Benign Cartilage Tumors of Bone

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Benign Cartilage Tumors of Bone

- Enchondroma
- Osteochondroma
- Chondroblastoma
- Chondromyxofibroma (CMF)
- Periosteal Chondroma
Enchondroma
General Information

- **Enchondroma** is a benign indolent intramedullary hyaline cartilage neoplasm
- Limited growth, most lesions are less than 5 cm in maximal dimension
- Bones grow from a cartilaginous growth plate that gradually lengthens and turns into bone as it lengthens. An enchondroma can be thought of as an island of cartilage within the bone that never transformed into normal bone.
- Most common tumor of a phalanx
Enchondroma

- Types:
  - Solitary Enchondroma
  - Multiple Enchondromas (Enchondromatosis/Ollier’s Disease)
  - Maffucci Syndrome (Multiple Enchondromas and Soft Tissue Hemangiomas)
Clinical Presentation

• **Signs/Symptoms:** Depends on location
  • Most long bone enchondromas are asymptomatic and found incidentally
  • Phalangeal tumors may be painful due to stress fractures

• **Age:** Range: 15 to 40 years of age

• **Sites:** 50% involve hands and feet (mostly phalanges)
  • Proximal humerus, Femur
  • Enchondromas of the pelvis, vertebrae and ribs are uncommon
  • Almost all hyaline cartilage tumors of the pelvis are considered malignant based on anatomic location no matter what the pathology shows. These tumors will ultimately grow locally and have the potential to dedifferentiate
Radiographic Presentation

- Localized, radiolucent defect usually with punctate calcifications
  - Calcifications are typical but not always present
  - Calcifications are stippled, punctate, popcorn like calcifications and “Ring and Arc” calcifications
- Central or eccentric
- Metaphysis most common for long bones
- Cortex may be scalloped and thinned in the phalanges
- MR better to see noncalcified chondroid lesions and full extent of lesions
Enchondroma

- MRI
  - Lobulated margin
  - Marked increased intensity T2 images
  - Calcified chondroid – low intensity all sequences
  - No cortical destruction; No soft tissue mass; No periosteal thickening
Enchondroma

- **CT Scan:**
  - Subtle calcifications not visible on plain X-rays
  - “Ring and Arc” calcification pattern which helps differentiate from a bone infarct
  - Cortical erosion and scalloping is best demonstrated with a CT scan
Plain X-Ray: Enchondroma of Proximal Phalanx

- Geographic lesion
- Stippled calcifications in lesion
- Phalanx is expanded
- Significant endosteal scalloping
- No cortical destruction
- No soft tissue extension

Cortex Scalloped and Expanded
X-Ray and MRI of Enchondroma of Middle Phalanx

- Geographic Lesion
- Bony Expansion
- Minimal Calcification
- Some Enchondromas do not calcify

T1: Intermediate Signal
- Lobular Growth
- No Soft Tissue Component

T2: High Signal
CT Scan: Enchondroma of Middle Phalanx

There was no mineralization detected in this enchondroma.
Bone Scan: Enchondroma of Middle Phalanx

- Enchondromas demonstrate increased uptake on a bone scan.
- Most benign cartilage tumors demonstrate uptake that is less than the normal uptake in the ASIS.
Plain X-Ray: Enchondroma of Proximal Humerus

- Metaphyseal Tumor
- Heavy Calcifications
- Ring and Arc Pattern of Calcifications
- Minimal Endosteal Scalloping
- No Cortical Destruction
- No Periosteal Reaction
- No Soft Tissue Component
X-Ray and CT Scan: Enchondroma of Femur

Ring and Arc Calcifications
Minimal Endosteal Scalloping
Cortex Intact
No Soft Tissue Component
No Periosteal Reaction
Pathology:
- Rests of hyaline cartilage with a lobular growth pattern
  - Cells are within lacunae
  - Hypocellular with cells spaced apart separated by matrix
  - Minimal pleomorphism
  - No mitotic figures
  - The matrix with ground glass basophilic appearance
- Variable amorphous calcification and enchondral ossification
Microscopic Pathology

- Enchondral ossification may occur around periphery of lobules and when calcified appear as “Rings and Arcs” on X-rays.
- No entrapment or destruction of trabeculae.
- Enchondromas of digits can have hypercellularity, bi and trinucleated cells and myxoid change and still be considered benign.
Microscopic Pathology: Enchondroma

- Cells in Lacunae
- Hypocellular
- Lobular growth
- No Pleomorphism
- Matrix Ground Glass
- Enchondral Ossification around Periphery
Microscopic Pathology: Enchondroma

- Hypocellular
- Cells are Bland and in Lacunae
- Occasional Binucleated Cells
- No Mitoses

Binucleated Lacunae
Microscopic Pathology: Enchondroma
Treatment

- Enchondromas are benign, indolent (not growing) tumors

- Indications for surgery:
  - Digits: Impending or actual pathological fracture
    - Intralesional curettage and bone graft
  - Long bones: Rare to fracture—usually observe
    - If grows it is considered chondrosarcoma and would recommend surgery accordingly
Prognosis

- Recurrence rate following curettage is <5%
- Recurrence of an enchondroma suggests malignancy
Enchondromatosis
(Multiple Enchondromas/Ollier Disease)

• Rare disorder
• Not hereditary
• Multiple intraosseous cartilaginous tumors or enchondromas.
• Clinical Data:
  • Variable severity
  • Marked skeletal deformity
  • May be predominantly unilateral or affect a single extremity/limb
  • Affected limb is often shortened and deformed and angulated
  • May become stable at puberty
  • Higher risk of malignant transformation to chondrosarcoma (5-50%) as opposed to an isolated enchondroma
Enchondromatosis
(Multiple Enchondromas/Ollier Disease)

- Enchondromas in enchondromatosis may involve the metaphysis, diaphysis, epiphysis and articular cartilage

- **Microscopic Pathology:**
  - *Hypercellular* with large number of binucleated chondrocytes
  - Higher degree of cellularity and atypia compared to isolated enchondroma
Enchondromatosis
(Multiple Enchondromas/Ollier Disease)

- Chondrosarcoma arising in enchondromatosis
  - Up to a 50% risk for developing a chondrosarcoma over a lifetime.
  - Pain, cortical destruction and an enlarging mass
  - Most are low grade chondrosarcomas but dedifferentiated chondrosarcomas can also occur
  - Most patients are between the ages of 30-60 when they develop a secondary chondrosarcoma
  - The most common sites to undergo malignant change are the scapula, rib cage and pelvis
Enchondromatosis
Enchondromatosis
Plain X-rays: Enchondromatosis

Enchondroma
Enchondromatosis
Enchondromatosis
Enchondromatosis
Maffucci Syndrome
(Described in 1881 by Maffucci as enchondromatosis with hemangiomas)

- **Clinical Data:**
  - Very rare; Nonhereditary
  - **Multiple Enchondromas**
  - **Cavernous Hemangiomas** anywhere in skin and subcutaneous tissues of body
    - May be unilateral or bilateral, may be only a few or many
    - Phleboliths (calcifications) usually seen in hemangiomas on X-ray
  - Hands and feet greatest involvement
  - Malignant Transformation: Chondrosarcoma (15-20%) Vascular Sarcoma (3-5%) Ovarian Malignancy, Glioma and Carcinoma
    Unrelated to bone or soft tissue
Plain X-Ray of Hand: Maffucci Syndrome

Enchondromas

Phleboliths
Plain X-Ray of Hand: Maffucci Syndrome

Hemangioma with Phleboliths
Osteochondroma
General Information

- Osteochondroma is a benign outgrowth of medullary and cortical bone
  - Bone covered with cartilaginous cap (exostosis)
  - May be secondary to a growth plate injury (Node of Ranvier injured)
- Solitary lesion or as multiple exostoses associated with a hereditary condition known as Multiple Hereditary Exostoses (MHE)
- Radiation exposure can also cause osteochondromas
- Most common benign bone tumor
General Information

- Osteochondromas likely arise from displaced cartilage through periosteal defect and grow at right angles to normal growth plate
- Lesions have self-limited growth that ceases after skeletal maturity
- Due to endochondral ossification, cartilage cap diminishes in thickness as age increases
Osteochondroma

(most common benign neoplasm of bone that leads to biopsy)

• **Radiographic Subtypes:**
  - Pedunculated
  - Sessile
Clinical Presentation

- **Signs/Symptoms:** Hard immobile swelling for many years
  - Mechanical symptoms from compression of adjacent structures such as tendons, nerve or blood vessels
  - An overlying bursa may form and result in a bursitis
  - Rare vascular injuries and arterial aneurysms from adjacent osteochondromas
  - Malignant Transformation: Solitary osteochondroma <1%

- **Sites:**
  - Femur (30%) Tibia (20%) Humerus (20%)
  - Surface of metaphyseal portions of long tubular bones
  - Knee area 35% of cases
Radiographic Presentation

• Plain X-rays:
  • Projects from bone with narrow (pedunculated) to broad (sessile) stalk
  • **Corticomedullary continuity**: Medullary bone continuous with that of osteochondroma and cortex blends with that of osteochondroma
  • Calcification in cartilaginous cap ("Ring and Arc" and stippled calcifications)
  • Long bones: arise from metaphysis, grows away from epiphysis toward diaphysis,
Plain X-ray: Osteochondroma of Proximal Tibia
Proximal Fibula Osteochondroma: Cortical-Medullary Continuity; Ring and Arc Calcifications

Calcifications in Cap

Cortical-Medullary Continuity
MRI of Proximal Fibula Osteochondroma Demonstrates Continuity of Medullary Canal of Fibula with Osteochondroma
MRI Demonstrates Stalk and Continuity with Underlying Fibula; Thin Cartilaginous Cap

Corticomedullary Continuity
Plain Xray/MRI: Distal Femur Sessile Osteochondroma

Thin Cartilage Cap
X-Ray/MRI: Distal Femur Osteochondroma
Osteochondroma of Proximal Femur

(Even though this was a large tumor, the cartilage cap was very thin)
Area between arrows: cartilage cap (approximately 1 cm thickness)
Osteochondroma vs. Secondary Chondrosarcoma

- Malignant transformation is suggested by:
  - Cartilaginous cap thickness greater than 2cm
  - Cortical destruction
  - Backgrowth of the cartilaginous cap into the stalk or medullary canal
  - Lysis of calcifications in cap
Plain X-ray: Secondary Chondrosarcoma of Proximal Femur
MRI: Secondary Chondrosarcoma of Proximal Femur: Thick Cartilage Cap (>2cm)
Pathology

- Pathology:
  - Medullary and cortical continuity with underlying bone
  - Hyaline Cartilage Cap with lobular growth similar appearance to growth plate
  - Cartilage cap involutes after growth
Microscopic Pathology
Replicates a Cartilaginous Growth Plate

- Hyaline cartilage cap overlying medullary bone
- Junction of cap and bone resembles epiphyseal plate
  - Enchondral ossification
Microscopic Pathology: Osteochondroma

Junction of Cap with Medullary Bone: Similar to Growth Plate

- Zone of Hypertrophy
- Zone of Provisional Calcification
- Enchondral Ossification
- Cells in Lacunae
- Ground Glass Hyaline Cartilage Matrix
Treatment

• Simple excision:
  • Cosmetic reasons
  • Impingement on tendons, nerves or blood vessels
  • Pain and limitation of motion
• For multiple exostoses, corrective surgery may be necessary due to secondary deformities
Prognosis

• Recurrence after excision is rare

• Rarely, osteochondromas may give rise to malignant chondrosarcoma
  • Solitary osteochondromas 1%-2%
  • Multiple osteochondromas 5%-25%

• Most common sites to undergo malignant change
  • Scapula, pelvis, ribs, proximal femur
Multiple Heredity Exostoses (MHE)

- **Clinical Data:**
  - Male predominance (3:1)
  - Autosomal Dominant inheritance
  - Mutation in **EXT1** or **EXT2** genes
  - Variable penetrance: variability in size and number of osteochondromas
  - MHE may be bilaterally symmetric
  - One side may predominate
  - Higher incidence of malignant transformation (10-20%) of osteochondromas that develop in MHE.
  - Radiographically: **undertubulation** (Erlermeyer Flask Deformity) of Metaphyses
  - Multiple deformities and shortening of extremities
X-ray: Multiple Hereditary Exostoses of Proximal Femur
X-ray: Multiple Hereditary Osteochondromas of Distal Femur and Proximal Tibia/Fibula
X-Ray: Multiple Hereditary Osteochondromas of Distal Fibula
X-Ray: Multiple Hereditary Osteochondromas of Scapula/Proximal Humerus
Dysplasia Episphysealis Hemimelica – Trevor Disease (Epiphyseal Osteochondroma)

- Male predominance (3:1)
- Very rare < 100 cases
- Swelling, pain and deformity
- Usually lower extremity, unilateral
- 65% multiple bone involvement: talus, distal femur, tibia
Dysplasia Epiphysealis Hemimelica – Trevor Disease

- Ankle and knee most common
- Lobular epiphyseal mass
- Histologically identical to an osteochondroma
- May produce deformity and secondary osteoarthritis
Chondroblastoma
General Information

- Benign neoplasm of **immature cartilage cell** (chondroblast) proliferation
  - Cells resemble chondrocytes/chondroblasts
  - Marked predilection for arising from the epiphysis
- Usually occurs in skeletally immature patients
- <1% of osseous neoplasms
- Benign Aggressive with high propensity for local recurrence
- Rare cases metastasize to lungs
Chondroblastoma

• **Location:**
  • *Almost all cases arise from the epiphysis of the bone*
Epiphyseal Lesions

• Differential Diagnosis of Lesions that tend to involve the epiphysis:
  • Chondroblastoma
  • Clear cell chondrosarcoma
  • Giant Cell Tumor (GCT)
  • Subchondral Cyst/Intraosseous Ganglion
  • Infection
  • Eosinophilic Granuloma (LCH)
  • Osteoid Osteoma
  • Osteoblastoma
  • Mets, myeloma, lymphoma
Clinical Presentation

- **Signs/Symptoms:** Mild Pain lasting from months to several years
  - Joint effusion and swelling with limitations in range of motion
- **Age:** 5 to 25
- **Sites:** Proximal femur, distal femur, proximal tibia & humerus
  - Calcaneus and talus
Radiographic Presentation

- **Geographic /defined/well circumscribed** oval/round lytic defect in epiphysis
  - Periosteal Reaction 30-50% of cases
    - Usually in Adjacent Diaphysis/Metaphysis
    - Calcifications often not visualized; CT better
Radiological Presentation

- MRI:
  - MRI
    - Intermediate Signal on T1
    - High signal on T2 mixed with low signal areas (low signal areas proposed to be secondary to lysosomal content of highly cellular areas)
    - Extensive Surrounding edema is common
Radiological Presentation

- **CT scan:**
  - Subtle mineralization that is not apparent on X-rays
  - Intact periosteum around any expansile soft tissue component that appears as a surrounding thin reactive shell of bone/mineralization (Egg Shell Rim of Calcification). This helps place the tumor in a benign category.

- **Bone Scan:** Chondroblastomas demonstrate intense increased uptake on a bone scan.
Plain X-Ray: Chondroblastoma of Proximal Humerus

- Geographic Lesion
- Epiphyseal Lesion
- Skeletally Immature
- Surrounding Sclerotic Rim
- No Mineralization Detected on Radiograph
CT: Proximal Humerus Chondroblastoma
Subtle calcifications detected on CT that were not detected on plain X-ray

Expansile Lesion: Periosteum Intact around Soft Tissue Component

Subtle Calcification in Tumor
CT Scan of Proximal Humerus
Chondroblastoma

- Calcifications
- Geographic
- Epiphyseal
- Expansile
- Benign
- Aggressive
- Tumor
MRI T2 Weighted Image: Lesion with Primarily High Signal with Low Signal in Many Areas and Extensive Surrounding Edema

Chondroblastomas are often associated with extensive surrounding edema.
Bone Scan: Increased Uptake in Chondroblastoma of Right Proximal Humerus
Plain X-ray: Proximal Tibia Chondroblastoma
CT Scan: Expansile Chondroblastoma of Proximal Tibia Epiphysis with Intact Periosteum
CT Scan: Expansile Chondroblastoma of Proximal Tibia: Subtle Calcifications within Tumor
MRI T1 Weighted Image
Proximal Tibia Chondroblastoma
Intermediate Signal on T1 Weighted Image
Chondroblastomas are primarily high signal on T2 with low signal areas in tumor.

There is also extensive Peritumoral Edema and a knee Joint effusion.

The tumor is well circumscribed.

The periosteum is intact around the expansile soft tissue component.
Bone Scan: Increased Uptake in Proximal Tibia Chondroblastoma
Plain X-Ray: Chondroblastoma of Toe Proximal Phalanx
Plain X-Ray: Chondroblastoma of Talus
Plain X-Ray: Chondroblastoma of Talus

Close Up Lateral
CT Scan: Chondroblastoma of Talus

Sagittal CT Preop
MRI T1: Chondroblastoma of Talus

Sagittal MRI T1 Preop
CT Scan: Distal Femur Chondroblastoma

- Geographic Tumor
- Epiphyseal Location
- Subtle Calcifications
MRI T1 and T2 Weighted Images
Chondroblastoma of Distal Femur

- T1 Weighted Image
- T2 Weighted Image
- Extensive Peritumoral Edema
- Knee Joint Effusion
Microscopic Pathology

- Chondroblast with distinct, thick cell membrane that gives it a "Chicken Wire Fence Appearance"
- Hypercellular
- Cytoplasm is plump, clear, eosinophilic
- Coffee Bean Shaped Nucleus with grooves/invaginations
- "Chicken Wire pattern of Calcification"
- S-100 positive
- None to minimal hyaline cartilage matrix
Microscopic Pathology: Chondroblastoma

- Tightly Packed Cells
- Dark, Thick Cell Membrane
- Bean Shaped Nuclei
- Abundant Cytoplasm
- No Pleomorphism
- No Atypical Mitotic Figures
Chondroblast: Prominent Indented Nucleus
Plump Eosinophilic Cytoplasm
Thick Cell Membrane
Uniform Appearance of Cells
Chicken Wire Pattern of Calcification

The calcium is deposited along the cell membranes and perimeter of the cells in a linear manner.
S 100 Protein Immunostain is Positive in Chondroblastoma
Cartilage stains positive for S-100
Treatment

- **Intralesional curettage resection and bone grafting** is the most common treatment.
  - High risk of local recurrence after curettage alone
Chondromyxofibroma
CMF
Chondromyxofibroma consists of lobules of spindle or stellate cells in an abundant myxoid and chondroid stroma.

Lobules are separated by fibrous type tissue containing spindle type cells and giant cells.

Benign aggressive tumor with high propensity for local recurrence following intralesional curettage. Tumor actively grows and destroys bone.
Clinical Presentation

- **Signs/Symptoms:**
  - Usually painful and may have mild swelling

- **Prevalence:**
  - CMF constitutes less than 1% of all bone tumors

- **Age:** 10 to 30 years of age

- **Sites:**
  - Proximal tibia is most common site
  - Femur
  - Ilium is the most commonly affected flat bone
  - May also occur in tubular bones of hands, feet, vertebrae, ribs
Radiology of Chondromyxoid Fibroma (CMF)

- **Imaging:**
  - Geographic, well circumscribed lesion
  - Eccentric metaphyseal location
  - Rare matrix calcification requires CT/Tomography usually for detection
  - Often has an indolent, sclerotic medullary border and expansive blown out exterior border
  - Internal trabeculations frequent
Radiographic Presentation

Chondromyxofibroma of Proximal Tibia

Geographic, Eccentric Lesion

Indolent, Sclerotic Medullary Border

Expansile Blown Out Border

Subtle Internal Trabeculations
Plain X-ray: Chondromyxofibroma of Right Posterior Ilium
Plain X-Ray: CMF of Posterior Ilium: Geographic, Expansile Lesion
MRI T2 CMF of Right Posterior Ilium
MRI T2: CMF of Right Posterior Ilium (High Signal Intensity)
CT Scan of Chondromyxofibroma of Right Posterior Ilium

- Subtle Mineralization
- Geographic Expansile Lesion
Plain X-ray: Chondromyxofibroma of Calcaneus
Plain X-ray: Chondromyxofibroma of Calcaneus

- Typically an eccentrically located, metaphyseal lesion
  - May extend into epiphysis
  - Long axes parallel to bone
- Usually sharply demarcated
  - Scalloped margins
- Intralesional calcified matrix rare
MRI T1 Chondromyxofibroma of Calcaneus
Intermediate Signal Similar to Muscle

Expansile Outer Border
Expanding Cortex
Indolent Border
Sharp Zone of Transition
MRI T2: Chondromyxofibroma of Calcaneus (Cartilage Tumors are Often High Signal on T2 Weighted Images)
Plain X-ray: Chondromyxofibroma of Right Femoral Neck
Xray: Chondromyxofibroma of Right Femoral Neck
CT: CMF of Right Femoral Neck

- Indolent Border
- Eccentric Lesion
- Geographic
- Well Circumscribed
- Expansile Border
- Periosteum Intact
CT: CMF of Femoral Neck

Expansile Outer Border with Egg Shell Rim of Calcification

Indolent Medullary Border

Sharply Circumscribed
Plain X-ray: Chondromyxofibroma of Proximal Phalanx of Toe

- Expansile Lesion
- Well Circumscribed
- Geographic
- No Mineralization Detected

(Mineralization is not always detected in CMF)
CT Scan (sagittal): CMF of Proximal Phalanx of Big Toe
MRI T2: CMF of Proximal Phalanx of Big Toe (High Signal)
Plain X-Rays of CMF of Proximal Right Tibia

Sclerotic/Indolent Medullary Border
CT Scan of CMF of Proximal Tibia
Calcifications within Lesion are Demonstrated
MRI T1 and T2 of CMF of Proximal Tibia
Chondromyxoid Fibroma (CMF)

• **Pathology:**
  • Myxoid, fibrous and chondroid tissue in various proportions with lobular growth pattern
  • Stellate chondroid cells are characteristic
  • Myxoid areas – central
  • Cellular areas – peripheral
Microscopic Pathology CMF

Chondromyxoid Area

Cellular Area
Chondromyxoid Area

Fibrous Area
Microscopic Pathology

Fibrous Areas Around Periphery of Myxoid Chondroid Areas

Myxoid Area with Chondroid Tissue with Stellate Types of Cells
High Power of Chondromyxoid Area with Stellate Types of Cells

Stellate Cells have Long Cytoplasmic Processes
Treatment

• Intralesion curettage and bone grafting
• Recurs 30%-60% of the time after intralesion curettage. It grows aggressively and destroys bone.
• En bloc resection may need to be considered for extremely large tumors and/or tumors that have recurred after being treated with previous intralesion surgeries.
Periosteal Chondroma
General Information

- Benign neoplasm
- Composed of mature hyaline cartilage
- Arises from surface of bone from inner layer of periosteum
- Erodes the outer table of the cortex
- Does not grossly extend into medullary cavity
- More cellular than an enchondroma
Periosteal Chondroma

- **Sites:**
  - Proximal humerus - Most common
  - Femur, tibia, phalanges are common sites
  - Pelvis, ribs, vertebrae less common
Radiographic Presentation

- Metaphysis (2/3 of tumors)
- Long bone lesions
  - Size: 2-3 cm in size, up to 6 cm
  - Over 6 cm is worrisome for periosteal chondrosarcoma
- Short tubular bones
  - Up to 3 cm in size, usually between 1 and 2 cm
  - Calcifications in a “Ring and Arc” manner and/or stippled calcifications
Xray/CT Scan: Periosteal Chondroma of Proximal Humerus
Microscopic Pathology

- Lobulated, hyaline cartilage tumor
- May be more cellular than an enchondroma with myxoid change of matrix
Microscopic Pathology: Periosteal Chondroma
Biological Behavior

- No metastasis
- No malignant change
- Exceedingly rare recurrence
- Non aggressive
Treatment & Prognosis

- Marginal excision without removal of surrounding tissue
  - Occasional rare recurrence
- En bloc excision
  - Invariable curative
Thank You!