MSK Onco

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Orthopedic Surgery

PHSU

Bone Tumors

- **Benign**
  - **1** Mostly hamartomas-Tumor like conditions.
    - NOF, UBC, enchondromas, Exostosis
  - **2** Some are neoplasms.
    - GCT, ABC, Chondroblastomas, osteoblastomas
- **Malignant**
  - **1** Secondary (Mets)
    - Elsewhere, foreign
    - **Common**
  - **2** Primary (Sarcomas)
    - Locals, natives
    - **Rare**

Malignant Bone Tumors

- Adenocarcinomas
  - **Metastatic bone disease**
    - 350,000/year USA
    - POBRELUKETY
  - **Multiple myeloma**
    - 36,000/year USA
  - Bone lymphoma
    - Several hundreds

- Sarcomas
  - **Bone sarcoma**
    - 3,260/year USA
    - Osa, CHsa, Ewing

Benign Bone Lesions

- Enchondroma
- Osteochondroma
- Unicameral Bone Cyst
- Non Ossifying Fibroma
- Fibrous Dysplasia
- Osteoid Osteoma
- Eosinophilic Granuloma

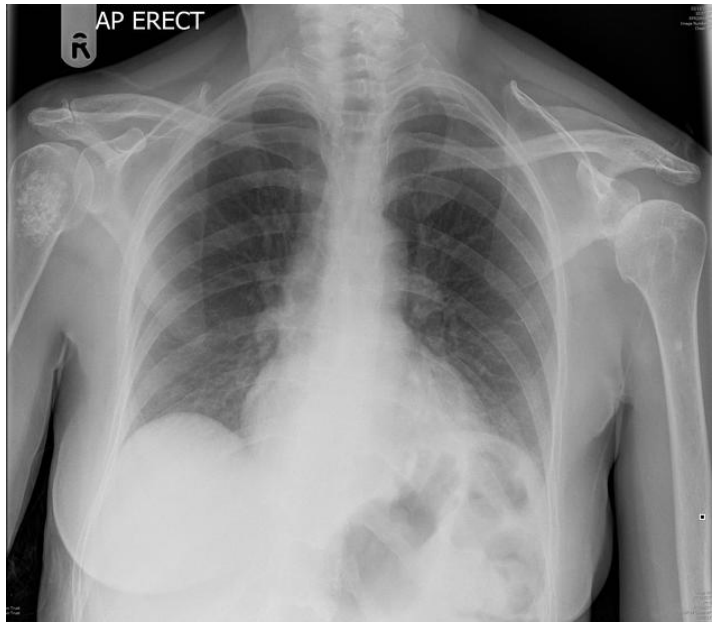
Enchondroma

Benign latent cartilaginous tumor

Often incidental finding

Most common primary bone tumor in the hand







Enchondroma Xray

Multiple Enchondromatosis (Ollier)

- ChSa
- Gliomas
- AML
- Juvenile granulosa cell tumors







Multiple Enchondromatosis & hemangiomas (Maffucci)

- ChSa
- Pancreatic & hepatic adenoCa
- Mesenchymal ovarian tumors
- Brain tumors
  - Glioma and astrocytoma
- Various sarcomas

Osteochondroma Xray



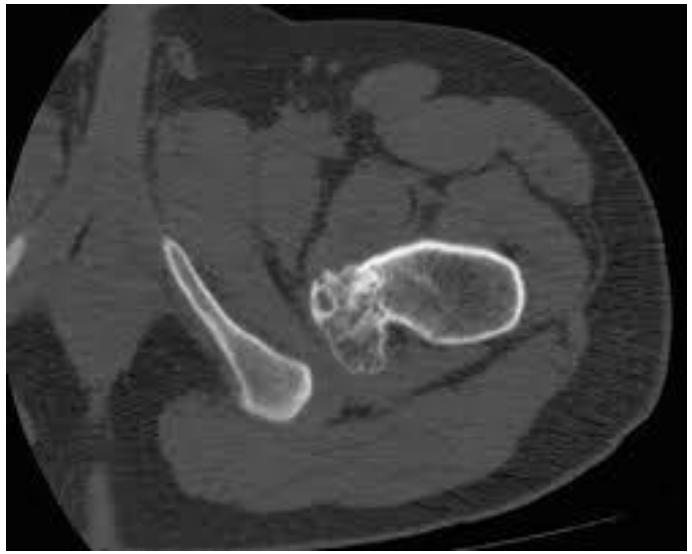




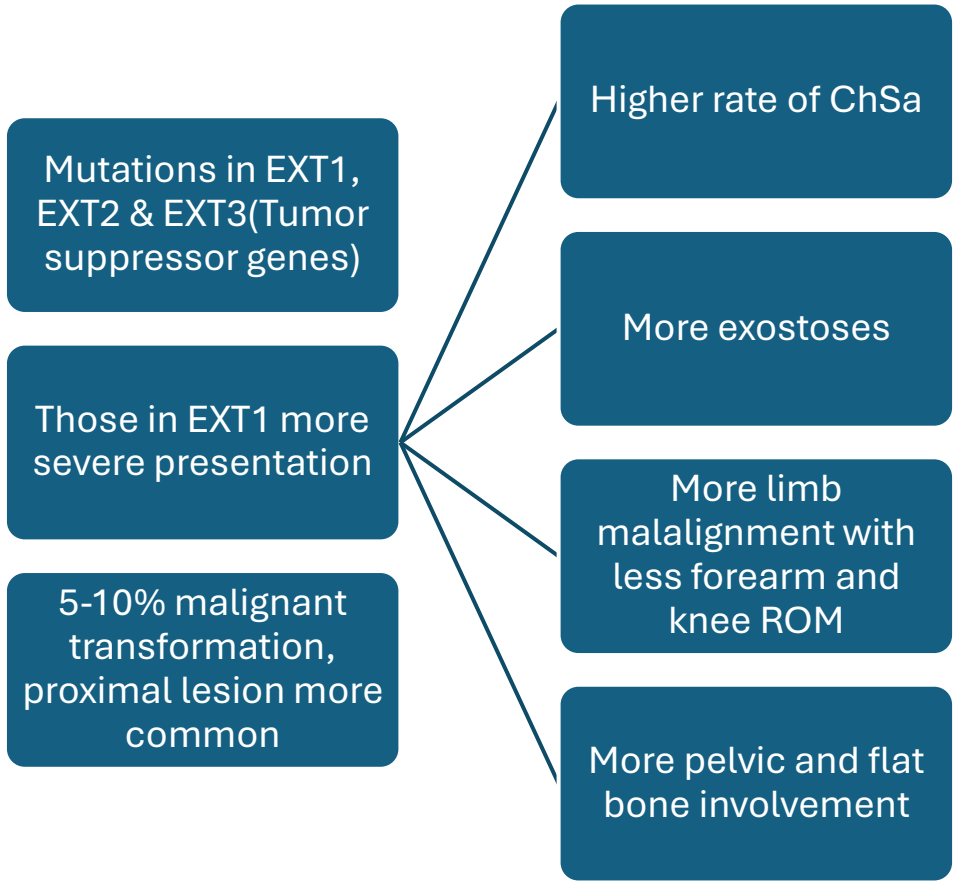
Osteochondroma CT







Multiple Hereditary Exostosis MHE



Unicameral Bone Cyst

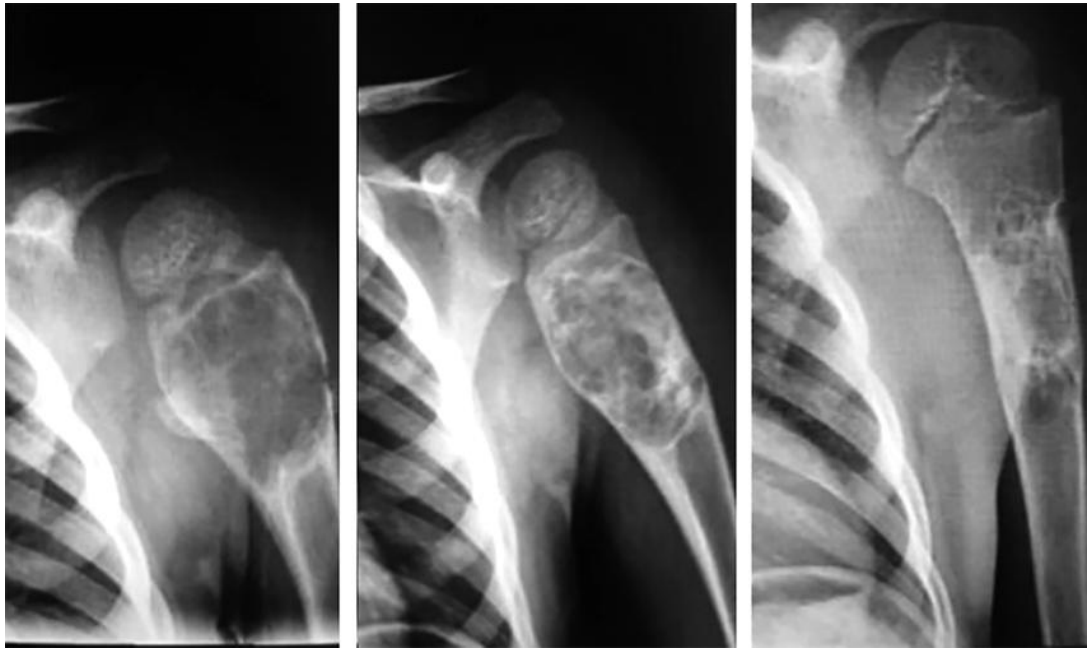
- Incidental or after path Fx before growth maturity
- Proximal humerus or femur
- Unknown etiology
- TX as needed





UBC Natural History

- Potential to resolve without intervention after skeletal maturity
- Low rates of complete healing, regardless whether surgical management is performed.
- Radiographic resolution does not seem to be required for a good functional outcome.
- A path fx was previously may stimulate cyst healing in less than 10% of patients.[4]



Non Ossifying Fibroma

- Benign fibrous lesions that result from dysfunctional ossification
- Metaphysis of long bones
- between 5 and 15 yo. **Incidental.**
- Metaphyseal eccentric "bubbly" lytic lesion surrounded by a sclerotic rim.
- Treatment is observation as most lesions resolve spontaneously.





Fibrous Dysplasia

Postzygotic activating mutations of the **GNAS** gene, encoding the α-subunit of the stimulatory G-protein **Gs (Gsα).**

- Failure of production of normal lamellar bone and arrest as woven bone.

Ptt's less than 30 years of age with an incidental finding.

Radiographs show a lesion with a ground glass appearance or a "punched-out" lytic lesion with a well-defined margin of sclerotic bone.

Tx is oral analgesics and bisphosphonates for pain control.

Surgery for

- Path real or impending fx
- Lesions leading to deformities
  - Coxa vara, scoliosis, and limb length discrepancy.



Fybrous Dysplasia

Osteoid Osteoma

Lower extremity (>50%)

- proximal femur > tibia diaphysis

usually found within the bone cortex

spine (10-15%)

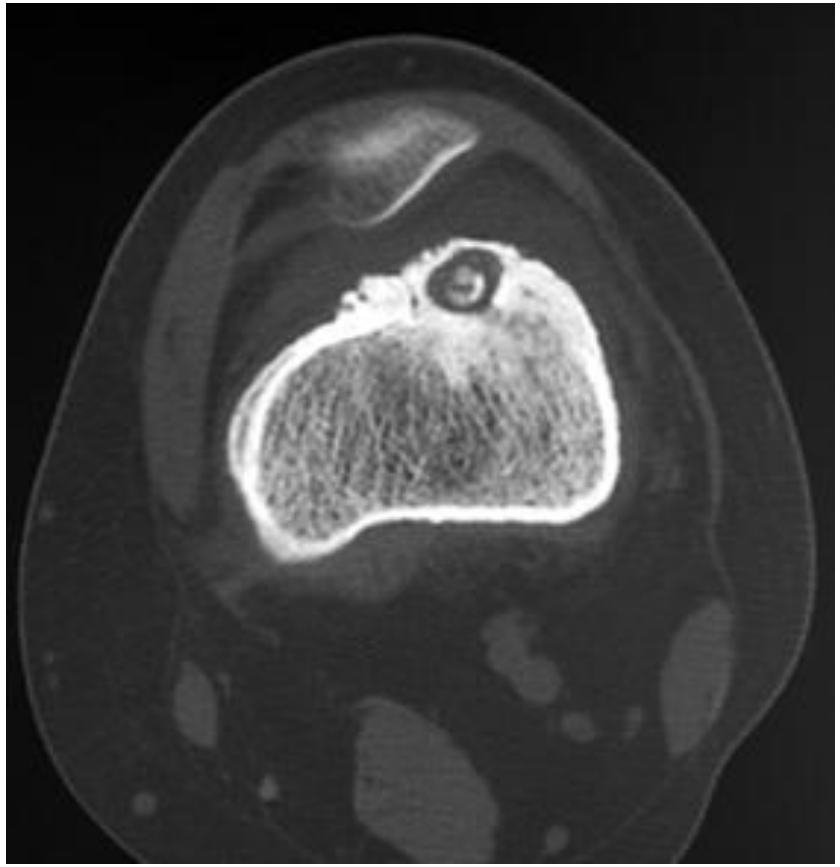
- Thoracic and lumbar regions > cervical and sacral
- Majority involve the posterior elements
- Usually on the side of concavity in scoliosis

Hand (5-10%)

- usually scaphoid and proximal phalanx

foot (<5%)

- predominantly talar neck



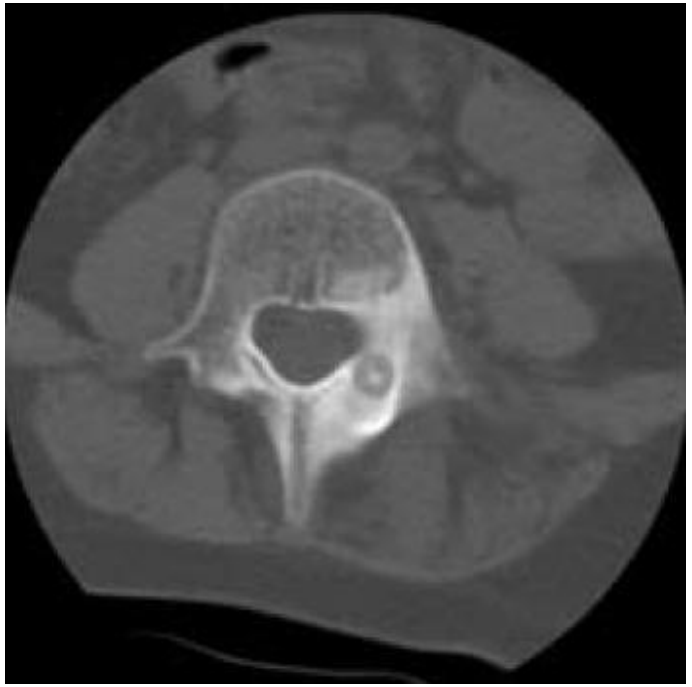
OO Radiographs

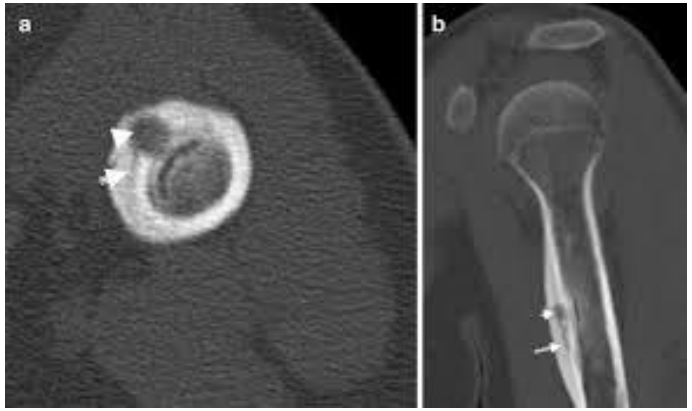


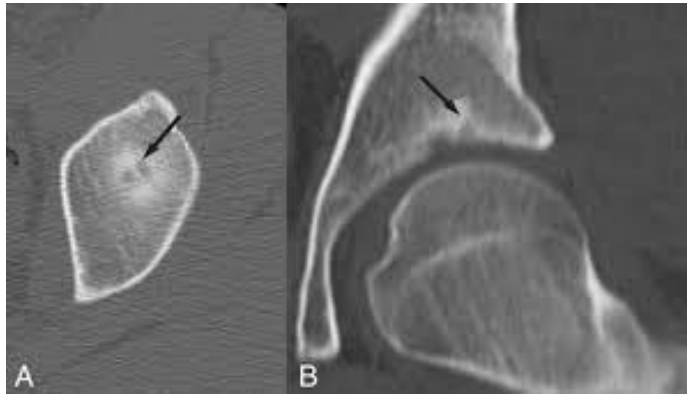


- Intensely reactive bone around a radiolucent nidus
- Because of intensely reactive sclerosis, may be possible to detect nidus only with CT or MRI

OO CT









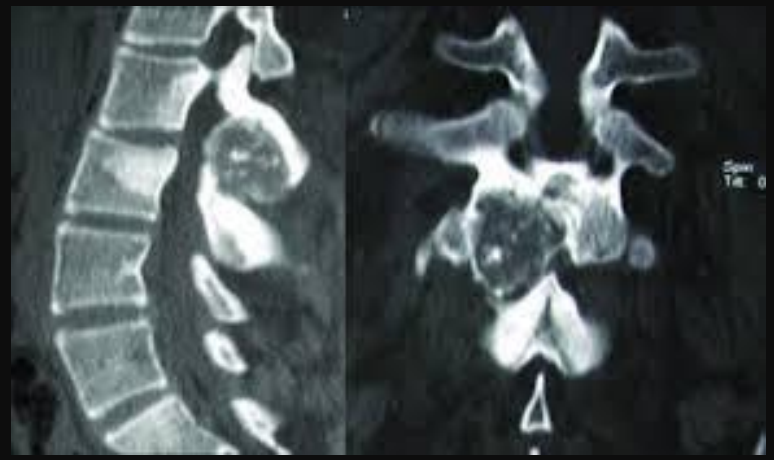
Benign??? Tumors of Bone

- Osteoblastoma
- Chondroblastoma
- GCT
- ABC

Osteoblastoma Images

- Lytic or mixed lytic-blastic lesion with radiolucent nidus 2-10cm
- 66% cortically based, 33% medullary based; rarely periosteal
- Often expansile with or without characteristic reactive bone formation seen in osteoid osteoma
- 25% appear very aggressive and often mistaken for malignant lesion given minimal secondary bone reaction





Chondroblastoma Images

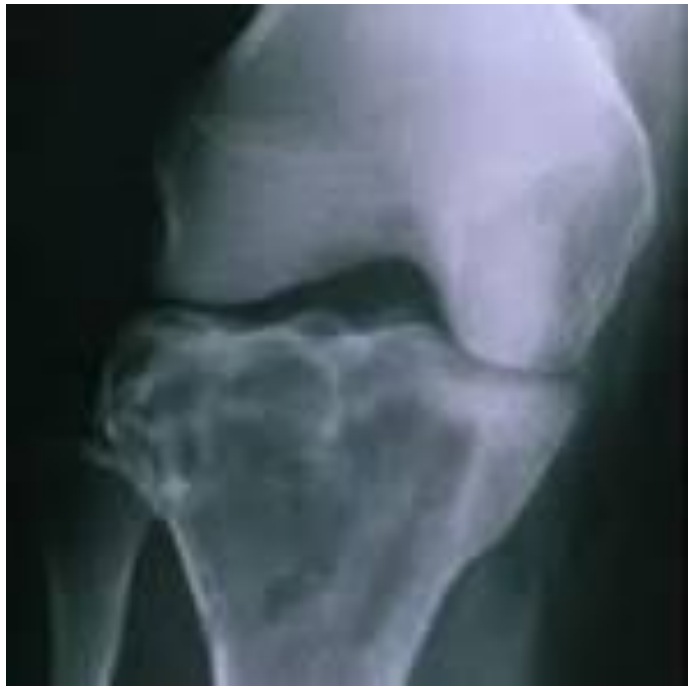






GCT Images

- **Appearance:** A lytic lesion that is well-defined but nonsclerotic, with a location near the joint space.
- **Aggressive features:** May show cortical expansion, destruction, or a soft-tissue component.
- **Fluid-fluid levels:** Can be seen in about 14% of cases due to secondary aneurysmal bone cyst formation.



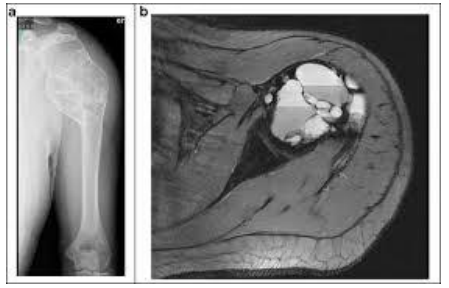




ABC Imaging

- **X-ray:** Radiolucent, expansile, and often eccentric.
- **MRI:** Shows characteristic "fluid-fluid levels".







Malignant Bone Tumors

- Metastatic Bone Cancer
  - **Metastases from AdenoCa"s**
    - 350,000/year USA

- Primary Bone Cancer
  - **Multiple myeloma**
    - 36,000/year USA
  - Bone lymphoma
    - Several hundreds
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Malignant Bone Tumors Presentation by **AGE**

Patient age less than 20	Ewing sarcoma Conventional osteosarcoma Periosteal osteosarcoma
Patient age between 20 and 40	Parosteal osteosarcoma Adamantinoma Fibrosarcoma Primary lymphoma of bone Conventional osteosarcoma Periosteal osteosarcoma
Patient age greater than 40	Metastatic disease (90%) Multiple myeloma (10% or 40%) Chondrosarcoma Osteosarcoma secondary to Paget disease or prior radiation

Metastatic Bone Disease

- Most common malignant cause of destructive bone lesions in the extremities of adult patients.
- Workup
  - H&P, XRays,
  - CT CAP
  - CBC, U/A, Alk Phase, Ca, SPEP,PSA
  - If not biopsy
  - **Finds 1ry in 85% of cases**
- Tx - Controlling pain, maintaining patient independence, and preventing fractures.



Epidemiology

Most common site for metastatic disease	•Lung •Liver •Bone
Solitary bone lesions > 40 yo are 500 times more likely to be metastatic carcinoma	
Anatomic location	•Spine > pelvis > proximal femur > humerus
Path Fx's secondary to metastatic disease	•Proximal femur, followed by the proximal humerus
50% in femoral neck, 20% pertrochanteric, 30% subtrochanteric	

Epidemiology

- Acral lesions are most commonly from lung
- Lung primary is the most common for occult metastatic disease
- Carcinomas that commonly spread to bone include
  - Prostate
  - Breast
  - Lung
  - Kidney
  - Thyroid



Antonio Luchetti - AFF, AEE, LUMA



Antonio S. Lucchetti	
Nacido	Antonio S. Lucchetti Otero 20 de enero de 1888 [1] Ponce, Puerto Rico
Fallecido	19 de diciembre de 1958 (70 años) San Juan, Puerto Rico
Nacionalidad	puertorriqueño
Educación	Ingeniero eléctrico
Alma máter	Universidad de Cornell
Conocido por	Padre de la Autoridad de Energía Eléctrica de Puerto Rico

Epidemiology

- Carcinomas that commonly spread to bone include
    - Mnemonic:
        - PoBreLuKeTy
        - BLT and a Kosher Pickle
        - PT Barnum Likes Kids





Frequency



PoBreLuKeTy

Blastic → **Lytic**

Lytic Lesions

- **Vicious circle**
  - Tumor cells secrete PTHrP which stimulates the release of RANKL from osteoblasts
  - RANKL then binds to the RANK receptor on osteoclast precursor cells
  - Differentiation to active osteoclasts occurs, which causes bony destruction
  - TGF-B, ILGF-1, and calcium are released from resorbed bone, which stimulates tumor cells to release more PTHrP
  - Tumor continues to grow through the release of growth factors, proinflammatory cytokines (IL-6 and IL-8), and VEGF





Blastic Lesions

Prostate and breast cancer mets

Due to tumor-secreted endothelin-1(ET-1)

Binds to endothelin A receptor (ETAR) on osteoblasts and stimulates osteoblasts

ET-1 decreases WNT suppressor DKK-1

Activates WNT pathway, increasing osteoblast activity

Metastatic Hypercalcemia

Medical emergency

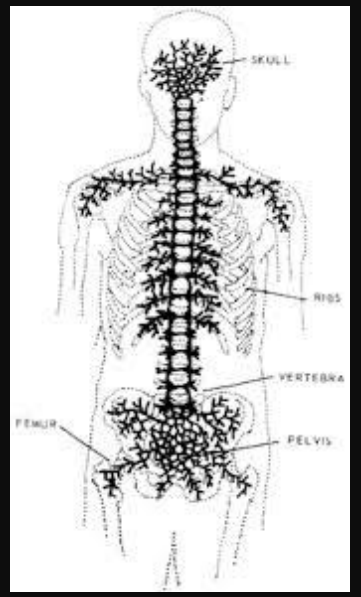
- muscle weakness
- polyuria & polydipsia
- nausea/vomiting
- Dehydration
- Confusion

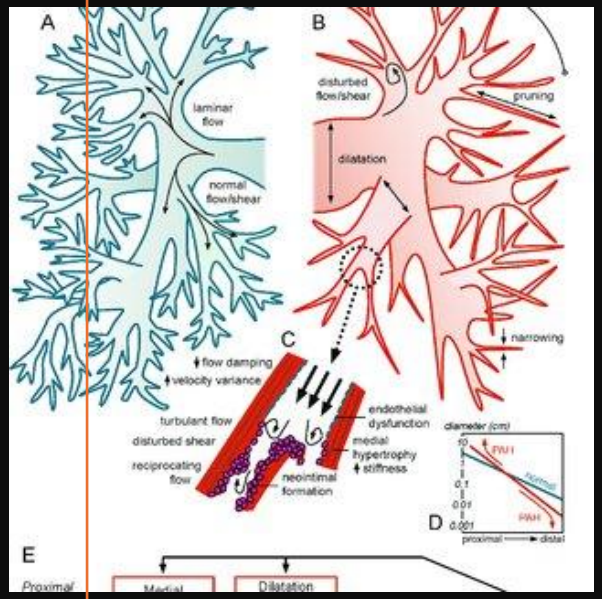
Treatment

- hydration (volume expansion)
- loop diuretics
- bisphosphonates

Anatomy

- **Vascular spread**
  - Batson's vertebral plexus
    - Valveless venous plexus of the spine that provides a route of metastases from lung, kidney, breast, prostate, or thyroid to axial structure including vertebral bodies, pelvis, skull, and proximal limb girdles
  - Arterial tree metastasis
    - mechanism by which lung and renal cancer spread to the distal extremities





Presentation

- Night pain
- weight loss
- pain with weight-bearing
- enlarging mass
- Mechanical pain due to bone destruction or tumorigenic pain
- pathologic fracture
  - occurs at presentation in 8-30% of patients with metastatic disease
- metastatic hypercalcemia
- neurologic deficits caused by compression of the spinal cord

Imaging

- **Xrays**
  - 90% of prostate CA are blastic
  - 60% of breast CA are blastic
  - lung, thyroid, and renal are primarily lytic
  - **PoBreLuKeTy**
  - Cortical metastases are common in lung cancer
  - Lesions distal to elbow and knee are usually from lung or renal primary
-

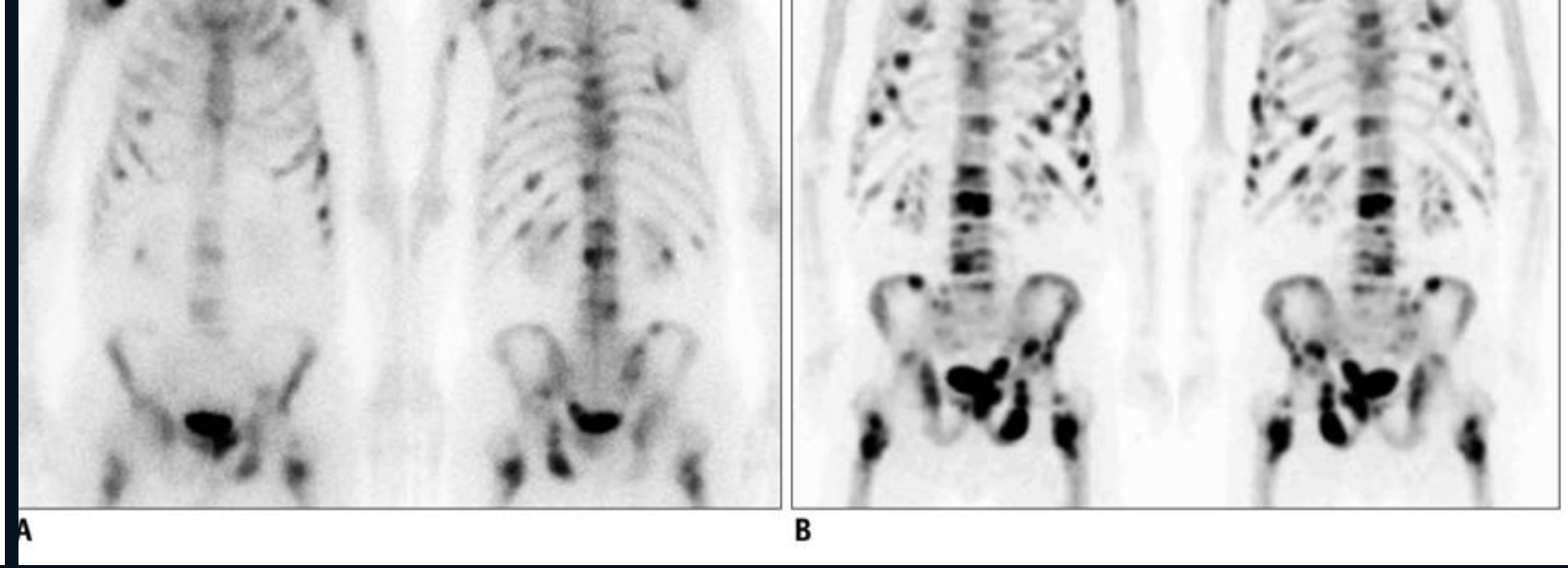
CT Scan

- CT of chest/abdomen/pelvis
- All patients >40-years-old with a lytic bone lesion of unknown origin
- CT of lesion mostly in path Fx





Nuclear Imaging

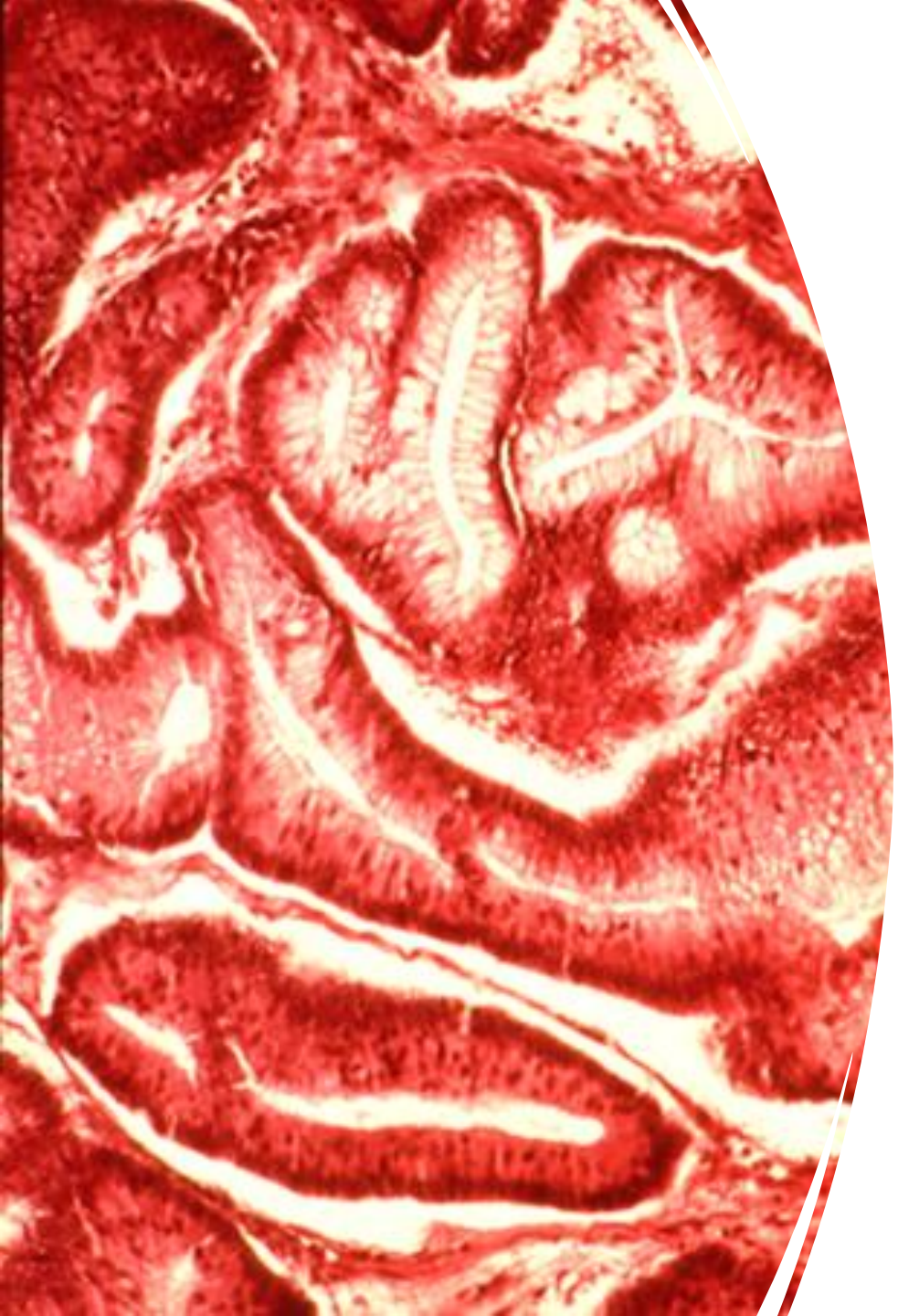




- Technetium bone scan
  - Used to identify other skeletal lesions
  - Myeloma and thyroid carcinoma are often cold on bone scan because it evaluates osteoblastic activity
    - evaluate with a skeletal survey
- Using bone scan and CT identifies the primary tumor about 50% of the time
- PET scan alone only has 30% specificity

Labs

CBC with differential	• Anemia
ESR	
CMP	• Liver
Ca, Phos,	
alkaline phosphatase	• Paget
serum and urine immunoelectrophoresis (SPEP, UPEP)	• MM
PSA	• Prostate
LDH	• lymphoma
U/A	• renal CA



Invasive

Biopsy

- in patients where a primary carcinoma is not identified
- Do not treat a bone lesion without tissue diagnosis
- Metastatic adenocarcinoma not identified by CT of the chest, abdomen, and pelvis is most likely from a small lung primary tumor
- Histology
  - epithelial cells in clumps or glands in a fibrous stroma
- immunostaining
  - Keratin
  - CK7 (breast and lung cancer)
  - TTF1 (lung cancer)

• Recommendation

- < 8: radiotherapy and observation
- 8: use clinical judgment
- > 8: prophylactic fixation
- scores of 9 had a 33% risk of fracture
- Sensitivity 80-90% and specificity 30-35%

•Mirels Criteria				
•Score	•Site	•Size (relative to shaft diameter)	•Radiographic appearance	•Pain
•1	•Upper extremity	•< 1/3	•Blastic	•Mild
•2	•Lower extremity	•1/3 to 2/3	•Mixed	•Moderate
•3	•Peritrochanteric	•> 2/3	•Lytic	•Functional (pain with weight-bearing)

Treatment

- **Nonoperative**
  - asymptomatic lesions
  - nondisplaced fractures (in the humeral shaft, pelvis, scapula)
  - non-weight-bearing bones
  - short life expectancy
  - tumors sensitive to systemic therapy
- **bisphosphonate therapy**
  - reduces rates of skeletally-related events
  - decreased lysis and associated hypercalcemia
- **Denosumab**
  - bone metastases from solid tumors and multiple myeloma
  - superior to zoledronic acid in preventing skeletally-related events
- **radiation therapy**
  - palliation of pain and local tumor control
  - reduces pain in 70% of patients at 2 weeks with complete pain relief in about 30%
  - renal cell carcinoma is less radiosensitive
- **chemotherapy, immunotherapy, and hormone therapy**

Operative

- **Stabilization of complete fracture, postoperative radiation**
  - fixation should not rely on bone healing
  - life expectancy > 6 months is the best predictor of fracture healing
  - all patients require postop radiation unless death is imminent or area has previously been irradiated
  - area of irradiation should include the entire fixation device (e.g. entire femur after intramedullary nailing of femoral lesion)

- **prophylactic stabilization of impending fracture, postoperative radiation**
  - more than 50% destruction of the diaphyseal cortices
  - permeative destruction of the subtrochanteric femoral region
  - >50-75% destruction of the metaphysis
  - persistent pain after radiation therapy
  - pain with weight-bearing

- **Prophylactic fixation** leads to greater likelihood of independent ambulation, discharge to home, and shorter hospital stays compared to fixation of completed fracture
- Wide resection of the bony metastasis provides lower risk local recurrence when compared to intralesional curettage

Other Considerations

- **preoperative embolization**
  - **renal cell carcinoma or thyroid carcinoma** these cancers are very vascular
  - palliative pain control in renal or thyroid cancer

- **other minimally invasive techniques**
  - radiofrequency ablation: thermal ablation of periosteal nerves
  - cementoplasty: cementing a lytic lesion to improve stability
  - cryoablation: freezes a lytic lesion
  - high-intensity focused ultrasonography: new and less available