

Orthopaedic oncology: Lumps, bumps, and heavy metal

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Oregon Health and Science University

AAPA Musculoskeletal Galaxy – June 8th, 2024

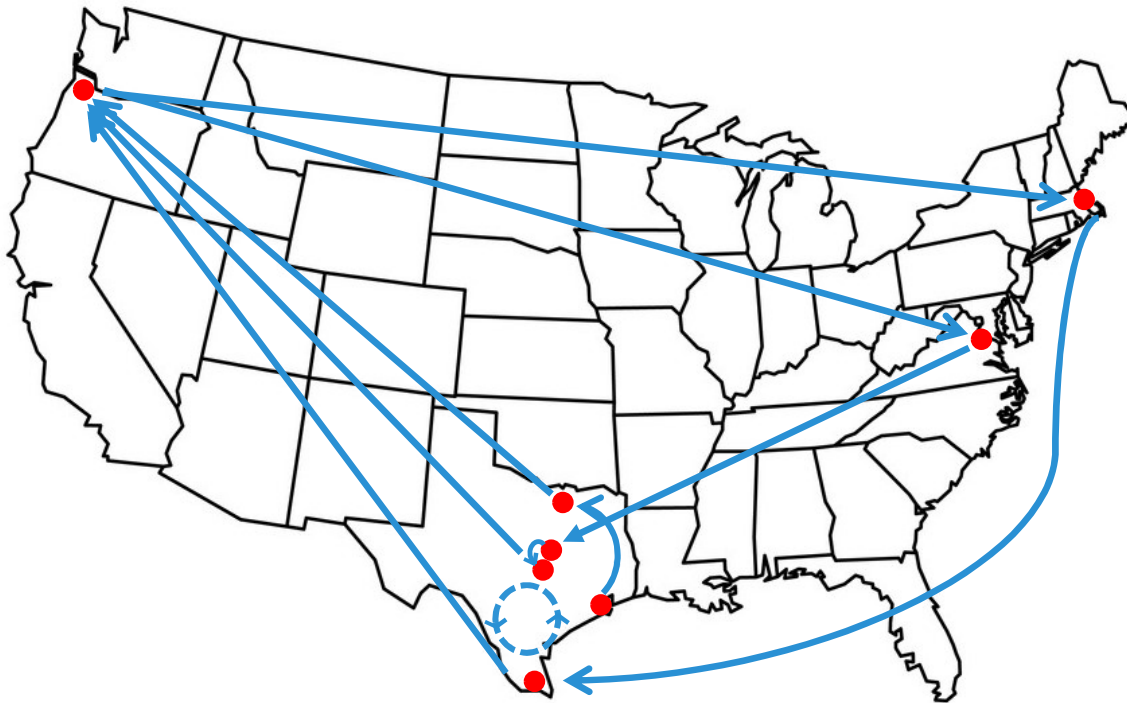


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I have no relevant relationships with ineligible companies to disclose within the past 24 months.



Past me



- **BA:** Reed College (math)
- **MS:** GMU (math)
 - Adjunct professor
 - Research scientist
- **MD, MPH:** UTHSCSA
- **Ortho:** OHSU
- **Ortho oncology:** Harvard/MGH
- **Faculty:** UT Rio Grande Valley
- **Faculty:** OHSU



Objectives

1. be familiar with basic diagnoses in MSK oncology.
2. initiate the diagnostic workup of MSK lesions.
3. understand the basic treatment strategies for MSK tumors.
4. **Know when to refer to orthopaedic oncology.**



The Plan

- What is this even
- Soft tissue sarcomas
- Bone sarcomas
- Benign tumors
- Benign vs malignant
- Metastatic disease of bone
- Cases



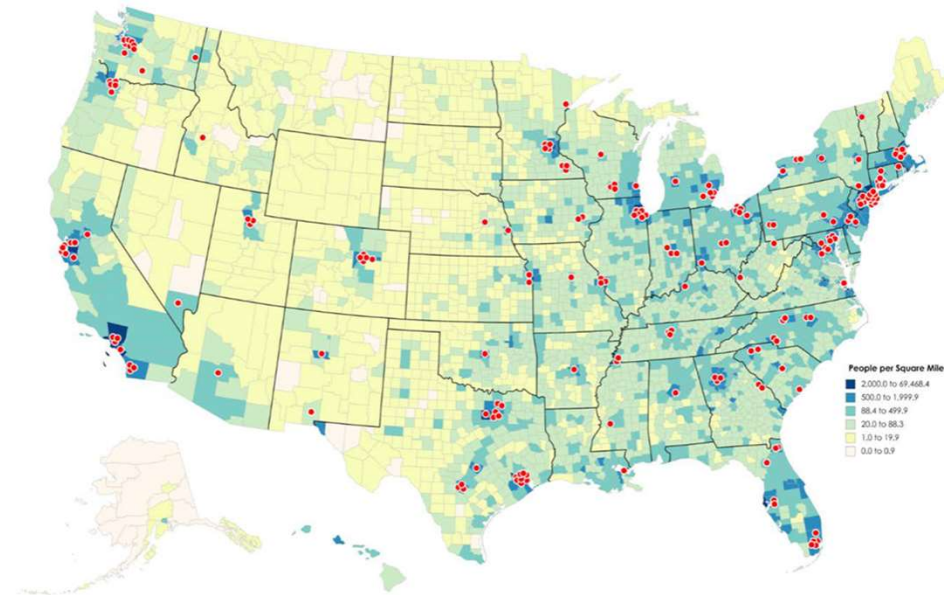
Orthopaedic oncology

Subspecialty of orthopaedics specializing in the **diagnosis and treatment of neoplastic processes involving the musculoskeletal system.**

- Bone sarcomas
- Soft tissue sarcomas
- Benign bone, soft tissue tumors
- Metastatic disease of bone

All ages

Extremities, pelvis, spine, or trunk.



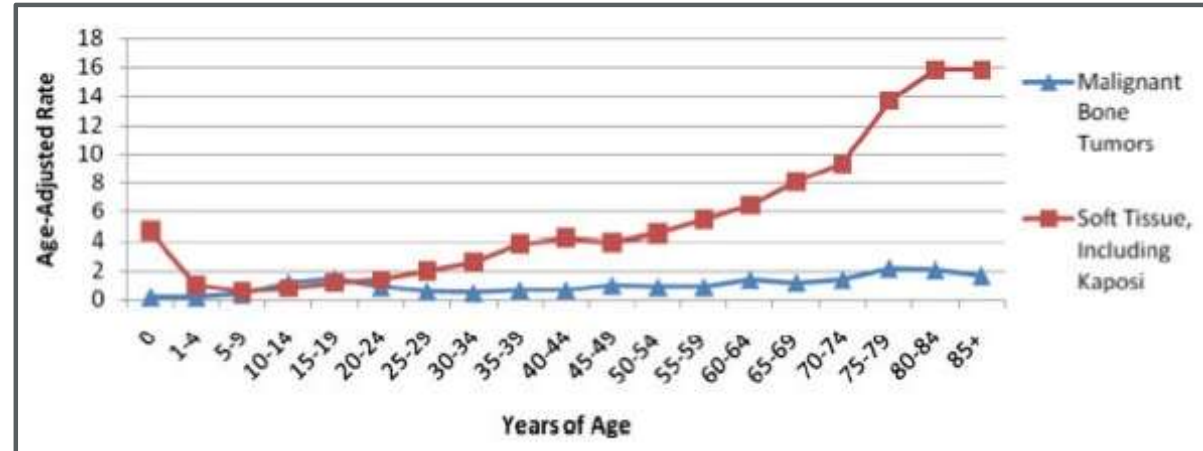
Orthopaedic oncology



Sarcomas

2% of all cancers

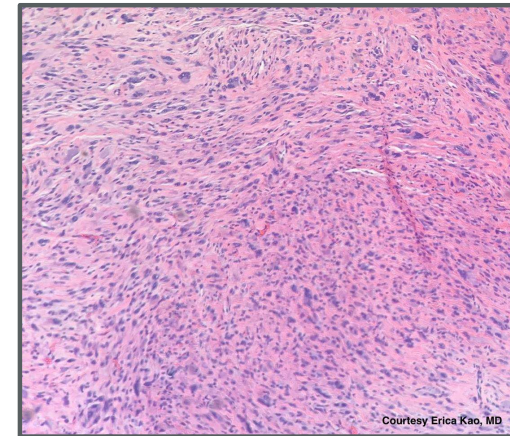
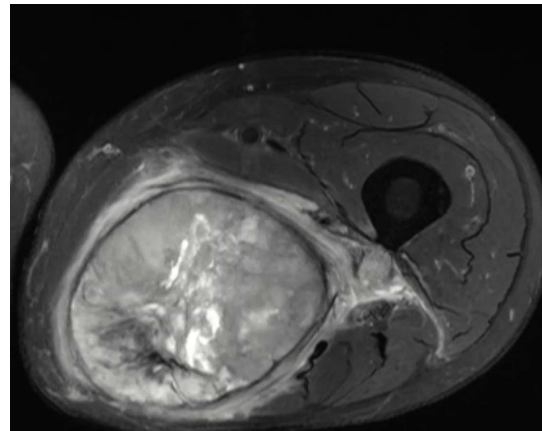
Mesenchymal origin



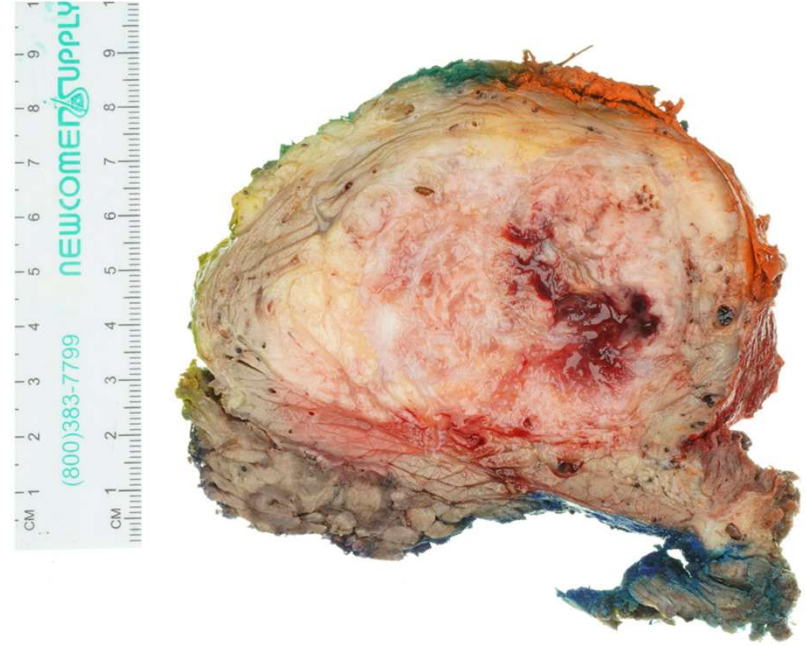
- Bone:
 - Osteosarcoma, Ewing sarcoma, chondrosarcoma
- Soft tissue:
 - Rhabdosarcoma, liposarcoma, fibrosarcoma, **+100 more**

Soft tissue sarcomas

Soft tissue sarcomas: σάρξ = “flesh”“fleshy excrescence.”



Soft tissue sarcomas



Soft tissue ~~sarcomas~~ masses

Q: lipoma or sarcoma?

Benign : malignant = 150 : 1

Worrying signs

- 5cm (golf ball rule)
- Deep (non-mobile)
- Growth

- NOT pain

Rule of Thumb:

Every **growing** soft tissue mass larger than a **golf ball** (> 5 cm) is **sarcoma** until proven otherwise.



Soft tissue ~~sarcomas~~ masses

What Is the Use of Imaging Before Referral to an Orthopaedic Oncologist? A Prospective, Multicenter Investigation

Benjamin J. Miller MD, MS, Raffi S. Avedian MD, Rajiv Rajani MD, Lee Leddy MD, Jeremy R. White MD, Judd Cummings MD, Tessa Balach MD, Kevin MacDonald MD

Unhelpful:

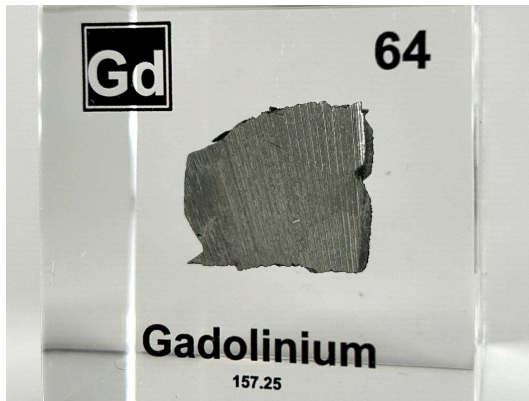
- 76% Ultrasounds
- 74% CT scans
- 62% Bone scans
- 50% PET scans

Utility of imaging studies

	Pro	Indicated?
Serial exams	Cheap, easy, non-invasive	Small, stable lesions
Ultrasound	Cheap, easy, non-invasive	1) Rule-in <i>small</i> lipoma 2) Get MRI approved
X-rays	Cheap-ish, easy, can help dx	1) Always acceptable 2) Gets MRI approved
MRI with contrast	Gold standard	If any worrying signs
CT, PET, bone scan	No	After diagnosis established



STS algorithm



H&P



Imaging



Biopsy



(Staging)



Treatment

Don't be fooled by "trauma."

- 1) X-rays
- 2) MRI **with contrast**

By the *definitive surgeon*

- 1) Radiotherapy
- 2) Surgery
- 3) +/- chemotherapy



Benign soft tissue masses

- Lipomas vs atypical lipomatous tumors

Lipomas vs atypical lipomatous tumors (ALTs)

	Lipoma	ALT
Benign	Benign	Benign (locally aggressive)
Size	Smaller, superficial	Larger, deep
Internal characteristics	Bland, homogeneous	Septations
Recurrence	~0%	~25%
Malignant transformation	0%	~5%



Desmoid tumor (fibromatosis)

- Benign aggressive soft tissue tumor
- Young adults, F>>M
- Hormone-related
- Familial adenomatous polyposis (FAP)

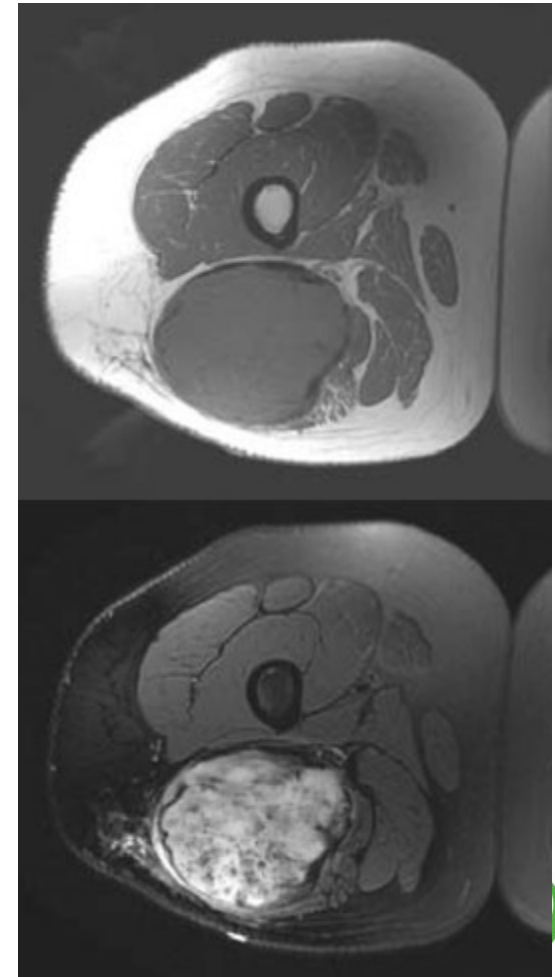
- Observation vs medical mgmt

The NEW ENGLAND JOURNAL of MEDICINE

RESEARCH SUMMARY

Nirogacestat, a γ -Secretase Inhibitor for Desmoid Tumors

Gounder M et al. DOI: 10.1056/NEJMoa2210140



Case 1: Soft tissue sarcoma

64M right-hand dominant

- 9 months of growing mass
- Past smoker, 30 pack-years

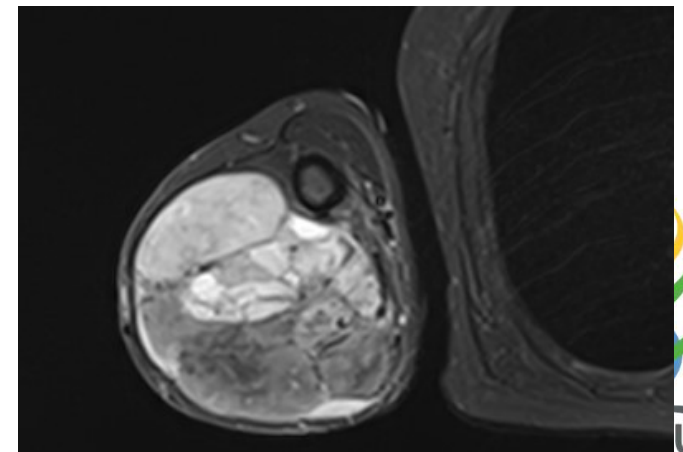
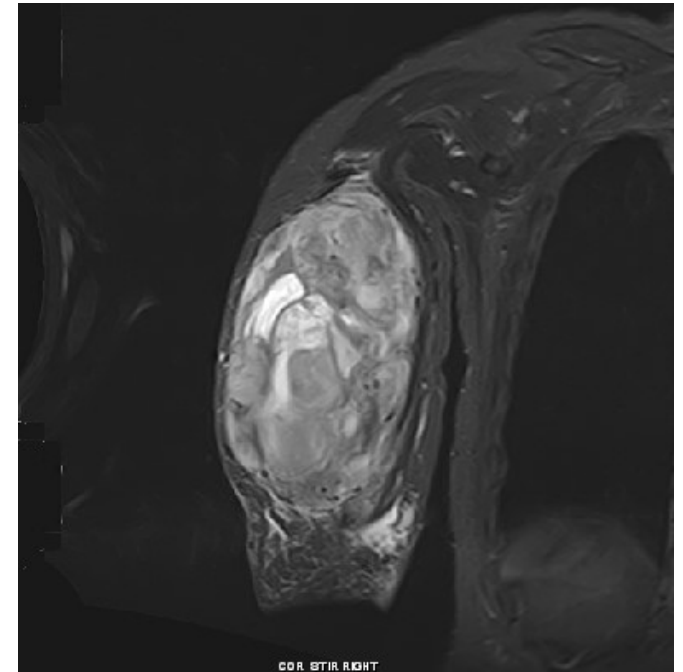
Worrying signs

- ✓ 5cm (golf ball rule)
- ✓ Deep (non-mobile)
- ✓ Growth



Case 1: Soft tissue sarcoma

1. H&P
2. X-rays
3. MRI with contrast
4. Percutaneous biopsy
 - Undifferentiated pleomorphic sarcoma
5. Staging studies (CT chest)
6. Treatment...



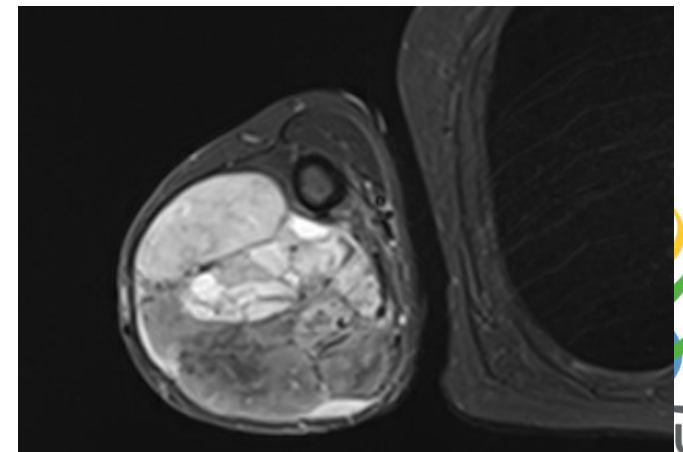
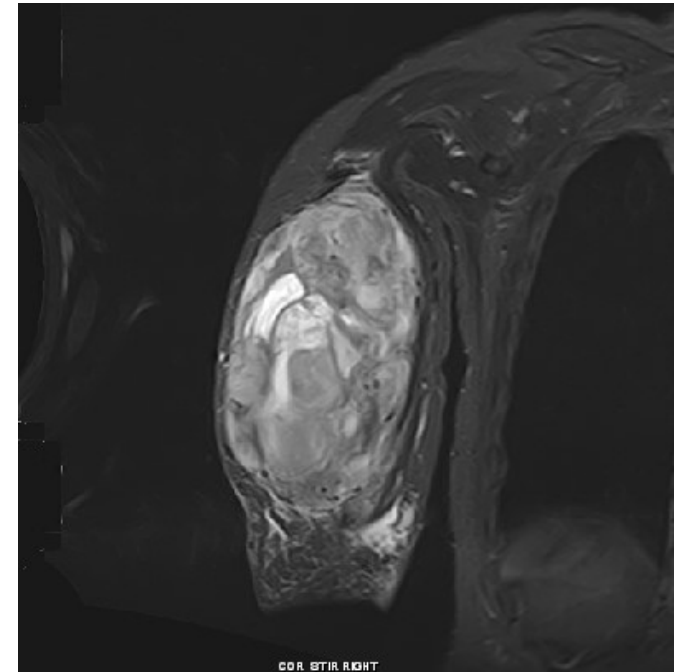
Case 1: Soft tissue sarcoma

Plan:

- 50Gy neoadjuvant radiation therapy
- Day 3: radial nerve symptoms

New plan:

- Surgery
- 60+ Gy adjuvant radiation therapy



Case 1: Soft tissue sarcoma



Whoops procedures

- “Unplanned intra-lesional procedure”
- 1/3 of sarcoma referrals in some centers

- Higher risk of positive margins on re-resection
- Higher morbidity
- Higher risk of eventual amputation

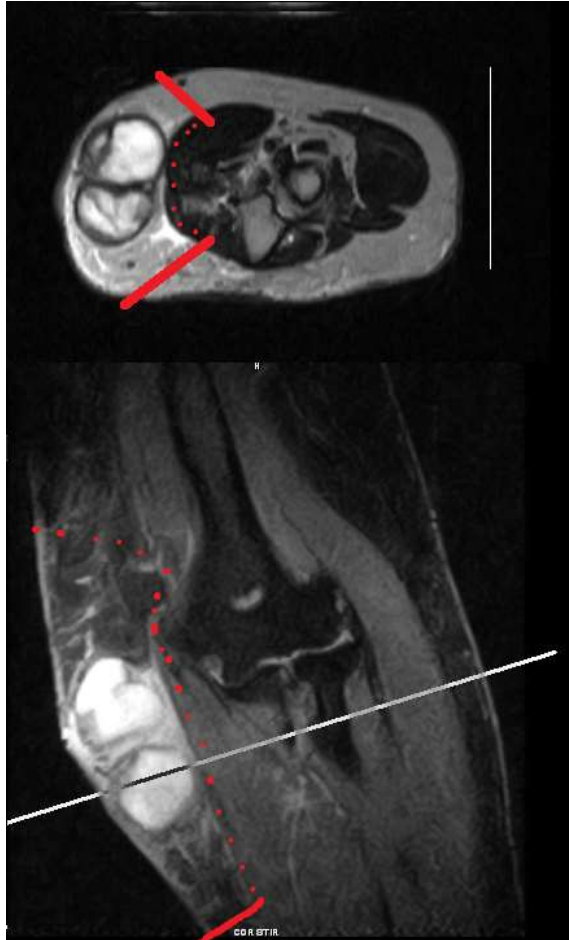
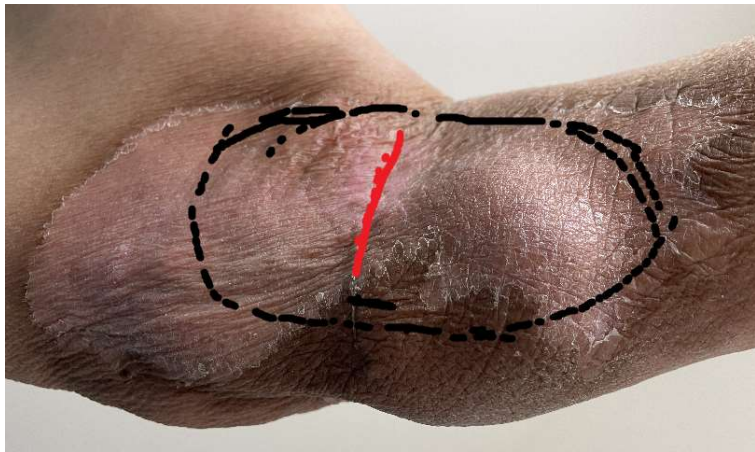


Whoops procedures

- 60's yo F
- American-healthy
- Partial excision of sarcoma by outside gen surg



Whoops procedures



Whoops procedures



Whoops procedures

Rule of Thumb:

Every **growing** soft tissue mass larger than a **golf ball** (> 5 cm) is **sarcoma** until proven otherwise.

- Refer, refer, refer
 - Preferably to **ortho oncology**
- Chronic hematomas (almost) don't exist



Bone ~~sarcomas~~ lesions



Worrying signs

- > 5cm
- Irregular borders
- Pain
- Swelling



1. X-ray whole bone
2. MRI **with contrast whole bone**

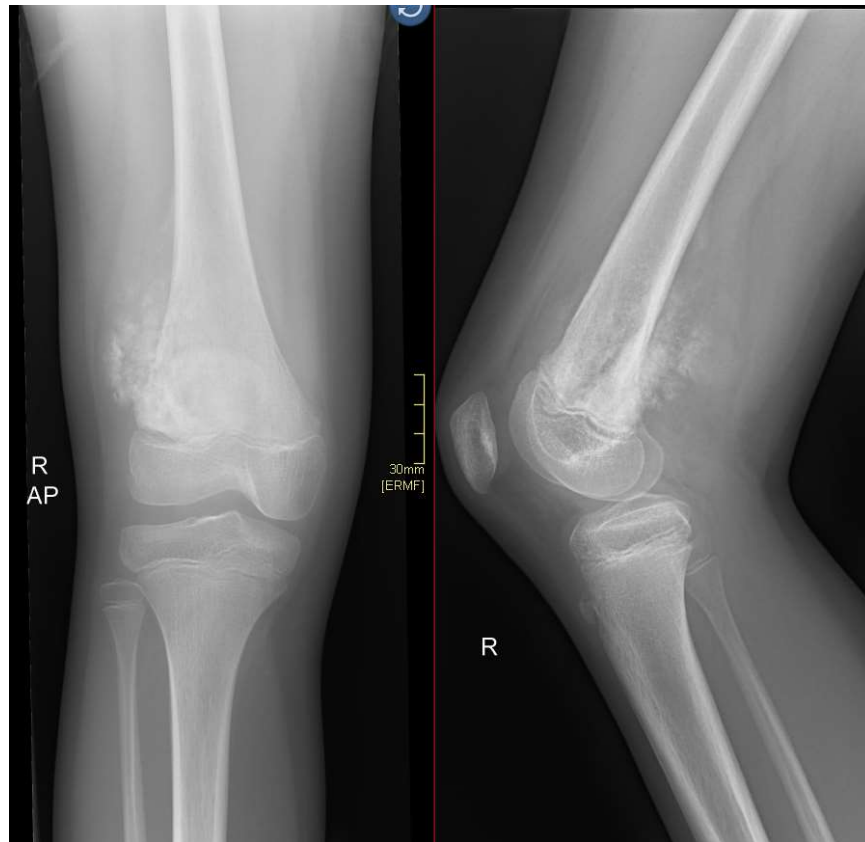
Benign vs malignant bone tumors

	Benign	Malignant
Transition zone	Narrow	Broad
What's tumor doing to bone?	Respects cortex	Cortical destruction
Periosteal reaction (What's bone doing to tumor?)	Smooth	Irregular (sunburst, onion-skin, Codman's Triangle)
Soft tissue mass	Rare	Common



Pediatric osteogenic sarcoma
Han Jo Kim^a, Peter N. Chalmers^b and Carol D. Morris^c

Benign, malignant, or normal?



Benign, malignant, or normal?

Unicameral bone cyst

- Benign
- Kids
- Not neoplastic



Benign, malignant, or normal?

Enchondroma

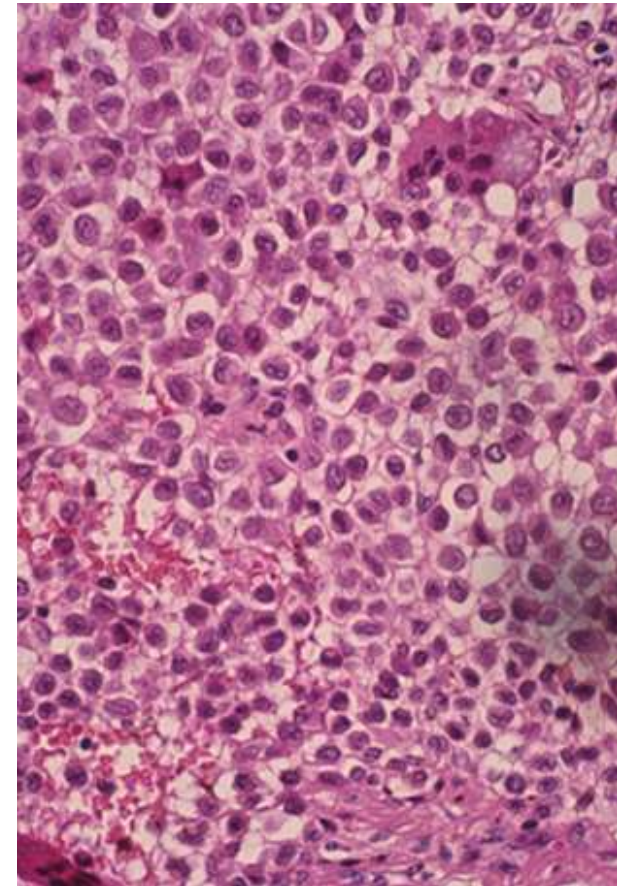
- Benign latent
- Adults



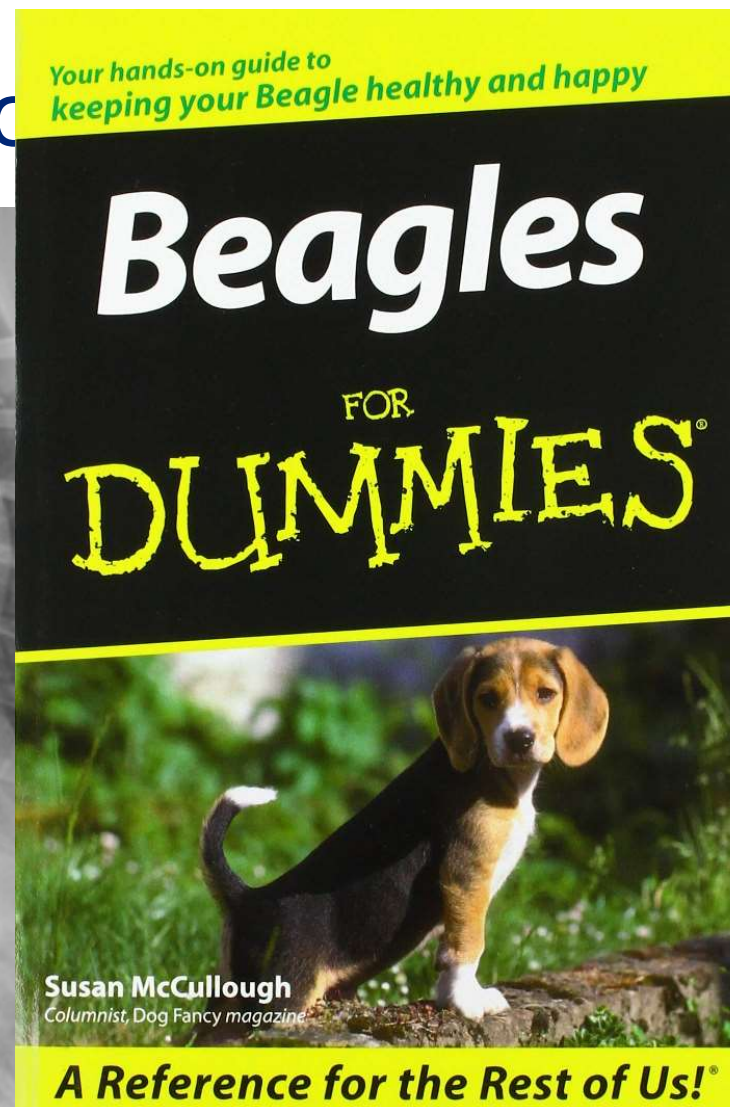
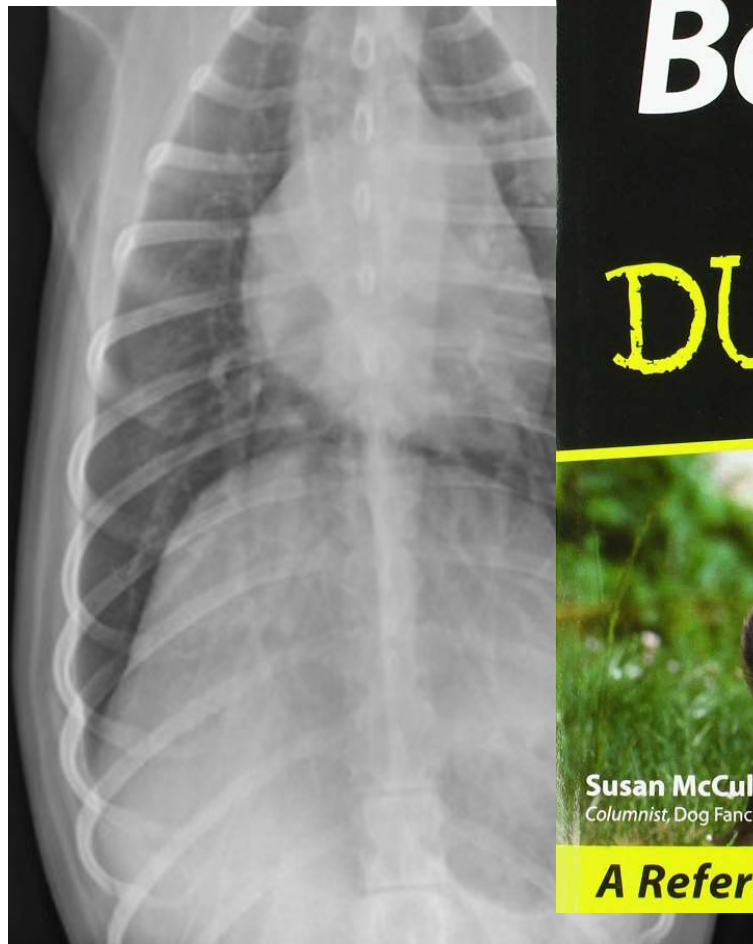
Benign, malignant, or normal?

Chondroblastoma

- Benign active
- Adolescents



Benign, malignant, or no



Benign bone tumors

- **Benign latent**

- Enchondromas
- Non-ossifying fibromas

Observe

- **Benign active**

- Osteoid osteoma
- Chondroblastoma
- Fibrous dysplasia

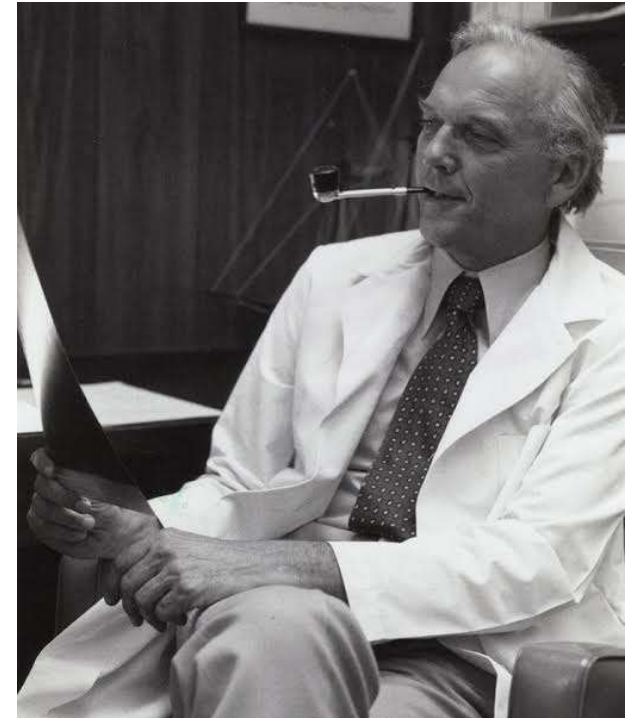
Observe vs curettage

- **Benign aggressive**

- Aneurysmal bone cyst
- Giant cell tumor of bone

Curettage

William Enneking



Benign latent bone tumors

Non-ossifying fibromas

- Skeletally immature
- Lytic, eccentric, metaphyseal, sclerotic rim
- Found incidentally
- **Resolve on their own***



Benign latent bone tumors

Enchondromas

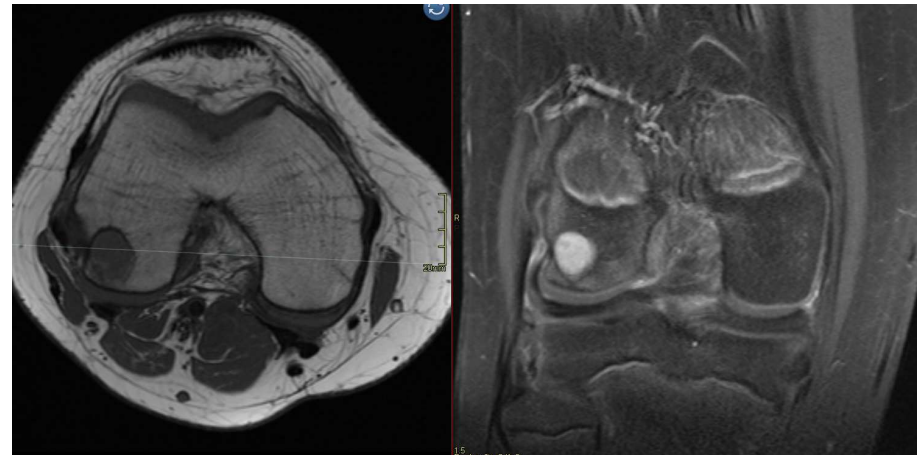
- Skeletally mature
- Metaphyseal, popcorn calcifications
- Found incidentally
- **Keep forever***



Benign active bone tumors

Chondroblastoma

- “Bar mitzva tumor”
- Lytic, epiphyseal
- **Curettage, bone graft**



Benign active bone tumors

Osteochondromas

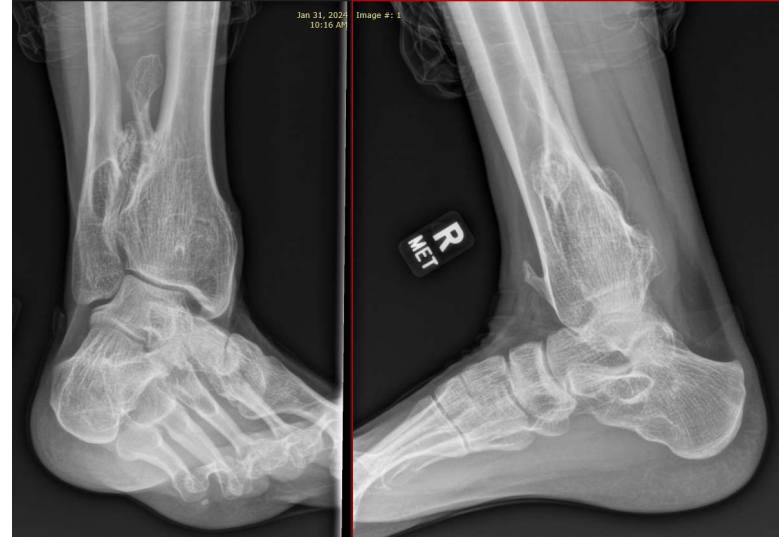
- Solitary or syndromic
 - multiple hereditary exostosis (MHE)
- Grow until skeletal maturity
- 1% risk of malignant transformation

- **Excise symptomatic lesions**



Multiple hereditary exostosis

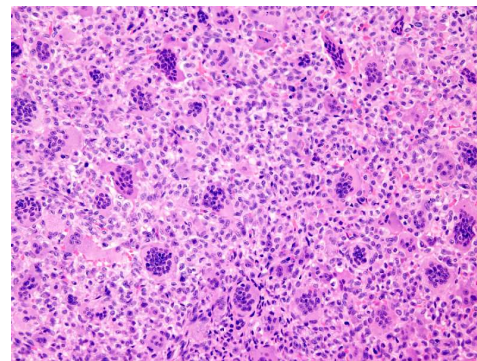
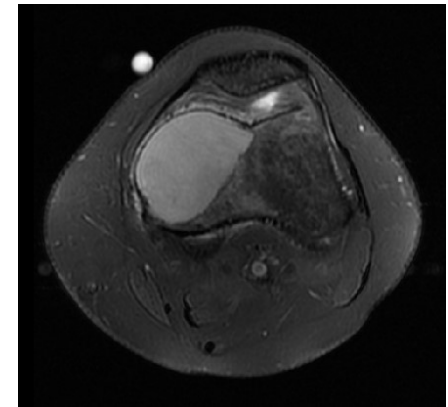
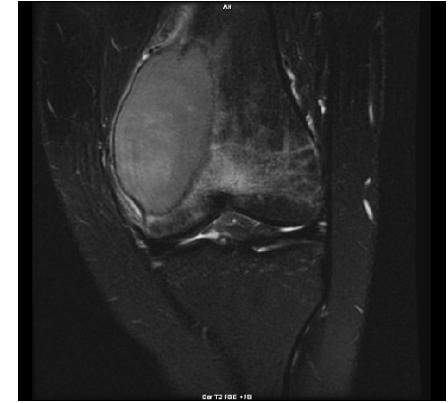
- aka osteochondromatosis
- 5% lifetime risk of chondrosarcoma
- Removal only of symptomatic lesions



Benign aggressive bone tumors

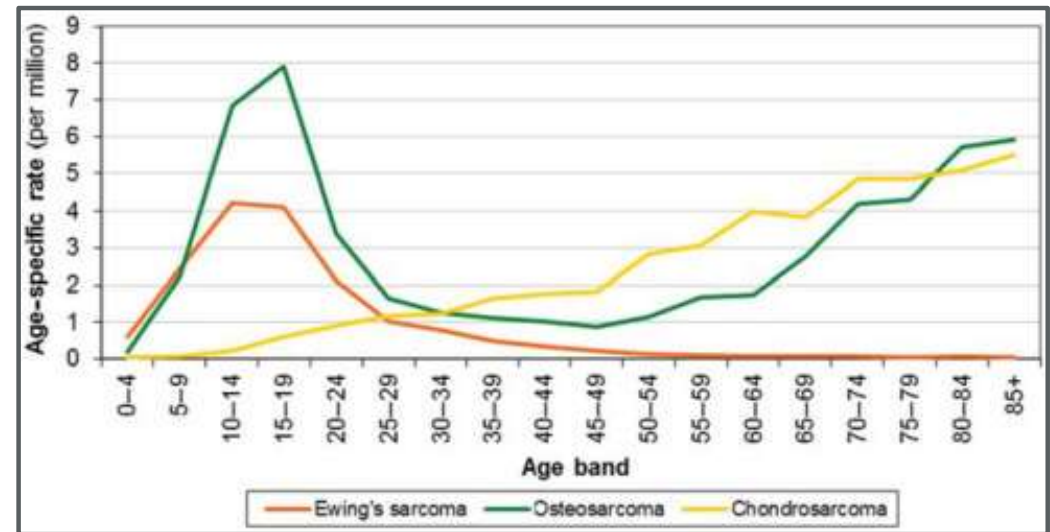
Giant cell tumor of bone

- Young adults
- Destructive
- 3-5% lung mets
- **Curettage, bone graft vs cement**



Bone sarcomas

- Osteosarcoma
- Ewing sarcoma
- Chondrosarcoma



Bone sarcomas

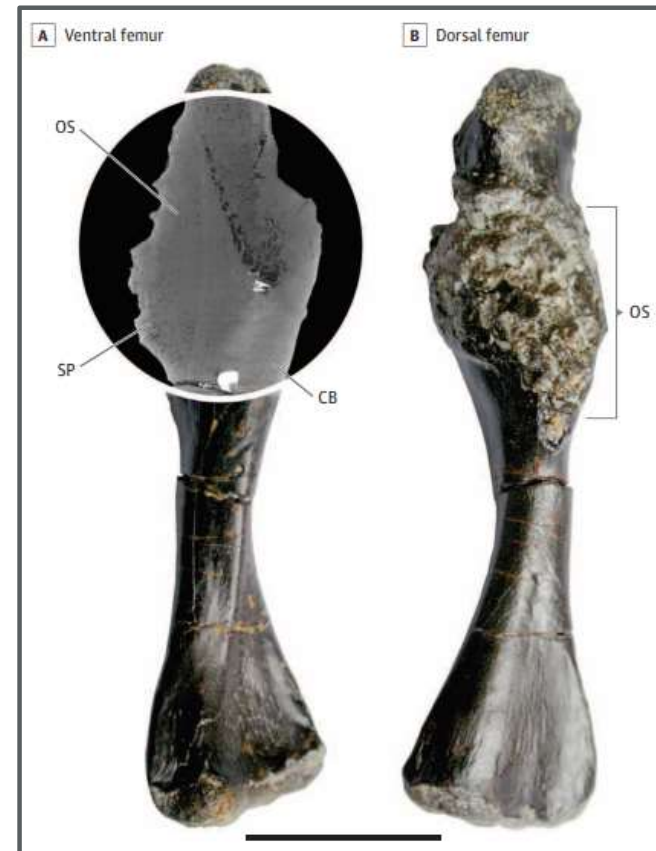
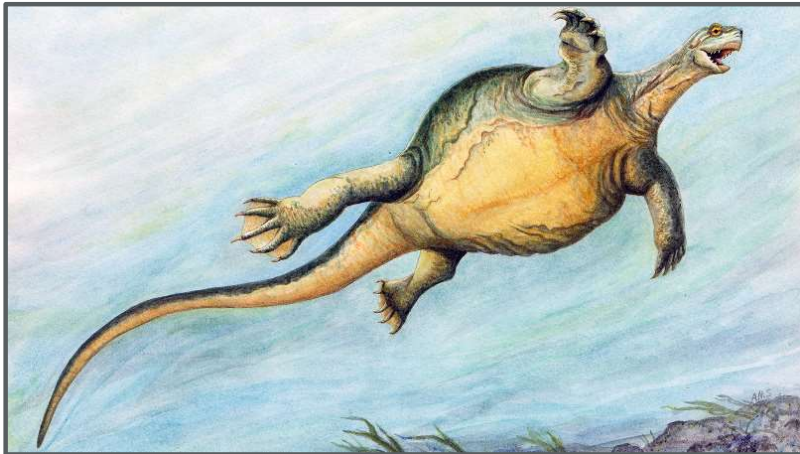
Research Letter

Triassic Cancer—Osteosarcoma in a 240-Million-Year-Old Stem-Turtle

Yara Haridy, MS; Florian Witzmann, PhD; Patrick Asbach, MD; et al.

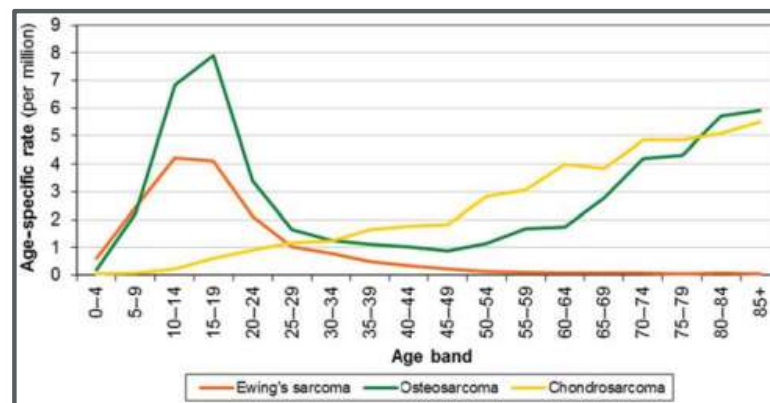
[Abstract](#) | [Full Text](#)

JAMA Oncol. Published online February 7, 2019. doi:10.1001/jamaoncol.2018.6766

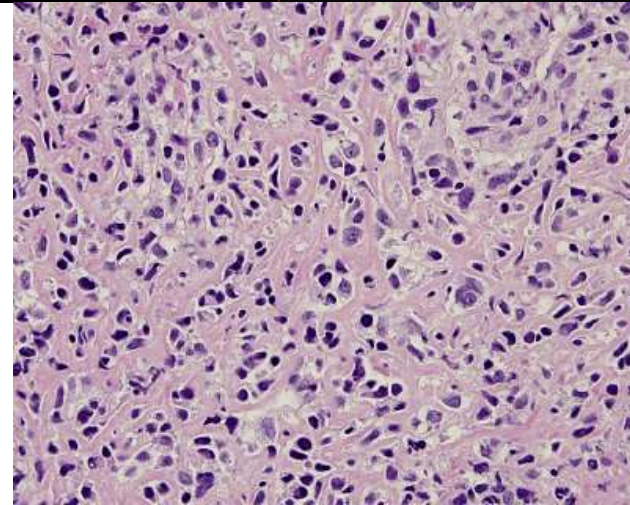
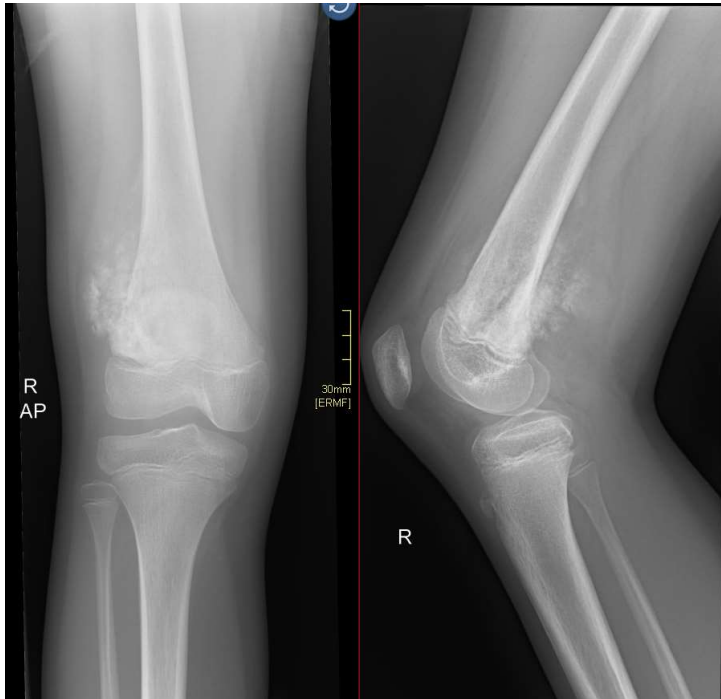


Bone sarcomas

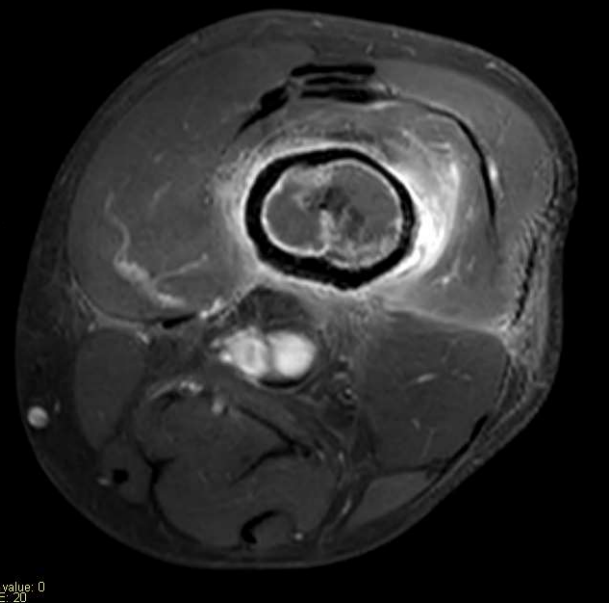
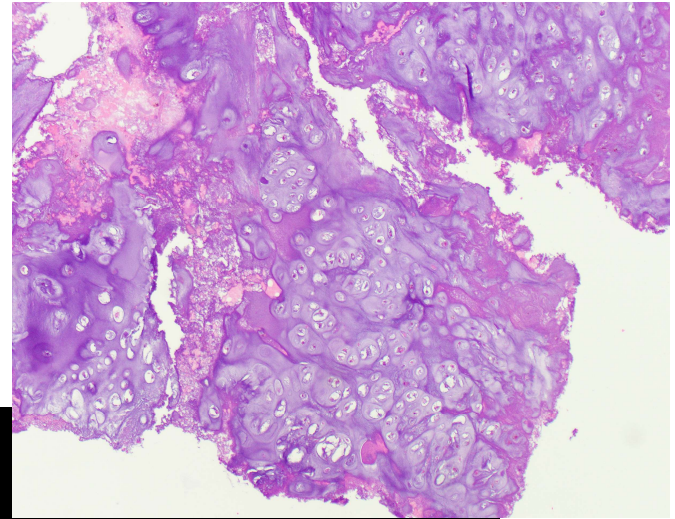
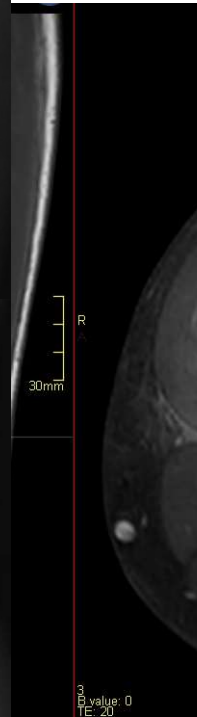
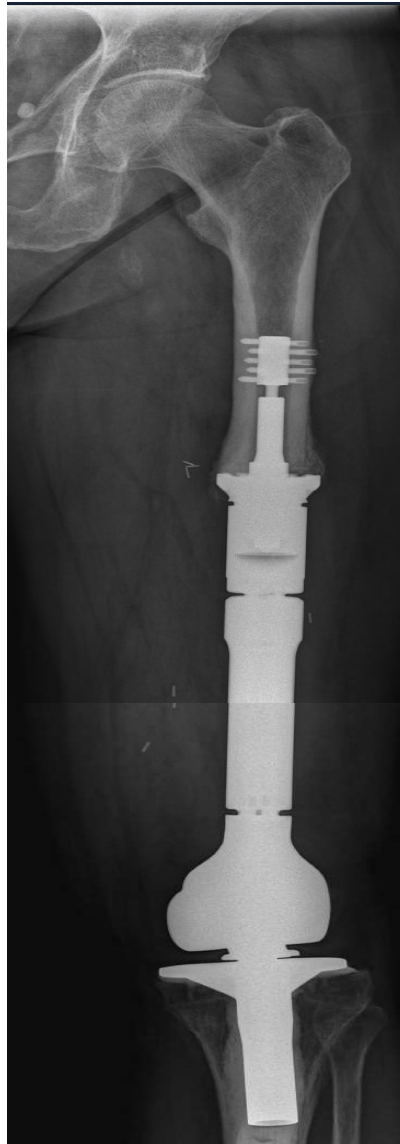
Osteosarcoma	Ewing sarcoma	Chondrosarcoma
Children/adolescents Older adults	Children/adolescents	Adults
Long bone metaphysis	Long bone diaphysis Flat bones/axial	Long > pelvis
Osteoid	Small, round, blue	Pastel chondrocytes
Chemo, surgery	Chemo, surgery Chemo, radiation	Surgery



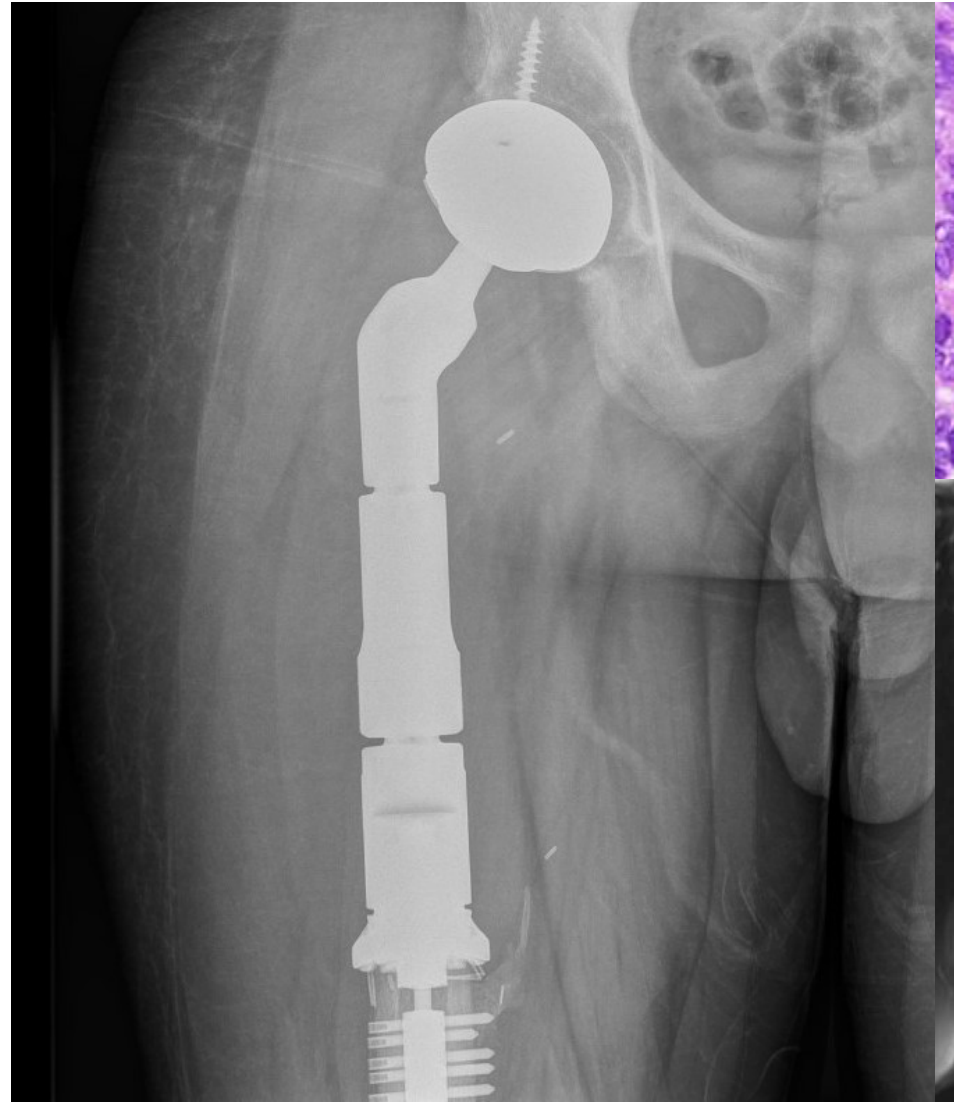
Osteosarcoma

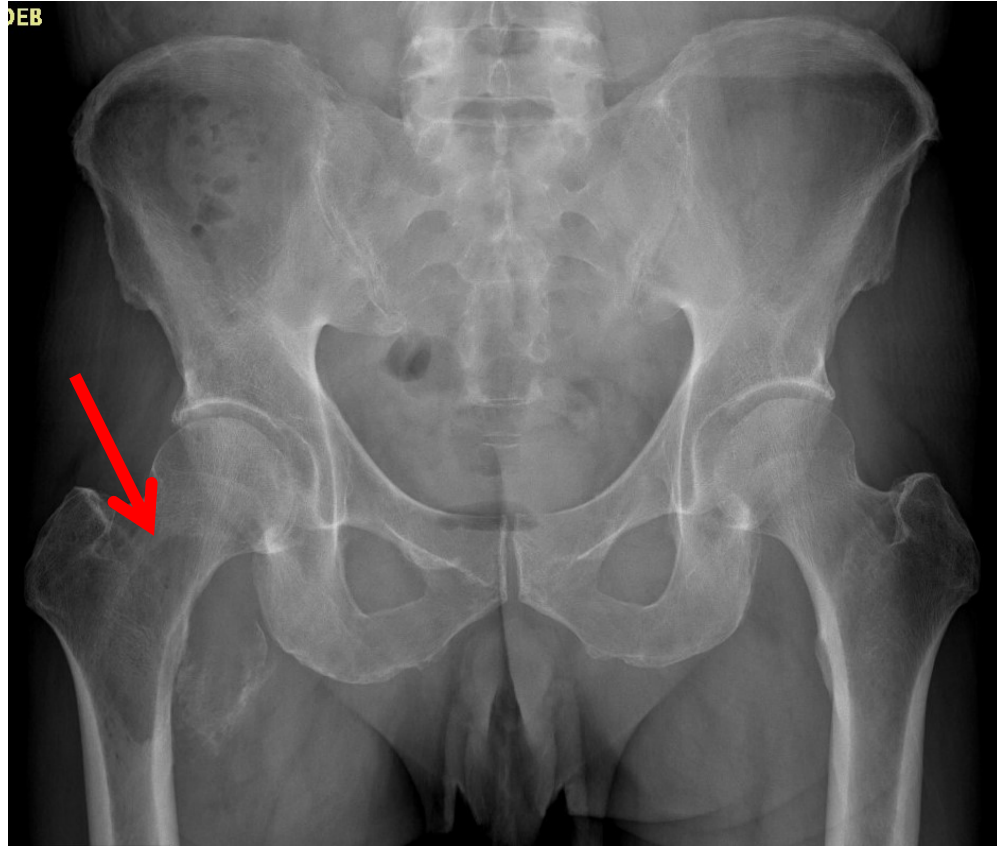


Chondrosarcoma



Ewing sarcoma





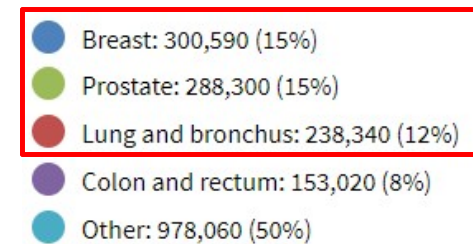
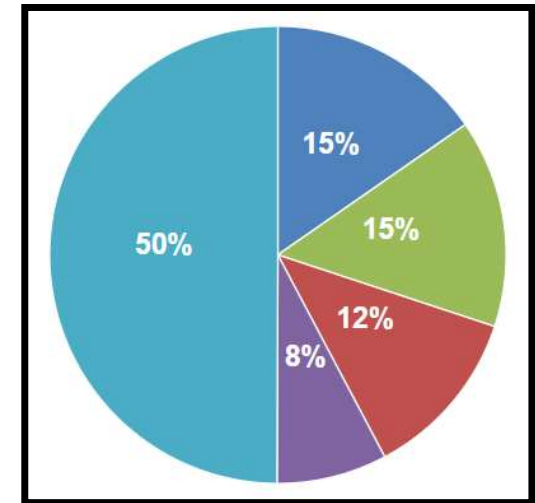
- 70M with right hip pain
- Decades-long smoker

Differential diagnosis:

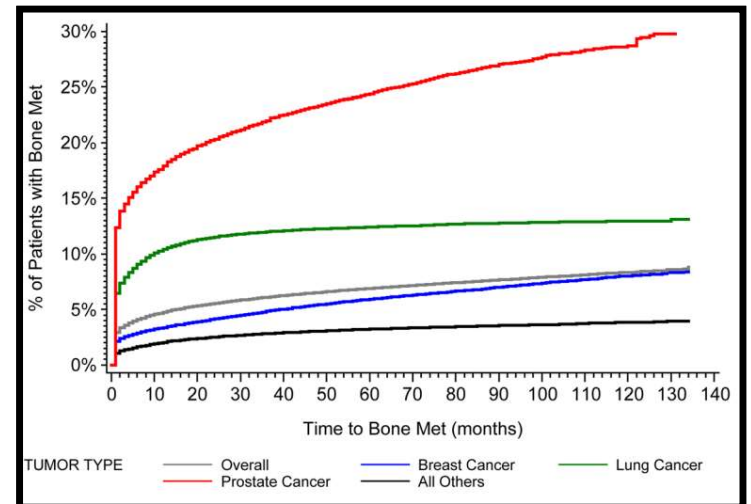
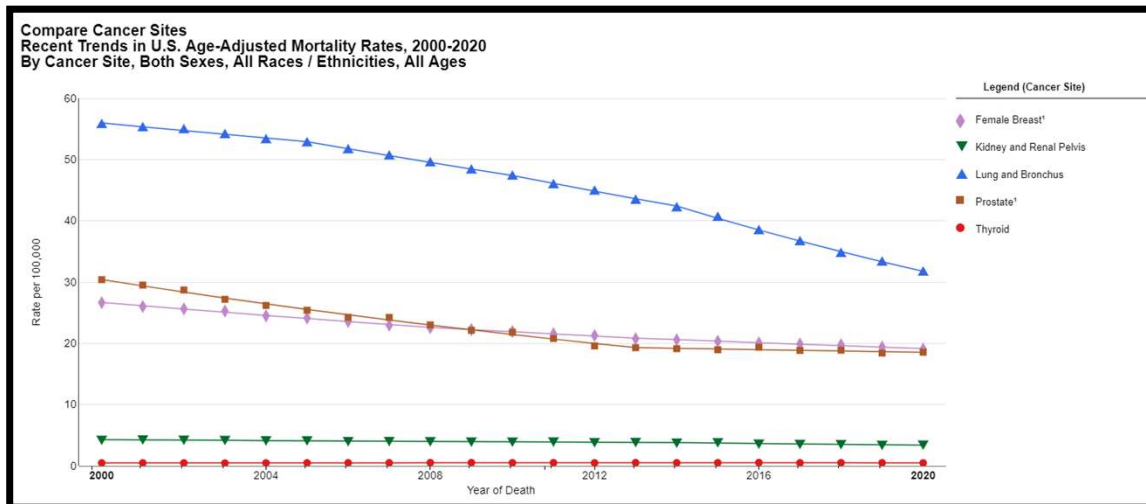
- Metastatic disease of bone
- Metastatic disease of bone
- Metastatic disease of bone
- Myeloma
- Lymphoma

Metastatic disease of bone

- 2 million new cancer diagnoses
- 45% are the Big 5 (900K!)
 - **Breast, prostate, lung, thyroid, kidney**
- 40% of bony mets are to pelvis



Metastatic disease of bone



Hernandez RK, Wade SW, Reich A, Pirulli M, Liede A, Lyman GH. Incidence of bone metastases in patients with solid tumors: analysis of oncology electronic medical records in the United States. BMC cancer. 2018 Dec;18:1-1.



Hole in (adult) bone

Skeletal metastases of unknown origin. A prospective study of a diagnostic strategy.

Rougraff, B T; Kneisl, J S; Simon, M A

[Author Information](#) ✓

The Journal of Bone & Joint Surgery 75(9):p 1276-1281, Sep 1993.

Labs

- CBC w/ diff
- CMP
- ESR/CRP
- SPEP, UPEP
- PSA
- TSH
- PTH
- LFTs
- **Ionized Calcium**

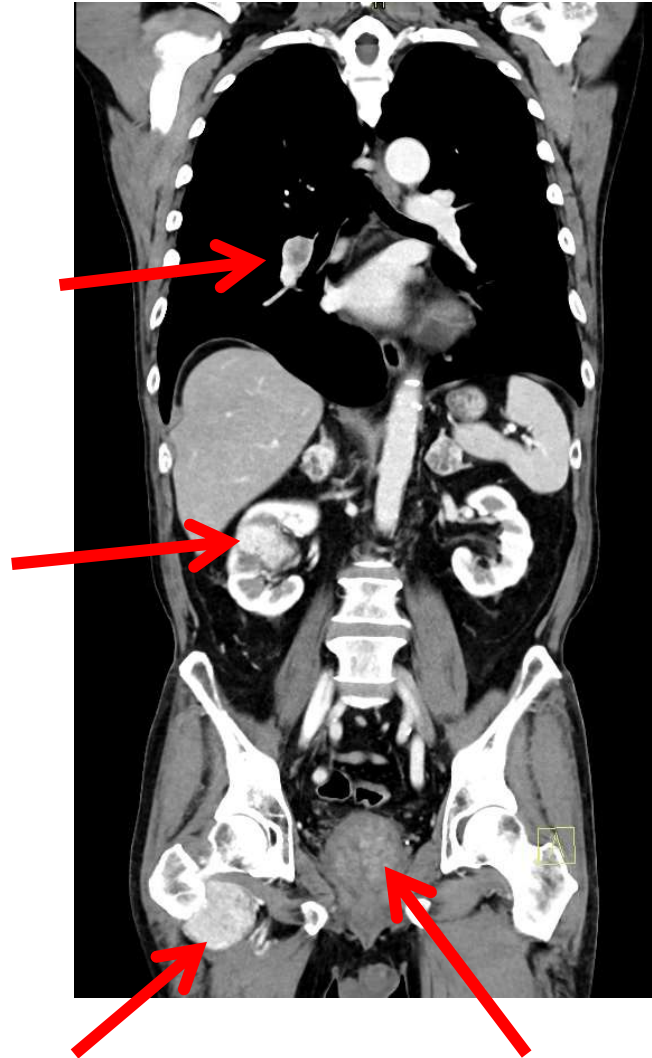
Imaging

- Xray of whole bone
- CT chest/abd/pelvis
- +/- Cross-sectional





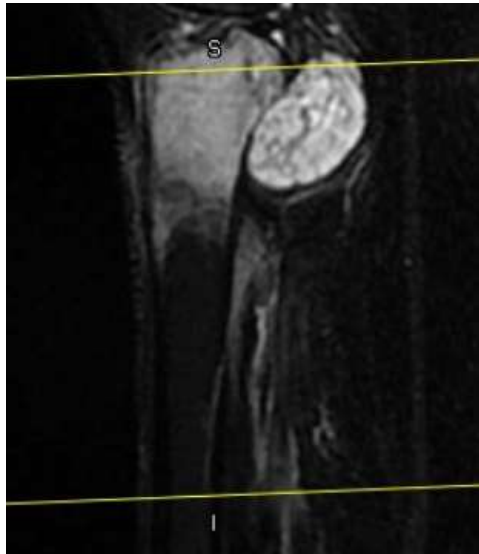
↑ PSA



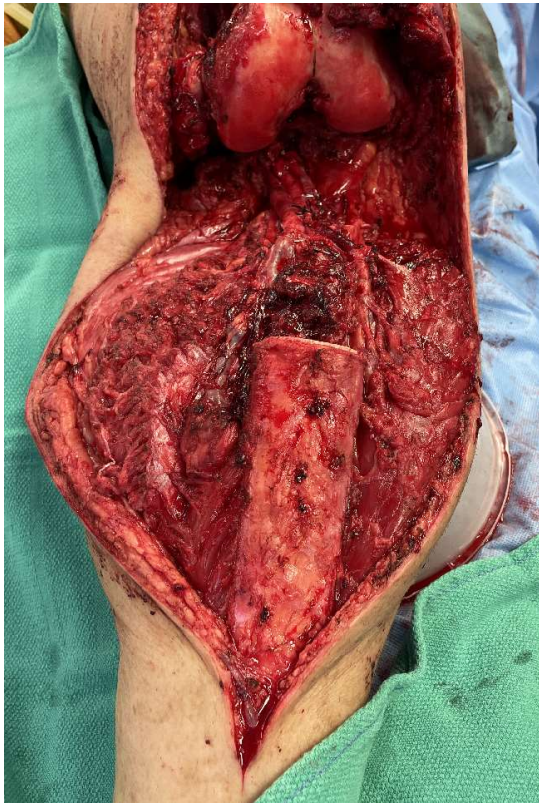
Case 3

45M, known osteochondromas

- DM, A1c = 11
- New onset left leg pain, swelling



Case 3

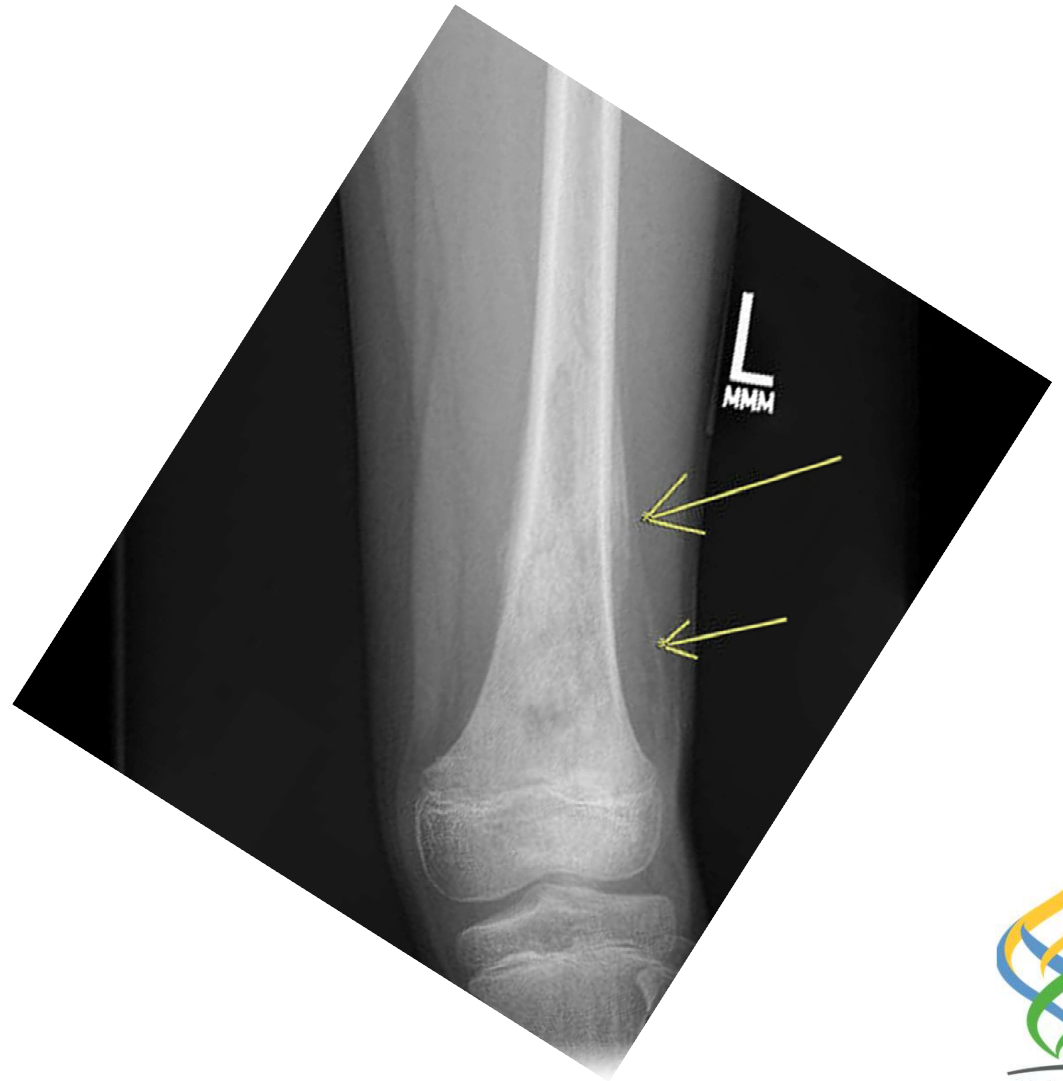


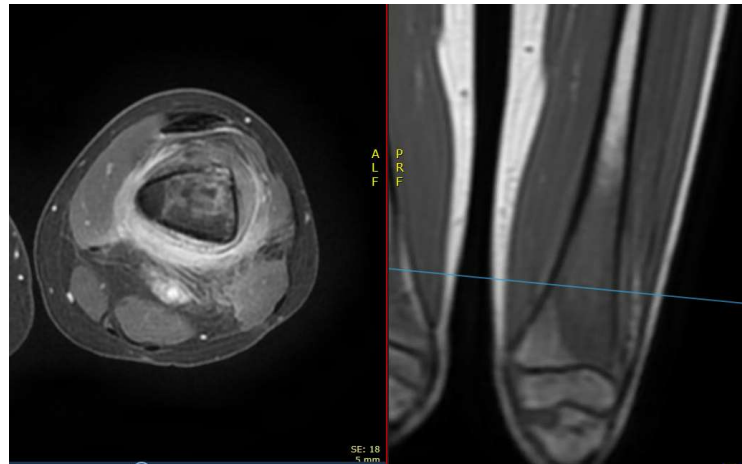
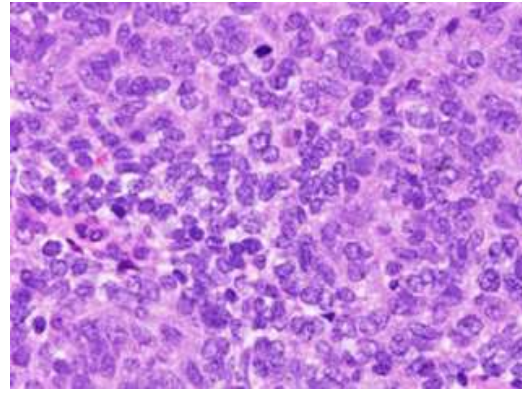
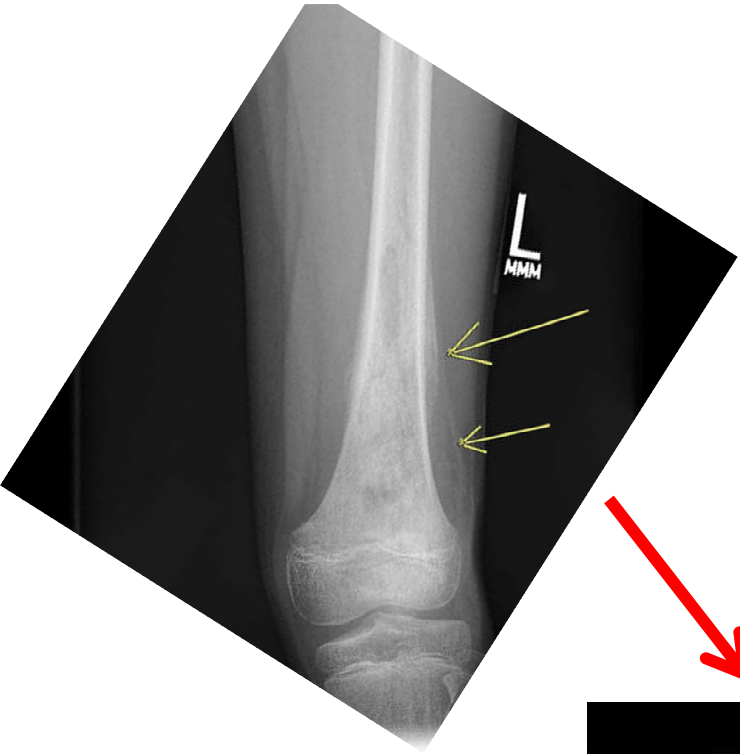
Case 3



Case 2

- 7yo girl, knee pain





Rotationplasty

- Procedure for lower extremity bone deficiency
- Biologic reconstruction
- Ankle replaces knee
 - Allows BKA prosthesis







Short story

Soft tissue masses

Worrying signs:

- 5cm (Golf ball)
- Growing
- Deep (non-mobile)

Work-up:

- X-rays
- MRI *with contrast*
- Refer

Bone masses

Worrying signs:

- 5cm
- Irregular
- Pain
- Swelling

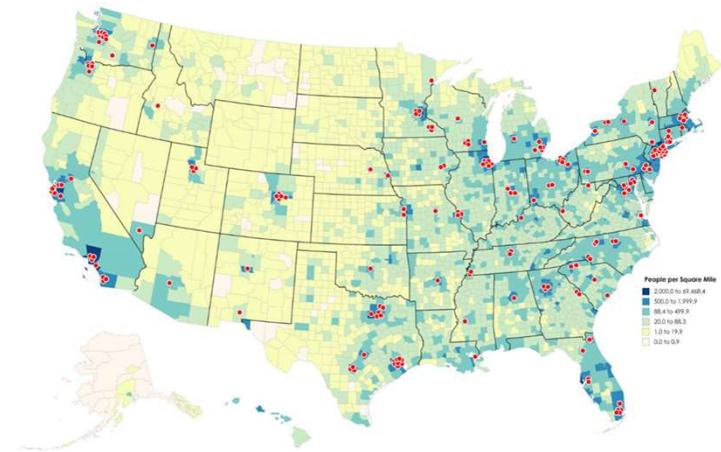
Work-up:

- X-rays
- MRI *with contrast*
- Refer



Shorter story

- Look up your local orthopaedic oncologist



- Reach out ramseyd@ohsu.edu – it's a small small world



References

- Aboulafia AJ, Kennon RE, Jelinek JS. **Benign bone tumors of childhood.** JAAOS-Journal of the American Academy of Orthopaedic Surgeons. 1999 Nov 1;7(6):377-88.
- Gounder M, Ratan R, Alcindor T, Schöffski P, Van Der Graaf WT, Wilky BA, Riedel RF, Lim A, Smith LM, Moody S, Attia S. **Nirogacestat, a γ -secretase inhibitor for desmoid tumors.** New England Journal of Medicine. 2023 Mar 9;388(10):898-912.
- Jang E, Danford NC, Levin AS, Tyler WK. **Intra-Articular Tumors: Diagnosis and Management of the Most Common Neoplasms Involving Synovial Joints.** JBJS reviews. 2018 Dec 1;6(12):e8.
- Rougraff BT, Kneisl JS, Simon MA. **Skeletal metastases of unknown origin.** A prospective study of a diagnostic strategy. JBJS. 1993 Sep 1;75(9):1276-81.
- Steffner RJ, Jang ES. **Staging of bone and soft-tissue sarcomas.** JAAOS-Journal of the American Academy of Orthopaedic Surgeons. 2018 Jul 1;26(13):e269-78.



