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Orthopaedic Tumors:

What the community provider should know

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Disclosures

- None

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Learning Objectives

- Ascertain whether a tumor is likely malignant vs benign
- Identify how to recognize conditions that need a referral to orthopaedic oncology
- Understand basics of treatment of primary bone sarcoma, metastatic disease, and myeloma
- Understand basics of treatment of common benign bone tumors

Plan

- Basics of H&P, Imaging, Diagnosis, Treatment
- 5 Case presentations intermixed to highlight important elements of the above features

- Focus more on bone tumors

Orthopedic oncology practice

- Patients with tumor: extremities or spine
  - Bone tumors
    - Benign
    - Malignant
    - Metastatic
  - Soft tissue tumors
    - Benign
    - Malignant
  - “Orthopedic weirdness”
    - Infections
    - Genetic abnormalities

Orthopedic oncology

- Aunt Minnie phenomenon
  - Pattern recognition
    - Little old lady, overweight, gray hair, always carries oversized purse

Orthopedic Oncology

- Diagnosis is a puzzle
  - History
  - Exam
  - Imaging
  - Pathology

Presentation

- History
  - Mass
    - How long present
    - Growing
  - Pain
    - How long
    - Changing
    - Night
    - Functional
    - Radiate

Presentation

- Past medical history
  - Cancer history
  - Surgical risk
- Family history
  - Primary relatives

Presentation

- History
  - Age
    - Skeletally immature
      - Chondroblastoma, osteochondromas, nonossifying fibroma
      - Ewings, osteosarcoma
      - rhabdomyosarcoma
    - young adult
      - Giant cell tumor of bone, fibrous dysplasia
      - Ewings, osteosarcoma
      - fibromatosis
      - Synovial cell sarcoma
    - older adult (>40)
      - Enchondroma
      - Chondrosarcoma, secondary osteosarcoma
      - Lipoma
      - Soft tissue sarcoma

Presentation

- Exam
  - Mass
    - Hard vs soft
    - Fixed
    - Superficial or deep
  - Skin
  - Distal extremity changes
    - Swelling
    - Vascular
    - Nerve
    - Range of motion
  - Other masses

Presentation

- Benign bone
  - Painless or painful
  - No mass
- Primary bone sarcoma or metastatic disease/myeloma
  - Painful
  - Sometimes mass
- Benign soft tissue
  - Painless or painful
- Soft tissue sarcoma
  - Painless unless large enough to cause pain from mass effect

Patient 1

- History
  - 24 yo male injured his knee playing soccer several months ago. Pain did not improve. Becoming too painful to walk. Mass slowly developed and is now growing.
- Past history, family history
  - None
- Exam
  - Large, hard, fixed mass on distal femur
  - Very painful any knee motion

- What are the key elements of history and exam?

Patient 1

- History
  - 24 yo male injured his knee playing soccer several months ago. Pain did not improve. Becoming too painful to walk. Mass slowly developed and is now growing.
- Past history, family history
  - None
- Exam
  - Large, hard, fixed mass on distal femur
  - Very painful any knee motion

- Key components
  - Young adult male
  - Mass that is growing
  - Pain progressive
  - Unable to bear weight
  - Large, hard, fixed mass

Patient 2

- History
  - 15 yo boy playing soccer and had a twisting injury with acute pain. No knee swelling. No previous history of pain. Stopped playing soccer and pain has gradually improved
- Past/family history
  - None
- Exam
  - Normal

- What are the key elements of history and exam?

Patient 2

- History
  - 15 yo boy playing soccer and had a twisting injury with acute pain. No knee swelling. No previous history of pain. Stopped playing soccer and pain has gradually improved
- Past/family history
  - None
- Exam
  - Normal

- Key components
  - Age: 15
  - Incidental finding
  - Probably not related to his pain
  - No exam findings

Patient 3

- History
  - 72 yo male with pain in his right hip/groin for a few months. Pain localized and progressive. Pain increased with weight bearing. Some difficulty with voiding. No blood in urine, cough, temperature issues.
- Past/Family history
  - Mother with breast
- Exam
  - Painful to hip motion

- What are the key elements of history and exam?

Patient 3

- History
  - 72 yo male with pain in his right hip/groin for a few months. Pain localized and progressive. Pain increased with weight bearing. Some difficulty with voiding. No blood in urine, cough, temperature issues,
- Past/Family history
  - Mother with breast
- Exam
  - Painful to hip motion

- Key components
  - Age > 40
  - Groin pain
  - Pain progressive
  - Functional pain
  - Difficulty voiding
  - Minimal exam findings

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Patient 4

- History
  - 51 yo male with year history of popping sensation in thigh when he crossed his legs. Recognized a mass 6 months ago. Mass has been growing. Over last month developed low grade constant pain that is a little worse with activity.
- Past/family history
  - none
- Exam
  - Large, soft mass, movable
  - Painful to palpation
  - Normal knee/hip motion

- What are the key elements of history and exam?

Patient 4

- History
  - 51 yo male with year history of popping sensation in thigh when he crossed his legs. Recognized a mass 6 months ago. Mass has been growing. Over last month developed low grade constant pain that is a little worse with activity.
- Past/family history
  - none
- Exam
  - Large, soft mass, movable
  - Painful to palpation
  - Normal knee/hip motion

- Key components
  - Age > 40
  - Growing mass
  - Low grade constant pain
  - Soft, freely movable mass

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Patient 5

- History
  - 22 yo male who had a fall on stairs 5 months ago. He had some knee soreness. Soreness progressed to knee pain. Pain localized to medial knee region. Pain constant. Minimal change with activity. No night pain.
- Past/family history
  - None
- Exam
  - Tender on medial distal femur
  - Normal knee

- What are the key elements of history and exam?

Patient 5

- History
  - 22 yo male who had a fall on stairs 5 months ago. He had some knee soreness. Soreness progressed to knee pain. Pain localized to medial knee region. Pain constant. Minimal change with activity. No night pain.
- Past/family history
  - None
- Exam
  - Tender on medial distal femur
  - Normal knee

- Key components
  - Age: young adult
  - Progressive pain
  - No functional pain
  - No mass
  - Minimal exam findings

Imaging

- X-ray for bone tumors
  - Location in bone
    - Epiphysis
    - Metaphysis
    - Diaphysis
    - Spine: anterior or posterior
  - Tumor appearance
    - Matrix
      - None
      - Calcification
      - Bone
      - Ground glass

Imaging

- X-ray
  - Tumor appearance
    - Lytic
    - Blastic
    - Mixed
  - Interaction with bone
    - Margin
    - Soft tissue extension
    - Periosteal changes

Imaging

- Benign vs Malignant
  - Margin of tumor
    - Well defined: benign
    - Poorly defined: malignant
  - Soft tissue extension
    - Present: malignant
  - Periosteal changes
    - Present: malignant
  - Multiple sites
    - Could be either

- 85% of bone tumors can be diagnosed from
  - Age patient
  - Location in bone
  - X-ray appearance

Imaging

- Metaphyseal region
- Wide zone transition
- Soft tissue extension
- Periosteal changes

- Metaphyseal region
- Well defined margin
- No periosteal changes
- No soft tissue mass

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- Metaphyseal region
- Calcifications
- Questionable margin
- No periosteal changes
- No cortical breakthrough or soft tissue extension

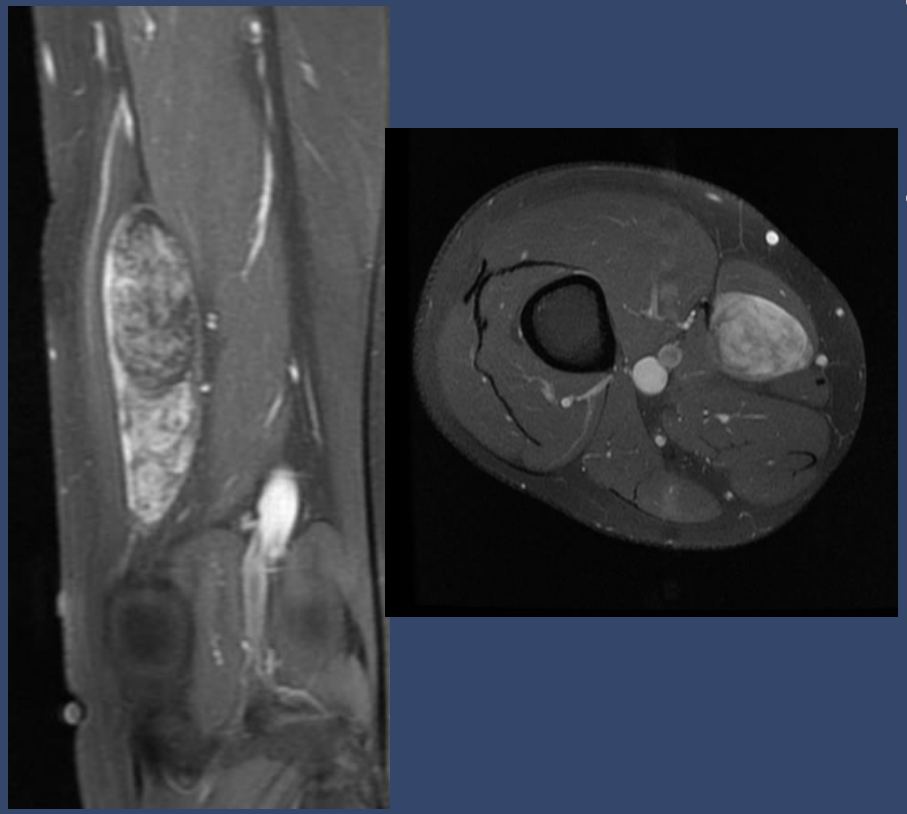
Imaging – other modalities

- Ultrasound
  - Best for small soft tissue tumors <5cm
  - Can evaluate: ganglions, lipomas
- MRI with contrast
  - Best used
    - Soft tissue masses >=5cm (golf ball)
    - Bone lesions concerning for malignancy (wide zone transition, soft tissue extension)
  - T1: Fat
  - T2: Water
  - Contrast images: vascularity/activity, necrosis
  - Whole bone vs just tumor

Imaging

- Soft tissue tumors
  - Many tumor types
  - Only small number can be diagnosed from MRI
  - Most look similar
- Benign vs malignant
  - Large: tends to be malignant
  - Necrosis: malignant

Imaging



- Large soft tissue mass
- Heterogeneous with contrast enhancement

Imaging

- Aunt Minnie applies to orthopedist and radiologist
- Mass involving the humerus. Mass has ….. This could represent a benign tumor such as enchondroma or osteoblastoma. Can not rule out sarcoma, eosinophilic granuloma, metastatic disease, or lymphoma. Recommend …..
  - Translation: I DON'T KNOW!

Patient 1

- A Benign
- B Malignant

Patient 1

- Mass in metaphyseal extending into epiphysis
- Wide zone transition
- Large soft tissue extension
- Bone formation in mass
- Periosteal changes

Patient 1

- A Benign
- B Malignant

Patient 2

- A Benign
- B Malignant

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Patient 2

- Tibia and femur
- Metaphyseal
- Well defined border
- No bone formation
- Involves cortex but no periosteal changes

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Patient 2

- A Benign
- B Malignant

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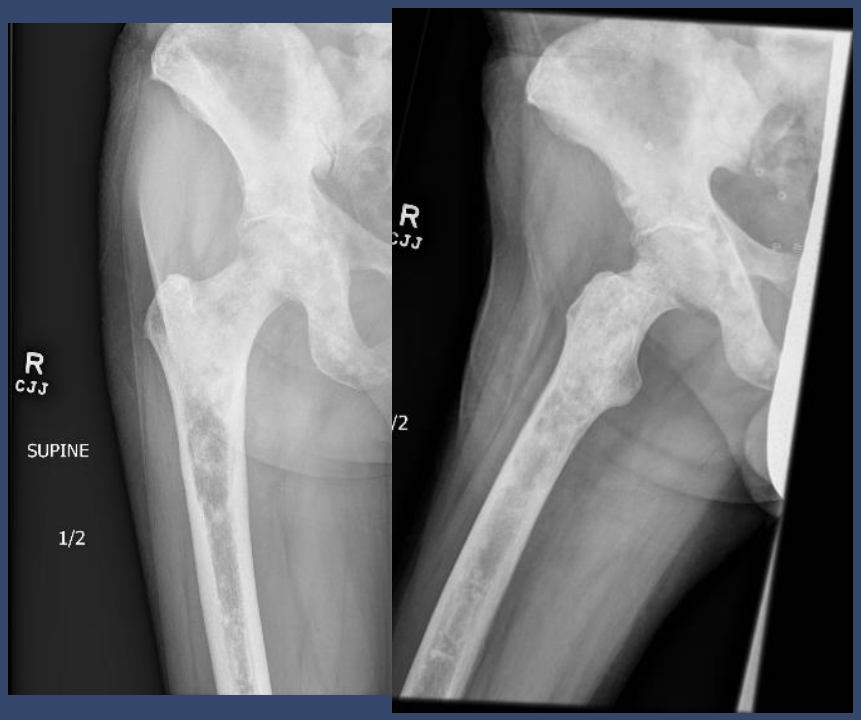
Patient 3



- A Benign
- B Malignant

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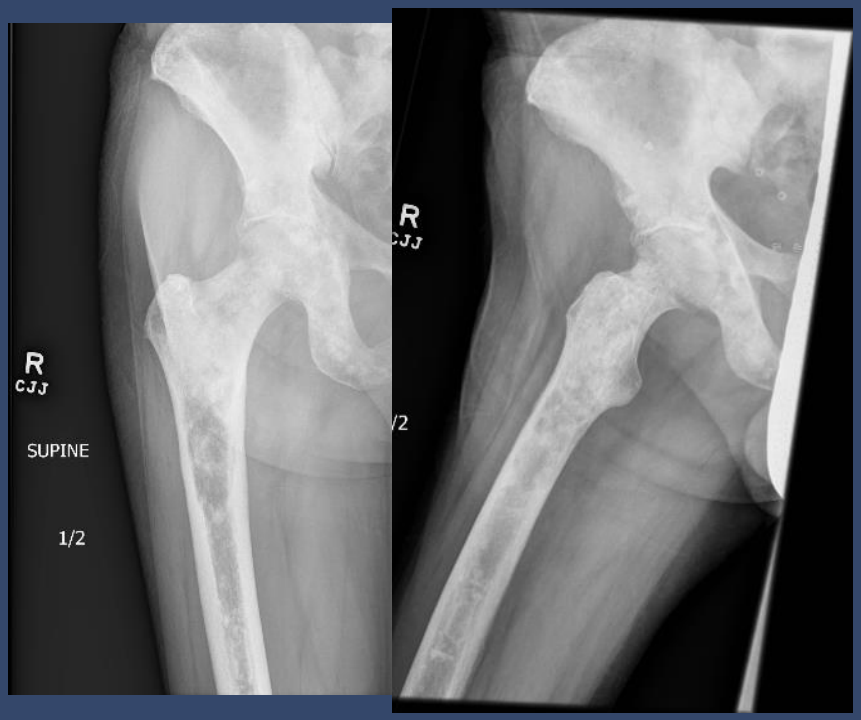
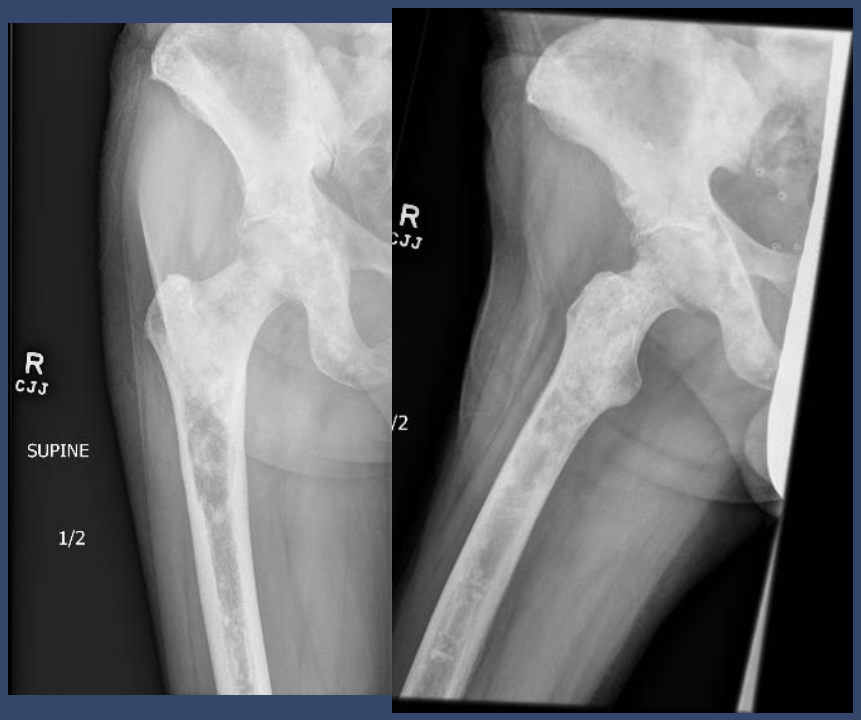
Patient 3



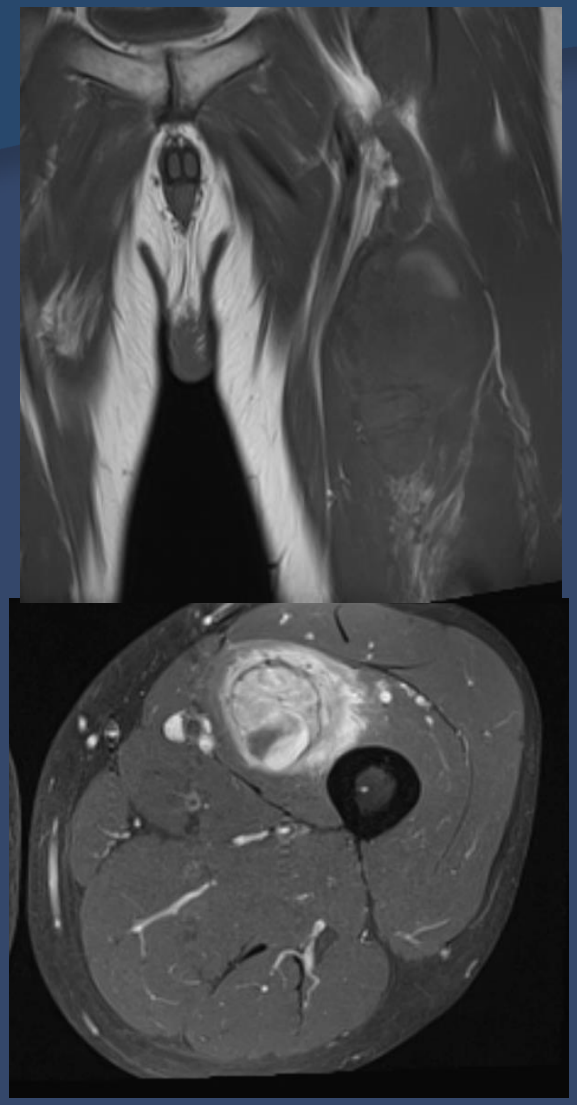
- Pelvis and femur
- Wide zone of transition
- Mixed blastic and lytic
- Femur with area of cortical thinning
- No periosteal changes

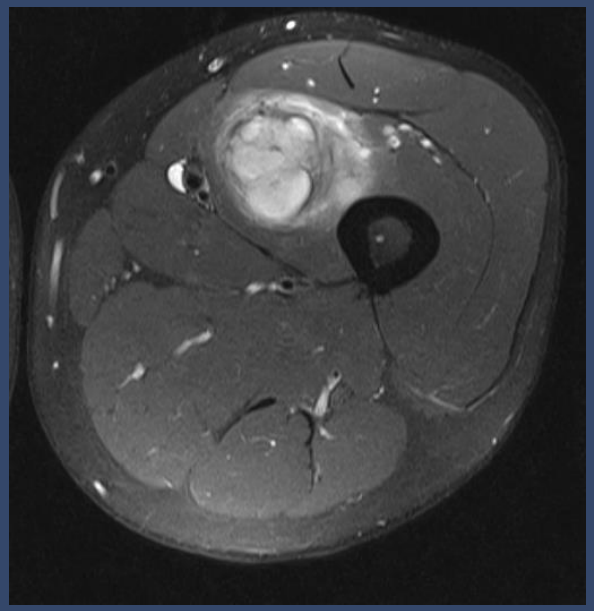
Patient 3



- A Benign
- B Malignant

Patient 4





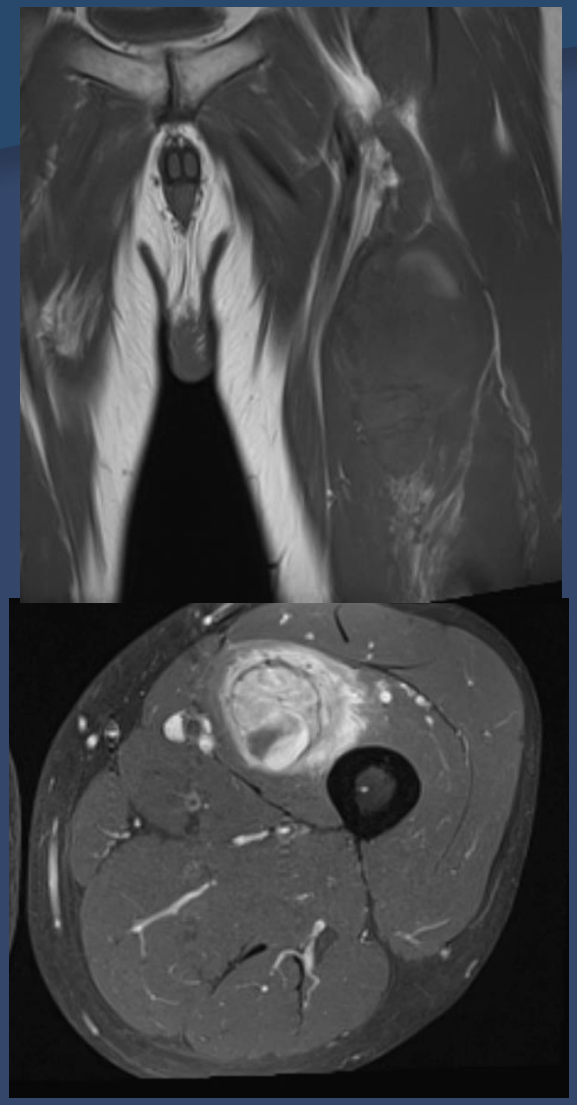
A Benign

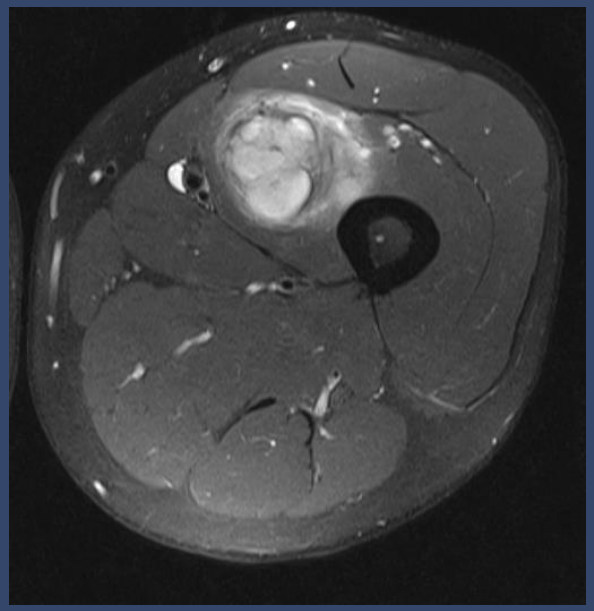
B Malignant

Patient 4

- Large soft tissue mass
- Deep (below fascial plane)
- Low T1, high T2 signals
- Heterogeneous on contrast

Patient 4





A Benign

B Malignant

Unclear, but more likely malignant

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Patient 5

- A Benign
- B Malignant

Patient 5

- Distal femur
- Epiphyseal into metaphysis
- Narrow zone transition
- Lytic lesion with no matrix

Patient 5

- A Benign
- B Malignant

Staging

- Benign tumors
  - Most (cysts, fibrous dysplasia, osteochondroma, etc)
    - Local imaging only
  - Giant cell tumor of bone, chondroblastoma
    - Low risk of developing pulmonary metastasis
    - Local imaging AND chest imaging
- Malignant primary bone tumors
  - Osteosarcoma, Ewing's sarcoma
  - Chondrosarcoma
    - MRI with/without contrast whole bone
    - CT chest, whole body bone scan
    - Biopsy prior to surgical treatment

Metastatic disease

- Metastatic tumors to bone
  - At autopsy 80-90 % of all adenocarcinomas have bone metastasis
  - PT Barnum Loves Kids or

    BLT w Kosher Pickle
    - Prostate, breast, lung, kidney, thyroid

Staging

- Metastatic tumors
  - Staging per recommendations of original tumor
  - Skeletal imaging
    - Bone scan
    - PET scan
    - Skeletal survey
  - Labs (Ca)
    - hyperCa: “Bones, stones, moans, groans”
  - If unknown primary:
    - CT chest/abdomen/pelvis will find in 85%
    - Labs;
      - Ca
      - CBC, CMS, UA, T3/T4, SPEP, UPEP, PSA

Staging

- Biopsy
  - Wait until all local imaging done
  - Who does biopsy
    - If suspect benign, surgeon does
      - Do frozen during procedure and may resect in same surgery
    - If suspect sarcoma
      - Treating physician should do
      - Biopsy tract should be completely excised
    - If suspect metastatic disease or lymphoid disease
      - CT or ultrasound guided needle

Patient 1

- Summary
  - Young adult
  - Large mass in distal femur
  - Mass growing
  - Mass painful
- Xray findings
  - Large soft tissue extension
  - Periosteal changes

- What imaging studies are needed

Patient 1

- Summary
  - Young adult
  - Large mass in distal femur
  - Mass growing
  - Mass painful
- Xray findings
  - Large soft tissue extension
  - Periosteal changes

- What imaging studies are needed

Patient 1

- Summary
  - Young adult
  - Large mass in distal femur
  - Mass growing
  - Mass painful
- Xray findings
  - Large soft tissue extension
  - Periosteal changes
- Additional studies
  - MRI with contrast of entire femur
  - CT chest
  - Bone scan
  - Biopsy

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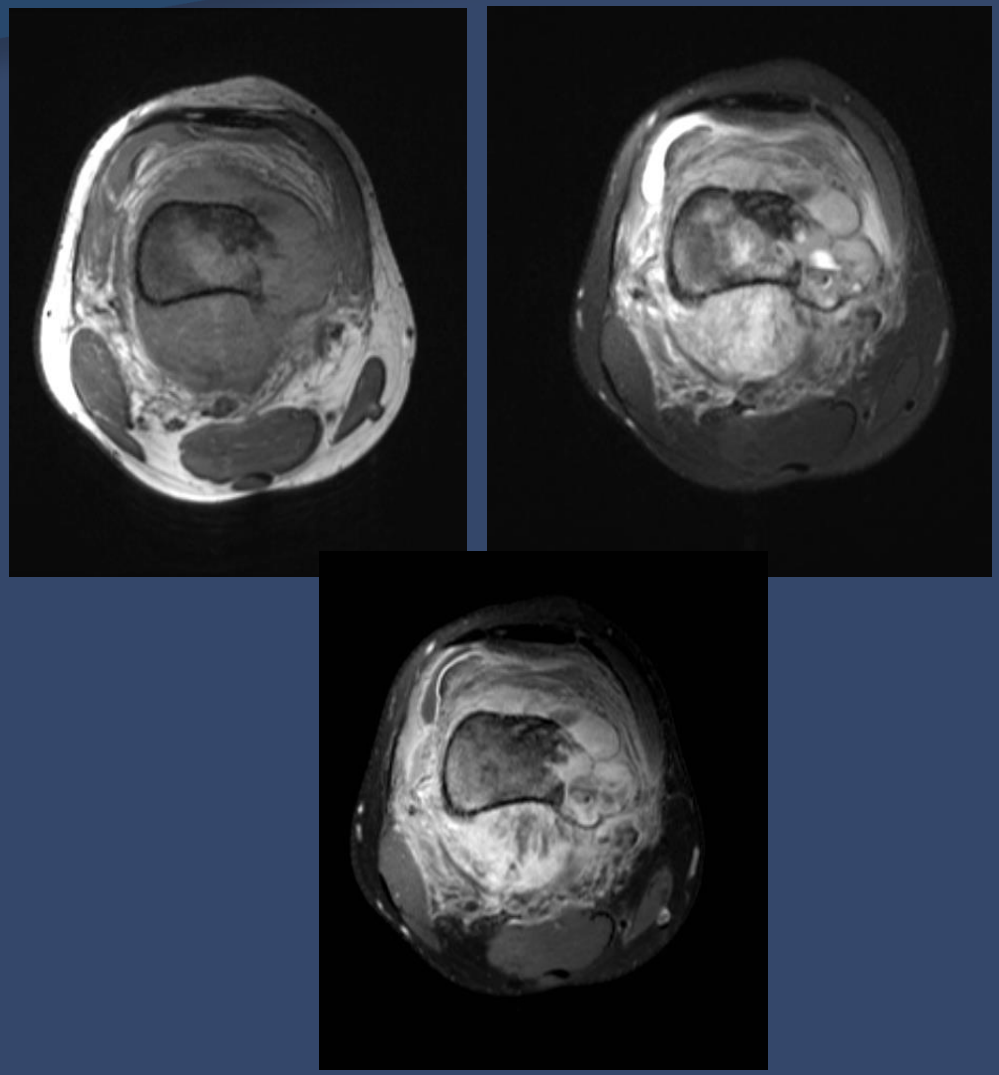
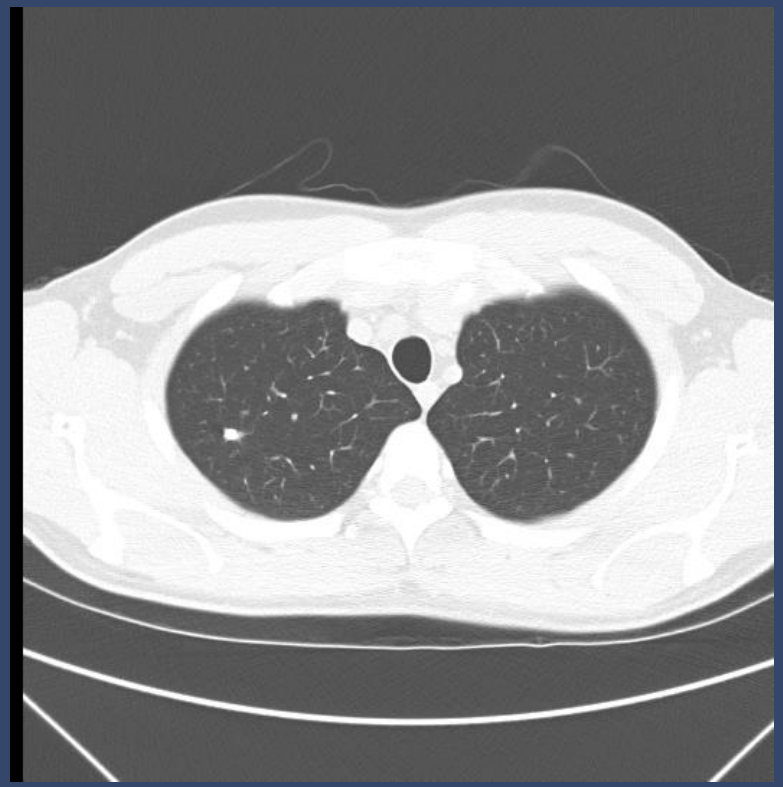
Patient 1



- Large soft tissue extension
- Cortical destruction
- Heterogeneous

Patient 1



- CT chest has a single 6 mm nodule

Patient 1

- Bone scan shows only the tumor

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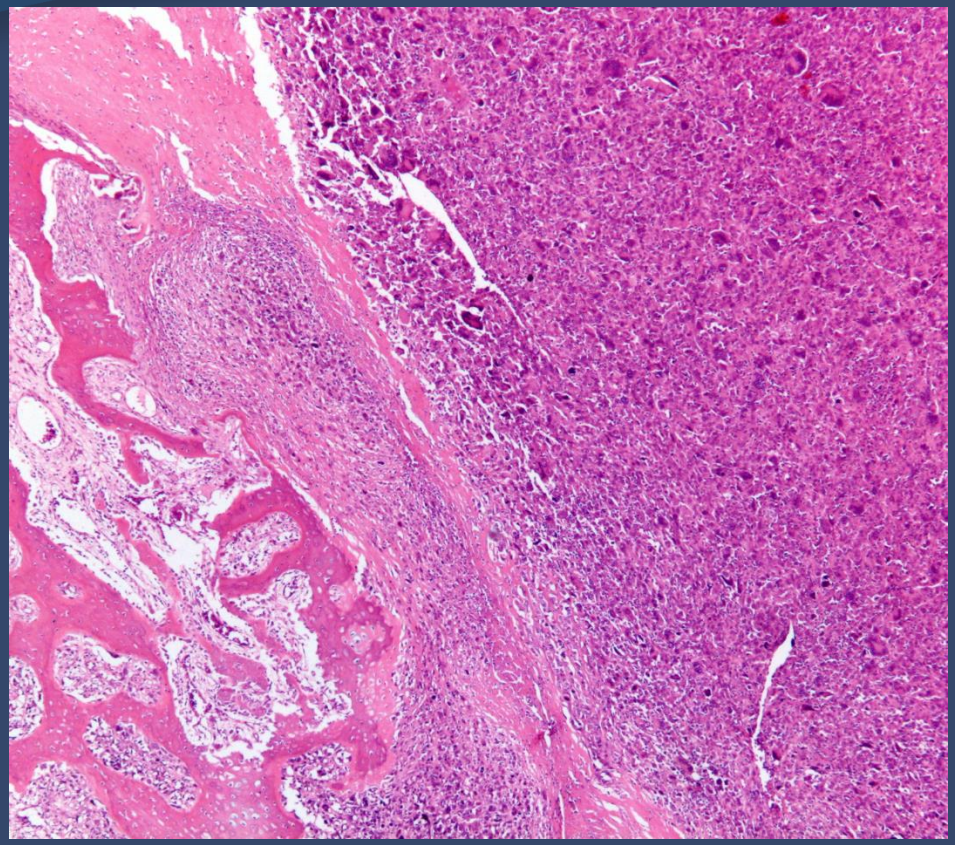
Patient 1



- Biopsy
  - Highly malignant cells with osteoid and bone
- Diagnosis: osteosarcoma

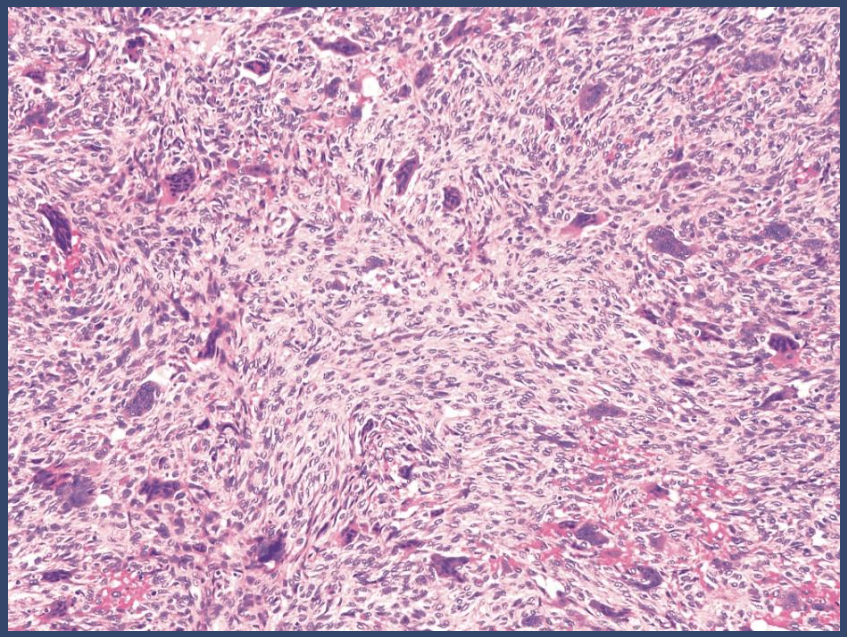
Patient 2

- Summary
  - 15 yo with an incidental finding of a mass in the proximal tibia and distal femur.
  - Xray
    - well defined margin
- What additional imaging studies are needed?

Patient 2

- Summary
  - 15 yo with an incidental finding of a mass in the proximal tibia and distal femur.
  - Xray
    - well defined margin
- Additional studies
  - None
  - X-ray diagnosis

Patient 2



- Pathology
  - Fibrous/stromal cells
  - Giant cells
- Diagnosis
  - Non ossifying fibroma

Patient 3

- Summary
  - Elderly male with progressive functional hip pain.
  - X-ray with mixed lesion in pelvis and femur
  - Trouble voiding
- What additional studies are needed?

Patient 3

- Summary
  - Elderly male with progressive functional hip pain.
  - X-ray with mixed lesion in pelvis and femur
  - Trouble voiding
- Additional Studies
  - Bone scan
  - CT chest/abdomen/pelvis
  - Labs
    - CMS
    - Ca
    - PSA

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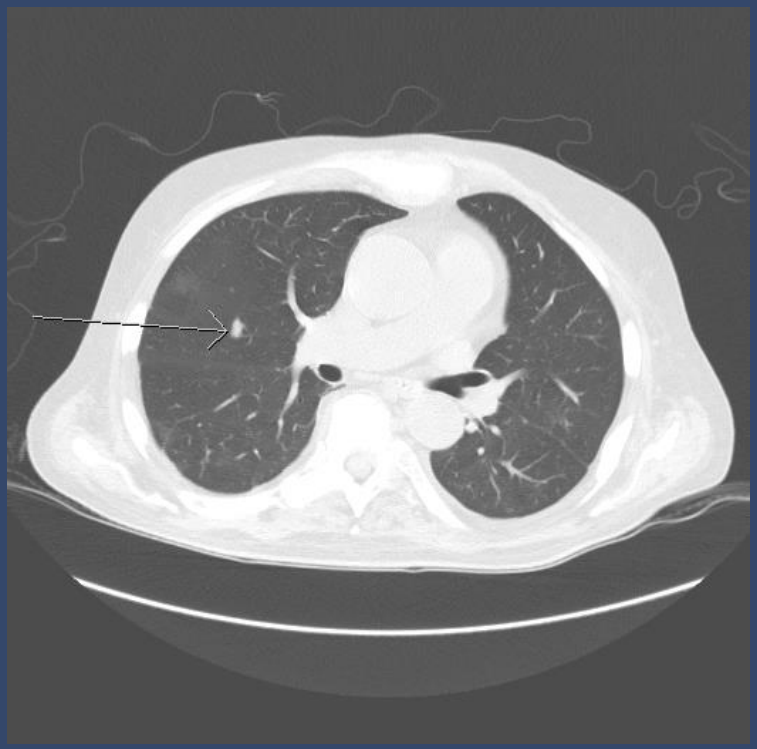
Patient 3



- CT chest
  - Single small mass

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Patient 3

- CT chest
  - Multiple vertebrae involved
  - Pathologic fracture T4 with extension into canal

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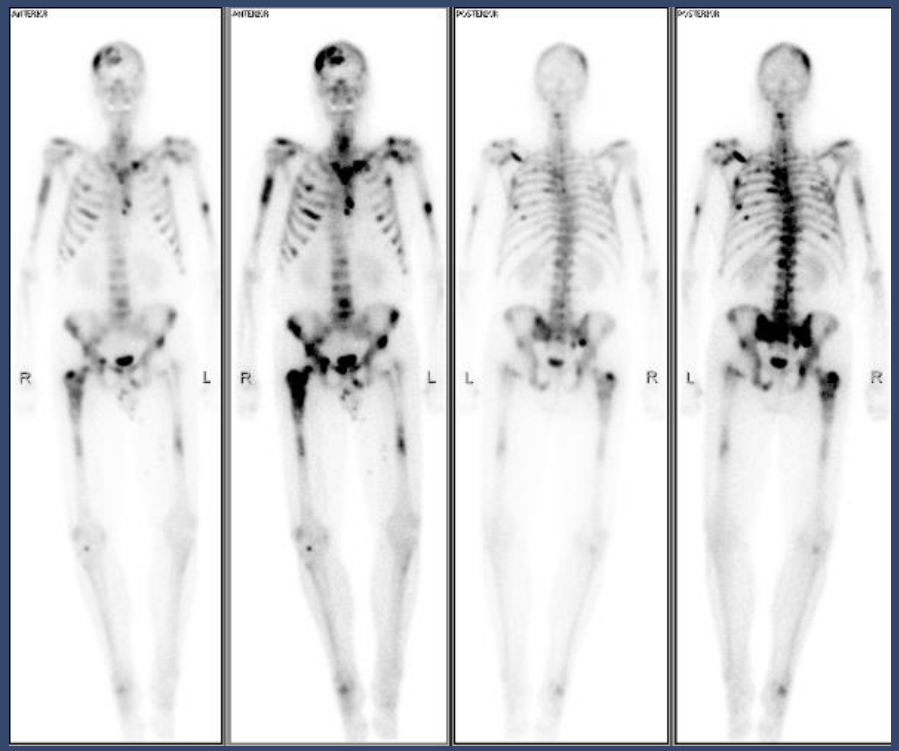
Patient 3



- Bone scan
  - Multiple bone mets

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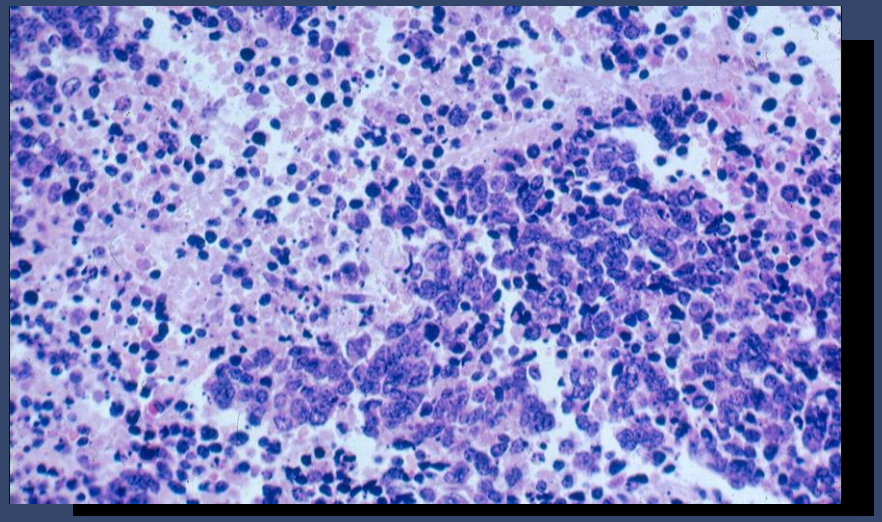
Patient 3

- Labs
  - Alb 2.0
  - Ca 11.1
  - Alk Phos 402
  - WBC 11
  - Hct 22

Patient 3



- Biopsy
  - Femur shows small cell lung
- Diagnosis
  - Metastatic lung carcinoma

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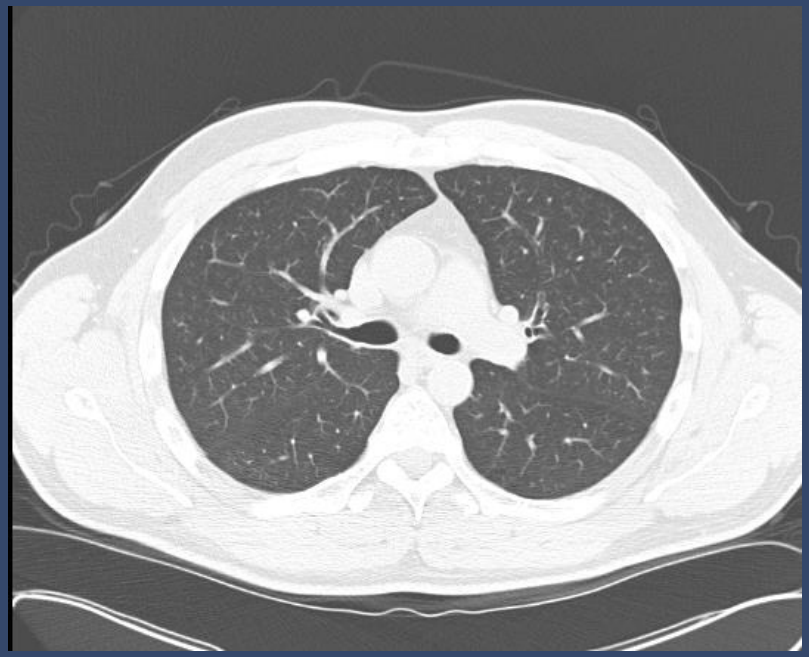
Patient 4

- Summary
  - Older adult
  - Large soft tissue mass
  - Growing
  - Now painful
  - MRI findings
    - Large soft tissue mass
    - Heterogeneous with contrast
- What imaging studies are needed?

Patient 4

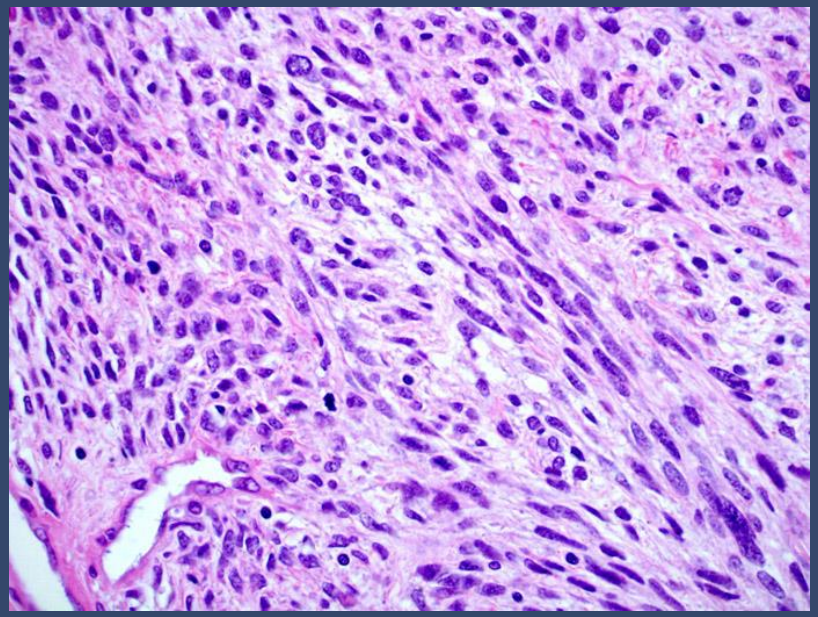
- Summary
  - Older adult
  - Large soft tissue mass
  - Growing
  - Now painful
  - MRI findings
    - Large soft tissue mass
    - Heterogeneous with contrast
- Additional studies
  - CT chest
  - Biopsy

Patient 4



- CT chest with no metastasis

Patient 4



- Pathology: pleomorphic cells, mitotic figures
- Diagnosis: Malignant Peripheral Nerve sheath tumor

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Patient 5

- Summary
  - Young adult
  - Progressive pain
  - No mass
  - X-ray
    - Lytic
    - Epiphyseal
    - Well defined margin
- What additional imaging studies are needed?

Patient 5

- Summary
  - Young adult
  - Progressive pain
  - No mass
  - X-ray
    - Lytic
    - Epiphyseal
    - Well defined margin
- Additional studies
  - MRI with contrast
  - Chest x-ray

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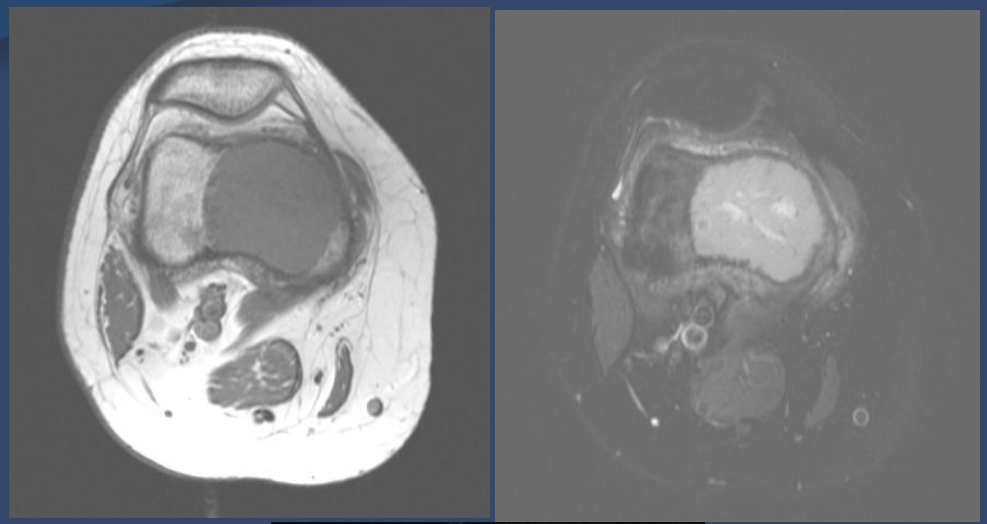
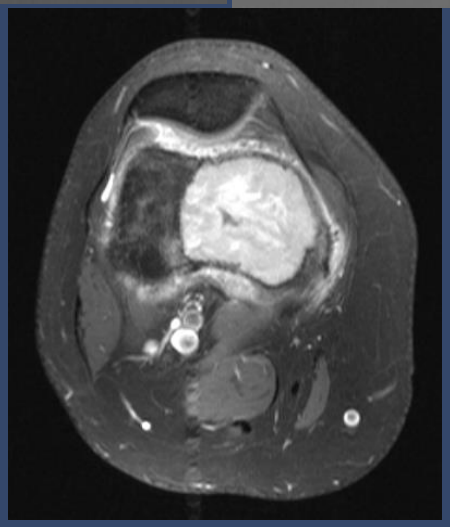
Patient 5





MRI

- Respects cortex
- No fat signal
- Bright fluid signal
- Brightly uniformly enhances

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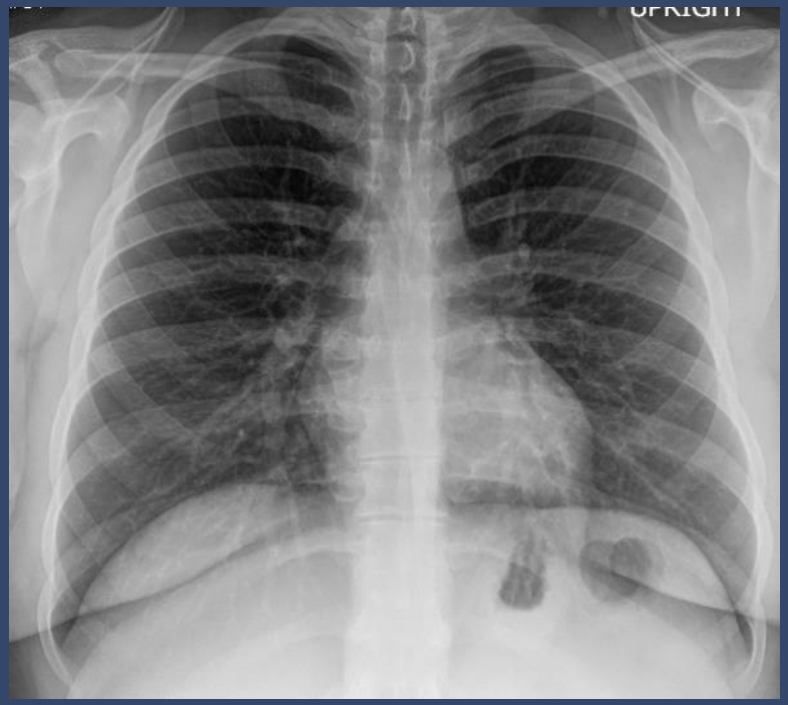
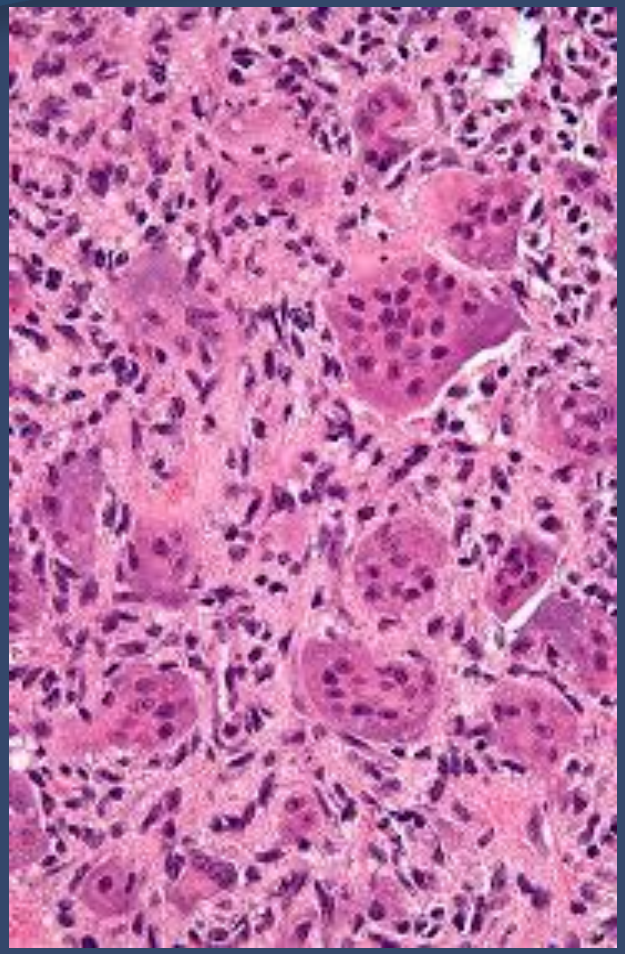
Patient 5



- CXR
  - normal

Patient 5



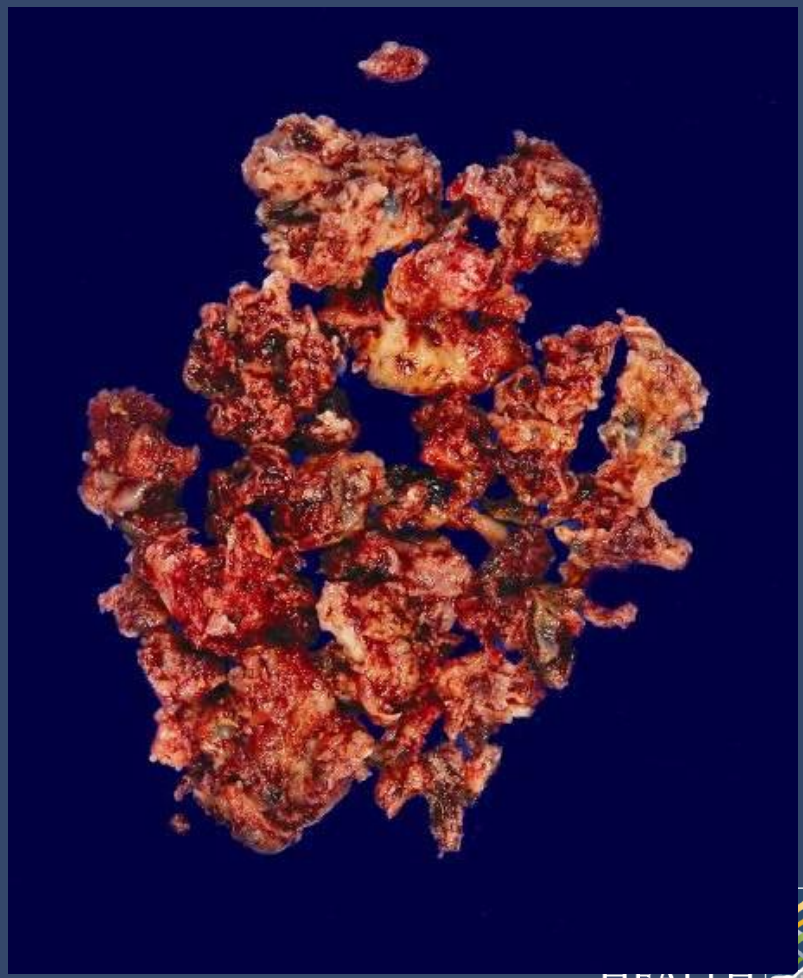
- Giant cell tumor of bone
  - Multiple large multinucleated giant cells
  - Background stromal cells

Treatment – Primary Bone Sarcoma

- Osteosarcoma
  - Chemotherapy
  - Surgery (wide excision)
- Ewing sarcoma
  - Chemotherapy
  - Surgery (wide excision) OR radiation
- Chondrosarcoma
  - Surgery (wide excision)

Treatment – Benign Bone Tumors

- Benign latent
  - Observation
- Benign active or aggressive
  - Surgery: Curettage and packing
  - Surgery: Wide excision
  - Some medications available for unresectable disease



Treatment – Metastatic Disease/Myeloma

- Treat primary tumor
- Osteoclast inhibition
  - Bisphosphonate OR
  - RANKL antibody
- Surgical stabilization for functional pain
- Radiation

Treatment – Soft Tissue Sarcoma

- Surgery (wide excision)
- Radiation
- Chemotherapy - controversy

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Patient 1

- Diagnosis:
  - Osteosarcoma
  - Possible pulmonary metastasis
  - No bone metastasis
- What are the elements of treatment?

Patient 1

- Diagnosis:
  - Osteosarcoma
  - Possible pulmonary metastasis
  - No bone metastasis
- Treatment
  - Chemotherapy
  - Wide surgical resection
  - Reconstruction of knee
  - 7 months duration
- Prognosis
  - 40- 50% 5 yr survival
  - 10 years surveillance

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Patient 1

Patient 2

- Diagnosis
  - Nonossifying fibroma
- What are the elements of treatment?

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Patient 2

- Diagnosis
  - Nonossifying fibroma
- Treatment
  - Unless very large, most will go away shortly after skeletal maturity.
  - If very large, curettage and grafting
- Prognosis
  - Normal growth
  - Normal function

Patient 3

- Diagnosis
  - Wide spread metastatic lung cancer

What are the elements of treatment?

Patient 3

- Diagnosis
  - Wide spread metastatic lung cancer
- Treatment
  - Stabilize fractures
  - Bisphosphonate
  - Radiation therapy
  - Chemotherapy
  - Palliative care/hospice
- Prognosis
  - Poor < 1 yr life expectancy

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Patient 3

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Patient 4

- Diagnosis
  - High grade soft tissue sarcoma
    - Malignant peripheral nerve sheath tumor
- What are the elements of treatment?

Patient 4

- Diagnosis
  - High grade soft tissue sarcoma
    - Malignant peripheral nerve sheath tumor
- Treatment
  - Possible chemotherapy
  - Radiation therapy
    - Pre or post operative
  - Surgical resection
- Prognosis
  - 50% 5 yr survival rate

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Patient 5

- Diagnosis
  - Giant cell tumor of bone
- What are the elements of treatment?

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Patient 5

- Diagnosis
  - Giant cell tumor of bone
- Treatment
  - Curettage, adjuvant therapy, packing
  - Possible stabilization
  - Xgeva (denusomab)
    - RANKL inhibitor
- Prognosis
  - 15-25 % recurrence rate
  - < 5% metastatic rate

Patient 5

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Take Home Points

- Many diagnoses
  - Depend on history, exam, images, biopsy
- Soft tissue tumors
  - 5cm (golf ball) or larger → MRI
  - Larger tumors more concerning for malignancy
- Benign vs Malignant bone tumors:
  - Well circumscribed vs wide zone of transition
  - Soft tissue extension and periosteal changes = malignant

Take Home Points

- Benign bone latent disease
  - Observation
  - Try not to over-image/over-treat
- Benign bone active/aggressive disease or primary bone sarcoma
  - Advanced imaging helpful (MRI)
  - Send to specialist
- Metastatic disease/myeloma
  - Look for hypercalcemia
  - Stage and treat for the original tumor type

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Additional sources

- Books
  - WHO Classification of Tumours of Soft Tissue and Bone. Fourth Edition, 2013
  - Pathophysiology of Orthopaedic Diseases (Great Educators)
  - Orthopedic Knowledge Update Musculoskeletal Tumors 3
  - Surgical Exposures in Orthopaedics: The Anatomic Approach, 2009

- Online sources
  - www.bonetumor.org
  - www.orthobullets.com
  - www.wheelessonline.com

Thank You

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