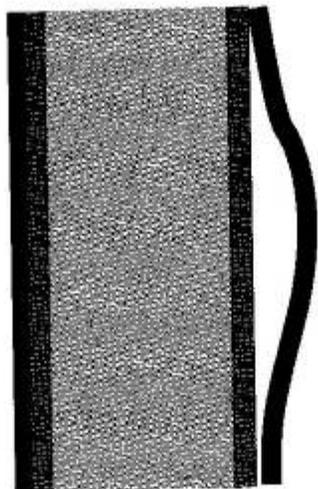


Dumbbell
Configuration

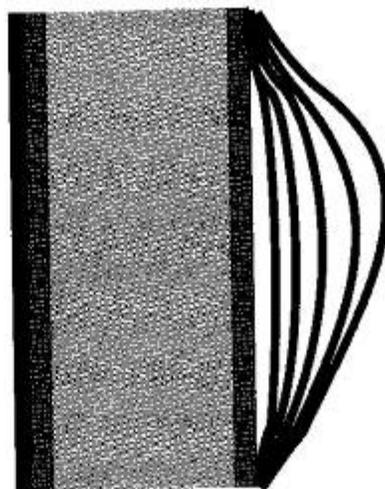
Continuous



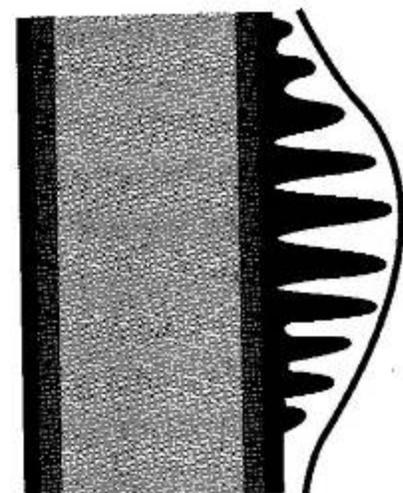
Solid



Single lamellar

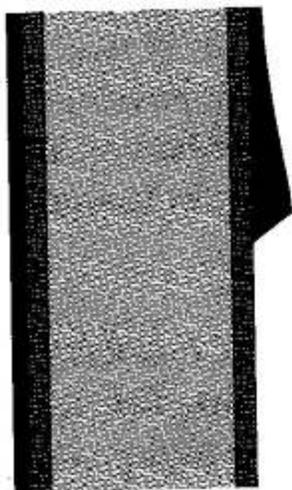


Onion-skin

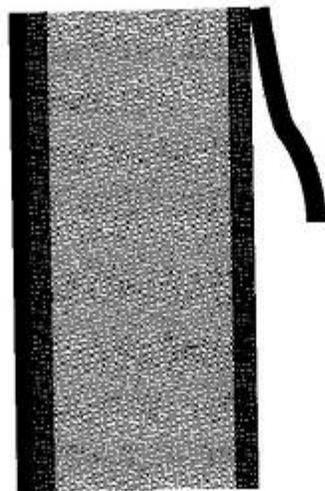


Spiculated

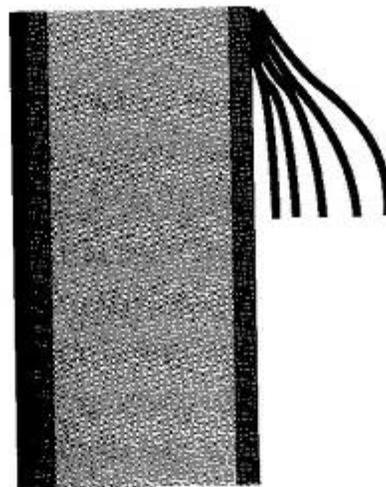
Interrupted



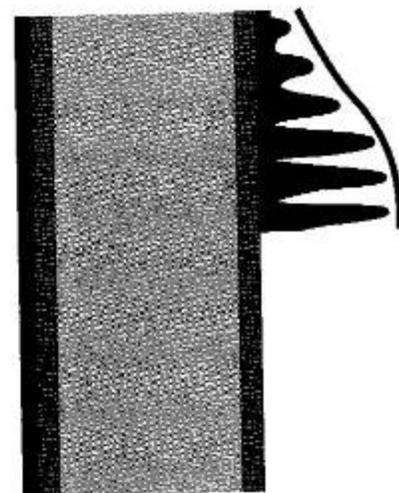
Buttress



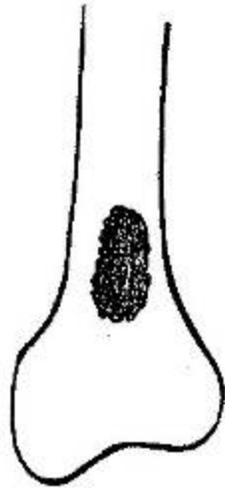
Codman's triangle



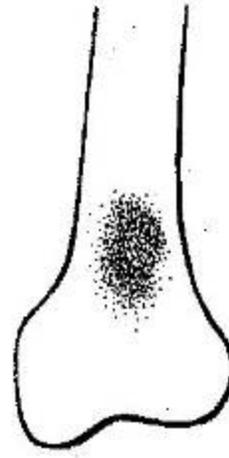
Lamellar



Spiculated



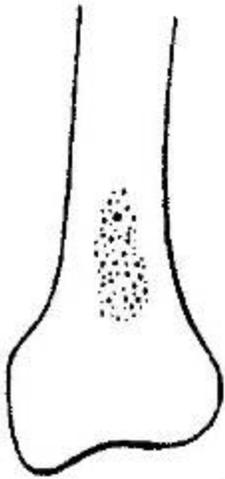
Solid



Cloud-Like



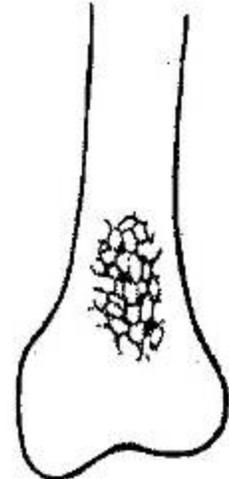
Ivory-Like



Stippled



Flocculent

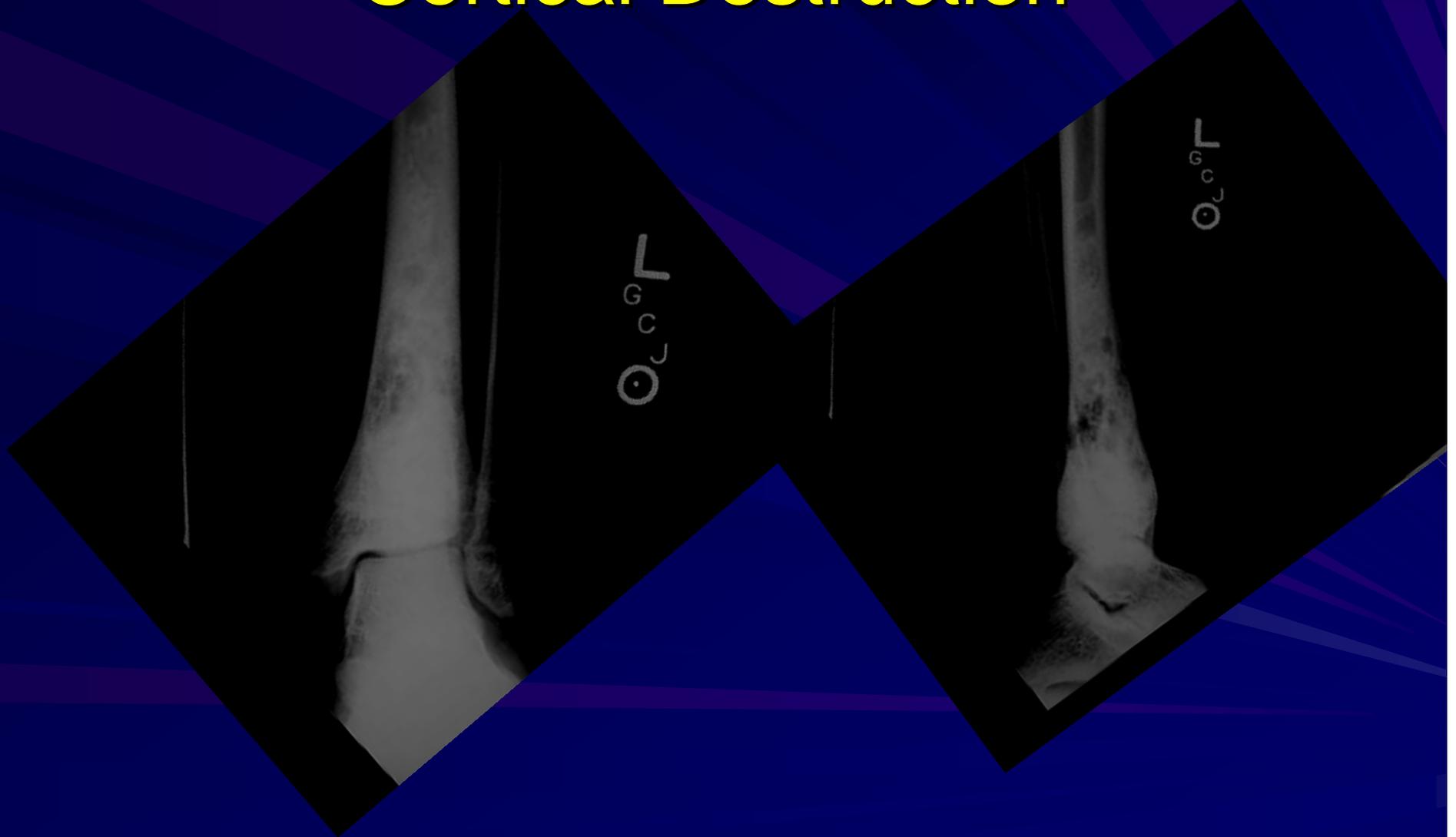


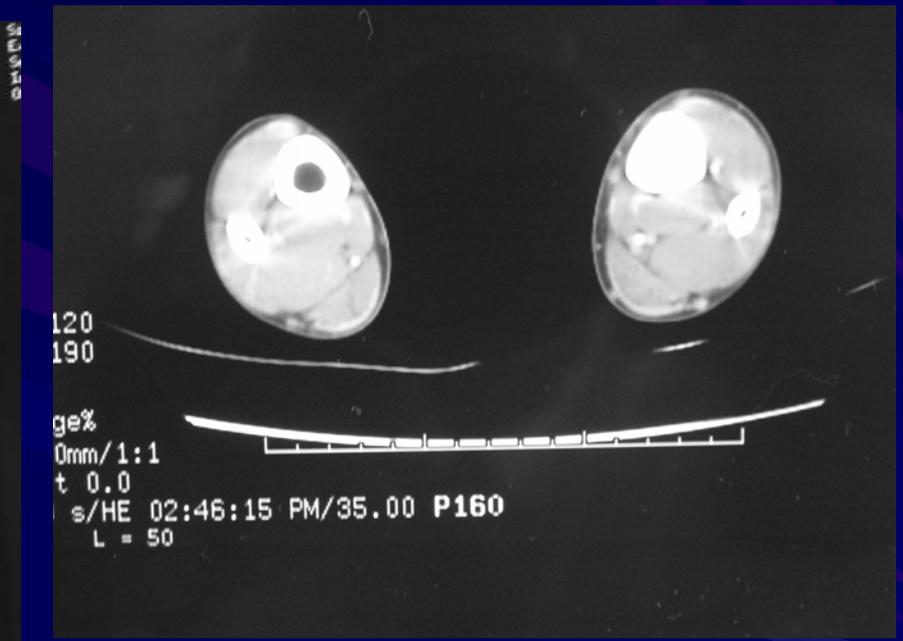
Rings and Arcs

Geographic



Permeative with Mineralization and Cortical Destruction





Geographioc



Permeative with Calcifications in a Ring and Arc-like Manner



Geographic Lesion



Permeative with Cortical Destruction and Soft Tissue Mass



Hair on End and Codman's Triangle Periosteal Reactions



Geographic Lesion Intracortical Continuous Periosteal Reaction/Cortical Thickening



Geographic Lesion



Geographic Lesion



2 DFOV 14.4cm
STND

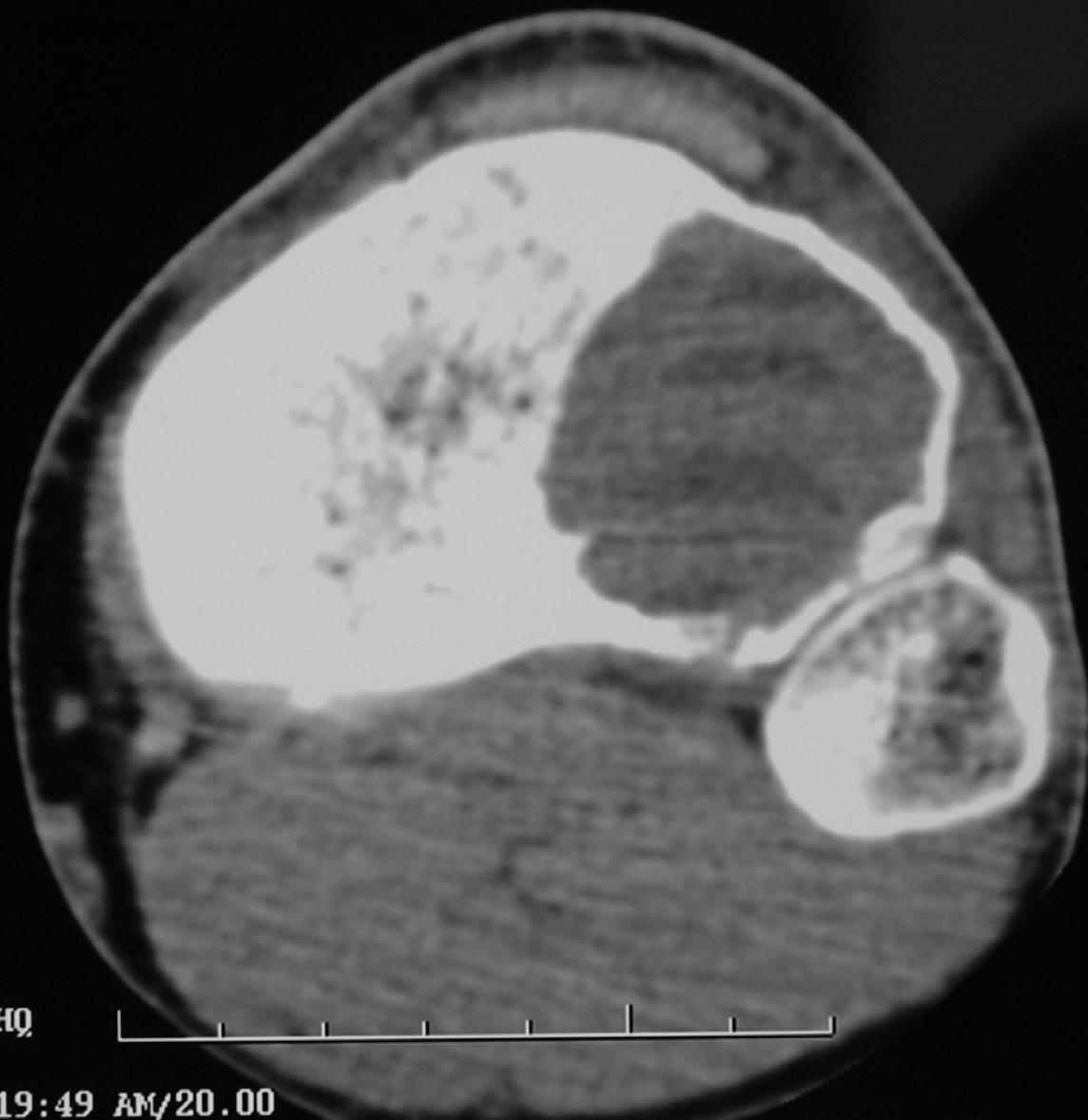
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Permeative Lesion with Ossification, Cortical Destruction and Codman's Triangle



MRI

- Evaluates entire bone and adjacent joint
- Best test for intraosseous extent and soft tissue extent
- Skip mets
- Proximity to vascular structures
- Occasionally helpful in diagnosis of bone or soft tissue tumors (experienced radiologist)

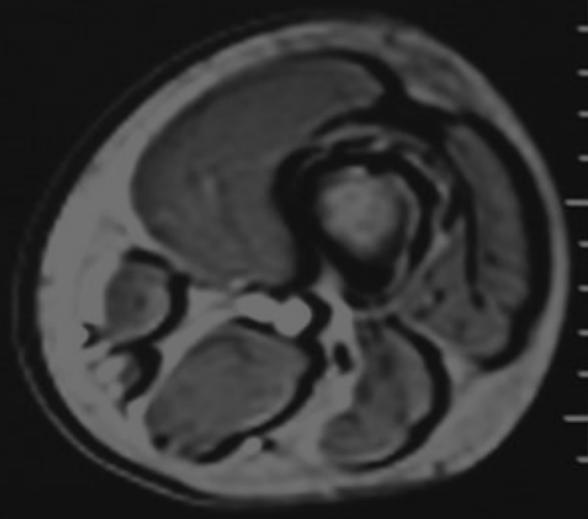
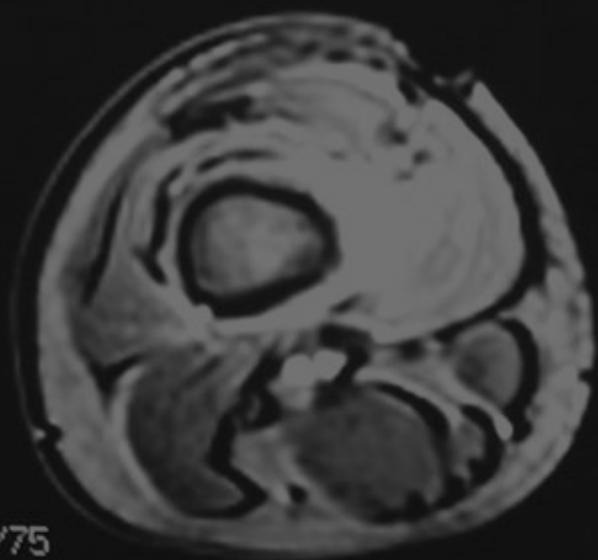




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BODY
FOV:35x35

CT

- Good for evaluating cortical details and destruction
- Subtle cortical erosions (endosteal;periosteal)
- Calcifications / ossification

2 DFOV 14.4cm
STND

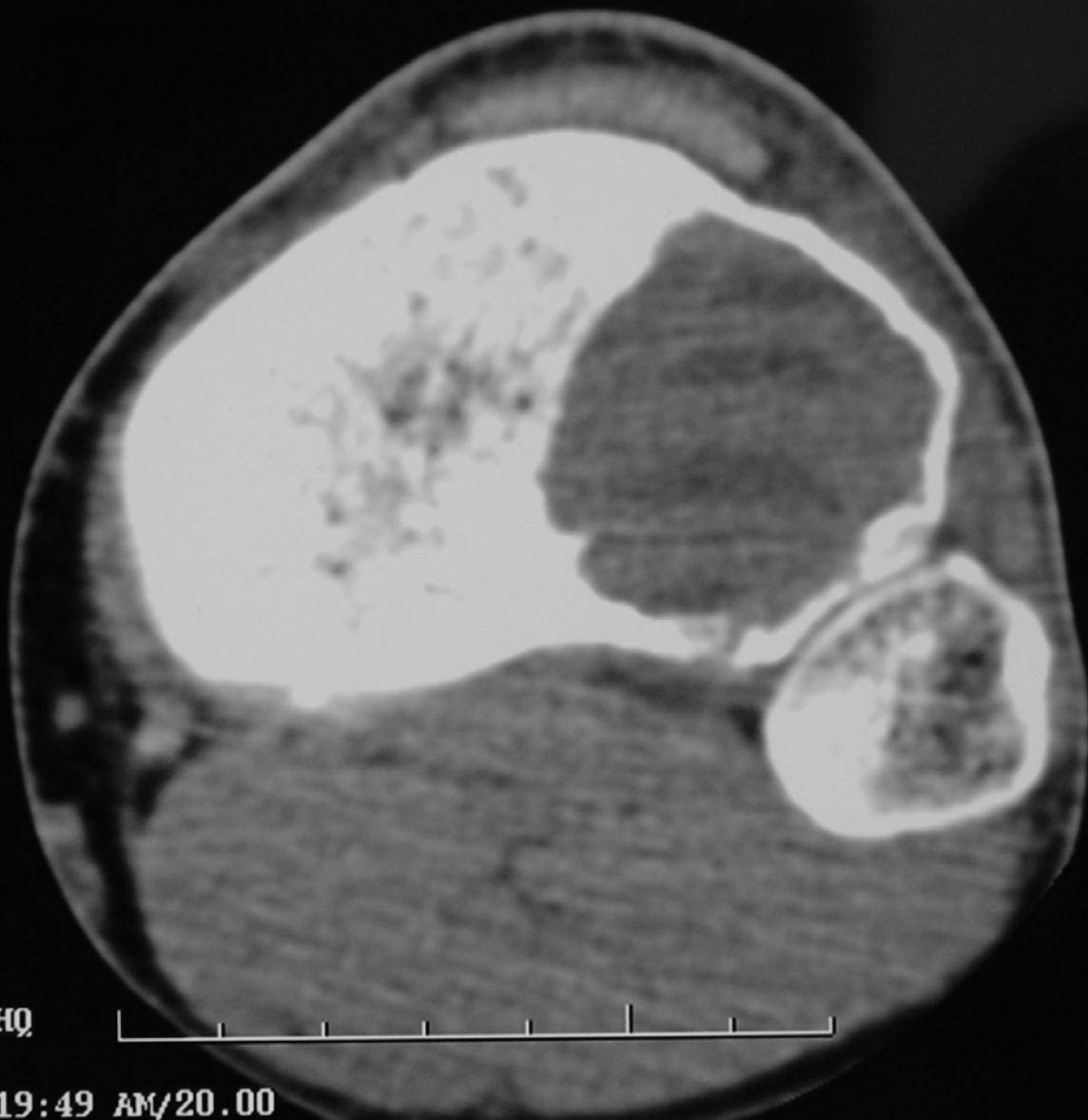
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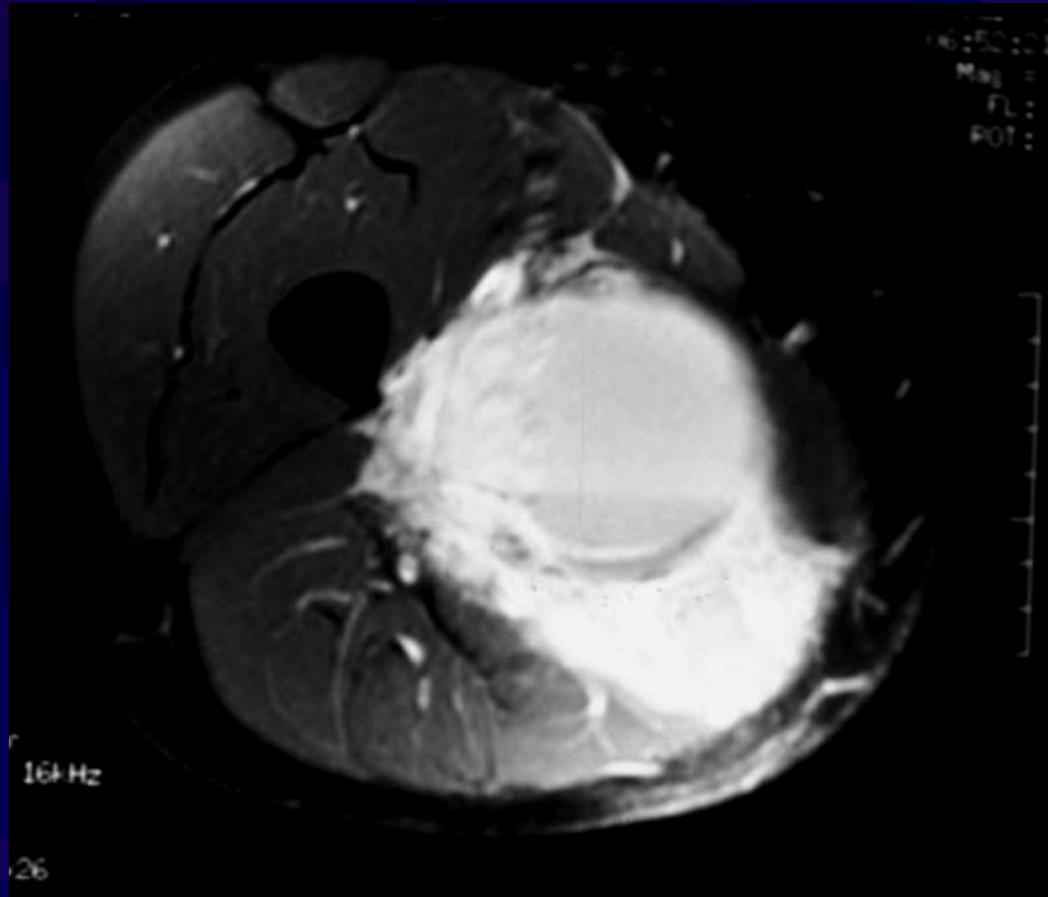
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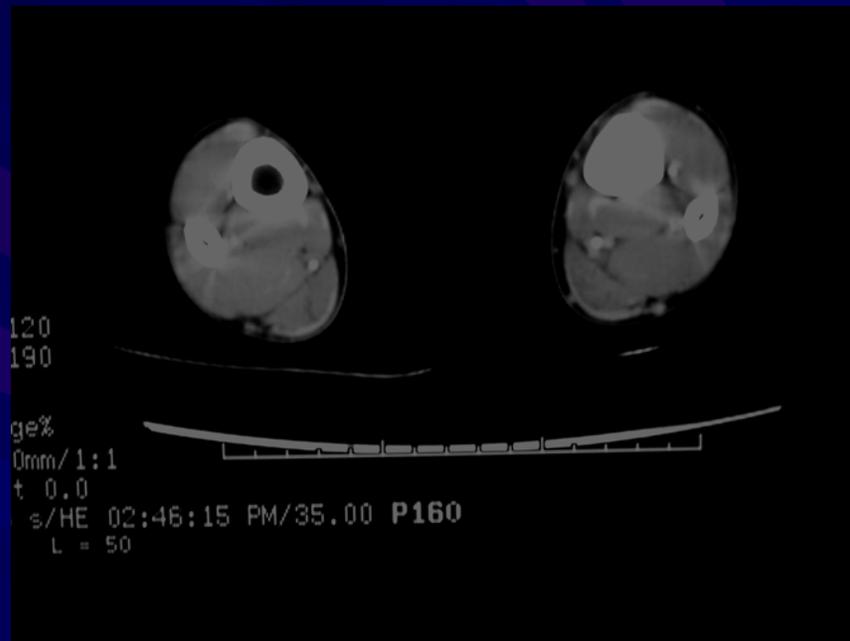


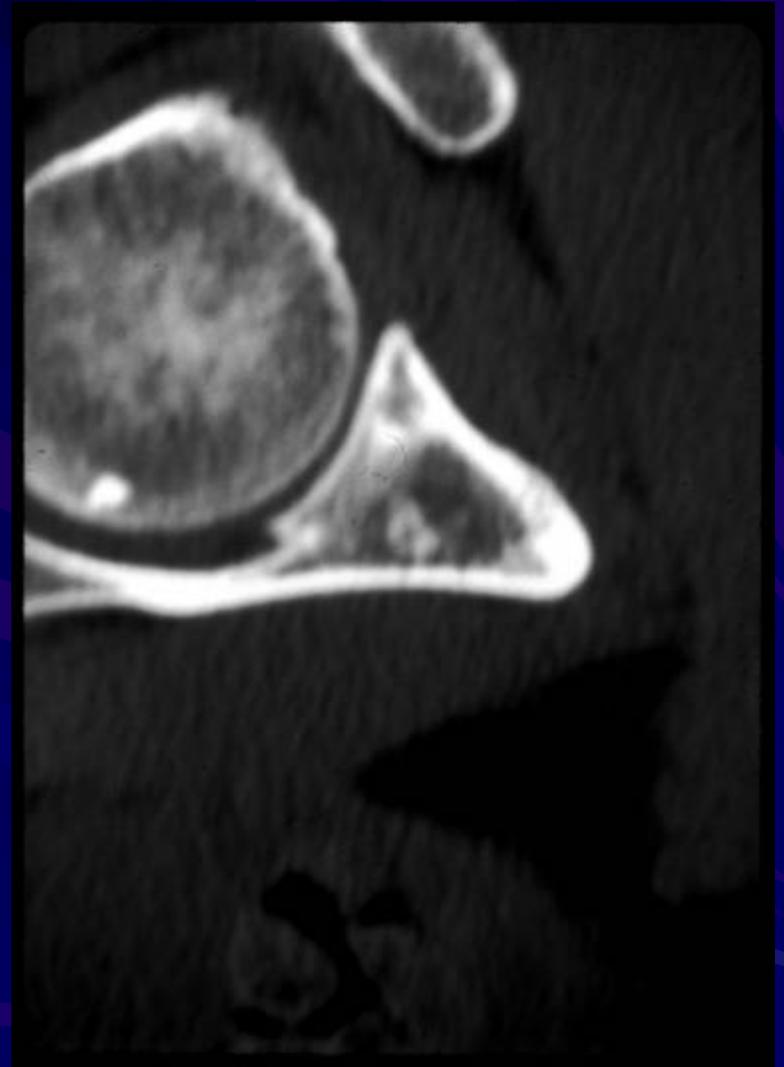
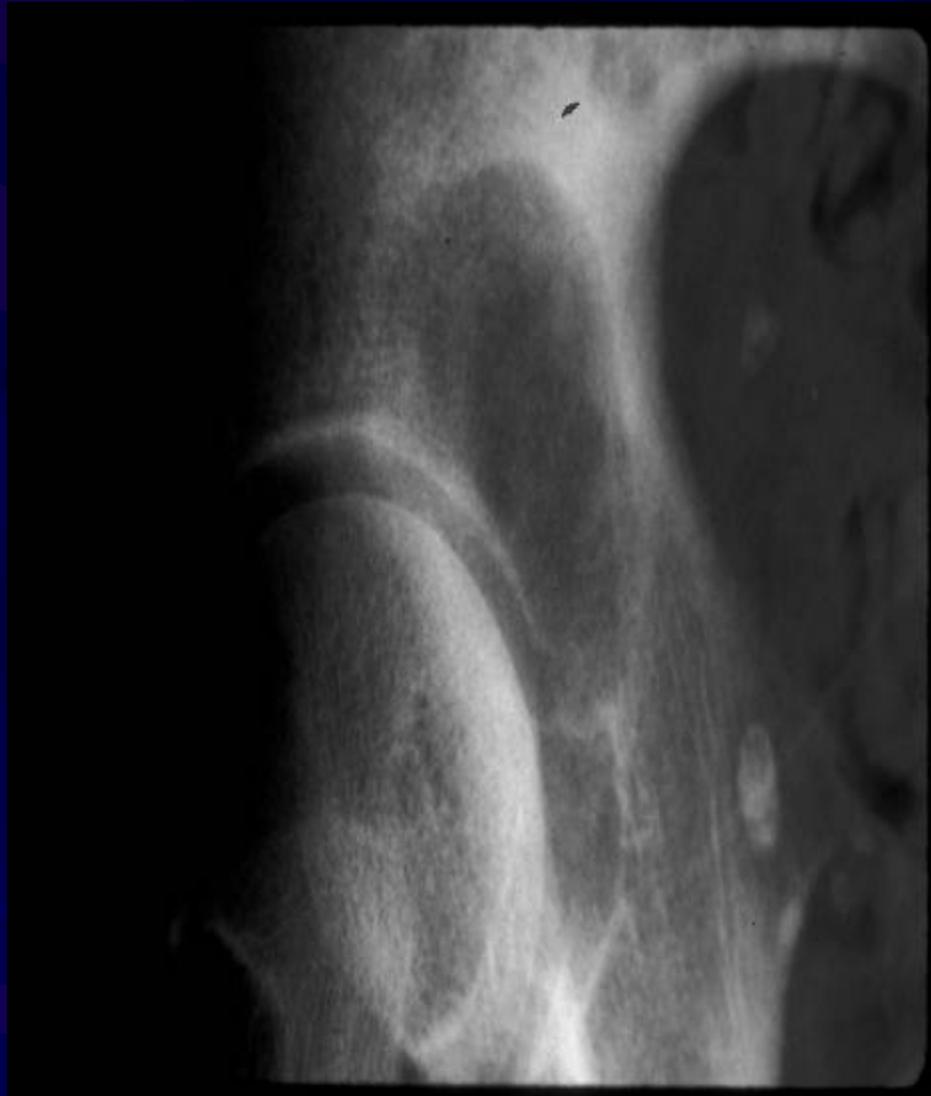
Fluid-Fluid Levels: Aneurysmal Bone Cyst Changes



Soft Tissue Extent and Fluid-Fluid Level







Bone Scan

- Whole body bone scan
- Sites of bony mets
- Active lesion??

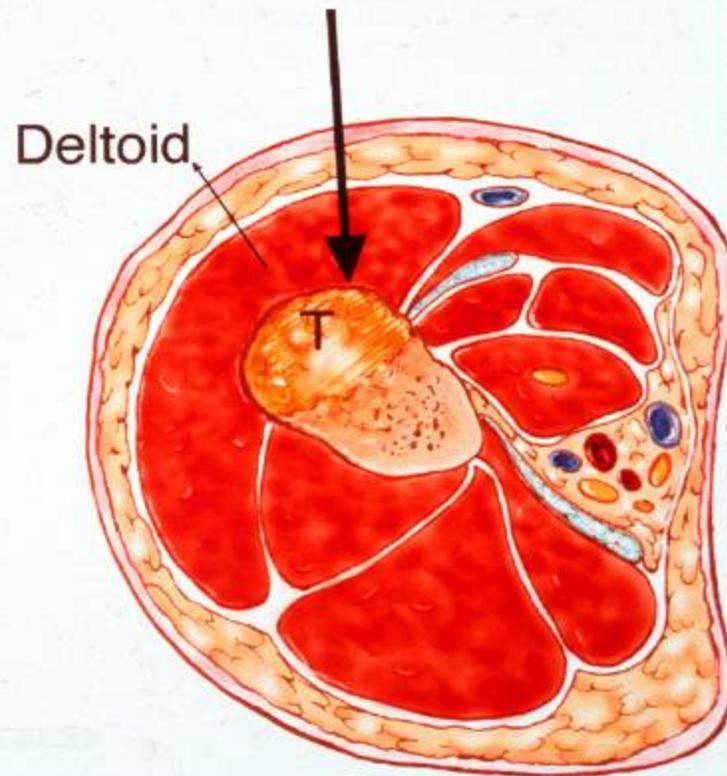
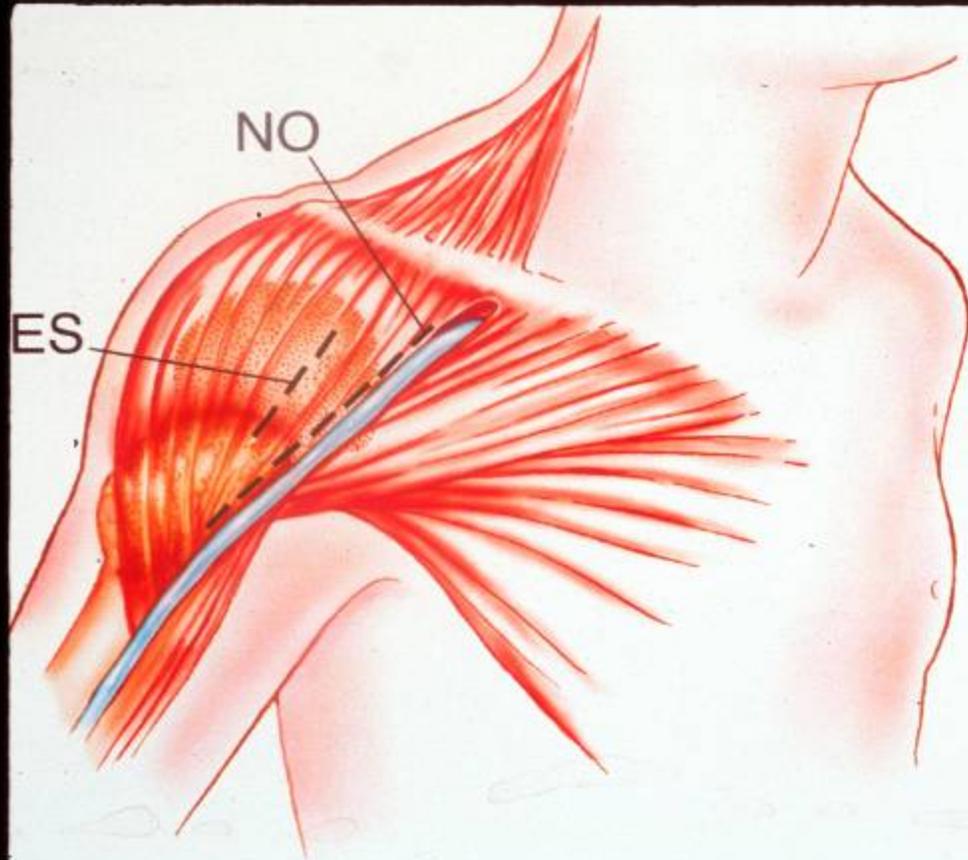
Chest CT

- Presence of metastatic disease



Biopsy

- CT guided or Open
- Through one compartment'
- Avoid neurovascular structures
- Biopsy soft tissue component
- Biopsy by surgeon who will perform procedure or by radiologist after communication with surgeon
- Tumors with necrosis and hemnorrhage



Staging

Benign Staging System (Enneking)

Stage 1: Latent

Grow slowly with growth of individual and then stop; tendency to heal spontaneously
(ex. NOF; UBC)

Stage 2: Active

Progressive growth

Stage 3: Aggressive

Staging

Malignant Bone Tumors

TNM Staging System (AJC)

<u>Stage</u>	<u>Grade</u>	<u>Tumor</u>	<u>Node</u>	<u>Mets</u>
IA	G1,2	T1	N 0	M 0
IB	G1,2	T2	N 0	M 0
IIA	G3,4	T1	N 0	M 0
IIB	G3,4	T2	N 0	M 0
III	Undefined for bone tumors			
IVA	Any G	Any T	N1	M 0
IVB	Any G	Any T	Any N	M 1

Staging

Enneking Staging System Malignant Bone Tumors

<u>Stage</u>	<u>Grade</u>	<u>Site</u>
IA	G1	T1
IB	G1	T2
IIA	G2	T1
IIB	G2	T2
III	Mets	Mets

(based on biological behavior)

Grading

- Biological Behavior / Natural History

G1

LG Chondrosarcoma
Chondrosarcoma

Secondary Chondrosarc
Osteosarcoma

Parosteal Osteosarcoma
Sarcoma/PNET

Adamantinoma

G2

High Grade

Conventional

Ewing's

MFH

Angiosarcoma

Staging

- Soft Tissue Sarcomas
- Important Prognostic Characteristics
 - Tumor Size (>5cm, worse prognosis)
 - Tumor Depth (Deep, worse prognosis)
 - Grade (High grade, worse prognosis)
 - Presence of Mets

Staging

Malignant Tumors

TNM Staging System (AJC)

Stage	Grade	Tumor	Node	Mets
IA	G1,2	T1a-b	N 0	M 0
IB	G1,2	T2a	N 0	M 0
IIA	G1,2	T2b	N 0	M 0
IIB	G3,4	T1a-b	N 0	M 0
IIC	G3-4	T2a	N 0	M 0
III	G3,4	T2b	N 0	M 0
IVA	Any G	Any T	N1	M 0
IVB	Any G	Any T	Any N	M 1

Grading

- Soft Tissue Sarcomas (Biological Behavior)
 - Tumors that are definitionally high grade
 - Ewing's Sarcoma
 - PNET
 - Rhabdomyosarcoma
 - Angiosarcoma
 - Pleomorphic Liposarcoma
 - Soft Tissue Osteosarcoma
 - Mesenchymal Chondrosarcoma

Grading

- Soft Tissue Sarcomas (Biological Behavior)
 - Tumors that are definitionally low grade
 - Well Differentiated Liposarcoma
 - Dermatofibrosarcoma Protuberans
 - Infantile Fibrosarcoma
 - Angiomatoid MFH

Grading

- Soft Tissue Sarcomas
 - Tumors not gradable but which metastasize often
 - Alveolar soft part sarcoma
 - Clear cell sarcoma
 - Epithelioid sarcoma
 - Synovial sarcoma
 - Low grade fibromyxoid sarcoma

Grading

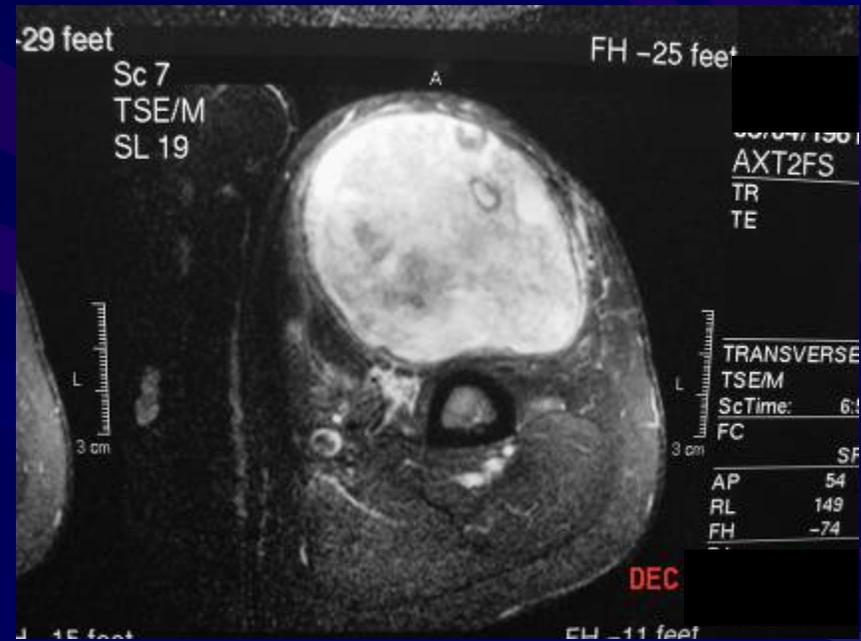
- Soft Tissue Sarcomas
 - Tumors of varying behavior—grading may be useful
 - Myxoid liposarcoma
 - Leiomyosarcoma
 - MPNST
 - Fibrosarcoma
 - Myxoid MFH

Grading

- Soft Tissue Sarcomas
 - Tumors of varying behavior—grading parameters not yet established
 - Hemangiopericytoma
 - Myxoid chondrosarcoma
 - Malignant granular cell tumor
 - Malignant mesenchymoma

Evaluating Response to Chemotherapy

Sarcoma of Biceps



Pseudocapsule after Chemotherapy

