An Imaging Approach to Focal Bone Lesions

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Bones? Did someone say bones?
FOCAL LESIONS

- Imaging Approach
- Analysis
- Pathology

IMAGING APPROACH

- Plain Radiographs
- CT
- MRI
- Nuclear Medicine
  - Bone Scan
  - PET Scan

Which modality is used for what?
APPROACH

- Diagnosis: Plain Radiographs >> MRI
- Staging:
  - CT
  - MRI
  - Nuclear Medicine
    - Bone Scan
    - PET Scan

PLAIN RADIOGRAPHS ARE ESSENTIAL TO MAKE THE DIAGNOSIS OF A FOCAL BONE LESION.
APPROACH

ALONG WITH APPROPRIATE CLINICAL INFORMATION – AT LEAST AGE, GENDER.

Sorry for screaming.

FOCAL LESIONS

- Imaging Approach
- Analysis
- Pathology
ANALYSIS

- Clinical
  - Age
  - Gender
  - Helpful:
    - Race
    - Onset
    - Pain History
    - Past Medical History (Prior H/O neoplasm).
    - Physical Exam
    - Laboratory

### Table 1: Bone Tumors and Tumor-Like Lesions: Typical Ages of Patients

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<th>TUMOR</th>
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<td>Parosteal Osteosarcoma</td>
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<td>Fibrosarcoma</td>
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<td>Fibrous Malignant Tumor</td>
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<td>Malignant Giant Cell Tumor</td>
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<td>Ewing's Sarcoma</td>
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<td>Enchondroma</td>
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<td>Hemangioma</td>
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<td>Giant Cell Tumor</td>
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<td>Neurofibroma</td>
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<td>Simple Bone Cyst</td>
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<td>Aneurysmal Bone Cyst</td>
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ANALYSIS

- Part of skeleton
  - Bone:
    - Membranous or Enchondral.
  - Location in bone, e.g.
    - Epiphysis, Metaphysis, Metadiaphysis, Diaphysis.
  - Centricity:
    - Eccentric or centric.
  - Part of bone:
    - Medulla, Cortex, Periosteal, Juxtacortical.

Madewell et al., 1981

ANALYSIS

- Type of Lesion:
  - Sclerotic
  - Lytic:
    - Geographic
    - Motheaten/Swiss cheese
    - Permeative
  - Mixed

Lodwick Classification

Geographic: IA  IB  IC
II: Motheaten  III: Permeative
ANALYSIS

- Lesion Edge:
  - Well demarcated/Sharp
  - Poorly defined
ANALYSIS

- Lytic Lesion Matrix:
  - None
  - Ground Glass
  - Chondrouss (circles and dots)
  - Osseous (spicules and cortex)

- Underlying bone:
  - Normal
  - Paget Disease
  - Radiation
  - Ollier's
ANALYSIS

- Lytic Lesion Margination:
  - None
  - Partial
  - Sclerosis – Thick rim of sclerosis.

- Bone Enlargement
  - Degree.
  - Consider bone affected.
ANALYSIS

- Cortical Destruction

ANALYSIS

- Soft Tissue Mass
ANALYSIS

- Pathologic Fracture

ANALYSIS

- Periosteal Reaction:
  - An attempt of a bone to heal itself.
  - Appearance related to competition between healing and destruction.
  - Types:
    - Solid
    - Laminar
    - Onion Skinning
    - Sunburst
  - Codman’s Triangle.
ANALYSIS

- Solid
- Lamellar
- Onion-skinning
- Sunburst

ANALYSIS

- Codman’s Triangle = Pathologic Breakthrough.
ANALYSIS

- Decision: Aggressive or Not Aggressive

- Same as Malignant or Benign but:
  NOT ALL FOCAL BONE LESIONS ARE NEOPLASTIC!

ANALYSIS

- Older patients: Metastasis and Myeloma/Lymphoma.

- Younger patients: Cell Lines (Mesoderm):
  - Fibrous
  - Cartilaginous
  - Osseous
  - Cystic
  - Angiomatous
  - Adipose
  - Infectious
  - Small Blue Cell

- Odd:
  - EG
  - Brown Tumor
  - Hematoma/Hemophilia
  - Tophus
  - Synovial Cell Sarcoma
  - Chordoma
ANALYSIS

- Using this approach:
  - The correct diagnosis can be reached in \( \sim 70 - 80\% \) of cases.
  - The correct diagnosis will be included in the differential in at least 90\% of cases.

FOCAL LESIONS

- Imaging Approach
- Analysis
- Pathology
PATHOLOGY: Metastasis to bone

- Over age of 40, the most common neoplastic lesion in bone.
- Includes:
  - Epithelial lesions.
  - Myeloproliferative lesions:
    - Multiple Myeloma
    - Lymphoma
    - Leukemia
PATHOLOGY: Metastasis to bone

- **Lytic**
  - Renal Cell - Common
  - Lung – Common
  - Breast – Common
  - GI – Uncommon
  - GU – Uncommon
  - MM – Common
  - NHL – Common
  - Neuroblastoma – Peds.

- **Blastic**
  - Prostate – Common
  - Breast – Common
  - Medulloblastoma – Peds.
  - Thyroid – Common
  - Carcinoid – Uncommon
  - Hodgkin’s – Common

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Most tumors that give rise to blastic metastases in peripheral skeleton are lytic in the skull.

- Perhaps related to membranous vs. enchondral bone formation?
PATHOLOGY: Lytic Metastasis

PATHOLOGY: Lytic Metastasis
PATHOLOGY: Lytic Metastasis

PATHOLOGY: Blastic Metastasis
PATHOLOGY: Blastic Metastasis

- Myeloproliferative lesions:
  - Multiple Myeloma
  - Lymphoma
  - Leukemia

- Lymphoma and leukemia younger age group than myeloma.

- Generally lytic, except Hodgkin’s which has a propensity to be blastic.
SMALL BLUE CELL LESIONS

PATHOLOGY: Myeloma/Leukemia

- Disease of RED marrow:
  - Lesions in axial skeleton >> Acral skeleton.
  - Late disease moves peripherally as new marrow is recruited.
**MYELOMA**

- **Imaging:**
  - Radiographs:
    - Osteopenia, not useful.
    - Permeative, unusual.
    - Lytic lesions, common.
    - Plasmacytoma.
  - Bone Scan: Only 50% of lesions positive.
  - MRI: Very sensitive.

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**MYELOMA: Lytic**

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MYELOMA: Plasmacytoma

MYELOMA: Infiltrative
MYELOMA: Lytic PET/CT

LEUKEMIA
LYMPHOMA, NON-HODGKINS

- Formerly, Reticulum Cell Sarcoma. (T cells)
- 5% of primary malignant bone tumors.
- Over 20% of patients with lymphoma have secondary bone involvement.
- Most intraosseous lesions are non-Hodgkin's.
- >20 yo in general.
- Radiographs show permeative to lytic lesions.
- MRI for staging.

69yo Man
LYMPHOMA, NON-HODGKINS

76yo Woman
LYMPHOMA, HODGKINS

- Involved: 9.2% - 30.3% of patients, rare as primary site. (B Cells)
- Lytic > sclerotic.

EWING'S SARCOMA
EWING'S SARCOMA

- Highly malignant primitive neuroectodermal tumor (PNET).
- 2nd most common malignant bone tumor in childhood.
- Most common in the first and second decade.
  - Cases age 2 to 80.
- Race: Whites >> Blacks and Asians.
- $\sigma:\varphi = 3:2$.

EWING'S SARCOMA

- Clinical Presentation:
  - Pain and swelling of weeks or months duration.
  - Erythema and warmth of the local area are sometimes seen.
  - Often confused for osteomyelitis because of intermittent fevers, leukocytosis, anemia and \^ESR.
EWING'S SARCOMA

- Differential Diagnosis:
  - Osteosarcoma, telangetatic.
  - Osteomyelitis.
  - EG.
  - Lymphoma.
  - Metastatic neuroblastoma.
  - Chondrosarcoma.

EWING'S SARCOMA

- Pathology:
  - Gross: Tumor is gray to white in color and poorly demarcated.
    - Areas of hemorrhage and necrosis are common.
EWING'S SARCOMA

Pathology:
- Histology:
  - Densely packed uniform small cells in sheets.
  - Scant cytoplasm without distinct borders.
  - Cells two to three times as big as lymphocytes.
- Glycogen is present within the cells causing a positive reaction to periodic acid-schiff (PAS) stain.

EWING'S SARCOMA

Pathology:
- Most Ewing’s sarcomas: + HBA-71 or 0-13 stain, an antibody to protein product of myc 2 gene on x and y chromosomes.
- + neuron-specific enolase and S-100 protein testing.
- Lymphoma excluded by reticulin stain.
- Metastatic neuroblastoma excluded by urine vanillylmandelic acid and homovanillic acid stain.
- Rhabdomyosarcoma ruled out by negative stain with desmin, myoglobin and actin.
EWING'S SARCOMA

Pathology:
- Thought to be neural origin:
  - Supported by electron microscope presence of pseudorosettes, neurites and dense-core granules.
  - Supported by choline acetyltransferase.
- Translocation t(11:22)(q24;ql2).
- Poorly differentiated end of the spectrum of PNET and has few organelles.
- Neuroepithelioma is an example of well differentiated PNET and has neurosecretory granules and neuritic processes.

Radiology
- Lesion is usually lytic and central.
- Permeative or "moth-eaten" appearance.
  - Spreads through Haversian canals.
- Large soft tissue mass.
- Endosteal scalloping often present.
- Periosteal reaction: "onion-skin"
- Sclerotic is 27% of cases.
- Pathological fracture.
EWING SARCOMA: Path Fx

26 yo man with sudden onset pain in femur.

EWING'S SARCOMA

Metastasizes:
- Bone.
- Lung.
EWING'S SARCOMA

- CT not specifically useful.
- MRI useful for staging.
  - TI-weighted images the tumor has low intensity
  - T2-weighted images the tumor is hyperintense
- Nuclear Medicine: ↑ activity on bone scan, PET scan.

21 yo male with pain and swelling, distal femur.
EWING'S SARCOMA

21 yo man with shoulder pain.

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EWING SARCOMA
EWING SARCOMA

FIBROUS-TYPE LESIONS
PATHOLOGY: Fibrous Lesions

- **BENIGN**
  - Fibrous Cortical Defect
  - Non-ossifying Fibroma
  - Benign Fibrous Histiocytoma
  - Fibrous Dysplasia
  - Ossifying Fibroma/Osteofibrous Dysplasia
  - Juxtacortical Desmoid
  - Desmoplastic Fibroma
  - Fibromatosis

- **MALIGNANT**
  - Malignant Fibrous Histiocytoma (Pleomorphic Sarcoma)
  - Fibrosarcoma
  - Adamantinoma

NONOSSIFYING FIBROMA

- Fibrous Cortical Defect
- Fibroxanthoma
- Benign Histiocytoma
- Jaffe Campanacci Syndrome
NONOSSIFYING FIBROMA (NOF)

- Very Common
  - Mostly children (75% occurring in the second decade).
  - Males more commonly females
  - May affect up to 35% of all children.\(^1\)
- Clinical:
  - Asymptomatic
  - Usually discovered incidentally on radiographs.

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NONOSSIFYING FIBROMA

- Pathology:
  - Dense fibroblasts are found in whorled and storiform patterns.
  - Xanthoma cells may be present.
  - No atypia & few mitoses.
NONOSSIFYING FIBROMA

- Fibrous Cortical Defect
  - Smaller lesion.
  - Younger kids.
- NOF
  - Any size – may be giant.
  - Generally teenagers to young adults.
- Fibroxanthoma
  - Obsolete nomenclature
  - Emphasizes that may lesion have xanthomatous component.
- Benign Fibrous Histiocytoma
  - Generally older individuals.
  - Usually not marginated.

NONOSSIFYING FIBROMA

- Benign fibrous lesions within METADIAPHYSIS of long bones.
  - Common in long tubular bones, especially the femur > tibia.
- Well defined lytic lesion.
- Marginated.
- Often “diamond” shaped.
- Eccentric and may thin cortex.
- Cortex not destroyed, except occasional blister.
- No periosteal reaction.
- Occasional pathological fracture.
- Heal spontaneously with sclerosis.
NONOSSIFYING FIBROMA

NONOSSIFYING FIBROMA: MRI
NONOSSIFYING FIBROMA:
Healed

NONOSSIFYING FIBROMA:
Bone Blister
BENIGN FIBROUS HISTIOCYTOMA

- Rare lesion
- Distribution:
  - Epiphysis or diaphysis of tubular bones
  - Ribs
  - Pelvis
- Nondescript lytic, loculated lesion
  - Well-defined margin.
  - Sclerotic margin +/-.
  - No cortical destruction.

BENIGN FIBROUS HISTIOCYTOMA

- Histology:
  - Fibroblasts
  - Mononuclear or multinucleated cells
    - Appearance of histiocytes.
  - “Foam cells”, lipid-filled cells with abundant vacuolated cytoplasm,
  - Masses of cholesterol.
  - No mitotic activity, cellular atypia, or pleomorphism is present.
  - HAM-56 + Histioyte cell line.
BENIGN FIBROUS HISTIOCYTOMA

49 year woman with knee pain.

JAFFE CAMPANACCI SYNDROME

- Multiple NOF’s
  - Resolve spontaneously.
- Café au lait spots.
- Hypogonadism.
- Mental retardation.
- Ocular problems.
- Ridiculously Rare.
FIBROUS DYSPLASIA

Monostotic
Polyostotic
Cherubism
McCune Albright
Mazabraud Syndrome

FIBROUS DYSPLASIA (FD)

- Exact incidence not established, but common.
- Four patterns:
  - Monostotic
  - Polyostotic
  - Cherubism
  - Endocrinopathy-related Syndromes:
    - McCune-Albright.
    - Others, particularly thyroid.
FIBROUS DYSPLASIA

- **Demographics**
  - M=F
  - All races.
- Initial manifestations aged 3-15 years.
- Pathology: Abundant fibroblasts is whorl pattern.

FIBROUS DYSPLASIA: Monostotic

- 70 – 80% of FD.
- Rib (28%) > Femur (23%) > Tibia > Craniofacial (10-25%) > Humerus.
- **Clinical:**
  - 20 or 30 years-old until may be symptomatic.
  - Increasing pain.
  - Pathologic fracture.
  - Enlarging soft tissue mass ⇒ Malignant change.
  - RARE: 0.4-1% in fewer than 50 reported cases.
FIBROUS DYSPLASIA: Polyostotic

- 20-30% of FD.
- 2/3 symptomatic before age 10 years.
- Femur (91%) > Tibia (81%) > Pelvis (78%) > Ribs > Skull and facial bones (50%) > UE’s > L-spine > Clavicle > C-spine.
- Bowing:
  - Associated osteomalacia.
    - S protein of c-AMP.
  - Severe coxa vara = Shepherd’s crook deformity.
FIBROUS DYSPLASIA: Shepherd’s Crook Deformity

FIBROUS DYSPLASIA: BS & CT
FIBROUS DYSPLASIA: MRI

T1

T2

T1FS p Gd
FIBROUS DYSPLASIA: Focal

- Autosomal-dominant disorder.
- M:F = 1:2.
- More severe in boys.
- FD of mandible.
- May regress after adolescence.

FIBROUS DYSPLASIA: Cherubism

- Autosomal-dominant disorder.
- M:F = 1:2.
- More severe in boys.
- FD of mandible.
- May regress after adolescence.
FIBROUS DYSPLASIA: Cherubism

- Precocious puberty in girls (McCune-Albright)
- Hyperthyroidism
- Hyperparathyroidism
- Acromegaly
- Diabetes mellitus
- Cushing syndrome

FIBROUS DYSPLASIA: Endocrinopathy-related

- 2-3% of FD
FIBROUS DYSPLASIA: McCune-Albright

- **Genetics:**
  - Mutations in GNAS1 gene.
  - Sporadic, not inherited.
- Sexual precocity in girls (95%).
- Polyostotic fibrous dysplasia.
- Café au lait:
  - Coast of Maine.
  - Ipsilateral to bone lesions.
  - Caution: Occurs in 50% of patients with polyostotic FD.
- Occasional hyperthyroidism as well.

FIBROUS DYSPLASIA: Craniofacial

- Facial Bones (Frontal, Sphenoid, Maxillary, and Ethmoid) > Occipital & temporal bones.
- Atypical appearance in skull:
  - Calvarium:
    - Mixed lytic-blastic.
    - May appear aggressive.
  - Facial Bones:
    - Sclerotic.
    - Enlarges bones ⇒ Ossium Leontasia.
FIBROUS DYSPLASIA: Craniofacial

- Hypertelorism
- Cranial asymmetry
- Facial deformity (ie, leontiasis ossea)
- Sphenoid/Frontal Bone: Visual impairment, exophthalmos, and blindness.
- Temporal bone: Vestibular abnormalities, tinnitus, and hearing loss.
- Cribiform plate: Anosmia
FIBROUS DYSPLASIA: Ossium Leontasia

FIBROUS DYSPLASIA: CT
FIBROUS DYSPLASIA: Monostotic (Simulating Paget)

FIBROUS DYSPLASIA: Focal Mistaken for Metastasis
FIBROUS DYSPLASIA: Focal Mistaken for Metastasis

Mazabraud Syndrome
OSSIFYING FIBROMA

Osteofibrous dysplasia

Benign fibrous tumor.
- Locally aggressive behavior.
- Not metastasize.
- First decade of life.
- Clinical: Painless, enlarging mass.
- Most common site in children is the tibia, followed by other long bones and pelvis.
- Ossifying fibroma and adamantinoma are on a continuum.
  - Osteofibrous dysplasia-like adamantinoma represents an intermediate between the two lesions.
OSSIFYING FIBROMA

- Histology:
  - Irregular spicules of trabecular bone lined by osteoblasts.
  - Osteoblasts produce rim of lamellar bone around centers of woven bone.
  - Stains positive for cytokeratin.

OSSIFYING FIBROMA

- Characteristic appearance:
  - Lytic lesion in the anterior cortex of the diaphysis or metaphysis of the tibia.
  - May cause anterior-posterior bowing.
  - Hot on BS.
OSSIFYING FIBROMA

- Differential Dx:
  - Adamantinoma
  - Fibrous dysplasia
  - Nonossifying fibroma
  - Osteoblastoma

JUXTACORTICAL DESMOID

- Tug Lesion
- Periosteal Desmoid
- Metaphyseal Fibrous Defect
- Cortical Irregularity Syndrome
JUXTACORTICAL DESMOID

- NOT A TRUE NEOPLASM:
  - Benign fibrous proliferation of the periosteum forming a tumor-like lesion.
- Demographics:
  - Early to mid teenager.
  - Often history of athletics.
- Nearly always located adjacent to the femoral condyle at insertion of:
  - Gastrocnemius, medial head.
  - Adductor magnus.
  - Can occur elsewhere.
- Reaction to trauma at musculotendinous insertion.

JUXTACORTICAL DESMOID

- Radiographs:
  - Soft tissue swelling.
  - Saucer-shaped cortical defect.
  - Adjacent sclerosis.
  - Focal periostitis.
- Key is to **not** mistake this lesion for a parosteal or periosteal osteosarcoma.
Juxtacortical Desmoid

- **Histology:**
  - Nonmalignant appearing reactive fibrous tissue, cartilage, and bone.
  - Poorly organized osteoid and hypercellularity can lead to a misdiagnosis of osteosarcoma.

- **Imaging:**
  - 1 – 3 cm. in size.
  - Irregular cortical lesion with lucency and sclerosis.
  - May be mineralization within the lesion.
  - Margin may irregular.
  - MRI may show the relationship of the muscle insertion to the lesion.
JUXTACORTICAL DESMOID

- Imaging:
  - CT Scan:
    - Help differentiate lesion from osteoid osteoma
    - Show the complex nature of the lesion:
      - Areas of cortical thinning and thickening.
      - Small lucent areas
      - Surrounding mild sclerotic bone reaction.
  - Bone Scan:
    - Activity ⇒ Inactivity as lesion involutes.

DO NOT CONFUSE WITH OSTEOSARCOMA.
DESMOPLASTIC FIBROMA

- Extremely rare (< 200 cases).
- Slowly progressing tumor:
  - Well-differentiated cells that produce collagen.
  - Benign but with aggressive local infiltration.
  - Diagnosis difficult to make radiologically.
- Demographics:
  - Most often in the first 3 decades.
  - M:F = 1:1
- Locations: Mandible > Femur > Pelvis.
DESMOPLASTIC FIBROMA

- Pathology:
  - Gross:
    - Grayish to yellowish white color.
    - Rubbery consistency.
    - Edges are irregular, round and blunt.
    - Occasional cystic foci with clear fluid.
  - Microscopic:
    - Interlacing bundles of dense collagen.
    - Low cellularity.
    - No atypia.
    - Nuclei are ovoid or elongated.
    - The edge may resemble fibrous dysplasia, but under polarized light lamellar structures are obvious.

- Clinical:
  - Pain late in the clinical course
  - Swelling.
  - Joint effusion if near a joint.
  - Pathologic fracture uncommon: (~12%).

- Radiographs:
  - Metaphysis aligned with the long axis of the bone.
  - Osteolytic, expansile, medullary lesion.
  - Well-defined sclerotic margins.
  - Thinned cortex
  - Fine intra-lesional trabeculae
  - “Soap Bubble” lobulated appearance.
DESMOPLASTIC FIBROMA

DESMOPLASTIC FIBROMA
DESMOPLASTIC FIBROMA

Differential Diagnosis:
- Nonossifying fibroma
- Giant cell tumor
- UBC
- ABC
- Fibrous dysplasia
DESMOPLASTIC FIBROMA

- Treatment
  - Marginal or wide surgical excision.

- Recurrence
  - 55-72% with curettage.
  - 17% with resection.


KELOIDS
KELOID

- Exuberant Scar Formation
  - Means "crab claw"
  - Occurs in 5-15% of wounds.
  - Lesions expand from the original scar into normal tissue.
- Demographics
  - Mean age at onset is 10-30 years
  - Blacks and Asians.
    - Women present more - cosmetic implications.
    - Tend to be familial

KELOID

- Keloid formation can occur within a year after injury.
- Dermal fibrotic lesions are a variation of the normal wound healing process.
- Areas of the body that are constantly subjected to high skin tension
  - Wounds on the anterior chest, shoulders, flexor surfaces of the extremities and anterior neck.
  - Wounds subjected to a prolonged inflammation.
**KELOID**

- **Management:**
  - Preoperative radiation therapy to the wound for prevention.
  - Occlusive dressings and compression.
  - Intralesion steroid injections.
  - Surgery: Excision.
  - Radiation, cryosurgery and laser therapy.
  - Interferon therapy.

- **Characteristic branching fibrous tissue.**
- **No mitoses.**
- **No atypia.**
KELOID

AX T1  AX T2

KELOID

COR T2  COR T1 GAD
MALIGNANT FIBROUS HISTIOCYTOMA

MFH
Pleomorphic Sarcoma

- Soft Tissue >> Bone
  - ~20-24% of soft tissue sarcomas.
  - Most common soft tissue sarcoma occurring in late adult life.
- Race: Caucasian > Negro or Asian.
- Sex M:F = 2:1
- Age: Broad Range but Peak is 40 – 60 years.
MALIGNANT FIBROUS HISTIOCYTOMA

- Location:
  - Extremities (70-75%, with lower extremities accounting for 59% of cases)
  - Retroperitoneum.

- Clinical presentation:
  - Enlarging painless soft tissue mass in the thigh,
  - Typically 5-10 cm in diameter.
  - Two thirds are intramuscular.

MALIGNANT FIBROUS HISTIOCYTOMA

- Postulated both histiocytic and primitive mesenchymal cell theories of origin.
- Contains both fibroblast-like and histiocyte-like cells in varying proportions.
- Five histological subtypes:
  - Storiform/pleomorphic - most common,
  - Myxoid,
  - Giant cell,
  - Inflammatory – usually retroperitoneal,
  - Angiomatoid – often more superficial than other types.
MALIGNANT FIBROUS Histiocytoma

- Pleiomorphic Storiform
- Immunoperoxidase Stain CD68 found in cytoplasm of histiocytes.

Survival correlated with Anaplasia (10 year survival rate):
- Low-grade – 90%
- Intermediate-grade – 60%
- High-grade – 20%
- Overall 5 year survival: ~40 – 50%

Survival correlated inversely with lesion size.

Metastasis:
- Lung (90%),
- Bone (8%),
- Liver (1%).
MALIGNANT FIBROUS HISTIOCYTOMA

- Imaging: MRI
  - Intramuscular mass with heterogeneous signal and enhancement consistent with areas of necrosis.

MFH
MALIGNANT FIBROUS Histiocytoma

HARDCASTLE SYNDROME
**HARDCASTLE SYNDROME**

- Rare – Five families described.
- Autosomal dominant bone dysplasia:
  - Symmetric long bone diaphyseal sclerosis.
  - Symmetric medullary stenosis.
  - Bones have appearance of "wormeaten wood".
  - Very high risk of development of malignant fibrous histiocytoma (~35%).
  - Prone to fracture with minor trauma.
  - Bone scan not show abnormal activity in all bones.

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**ADAMANTINOMA**
ADAMANTINOMA

- Fischer first described the tumor in 1913.
- Only around 200 reported.
- Age: 20–50 years.
- M:F = 1.3:1.

Location:
- Tibia in 80% of cases
- Anterior cortex
- Middle third of the diaphysis.
- Typically eccentric.

Radiographs:
- Multilocular or slightly expansile osteolytic lesion,
- May be locally aggressive.
- Size: 3.0–15.0 cm
ADAMANTINOMA

- Metastasize:
  - Lungs
  - Nodes
  - Abdominal organs (hematogenous and lymphatic routes).

- RX:
  - Wide surgical excision.
  - Insensitive to radiation.
  - Chemotherapy not useful.

- Survival: 10yr = ~10%

Bonetumor.org
FIBROSARCOMA

- Malignant tumor of soft tissue or bone.
  - Tissue derived from collagen-producing fibroblasts.
- Age: 30 to 60 years old.
**FIBROSARCOMA**

- **Histology:**
  - Ranges from being poorly differentiated to well differentiated,
  - Does not produce cartilage, osteoid, or bone.
  - May be subtype of MFH.
- **Prognosis is poorer for fibrosarcomas of bone c/w soft tissue.**
  - Recurrence after surgery is common.
- **Infantile form: Better prognosis.**

---

**FIBROSARCOMA**

- **Imaging:**
  - Large, destructive, infiltrating tumor of long bones:
    - Commonly around the knee followed by the pelvis.
  - Lytic with bone destruction of the moth-eaten, geographic or permeative pattern.
  - Little osteosclerosis or periostitis.
  - May have sequestrum of dead native bone within.
FIBROSARCOMA: Bone

- Differential diagnoses:
  - Malignant fibrous histiocytoma.
  - Osteosarcoma.
  - Metastasis.
  - Lymphoma.

FIBROSARCOMA: Multifocal
FIBROSARCOMA: Radiation Induced

FIBROSARCOMA: Bone
CARTILAGE-TYPE LESIONS

PATHOLOGY: Cartilaginous Lesions

- BENIGN
  - Osteochondroma
    - Multiple Hereditary Exostoses
  - Enchondroma
    - Ollier’s Disease
    - Maffucci’s Syndrome
    - Metachondromatosis
  - Chondroblastoma
  - Chondromyxoid Fibroma
  - Juxtacortical Chondroma

- MALIGNANT
  - Chondrosarcoma
    - Peripheral
    - Central
OSTEOCHONDROMA

Exostosis
Multiple Hereditary Exostoses

- Cartilage capped bone projection:
  - Accounts for 20-50% of benign bone tumors and 10-15% of all bone tumors.¹
  - Sessile or pedunculated.
- Male:Female = 1.5 to 1.
- First two decades of life.
- Location:
  - Most common in long bones.
  - 40% of osteochondromas occur around the knee.²
OSTEOCHONDROMA

- Not a true neoplasm:
  - Occur only in bones that develop from cartilage (endochondral ossification).
  - Congenital defect or trauma to the perichondrium.
  - Herniation of a fragment of the epiphyseal growth plate through the periosteal bone cuff.

OSTEOCHONDROMA

- Behavior:
  - Migrate away from growth plate.
  - After the close of the growth plate, growth ceases.
  - Cartilage cap normally 1-6 mm thick.
  - May decrease in thickness with age.
OSTEOCHONDROMA

- Clinical:
  - Pain due to mechanical irritation
    - Occasionally fracture through the stalk.
    - Neurovascular compression.
  - Mass.
- Malignant Transformation:
  - Chondrosarcoma from cap.
  - ~1% for a solitary lesion.
  - Suspect with:
    - Growth after physeal plate closure.
    - Pain.
  - Look for thick cartilage cap on MR:
    - Malignancy reported at various depths: 1 – 3 cms.

OSTEOCHONDROMA

- Gross Pathology: Stalk of normal appearing bone with gray cartilage cap.
OSTEOCHONDROMA

- **Histology:**
  - See normal marrow
  - Has normal appearing growth plate with rows of chondrocytes.
  - Cartilage is more disorganized than normal,
  - Binucleate chondrocytes in lacunae, and is covered with a thin layer of periosteum.

OSTEOCHONDROMA

- **Diagnostic Imaging:**
  - Marrow cavity of the bone is contiguous with the lesion.
  - Move away from growth plate as bone grows.
  - Pedunculated lesions point away from joint.
  - Sessile lesions have wide base
    - Cause metaphyseal widening
    - "Trumpet shaped deformity".
  - **CT:**
    - Useful to evaluate continuity of cortex in difficult lesions.
  - **MRI:**
    - Visualize relationship to other structures.
    - Evaluate the thickness of the cartilage cap.
OSTEOCHONDROMA: Pedunculated

OSTEOCHONDROMA: Sessile
OSTEOCHONDROMA

OSTEOCHONDROMA
OSTEOCHONDROMA

- Bursae may develop over cap.

OSTEOCHONDROMA

- Therapy:
  - No treatment necessary unless symptoms.
  - If symptomatic:
    - Excised at the base.
    - No recurrence if entire cartilage cap is removed.
MULTIPLE HEREDITARY EXOSTOSES

- Autosomal dominant.
- The lesions may occur on different bones or on the same bone, and symptoms present in the first decade of life.
- Malignant transformation:
  - Chondrosarcoma.
  - Risk unknown, but felt to be additive.
  - Risk increases with number and size.
MULTIPLE HEREDITARY EXOSTOSES

[Radiographic images of bones with exostoses]

MULTIPLE HEREDITARY EXOSTOSES

[Radiographic images of bones with exostoses]
ENCHONDROMA

- Ollier’s Disease
- Maffucci’s Syndrome
- Metochondromatosis

ENCHONDROMA

- Benign cartilaginous neoplasm in bone:
  - Solitary benign lesion in intramedullary bone.
  - Ectopic hyaline cartilage rests in intramedullary bone.
  - Matrix:
    - Calcifications throughout the lesion.
    - Rings, dots and commas.
  - 12-14% of benign bone neoplasms.
  - 50% of solitary enchondromas in the hands:
    - Typically in mid and distal portions of the metacarpals and proximal portions of the phalanges.
ENCHONDROMA

- Age: 20-40 years.
- Metabolically active:
  - Grows and evolves throughout patient’s lifetime.
  - Calcified matrix is progressive.
- Small incidence of malignant transformation:
  - Loss of calcification in a focal region suggests malignant degeneration.
  - In a patient with enchondromatosis, the incidence of chondrosarcoma is 50% higher.
ENCHONDROMA

Radiological Differential Diagnosis:
- Lesion with matrix:
  - Bone infarct.
  - Chondrosarcoma.
- Lesion without matrix (purely lytic):
  - Nonossifying fibroma.
  - Unicameral bone cyst.
  - Fibrous dysplasia.
  - Eosinophilic granuloma.
  - Clear cell chondrosarcoma.

X-ray:
- Classic pattern of calcifications, described as rings, arcs and dots, corresponding to calcification around lobules of cartilage.
- Calcifications may be difficult to distinguish from the dystrophic calcifications seen in bone infarction.

Bone Scan:
- Increased activity in region of lesion.
ENCHONDROMA

- CT Scan:
  - Endosteal lesions with a lobular morphology and variable matrix.

- MRI:
  - Lobulated lesion with high-signal-intensity foci on T2W.
  - Absent signal where extensive matrix.

After Therapy
ENCHONDROMA

- Multiple enchondromas in three distinct disorders:
  - Ollier disease – Multiple enchondromas with a predilection for unilateral distribution.
  - Maffucci syndrome - Multiple hemangiomas and enchondromas
  - Metachondromatosis - Multiple enchondromas and osteochondromas.
    - Autosomal Dominant.

OLLIERS DISEASE
OLLIERS DISEASE

MAFFUCCI SYNDROME
CHONDROBLASTOMA

Codman’s Tumor

CHONDROBLASTOMA

- Rare cartilaginous neoplasm:
  - <1% of all primary bone tumors.
  - Benign slow-growing.
  - Most common site is the lower extremity.

- Demographics:
  - Age 10-30 years, but mostly before physeal closure.
  - Male-to-female ratio is 2-3:1.
CHONDROBLASTOMA

Pathology:
- Derived from the epiphyseal cartilage plate
- Uniform, closely packed polygonal cells.
- Primitive cells
- Abundant cytoplasm.
- Scant mitotic activity.
- Giant cells are often present.
- Chicken Wire appearance.
CHONDROBLASTOMA

Radiographs:
- Epiphysis or apophysis of long tubular bones.
  - Generally eccentric position in the epiphysis.
- Geographic, lucent lesion with sharply margined borders:
  - 40% uniformly lucent.
  - 60% have mottled opacity due to amorphous calcification or peripheral septa.
  - May extend into the metaphysis.
  - May cause endosteal scalloping.
  - Margination is variable.
CHONDROBLASTOMA

- Differential Diagnosis:
  - Brodies Abscess.
  - EG.

CHONDROBLASTOMA

- MRI
  - Extension to the metaphysis.
  - Low signal on T1W images and variable signal intensity on T2W images.
  - Evaluation of aggressive or recurrent tumors.
  - Soft tissue extension.

- CT
  - Minimal role.
CHONDROBLASTOMA

- Treatment: Surgical curettage and packing with a bone graft or polymethylmethacrylate.
- Risk factors for recurrence:
  - Rate: low.
  - Lesion larger than >3.7 cm.
  - Secondary aneurysmal bone cyst.
  - Location in the proximal femur or pelvis.
CHONDROBLASTOMA

Chondroblastoma & . . .
Chondroblastoma & Stress FX

CHONDROBLASTOMA
CHONDROMYXOIDFIBROMA

Extremely rare (<1% of all bone tumors).
Location: Metaphyses around the knee (proximal tibia, proximal fibula, or distal femur).
Clinical:
- Second to third decade
- Male:Female = 2 to 1.
- Usually chronic pain, swelling and possibly a palpable soft tissue mass or restriction of movement.
- Pathological fracture unusual presentation (<5%).
CHONDROMYXOIDFIBROMA

- Radiology:
  - Eccentric metaphyseal lobulated, lytic lesion
  - Well defined, sclerotic margins.
  - Matrix unusual.
  - False trabeculae from scalloping ridges and grooves occurring in the margins.

- Bone Scan:
  - Increased activity.

- CT:
  - Helps define cortical integrity.
  - Confirms absence of mineralization of the matrix.

- MRI:
  - Similar appearance on MRI as other cartilage tumors
  - ◇ Signal on T1W.
  - ▲ Signal on T2W.
  - MRI is helpful in preoperative planning and staging.
CHONDROMYXOID FIBROMA

Radiological Differential diagnosis:
- Giant cell tumor.
- Aneurysmal bone cyst.
- Unicameral bone cyst.
- Chondroblastoma.
- Fibrous dysplasia.

Pathology:
- Histologically very similar to chondrosarcoma.
  - Often radiology helps to make the final diagnosis.
- Zonal architecture and lobular pattern.
- Nodules of cartilage intermixed with fibromyxoid areas.

CHONDROMYXOID FIBROMA

Stacy, G.. Chondromyxoid Fibroma, eMedicine, 2003

JUXTACORTICAL CHONDROMA

Periosteal Chondroma
JUXTACORTICAL CHONDROMA

- Rare benign surface lesion composed of cartilage:
  - 2.2% of benign tumors and 0.5% of all tumors in the Mayo Clinic series (Unni, 1996).

- Demographics
  - Age: 10 - 30.
  - Slight male predominance.

- Clinical:
  - Local swelling.
  - Pain.

- Pathology:
  - Benign cartilage:
    - Typical chondrocytes in scattered lacunae.
    - Noneoplastic bone being formed by enchondral ossification.
    - No neoplastic cartilage.
JUXTACORTICAL CHONDROMA

- Radiology:
  - Well-defined small lesion (2 and 4 cm).
  - Epicenter in the cortical surface of the bone, typically adjacent to the metaphysis.
    - Proximal humerus most frequently involved.
  - The cortex may be involved to a variable degree, but the lesions do not involve the medullary space.
  - Small soft tissue mass extends beyond the confines of the bone and may contain amorphous calcification.

- Radiology (Cont):
  - Pressure from mass causes “J”-shaped lesion without no stalk or peduncle:
    - No periosteal reaction.
    - May lift the periosteum and create buttresses of mature periosteum at each or one end of the mass out from the bone.

- Nuclear Medicine:
  - Bone scan shows increased activity.
  - CT shows juxtacortical location and amorphous Ca++.
  - MRI shows soft tissue mass but is otherwise not helpful.
JUXTACORTICAL CHONDROMA

- Differential Diagnosis:
  - Periosteal Chondrosarcoma.
  - Surface Osteosarcoma.
  - Periosteal Osteosarcoma.

- Potential for confusion with periosteal or parosteal osteosarcoma ⇒ Should biopsy but not in vacuo.

CHONDROSARCOMA
CHONDROSARCOMA

- Second most frequent primary malignant tumor of bone.
- 7.8% of all primary osseous neoplasms.
- 25% of all primary osseous malignant neoplasms.
- Demographics:
  - >40 years and increases with age.
  - Male-to-female ratio of 1.5-2:1.
  - No racial predilection.
- Slow-growing non-metastasizing lesions to highly aggressive metastasizing sarcomas.

CHONDROSARCOMA

- Types:
  - Central – Originating within the medullary canal.
  - Peripheral – Arising from osteochondroma.
- Primary - Arise de novo.
- Secondary (age 20-40)- within a preexisting lesion
  - Enchondromatosis syndromes (Large Risk).
  - Hereditary multiple exostosis (Mild Risk).
CHONDROSARCOMA

Clinical:
- Chronic dull pain – average duration of symptoms prior to presentation is 1-2 years.
- Occasional presentation with path fracture.
- Metastases: Lung and pleura – may cause PTX.

Prognosis:
- Size of the lesion: Larger is worse.
- Anatomic location: Axial worse than acral.
- Histologic grade: Higher grade is worse.
  - Less ca++ matrix is worse.
CHONDROSARCOMA

- Histologic Grading:
  - Grade 1 – least aggressive.
    - Not metastasize.
    - 95% 5-year survival.
  - Grade 3 – the most aggressive.
    - Metastases – lung.
    - ~30% 5-year survival.

- Pathological Classification:
  - Conventional (Majority).
  - Clear cell. (Low grade, >90% Epiphyseal, Younger patients)
  - Myxoid.
  - Mesenchymal. (High grade)
  - Dedifferentiated. (Highest grade, >60 years old)

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CHONDROSARCOMA

- Pathology
  - Typical: Very similar to enchondromas and osteochondromas, making differentiation difficult.
  - Occasional binucleated cells.
  - High grade lesions:
    - Increased cellularity.
    - Atypia.
    - Mitoses.
CHONDROSARCOMA

Radiological Evaluation:
- Radiographs are essential for diagnosis.
- CT supplement to radiographs to detect subtle matrix if needed.
- MRI for staging and operative planning.
- Bone Scan:
  - No role in diagnosis.
  - For metastases if high grade and not recognized as chondrosarcoma initially.
CHONDROSARCOMA

- Radiology:
  - Lytic lesion (grade dependent):
    - Varying edges.
    - Varying cartilaginous matrix.
    - Endosteal scalloping, often deeper than with benign enchondroma.

CHONDROSARCOMA: Low Grade
CHONDROSARCOMA: High Grade

CHONDROSARCOMA: ST Mass
CHONDROSARCOMA: Ollier’s

- Low Grade.
- Ends of long bones.
- Rare in flat bones.

CHONDROSARCOMA: Clear Cell
CHONDROSARCOMA: Mesenchymal

CHONDROSARCOMA: Multifocal Mesenchymal
MESENCHYMAL CHONDROSARCOMA

- Rare malignant neoplasm
- Second and third decades
- More in the head and neck
- Increased tendency to occur as entirely extraskeletal lesions
- Skeletal lesions are primarily lytic and destructive with poor peripheral margins
- More metastases than other cartilaginous malignancies

X-ray and CT

- Skeletal lesions are primarily lytic and destructive with poor peripheral margins
- Cortical destruction and extraosseous extension
- Endosteal scalloping may be present
MESENCHYMAL CHONDROSARCOMA

- MRI
  - Useful in assessing the thickness of the cartilage cap of osteochondromas to identify chondrosarcoma transformation
  - Clarifying the intramedullary and extraosseous extent of a chondrosarcoma
  - Lobulated lesions of high signal on T2W images

- Treatment
  - Radical resection of the lesion
  - Adjuvant radiation therapy for some high grade lesions
  - Poor long-term prognosis
## OSSEOUS LESIONS

## PATHOLOGY: Osseous Lesions

### BENIGN
- Osteoma
- Osteoid Osteoma
- Osteoblastoma

### MALIGNANT
- Osteosarcoma
  - Medullary
  - Intracortical
  - Surface:
    - Parosteal
    - Periosteal
    - High-grade Surface
  - Telangectatic
- Osteosarcomatosis
OSTEOMA

- Developmental and increase with age.
- Generally arises from membranous bones:
  - Skull and facial bones.
  - Predilection for sinuses.
    - Frontal and ethmoid most often.
- Composed of dense, compact osseous tissue.
  - Indistinguishable pathologically from bone island, insula compacta, enostosis.
GARDNER’S SYNDROME

- Autosomal dominant.
- Syndrome:
  - FAP
    - 90% have hyperplastic gastric polyps.
    - Adenomatous polyps of colon.
  - ST Desmoids.
  - Osseous & soft tissue osteomas.
  - Epidermoid cysts.

OSTEOID OSTEOMA
OSTEOID OSTEOMA

- 5-55 yrs.
  - Peak: 10 – 30’s
- $\text{♂} : \text{♀} = 2:1$
- Mayo Clinic:
  - 12.1% of benign tumors.
  - 2.9% of all tumors.
- Location: 1arily Long bones.

OSTEOID OSTEOMA

- 80% of cases involve the cortical bone, remainder medullary or juxtacortical.
- Nidus may Ca++ $\rightarrow$ Central necrosis.
- Usually elicit significant surrounding sclerotic reaction.
  - Exception: Intracapsular lesions.
- Pain at night relieved by NSAIDS.
- Will resolve in 2-4 years time with medical therapy.
OSTEOID OSTEOMA

- Histologically indistinguishable from osteoblastoma.
  - But nidus <1.5 cm diameter.
- Tumor Nidus: Contiguously joined trabeculae of well-calcified, woven bone (N).
- Host: Lamellar trabeculae (H).
- Interface: Demarcated by a sheet of loose fibrovascular tissue (F).

OSTEOID OSTEOMA
OSTEOBLASTOMA

- 80% of patients < 30 years.
- ♀:♂ = 2-3:1
- 1% of all bone tumors / 3% of benign bone tumors.
- Pain is the most common symptom.
  - Not relieved by NSAID
- Scoliosis has been reported frequently.
- Common locations:
  - Vertebrae & Flat bones (Ribs) >> Long bones (diaphysis).
  - Spine is location of 10% of all osteoid osteomas and 36% of osteoblastomas:
    - C-spine most frequently affected.
    - Two thirds involve the posterior elements
    - One third the vertebral body.
OSTEOBLASTOMA

- Bone-forming lesion.
  - May be found within the cortex, medullary canal, or periosteal tissues.
  - Cortical expansion is often present, but outer rim of tumor is always covered by periosteum and thin rim of reactive bone.

OSTEOBLASTOMA

- Staging:
  - Stage 1 – Latent
  - Stage 2 – Active
  - Stage 3 – Aggressive
  - Most are stage 2 lesions
OSTEOBLASTOMA

Radiographs:
- Well-circumscribed radiolucent lesion containing a thin shell of peripheral new bone.
  - Occasionally may be lytic with destruction of the cortex and expansion into the surrounding soft tissues.
- Larger than 2 cm in diameter and without a large reactive zone of surrounding bone, unlike osteoid osteoma.
- Two thirds of osteoblastomas in tubular bones occur within the cortex.
- Within the vertebral column - mostly in the posterior elements.

OSTEOBLASTOMA

Computed tomography:
- Defines the extent of osseous involvement and tumor delineation when the lesion is within the cortex of bone.

Magnetic resonance imaging:
- To evaluate changes in the surrounding soft tissue and marrow extension or involvement.
OSTEOBLASTOMA

TREATMENT - Surgical
- Stage 1 and 2 lesions - Extensive intralesional excision
- Stage 3 lesions - Wide resection
- Recurrence - 10-20%

OSTEOBLASTOMA

Neurological deficits higher in osteoblastoma than in osteoid osteoma of spine.
- Histologically equivalent to osteoid osteoma.
  - Distinction:
    - Size.
    - Absence of surrounding sclerosis.
OSTEOBLASTOMA

- Lucent expansile lesion ± matrix.
  - Occasionally sclerotic.
  - 25% may have features suggestive of a malignant process.
OSTEOBLASTOMA

AX T1

AX T2
OSTEOSARCOMA

Medullary
Intracortical
Surface: Parosteal, Periosteal, High-grade Surface
Telangectatic
Osteosarcomatosis
OSTEOSARCOMA: General

- Most common primary malignant neoplasm of bone.
- Malignant mesenchymal sarcoma characterized by formation of bone or osteoid.
- Primarily teen to young adult & small second peak related to Paget disease.
- ♂ >> ♀.
- Highly aggressive.
  - NOW, with chemotherapy 5-year survival > 75%
- Location:
  - Most common around the knee.
  - May occur anywhere.

OSTEOSARCOMA: Medullary

- Most common form of osteosarcoma: 75% - 85%
- Predominantly metaphyseal.
  - Tendency to develop skip lesions.
- Most common sites = most rapid bone growth centers:
  - Distal femur > Proximal tibia > Proximal humerus > Pelvis.

Huvos, A., Bone Tumors: Diagnosis, Treatment and Prognosis, W.B. Saunders, Co., 1991.
OSTEOSARCOMA: Medullary

Clinical:
- Adolescents are most affected – occurs before the growth plate is closed – with peak between 10 – 20 years.
- Second peak in 7th decade: Paget disease.
- $\sigma:\varphi = 3:2$
- Symptoms: rapidly progressing pain, swelling and fever.
- Alkaline phosphatase may be 2 - 3x NL.

Pathology:
- Medullary osteosarcoma is generally a high grade tumor with nuclear atypia, hyperchromasia and a high mitotic rate.
- Three histological variants of conventional osteosarcoma:
  - Osteoblastic osteosarcoma has abundant osteoid ranging from a lace like matrix to thickened trabecular bone.
  - Chondroblastic osteosarcoma has cartilage production.
  - Fibroblastic form has a spindle cell stroma with focal osteoid.
- Microscopically, osteoid production is essential for diagnosis.
OSTEOSARCOMA: Osteoblastic

- Highly cellular.
- Pleomorphic nuclei with prominent nucleoli.
- Stringy dense pink matrix c/w osteoid.

OSTEOSARCOMA: Chondroblastic

- Pink staining c/w osteoid.
- Bluish-grey staining c/w proteoglycans.
- Pleomorphic nuclei.
OSTEOSARCOMA: Medullary
Diagnostic Imaging

- Radiographs:
  - May be sclerotic, lytic or mixed.
  - Cortical destruction
  - Soft tissue mass.
  - Periosteal reaction and Codman’s triangles.
    - Typically described as sunburst.

- Bone scan for skip lesions.
- CT scanning has limited role.
- MRI for staging:
  - Delineate soft tissue extension.
  - Cellular areas, and necrosis.
  - Neurovascular bundle involvement.
  - Joint involvement.
  - Muscle involvement.
- Chest CT for metastases.
OSTEOSARCOMA: Medullary

- Differential Diagnosis:
  - Ewing sarcoma.
  - Infection.
    - Chronic osteomyelitis > Acute osteomyelitis.
    - Chronic granulomatous osteomyelitis.
  - Langerhans cell histiocytosis.
  - Chondrosarcoma.
  - Fibrosarcoma.

- Treatment:
  - Surgery – Limb salvage is possible.
  - Chemotherapy – Generally presurgical.
OSTEOSARCOMA: Medullary

- 14yo girl
OSTEOSARCOMA: Medullary

- 42yo woman

OSTEOSARCOMA: Medullary

- 30yo man
PAGET DISEASE: Malignant Transformation

- Second peak of Osteosarcoma.
  - Usually in late 7th decade.
- Also may see:
  - Chondrosarcoma.
  - MFH.
- Clinical Indicators:
  - Increase in pain.
  - Change in size.

PAGET DISEASE: Osteosarcoma

(http://www.medicopaedia.com, Amersham Health)
OSTEOSARCOMA: Variants

<table>
<thead>
<tr>
<th>Tumor</th>
<th>Frequency, %</th>
</tr>
</thead>
<tbody>
<tr>
<td>Telangiectatic</td>
<td>3.5-11</td>
</tr>
<tr>
<td>Parosteal</td>
<td>3-4</td>
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<tr>
<td>Periosteal</td>
<td>1-2</td>
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<tr>
<td>Gnathic (Mandible)</td>
<td>6-9</td>
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<tr>
<td>Small cell</td>
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<tr>
<td>Intraosseous, low grade</td>
<td>&lt;1</td>
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<tr>
<td>Surface, high grade</td>
<td>&lt;1</td>
</tr>
<tr>
<td>Secondary</td>
<td>5-7</td>
</tr>
</tbody>
</table>

OSTEOSARCOMA: Telangiectatic

- ~5% of osteosarcoma.
- Rare and aggressive variant.
- Metaphysis and diaphysis.
- Similar epidemiology to conventional osteosarcoma.
OSTEOSARCOMA: Telangiectatic

Pathology:
- Characteristic gross: Multicystic "bag of blood".
- Microscopically:
  - Large blood filled spaces and thin septation.
  - Within the septa:
    - Scanty osteoid production.
    - Pleomorphic malignant cells.
- May be confused with ABC.

Radiographs:
- Typically entirely lytic, i.e. no osteoid matrix.
- Cortical destruction.
- Periosteal reaction and Codman’s triangles.

MRI:
- Useful for staging.
- Diagnostic Hint:
  - T1W has high intensity.
  - Fluid-fluid levels.
OSTEOSARCOMA: Telangectatic

- 12yo girl

OSTEOSARCOMA: Parosteal

- Most common form of surface osteosarcoma.
- 5% of osteosarcoma.
- Older age than standard osteosarcoma: Median age 27yo.
- Metaphysis of long bones:
  - Typically, posterior distal femur.
  - Not to be confused with juxtacortical desmoid.
- Arises from the surface of the bone
- Encircles bone.
- Slow growing mass.
- Milder symptoms: Vague pain and limited ROM.
- Good prognosis.
OSTEOSARCOMA: Parosteal

Pathology:
- Originates from the outer fibrous layer of periosteum.
- Low-grade tumor microscopically.
- Well-differentiated spindle cell stroma, minimal atypia and a low mitotic rate.

Radiology very characteristic.
- Prolific Ca++ tumor bone that wraps around the bone.
- Lesion merges with the cortex:
  - Stalk early in the disease → Broad base later.
  - Rare to see medullary involvement by the tumor.
- CT show radiolucent zone of periosteum and fibrous tissue trapped between tumor and cortex.
- MRI tumor extension and staging.
OSTEOSARCOMA: Parosteal

- 27yo woman
OSTEOSARCOMA: Periosteal

- 1 – 2% of osteosarcoma.
- Juxtacortical.
- Diaphyseal.
- Radiolucent fusiform mass.
- May erode cortex.
- Periosteal reaction: sunburst.
- Pathology: Similar to chondroblastic osteosarcoma.
OSTEOSARCOMA: Surface

- < 1% of osteosarcoma.
- High-grade and aggressive form of periosteal osteosarcoma.
- Location like periosteal osteosarcoma.
- Bone marrow infiltration is visible on MRI.
- Pathology: Identical to conventional osteosarcoma microscopically.
- Treated similarly and have the same prognosis.
OSTEOSARCOMATOSIS

- Rare aggressive form of osteosarcoma.
- Typical osteosarcomas arising in multiple locations:
  - Synchronous.
  - Metachronous.
- Typical in young teenage years.
- Rapidly fatal.

- 12yo girl
CYSTIC LESIONS

Unicameral Bone Cyst – UBC
Aneurysmal Bone Cyst – ABC

PATHOLOGY: Cystic

- BENIGN
  - Unicameral Bone Cyst
  - Aneurysmal Bone Cyst

- MALIGNANT
UNICAMERAL BONE CYST

Simple Bone Cyst
UBC

Common, benign, fluid-containing lesion, usually occurring in the metaphysis of long bones.

- ~3% of all primary osseous neoplasms.
- Typically proximal humerus (55-65%) > proximal femur (25-30%).

Bimodal peak:
- Children 5 – 15 years: Humerus & Femur.
- Adults >20 years: Ilium and Calcaneus.

♂:♀ = 2:1
UNICAMERAL BONE CYST

- Etiology:
  - Unknown but agreed to be abnormality of growth and not a true neoplasm.
  - Evidence that temporary venous obstruction and blockage of interstitial fluid drainage, in an area of rapidly growing and remodeling cancellous bone, may lead to bone resorption.
- Form at physeal plate and move away as bone grows.

- Pathology:
  - Contain clear fluid resembling synovial fluid.
  - Flat, cuboidal mesothelial cells line simple bone cysts.
UNICAMERAL BONE CYST

- **Clinical:**
  - Asymptomatic.

- **Radiographs:**
  - Metaphyseal to diaphyseal.
  - Central
  - Lytic
  - Geographic
  - Well-defined edges.
  - May thin cortex.

- 50% present with path fracture.
  - Fallen fragment sign.

- 10% develop growth arrest in the affected bone.
UNICAMERAL BONE CYST

- 50% present with path fracture.
  - Fallen fragment sign.
- 10% develop growth arrest in the affected bone.
UNICAMERAL BONE CYST

CT and MRI occasionally useful.
**UNICAMERAL BONE CYST**

Differential Diagnosis:
- Aneurysmal Bone Cyst
- Chondromyxoid Fibroma
- Enchondroma
- Fibrous Dysplasia
- Giant Cell Tumor
ANEURYSMAL BONE CYST

ABC

- Common, solitary, eccentric, highly expansile, lytic metaphyseal lesion.
- Age: 10-30yo with peak at ~15 years.
  - 75% < 20yo.
  - ♂:♀ 1:2.
- Location: Any bone in the body.
  - Most common in the lower extremity (40%) > upper extremity (20%).
  - Vertebra & posterior elements (16%).
  - ~50% of flat bone lesions occur in the pelvis.
ANEURYSMAL BONE CYST

- **Etiology:** Theory that primary ABCs arise as a result of increased venous pressure ➔ Leads to hemorrhage ➔ Osteolysis ➔ Leads to more hemorrhage ➔ Osteolysis.
- ~30% ABC’s secondary to underlying lesion:
  - Giant Cell Tumor (20-40%).
  - Osteoblastoma.
  - Angioma.
  - Chondroblastoma
  - Non-ossifying fibroma.
  - UBC’s.
  - Chondromyxoid fibroma.
  - Fibrous dysplasia.
  - Trauma.

The association with other primary lesions is so strong that the lesion should be examined microscopically in several places to eliminate the possibility of a primary lesion.

ANEURYSMAL BONE CYST

- Clinical:
  - Pain of rapid onset and increase in severity.
  - Swelling,
  - Tenderness.

- Pathological fractures are unusual because of eccentric location of lesions.
  - Infractions are common.

ANEURYSMAL BONE CYST

- Differential diagnosis:
  - Giant cell tumor.
  - Osteoblastoma.
  - Enchondroma.
  - UBC.
  - Chondromyxoid fibroma.
  - Telangiectatic osteosarcoma.
  - Brown Tumor.
ANEURYSMAL BONE CYST

- Pathology:
  - Gross: A blood filled sponge with a thin periosteal membrane and fibrous septae.

ANEURYSMAL BONE CYST

- Pathology:
  - Microscopic:
    - Cystic spaces filled with blood.
    - Fibrous septa:
      - Immature trabeculae of woven bone.
      - Macrophages filled with hemosiderin.
      - Fibroblasts.
      - Capillaries.
      - Giant cells.
ANEURYSMAL BONE CYST

- Radiographs:
  - Eccentric in the metaphysis.
  - Lytic.
  - Periosteal elevation.
  - Cortex is eroded to a thin margin.
  - A "blow-out" or "soap bubble" appearance.
ANEURYSMAL BONE CYST

- CT: May be useful for pelvic or spine lesions.

ANEURYSMAL BONE CYST

- Bone Scan: Increased activity around lesion.

- 10 yo boy kicked in the knee, evaluate for fracture.
ANEURYSMAL BONE CYST

MRI:
- Multiple fluid-fluid levels.
- T1 and T2 show thin low signal rim from cortex.
ANEURYSMAL BONE CYST: Atypical

43yo Woman

ANEURYSMAL BONE CYST: Atypical

25 yo man with chest pain.
ANEURYSMAL BONE CYST

Therapy: Curettage and Packing.

Recurrence rate:
- ~10%
- Should trigger reevaluation for underlying primary lesion.
ANGIOMATOUS LESIONS

Hemangioma
Lymphangioma
Angiosarcoma
Gorham’s Disease

PATHOLOGY: Angiomatous

- BENIGN
  - Hemangioma
  - Lymphangioma
    - Gorham’s Disease
  - Glomus Tumor

- MALIGNANT
  - Hemangiopericytoma
  - Hemagioendothelioma
  - Angiosarcoma
HEMANGIOMA

- Common benign vascular tumor.
  - ~10% of autopsy cases have vertebral hemangiomas.
- Clinical: Typically, asymptomatic lesions discovered on x-ray or autopsy.
- Location in bone:
  - Vertebral bodies (thoracic especially) ~50%.
  - Calvarium ~20%.
  - Tibia, femur and humerus.
HEMANGIOMA

Pathology:
- Four types of hemangiomas:
  - Capillary
  - Cavernous
  - Arteriovenous
  - Venous.
- Gross:
  - Cystic, dark red, cavities.
  - Well demarcated.
  - Coarse trabeculae within.

Microscopic:
- Most common in bone: Capillary and cavernous lesions
- Capillary hemangiomas:
  - Capillary size vessels lined by flattened endothelial cells.
- Cavernous hemangiomas:
  - Calvarium
  - Large dilated vessels with flattened endothelium.
- Non-vascular components:
  - Fat, smooth muscle, fibrous tissue, bone, hemosiderin and thrombus.
HEMANGIOMA

- Radiologic Appearance:
  - Typical and diagnostic appearance.
  - Vertebral lesions have thickened vertebral trabeculae giving a corduroy appearance.

CT scan:
- Vertebral lesions: “polka dot” appearance.
- Calvarial lesions: “spoke wheel” appearance.
- Other lesions: “Irish lace” appearance.
HEMANGIOMA: Irish Lace

MRI

Three essential imaging findings:

- Fat.
- Spicules.
- Slow flow.

T1W                   T2W
HEMANGIOMA

• 14 yr old wrist pain on motion

HEMANGIOMA: Soft Tissue
GORHAM’S DISEASE

- First reported in 1838.
- >100 cases described.
- Age < 40 at onset.
- 50% have history of trauma.
- Location:
  - Upper arm.
  - Shoulder.
  - Mandible.
- Pathology: Characterized by proliferation of thin-walled, endothelial-lined, anastomosing vascular channels, resembling hemangioma or lymphangioma.

14 months later.
Glomus Tumor

- Glomus apparatus is paraganglioma:
  - Ends of digits may involve bone.
  - Occur adjacent to carotid bulb and at jugular foramen among other locations.
- Baroreceptor whose role is moderate blood flow.
- Adults in 4th 5th decades of life.
- Clinical: Joint tenderness and pain.
- Location: Dorsal surface of the finger, either medially or laterally.
- MRI: Very high signal on T2W.
Glomus Tumor

ANGIOSARCOMA
ANGIOSARCOMA

- Rare aggressive sarcoma.
- Older adult.
- Any bone.
  - 33% in the axial skeleton.
  - 33% in long tubular bones.
- Often multifocal:
  - Same bone with multiple lesions.
  - Multiple bones of the same extremity.
  - Single rapidly progressive lesion.

ANGIOSARCOMA

- Clinical:
  - Nonspecific symptoms.
  - Pain/tenderness is common.
  - Occasional swelling and increased size of the affected limb.
  - Pathologic fractures occur in 10% of patients.
ANGIOSARCOMA

- Radiology:
  - Cannot distinguish angiosarcoma, hemangioendothelioma and hemangiopericytoma radiographically.
  - All give lytic “swiss cheese” appearance.

ANGIOSARCOMA: Low Grade
Hemangioma with malignant transformation
ANGIOSARCOMA: High Grade

35yo Man – Died 1yr p presentation.

HEMANGIO-ENDOTHELIOMA

- Intermediate aggressiveness.
  - Can be either benign or malignant.
- Typically young patients.
- Involves soft tissue more commonly than bone.
HEMANGIOPERICYTOMA

- Malignant.
  - 0.1% of primary bone tumors.
  - 0.08% of all bone tumors.
- Thought to be derived from pericytes.
- Rarely primary intraosseous lesion.
- No distinguishing radiological characteristics.


69yo Man

ADIPOSE LESIONS
PATHOLOGY: Adipose

- **BENIGN**
  - Lipoma
  - Lipoblastoma
  - Liposclerosing Myxofibrous Tumor

- **MALIGNANT**
  - Liposarcoma
LIPOMA

- Most common soft tissue tumor (~50%).
  - As many as 2.1% of individuals have a lipoma.
  - Etiology: Neoplasm vs. Focal Hyperplasia.
  - Classification related to fascia:
    - Superficial (common)
    - Deep (Very uncommon).

LIPOMA

- Usually asymptomatic doughy mass.
- No gender predominance.
- Age: 20’s on.
- Multiple in 5%–15% of patients, more often men.
LIPOMA: Superficial

83yo Woman
LIPOMA: Deep

- Osseous:
  - Most frequent adipose lesion of bone.
    - Intraosseous >> cortical or parosteal.
  - Prevalence of about one per 1000 cases of 1° bone tumors.
  - Pain reported in up to 2/3 of cases.
  - Frequent locations:
    - Proximal femur (34% of cases), Calcaneus (8%), Ilium (8%), Tibia (13%), Fibula (10%), Humerus (5%), Ribs (5%).
  - Long bone lesion tend to be in metaphysis.
  - Areas of necrosis.
LIPOMA

31yo Woman

LIPOMA: Mature Fat Necrosis
LIPOMA: CT

LIPOMA
PAROSTEAL LIPOMA

- May be simple or have a bony stalk.
- Stalk may mimic osteochondroma.
- 60% have reaction in underlying bone.

52 yo woman

LIPOSCLEROSING MYXOFIBROUS TUMOR

Polymorphic fibro-osseous tumor of bone
LIPOSCLEROSING MYXOFIBROUS TUMOR

- Usually in the proximal femur (85%).
- Pain presenting symptom in ~50%.
- Diagnosis based on combination:
  - Location.
  - Appearance.
  - Predominant histological pattern.
- The age range is broad, usually adults, The tumors probably arise in childhood.
- Appearance may evolve slowly over time

LIPOSCLEROSING MYXOFIBROUS TUMOR

- Pathology:
  - Crudely woven bone that may have a pagetoid appearance surrounded by fibrous tissue.
  - Fat and myxoid change may be present.
  - May resemble fibrous dysplasia.
- Biopsy required for diagnosis.
- Therapy is curettage.
- Malignant transformation 10%
  - Must follow periodically.
LIPOSCLEROSING MYXOFIBROUS TUMOR

57yo Man with hip pain.

LIPOSARCOMA
LIPOSARCOMA

- Most common soft tissue sarcoma in the adult.
- Rarely arise from preexisting lipoma.
- Location: Thigh, gluteal region, retroperitoneum, leg, and shoulder area.
- Clinical: Most commonly a painless, palpable mass.
- Types:
  - Myxoid: Most common, intermediate differentiation.
  - Well-differentiated: Next most common.
  - Round cell
  - Pleomorphic: Least common, most dedifferentiated.

MRI:
- Entire lesion enhances.
- Areas of necrosis.
- Soft tissue elements: The more the more malignant.
- May be hyperbright.

DDX:
- Angiolipoma.
- Leiomyosarcoma.
- Desmoid tumor.
- MFH.
- Round cell type - Ewing sarcoma and lymphoma.
LIPOSARCOMA

- Therapy:
  - Excision followed by radiation therapy (60-70 Gy).
  - Reduces local recurrence compared with excision alone.

Myxoid Liposarcoma

56 yo man with new claudication in rt thigh.
PATHOLOGY: Other

- **BENIGN**
  - Giant Cell Tumor
  - EG
  - Brown Tumor
  - Tophus/Gout
  - Intraosseous Hematoma (Hemophilia)
  - Ameloblastoma (Gnathic)

- **MALIGNANT**
  - Synovial Cell Sarcoma
  - Chordoma

**GIANT CELL TUMOR**
GIANT CELL TUMOR

- 5 – 9% of primary bone tumors.
  - Possibly most common bone tumor in young adults 25 to 40.
- Women > Men.
- Location: Distal femur > Proximal tibia > Distal radius.
  - Most common tumor of distal phalanges.
- Occur essentially only after physeal plates have closed.

Benign lesion with variable behavior.
- Rare cases shows metastasis to the lungs but lung lesions may behave indolently and even require no treatment.
- Chest CT recommended as a precaution.

Clinical:
- Present with slowly progressive focal pain.
- +/- mass.
- Pathologic fracture relatively common (~10%).
GIANT CELL TUMOR

Radiologic Findings:
- Lytic lesion at the end of a long bone (85%) with characteristic subarticular extension.
- Sacrum and vertebrae common sites (5%).
  - Vertebral lesions in younger age group.
- Geographic appearance with sharp margins and occasional septation.
- Cortex is typically thinned and bone enlarged.
  - Eggshell cortex remaining.
- MRI for staging.

GIANT CELL TUMOR

Pathology:
- Gross: Plain, firm, and homogeneous.
- Foci of hemorrhage or necrosis.
- Microscopic: Numerous multinucleated giant cells.
- Stromal cells are homogeneous mononuclear cells with large nuclei and indistinct nucleoli.
- The nuclei of the stromal cells are identical to the nuclei in the giant cells, a feature that distinguishes giant cell tumors from other lesions with giant cells.
GIANT CELL TUMOR

- **Differential Diagnosis:**
  - Aneurysmal Bone Cyst.
  - Brown Tumor.
  - Chondroblastoma.
  - Telangiectatic osteosarcoma.
  - Malignant fibrous histiocytoma.
  - Metastasis.
  - Plasmacytoma.

- **Therapy:**
  - Surgical resection.
  - Chemical cautery.
  - Methacrylate (heat) or allograft packing (support).
  - NO RADIATION THERAPY.
  - Recurrence rate ~10%
GIANT CELL TUMOR

28 yo man presenting with right knee mass.

GIANT CELL TUMOR

20 yo woman.
GIANT CELL TUMOR

35 yo man.

GIANT CELL TUMOR

20 yo man with left hip pain
GIANT CELL TUMOR

36 yo woman.
GIANT CELL TUMOR

51 yo male with 5 years of low back pain.
EOSINOPHILIC GRANULOMA (EG)

Langerhans Cell Histiocytosis

EOSINOPHILIC GRANULOMA

- Least severe form of a spectrum of Langerhan’s cell histiocytosis:
  - Listerer Siwe Disease (10% of cases, children < 3yo, rapidly fatal.)
  - Hans Schuller Christian Disease (diabetes insipidus, exophthalmos and skull lesions).
  - Eosinophilic Granuloma (60-80% of all cases).
- Etiology: Unknown.
**EOSINOPHILIC GRANULOMA**

- Solitary, non-neoplastic proliferation of histiocytes.
  - Affects bone or lung:
    - Location in bone:
      - Skull.
      - Mandible
      - Spine
      - Long bones.
  - EG can convert to systemic forms of the disease.
  - Bone lesions occur most commonly in children aged 5 to 10.
  - Uncommon in blacks.
  - $\sigma:\varphi = 2:1$.

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**Clinical:**

- Normally symptomatic:
  - Local pain and tenderness.
  - Swelling.
- ESR may be elevated.
EOSINOPHILIC GRANULOMA

Pathology:
- Gross:
  - Soft, granular or gelatinous mass.
  - Gray red to brown with flecks of yellow.
- Histology:
  - Sheets of Langerhan’s cells:
    - Derived from mononuclear cell and dendritic line precursors in the bone marrow.
    - Electron microscopy: Langerhan’s cell has racket shaped cytoplasmic inclusion bodies called Birbeck’s granules.

Radiologic appearance is non-specific:
- Lytic lesion.
- Relatively sharp margin.
- Periosteal reaction is common.
EOSINOPHILIC GRANULOMA

- Long Bones:
  - Diaphysis or metaphysis.
  - Centered in the medullary cavity.
  - May cause endosteal scalloping
  - Periosteal reaction.
- Bone scan is not useful.
- CT cortical penetration.
- MRI staging.

Case courtesy of Akbar Bonakdarpour MD
EOSINOPHILIC GRANULOMA

- Skull lesions have sharp, punched out borders that affect both inner and outer table at different points causing "beveled edge".

EOSINOPHILIC GRANULOMA

- Spine: Normally found in the vertebral body.
- Vertebra Plana:
  - Not specific but EG most common cause in young.
  - Will remodel entirely and is curative.
  - Older patients:
    - Metastasis.
    - Myeloma.
EOSINOPHILIC GRANULOMA

- Differential Diagnosis (Broad):
  - Osteoblastoma.
  - Brown Tumor.
  - Malignant fibrous histiocytoma.
  - Telangiectatic osteosarcoma.
  - Metastasis.
  - Plasmacytoma.
  - Osteomyelitis.

BROWN TUMOR
HYPERPARATHYROIDISM: 1° Brown Tumor

- Well-demarcated lytic lesion.
- Path: Osteoclasts and fibroblasts with hemorrhage.
HYPERPARATHYROIDISM: 1°
Brown Tumor & Osteitis Fibrosa Cystica

HEMOPHILIC PSUEDOTUMOR
HEMOPHILIA: Intraosseous Pseudotumor

INTRAOSSEOUS TOPHUS
INTRAOSSEOUS TOPHUS

- 36yo man with gout.
- Lytic lesion with well-defined, sclerotic margins
- Occur around joints and in flat bones primarily.
AMELOBLASTOMA
(Gnathic)

Adamantinoma (Gnathic)

Peak 4th and 5th decades.
- Wide age range.
- $\sigma:\varphi=1:1$
- Benign and locally aggressive.
- 3 clinical subtypes:
  - Common (polycystic).
  - Peripheral (extraosseous).
  - Unicystic.
AMELOBLASTOMA

- Clinical: Usually asymptomatic, locally invasive tumor.
- Classified into four categories based on clinical behavior, anatomic location, radiographic appearance, and/or histologic features:
  - Solid (multicystic).
  - Unicystic.
  - Desmoplastic.
  - Peripheral.
- Believed to arise from remnants of odontogenic epithelium or developing enamel organ.

AMELOBLASTOMA

- Histology:
  - Mixture of ameloblasts and epithelial cells that try to duplicate the enamel organ.
  - Characterized by a jigsaw-like configuration of the ameloblasts:
    - Exhibit a reverse polarity of cells (Vickers-Gorlin change) = Subnuclear vacuolization away from the basement membrane.
    - Form columnar epithelial cells palisaded about the periphery of the tumor nests.
  - Stellate reticulum in the center.
AMELOBLASTOMA

- Radiographic Appearance:
  - Expansile, circumscribed, unilocular or multilocular radiolucent with corticated margins:
    - Multilocular "soap bubble" appearance.
    - Root resorption and destruction of the cortical plates.
AMELOBLASTOMA

AMELOBLASTOMA
AMELOBLASTOMA

- Treatment:
  - Surgery: Marginal resection approximately 1 cm past the radiographic boundary.
  - Recurrences (up to 25%) usually occur within a year of the initial surgery.
SYNOVIAL SARCOMA

- Uncommon:
  - 8-10% of all sarcomas.
  - 800 new cases per year in the United States.
  - Adults 20 - 50yo.
  - $\varphi$ : $\varphi$ = 3 : 2.
- Cell type: Unknown.
- Malignant:
  - Often misdiagnosed as benign because of
    - Small size.
    - Slow growth.
    - Well-defined appearance.
  - Lower extremities most common: Knees and feet.
  - Intimately related to tendons, tendon sheaths, bursae and fascial aponeuroses.
SYNOVIAL SARCOMA

- **Clinical:**
  - Local pain and mass.
  - Average of ~ 2.5 years before presentation.
  - 5-year survival rate: 23.5 - 64%

- **Pathology:**
  - Gross: Well-demarcated, pink, fleshy masses
    - Calcification noted.
    - Heavy calcification ⇒ Less aggressive behavior.
  - Histology:
    - Large polygonal cells (epithelioid):
      - Secrete hyaluronic acid.
      - Organization suggestive of microscopic joint spaces.
    - Spindle cells:
      - Simulate subsynovial mesenchymal cells.
  - Cytogenetics:
    - 90% have t(X;18) translocation mutation.
    - Unique to synovial cell sarcoma.
SYNOVIAL SARCOMA

- Radiographs:
  - ± Soft tissue mass.
  - 30% calcification ≠ NOT PVNS.
  - Occasional bone erosion or invasion.
- CT: Nonspecific.
- MRI:
  - Soft tissue mass: T1- Intermediate, T2- High.
  - Ill-defined margins.
  - Gd mixed enhancement.

85yo woman with pain posterior to knee.
SYNOVIAL SARCOMA

21yo man with a mass posterior to his knee.

- Note bone marrow signal and cortical irregularity.
SYNOVIAL SARCOMA

11 yo boy with lump on chest.
SYNOVIAL CELL SARCOMA

- Differential Diagnosis:
  - PVNS.
  - Chondrosarcoma.
  - Liposarcoma.
  - MFH.
  - Osteosarcoma, Soft tissue.
  - Ewing Sarcoma, Soft tissue.

CHORDOMA
CHORDOMA

- Rare malignant tumor (1 - 4% of all bone tumors).
- Arises from notochord remnants ⇒ Occur in the mid-line of the axial skeleton.
- Clinical:
  - ♀:♂ = 2:1
  - Highest prevalence 40’s to 60’s.
  - Younger patients at superior end of spine.
- Location:
  - Sacrum: 50%
  - Clivus: 33%
  - C2 and then spinal axis.

CHORDOMA: Clinical

- Presentation depends on the location of the tumor.
  - Sacrococcygeal tumors:
    - Low back pain.
    - Bowel and bladder dysfunction.
    - Sacral tumors are often large at presentation.
  - Clival tumors may present with headaches.
  - Cervical tumors can present as dysphagia and/or neurological deficits.
- Recurrence rate is high.
- Survival is low.
CHORDOMA: Pathology

- Embryology:
  - 4th to 6th week of gestation: Mesenchymal cells from individual sclerotomes merge to surround the notochord and form the vertebral bodies.
  - Notochord normally degenerates and remnants form the nucleus pulposus of the vertebral disc.
  - Theory: Notochord fails to degenerate and undergoes malignant transformation.
    - Fly in the ointment: Normal notochord remnants have never been observed.

CHORDOMA: Pathology

- Gross:
  - Soft, blue-gray, lobulated tumors.
  - Gelatinous translucent areas.
  - Pseudocapsule is present.
  - Lesion tracks along nerve roots in the sacral plexus or out the sciatic notch in planes of least resistance.
CHORDOMA: Pathology

- Histology:
  - Lobules and fibrous septa.
  - Malignant cell has eosinophilic cytoplasm.
  - Prominent vacuoles of mucus push the nuclei to the side resulting in "physaliphorous" cells (Greek: bubble or drop).

CHORDOMA: Clinical

- Radiographs:
  - Solitary mid-line lesion with bony destruction.
  - Often an accompanying soft tissue mass.
  - 50% have focal calcifications.

- CT and MRI:
  - Necessary for optimal staging and possibly visualization.
CHORDOMA

Calcification in ~50%

66yo man with sacral mass: CT.
CHORDOMA

- Generally low on T1.
- Mixed heterogeneous signal on T2.
- Variable enhancement with Gd.

66yo man with sacral mass: MRI(T2FS).
CHORDOMA

76yo man with 40 years of back pain, now with constipation.

INFECTIOUS LESIONS
PATHOLOGY: Infectious

- Brodie’s Abscess
- African Histoplasmosis
RADIOGRAPH: Brodie Abscess

- Formation of a walled off abscess within the medullary cavity:
  - More common in younger patients.
  - Well-defined lytic lesion with sclerotic margins.
  - Generally shows increased activity on bone scan or indium 111 leukocyte scan.
  - Staphylococcus aureus most common organism.
  - 50% are sterile at aspiration.
  - If small, may appear identical to osteoid osteoma.
RADIOGRAPH: Brodie Abscess

BRODIE ABSCESS
REFERENCES

Wow! Bones!
And I thought they were just good eats!