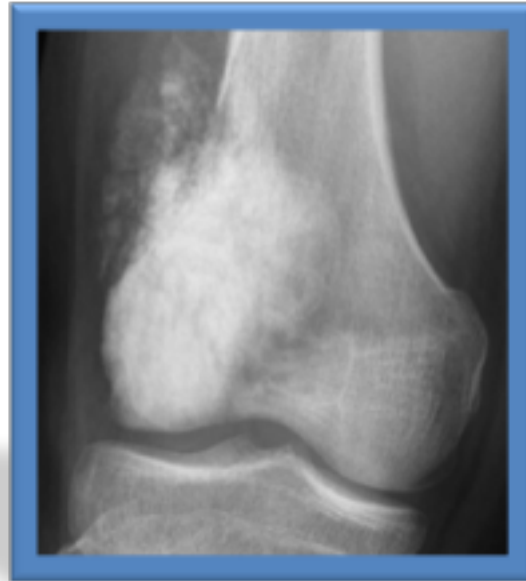


Podcast (Video Recorded Lecture Series):
Bone Tumors, Sarcomas for the USMLE Step One Exam



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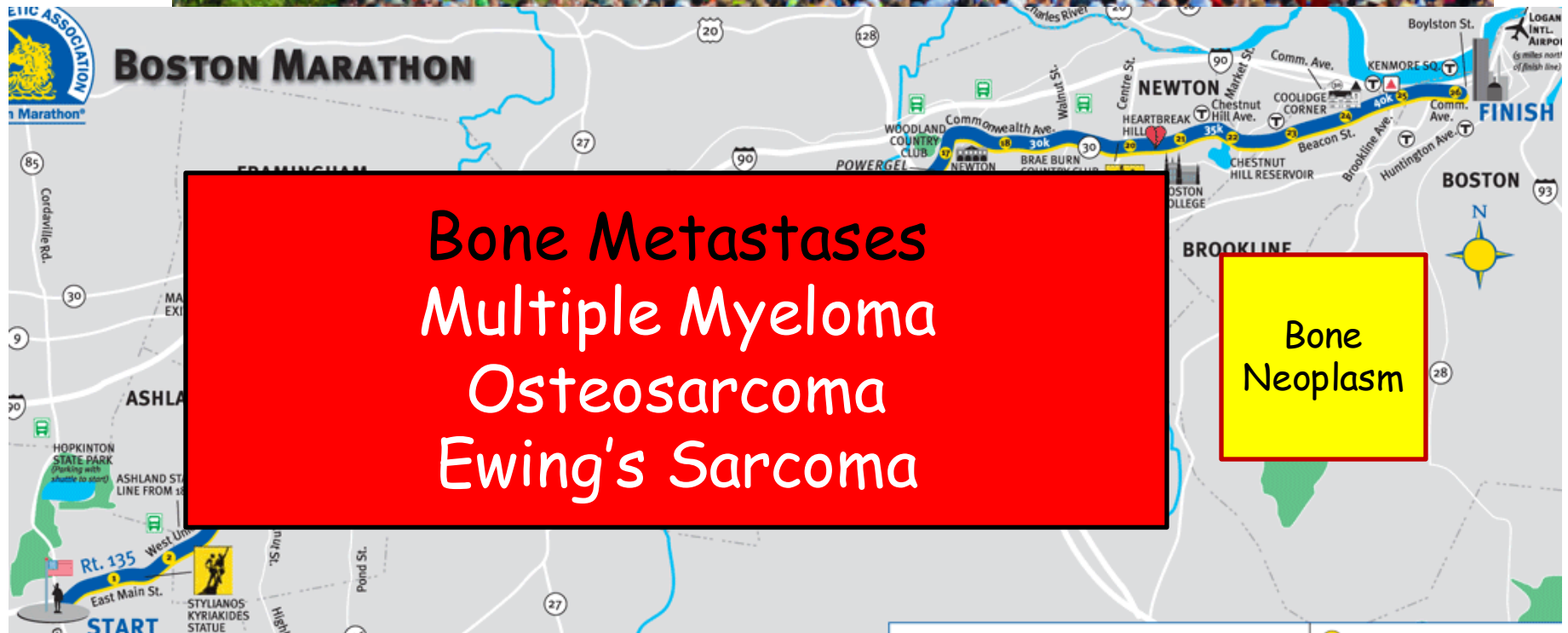
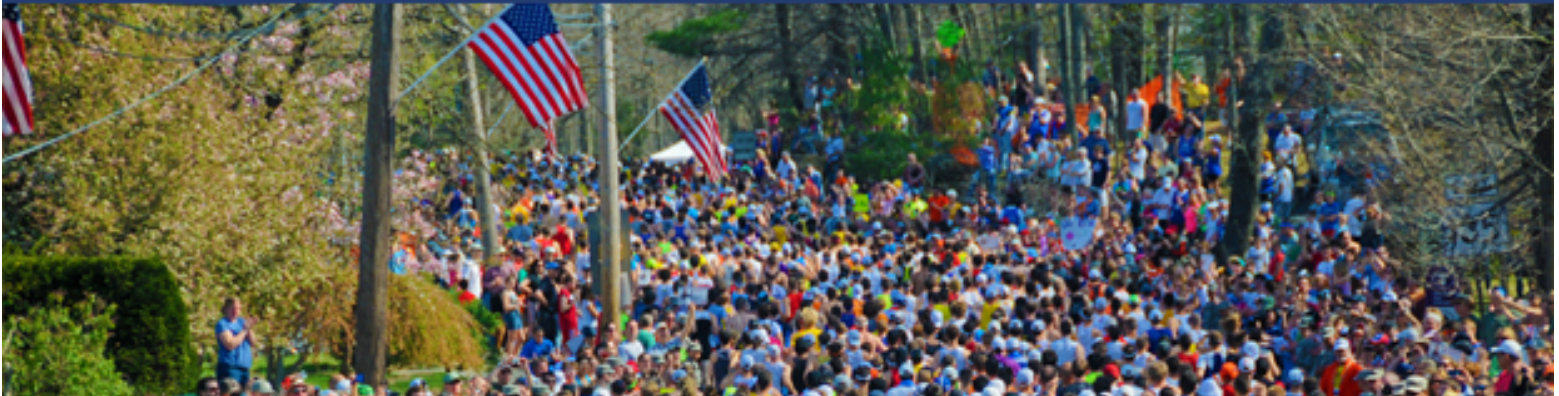
Tutorial Services
(check website for details)

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**117TH BOSTON
MARATHON**

Rheumatology Review



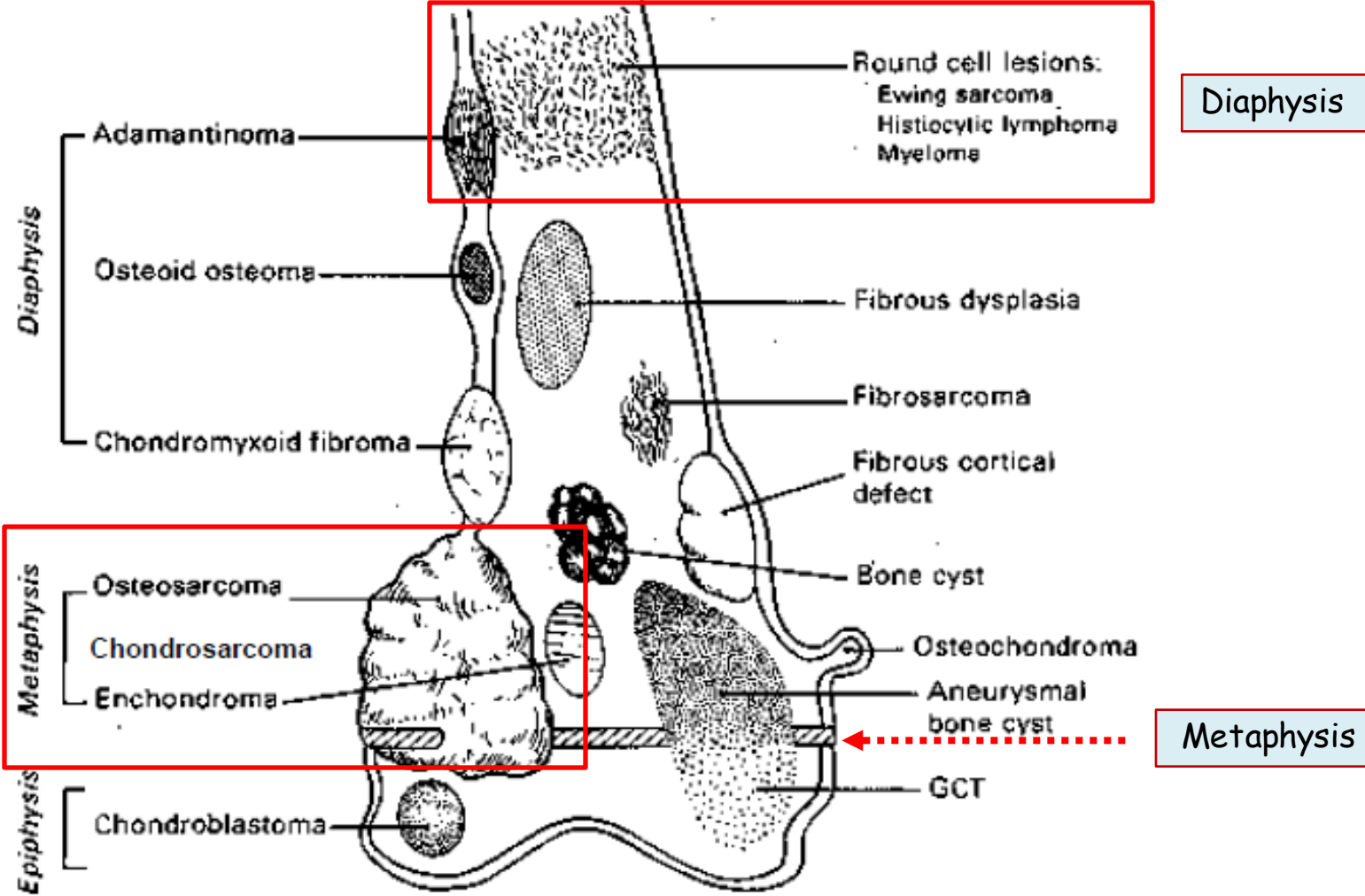
Bone Tumors

Normal Tissue	Benign Tumor	Malignant Tumor
Bone	Osteoma Osteoid Osteoma Osteblastoma	Osteosarcoma - Osteoblastic - Chondroblastic - Telangiectatic - Dedifferentiated
Cartilage	Enchondroma Osteochondroma Chondromyxoid fibroma Chondroblastoma	Chondrosarcoma - Conventional - Clear cell - Mesenchymal - Dedifferentiated
Fibro-osseous	Fibrous dysplasia Osteofibrous dysplasia	Adamantinoma
No normal counterpart	Giant cell tumor Aneurysmal bone cyst	Malignant giant cell tumor Ewing's sarcoma

Bone Tumors

Normal Tissue	Benign Tumor	Malignant Tumor
Bone	Osteoma Osteoid Osteoma Osteoblastoma	Osteosarcoma - Osteoblastic - Chondroblastic - Dedifferentiated
Bone formation by tumor cells is 'diagnostic'		
Cartilage	Enchondroma Osteochondroma Chondromyxoid fibroma Chondroblastoma	Chondrosarcoma - Conventional - Clear cell - Mesenchymal - Dedifferentiated
Fibro-oss	Unknown cell of origin: sheets of primitive, uniform small round cells w/o differentiation	
No normal counterpart	Giant cell tumor Aneurysmal bone cyst	Malignant giant cell tumor Ewing's sarcoma

BONE TUMORS

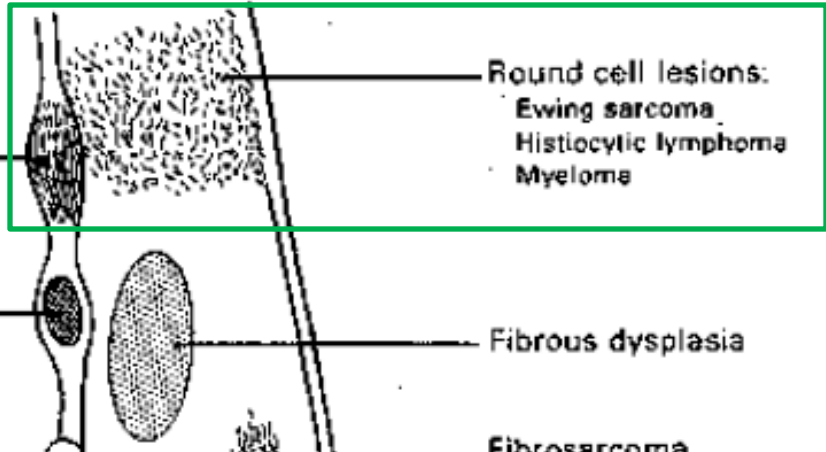


Specific types of bone tumors target certain **age groups** and **anatomic sites** in the bone.

BONE TUMORS



Onion skin periosteal rxn



- Metaphysis**
- Osteosarcoma
 - Chondrosarcoma
 - Enchondroma

- Epiphysis**
- Chondroblastoma

Specific types and anatomic



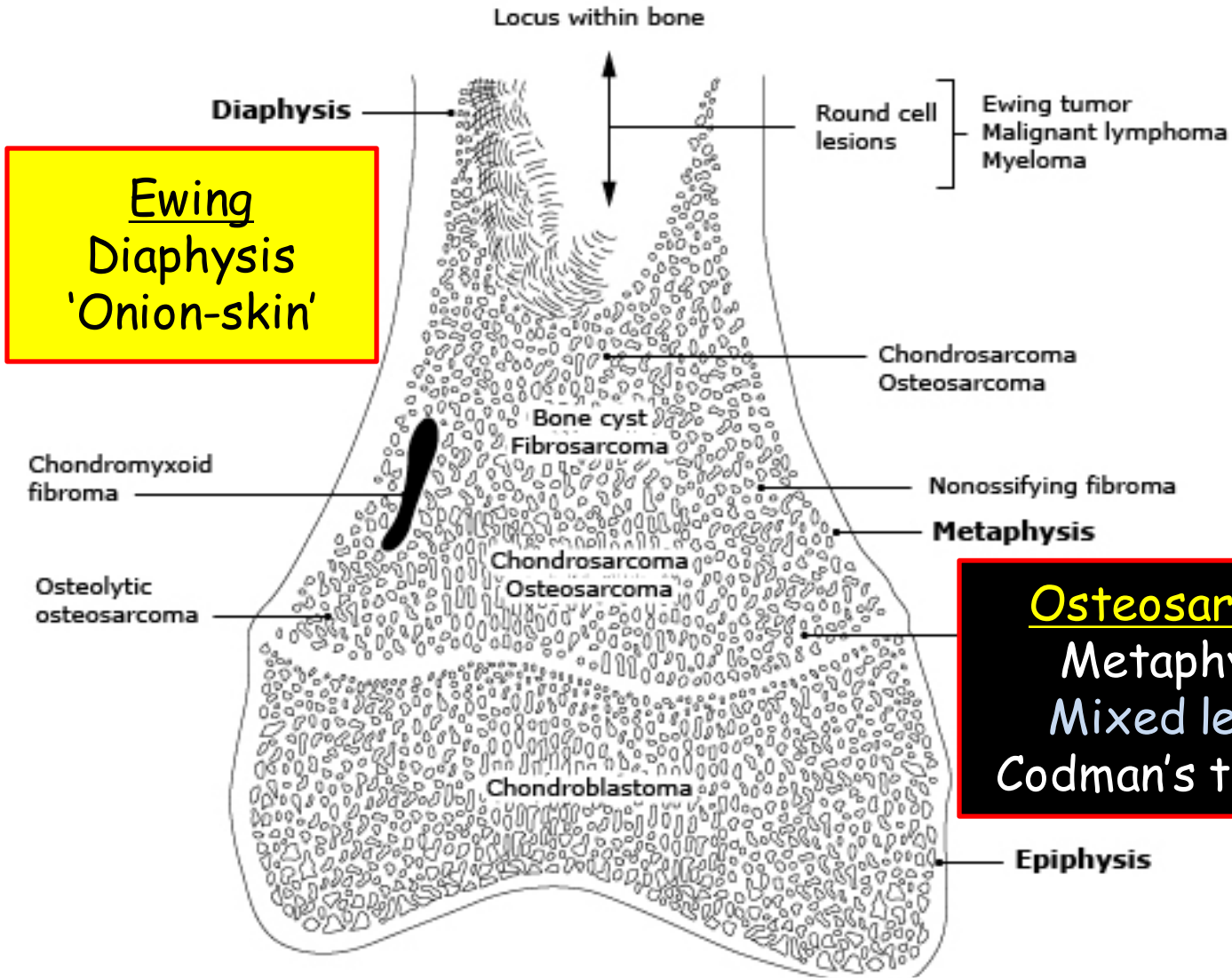
Osteoblastic = bone formation

- Fibrosarcoma
- Fibrous cortical defect
- Bone cyst
- Osteochondroma
- Aneurysmal bone cyst
- GCT

in age groups

Primary Bone Tumors (‘by cell of origin’)

- Cartilage
 - Benign: Osteochondroma
 - Malignant: Chondrosarcoma
- Bone
 - Malignant: **Osteosarcoma**
- Unknown
 - **Ewing sarcoma**



Ewing
Diaphysis
'Onion-skin'



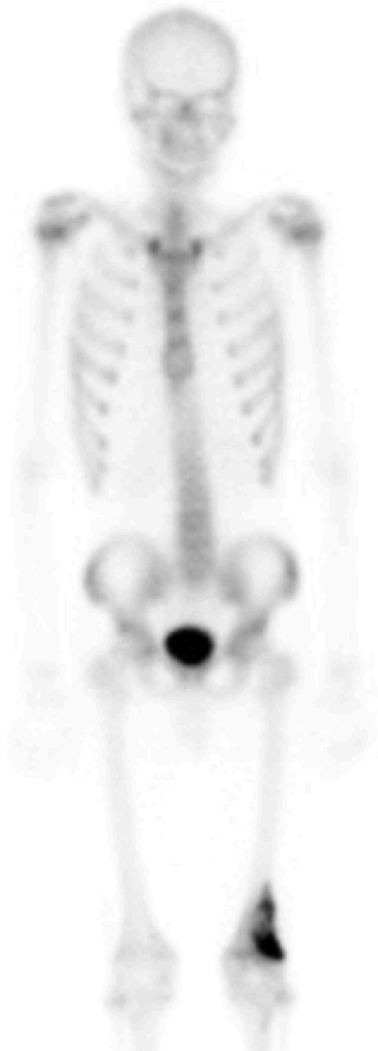
Center

Ewing tumor
Malignant lymphoma
Myeloma

Chondrosarcoma
Osteosarcoma

Bone cyst





16 y.o. w/ knee pain seen by
pediatrician who ordered film

FINDINGS:

There is a large, approximately 7 x 3 cm, predominately lytic lesion in the distal lateral femoral metaphysis. It is heterogeneous with a moth-eaten appearance, ill-defined margins and destruction of the lateral cortex. There is some irregular periosteal new bone in a Codman's triangle configuration. The lesion extends into the adjacent soft tissues. Alignment is anatomic. Joint spaces are maintained. No knee joint effusion.

Given the size of the lesion, the patient is at risk for pathologic fracture.

Osteosarcoma

- Background
 - Majority occur in young adults (peaking during **growth spurt**)
 - Key Point: arise in the metaphysis with majority at knee (distal femur/proximal tibia) - region of **fastest growth**.
 - Older adults with predisposing condition such as **Paget's**



Osteosarcoma

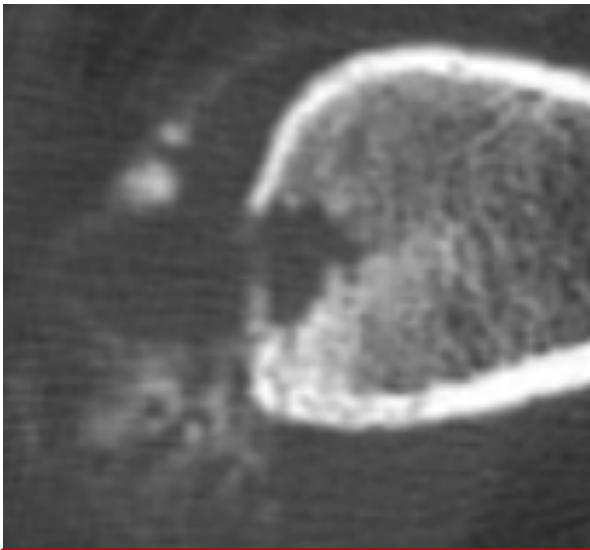
- Background
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 - Key Point: arise in the metaphysis with majority at knee (distal femur/proximal tibia) - region of fastest growth.
 - Older adults with predisposing condition such as Paget's
- Pathogenesis
 - Majority with mutations of suppressor or oncogenes
- Pathology
 - Different types but majority: intramedullary, osteoblastic and high grade
 - Bulky & aggressive: they destroy cortex and produce soft tissue masses
 - Bone formation by tumor cells is diagnostic

Blastic



Pathology

- Different types but majority: intramedullary, osteoblastic and high grade
- Bulky & aggressive, they destroy cortex and produce soft tissue masses
- Bone formation by tumor cells is diagnostic



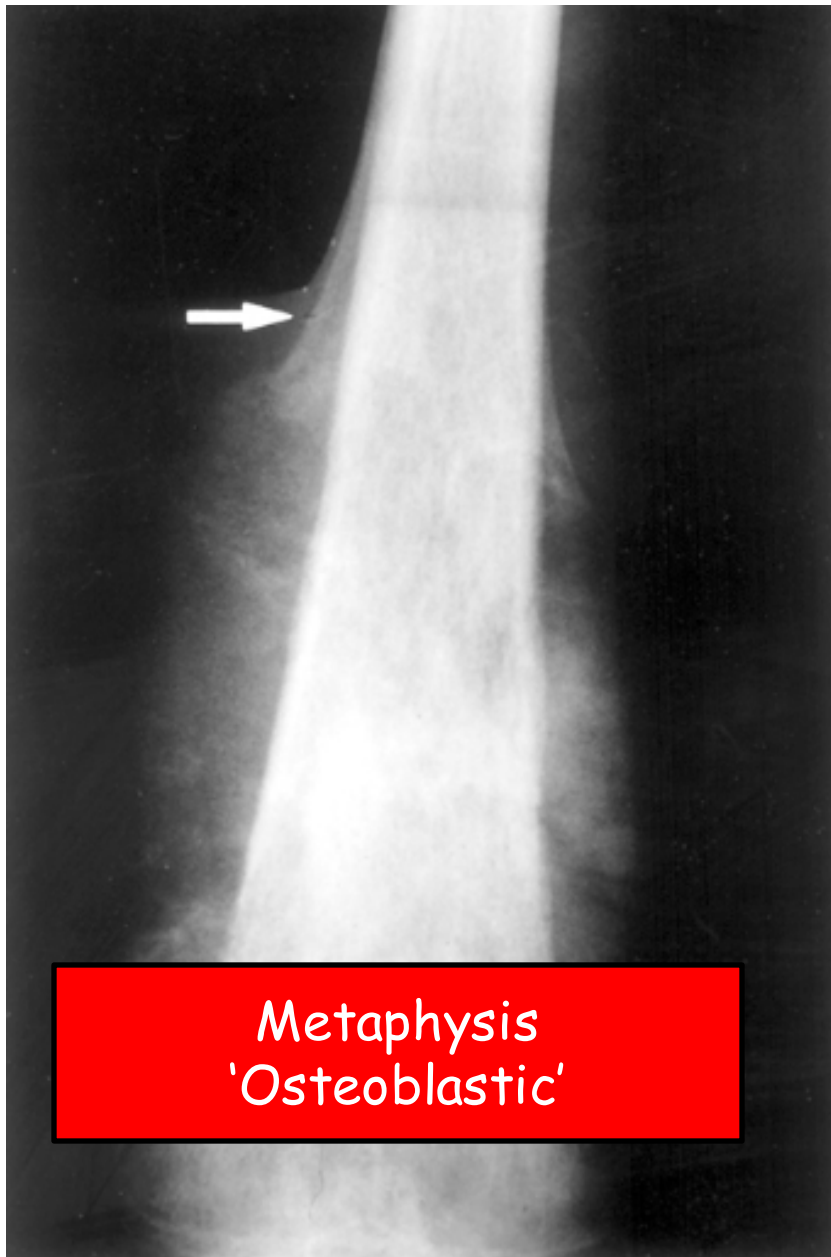
Destroy cortex
Soft tissue mass
Bone formation



Osteoblastic = new bone formation

Osteosarcoma

- Background
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 - Different types but majority: intramedullary, osteoblastic and high grade
 - Bulky & aggressive, they destroy cortex and produce soft tissue masses
 - Bone formation by tumor cells is diagnostic
- Clinical
 - **Painful**, progressively enlarging mass → pathologic fx
- Diagnostics
 - Radiograph: destructive **mixed** (blastic/lytic) lesion with infiltrative margins



Tumor breaks through cortex
and lifts the periosteum



Reactive periosteal bone
formation

Triangular shadow called
Codman triangle



'Sunburst Pattern'

Periostitis

Periosteum and USMLE:

Osteosarcoma: Codman Δ , periostitis

HyperPTH: subperiosteal bone resorption

Hypertrophic Osteoarthropathy (AdenoCa):

Marked periosteal new bone formation

Osteosarcoma

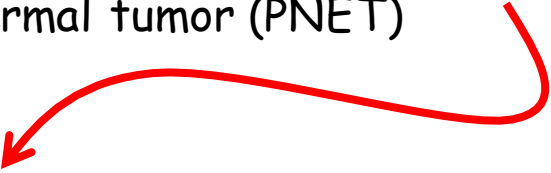
- Diffuse areas of lytic destruction
- Other areas of sclerotic bone with increased radiodensity
- Permeative, invasive, destructive, no distinct borders
- Several histological subtypes



Osteosarcoma Question...

Option: Anaplastic cells that form **osteoid matrix**
(i.e. new bone formation)

Ewing sarcoma (family of tumors)

- Background
 - Peak during adolescence most commonly involving the **diaphysis of long bone** (tibia, femur) and the flat bones of the pelvis
 - Pathogenesis
 - **Unknown cell of origin**
 - Mesenchymal v **primitive** neuroectodermal tumor (PNET)
 - Pathology
 - Sheets of primitive, uniform small round cells without differentiation
 - Homer-Wright rosettes (see notes)
 - Scant cytoplasm, rich in glycogen (clear appearance)
 - **Arise in the medullary canal** and invade cortex, periosteum and soft tissue
- 

Ewing sarcoma (family of tumors)

- Clinical (Pain and Swelling for weeks-months)
 - Painful, enlarging mass; may be warm and swollen
 - Nonspecific constitutional symptoms (fever, fatigue, wt loss)
- Diagnostics
 - Radiograph: destructive lytic tumor that extends into surrounding soft tissues.
 - Periosteal reaction → reactive bone deposited in onion skin appearance

Ewing sarcoma (family of tumors)

Periostitis: Onion skin, **diaphysis**, Homer-Rosettes (neuroectoderm)

versus

Periostitis: Sunburst/Codman's, **metaphysis**, new bone formation

- **Diagnostics**

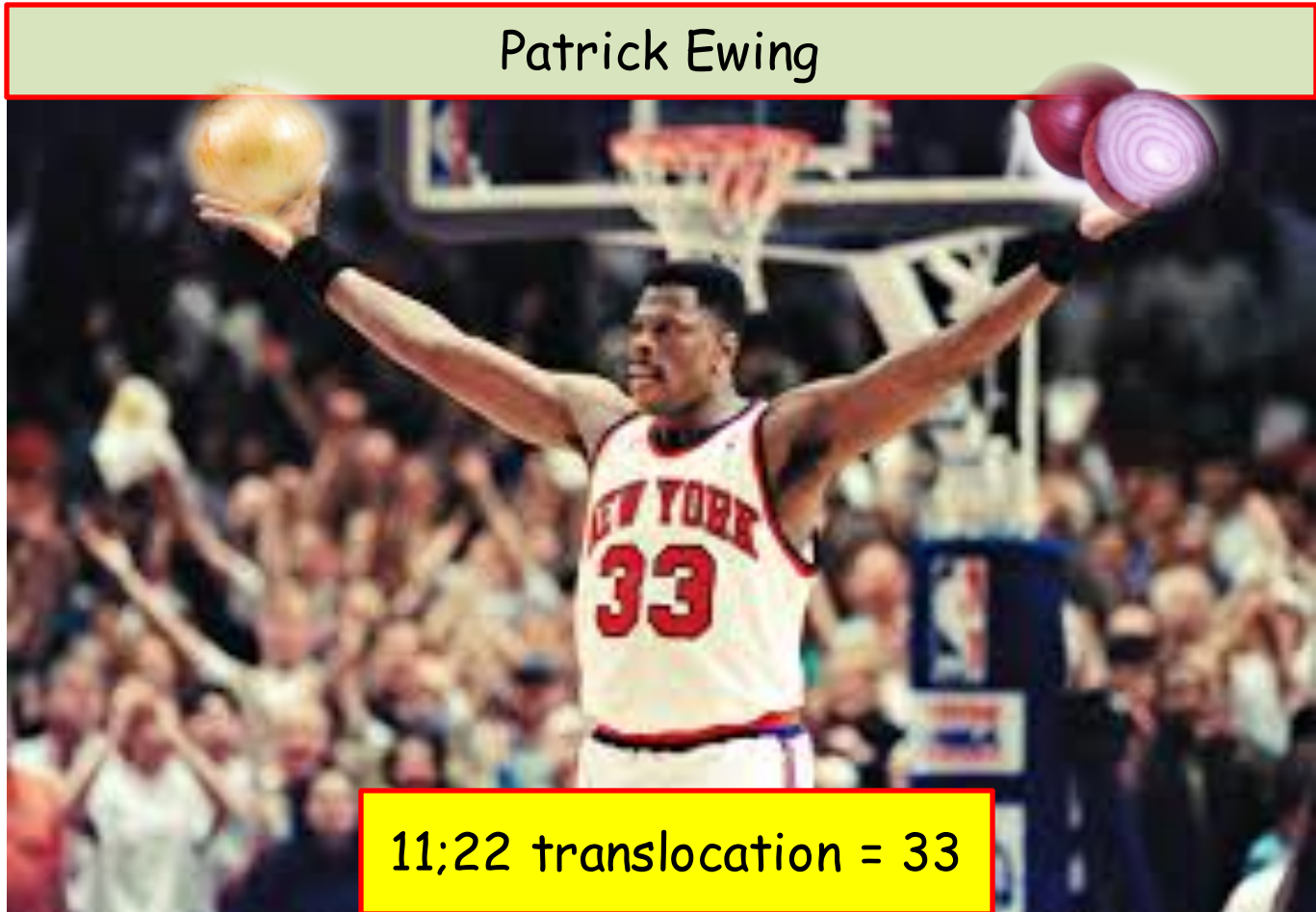
- Radiograph: destructive lytic tumor that extends into surrounding soft tissues.
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Ewing sarcoma (family of tumors)

- Clinical (Pain and Swelling for weeks-months)
 - Painful, enlarging mass; may be warm and swollen
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- Diagnostics
 - Radiograph: destructive lytic tumor that extends into surrounding soft tissues.
 - Periosteal reaction → reactive bone deposited in onion skin appearance
- Notes
 - Homer-Wright rosettes: tumor cells are arranged in a circle about a central fibrillary space, indicative of neural differentiation (derive from neuroectoderm?)

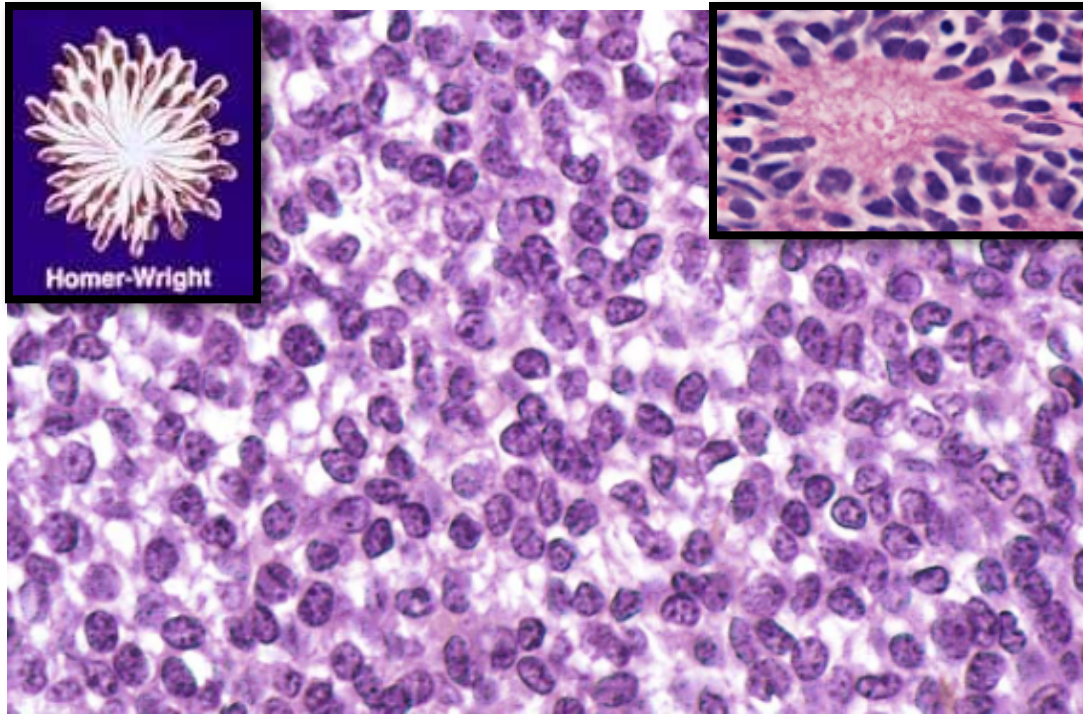


Patrick Ewing

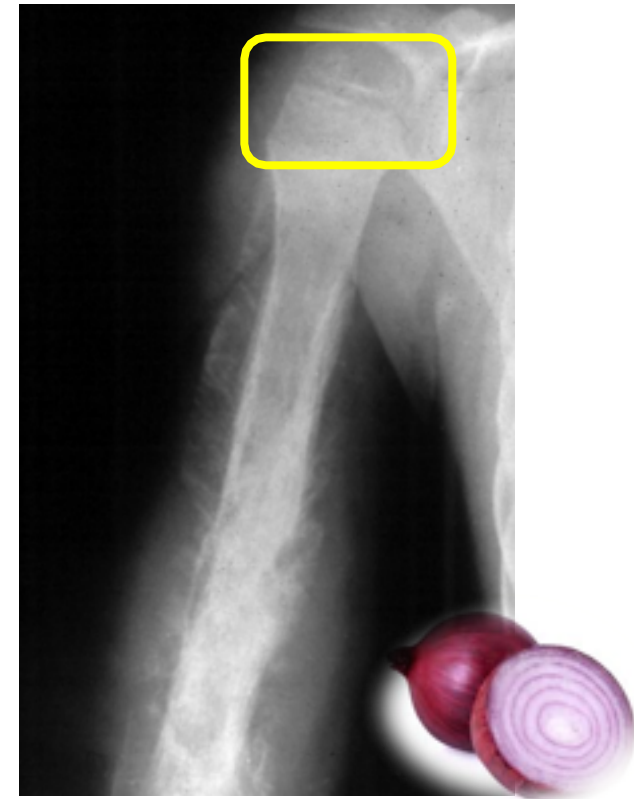


11;22 translocation = 33

Ewing Sarcoma



1. Sheets of primitive, uniform small round cells
2. No differentiation
3. Scant cytoplasm, rich in glycogen (clear appearance)
4. Homer-Wright rosettes (neuroectoderm)



Reactive bone deposited
in onion skin appearance

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