# Benign Cartilage Tumors of Bone

James C. Wittig, MD Orthopedic Oncologist Sarcoma Surgeon www.TumorSurgery.org



## Benign Cartilage Tumors of Bone

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



# Enchondroma



### **General Information**

- <u>Enchondroma</u> 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
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### 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 *Wittig Orthopedic Oncols*

### 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

T1: Intermediate Signal **T2: High Signal** Lobular Growth No Soft Tissue Component **Geographic Lesion Bony Expansion** 10B **Minimal Calcification** ະສຸດກັ TRAT KART 000 Some Enchondromas do not calcify Orthopedic Onco

#### **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



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#### Plain X-Ray: Enchondroma of Proximal Humerus



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#### X-Ray and CT Scan: Enchondroma of Femur



Ring and Arc Calcifications Minimal Endosteal Scalloping Cortex Intact No Soft Tissue Component No Periosteal Reaction R

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### Enchondroma

### 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



#### Microscopic Pathology: Enchondroma





#### Microscopic Pathology: Enchondroma



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### 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





- 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 *Wittig Orthopedic Oncols*

### 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

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### Plain X-rays: Enchondromatosis















### Maffucci Syndrome

(Described in 1881 by Maffucci as enchondromatosis with hemangiomas)

#### • Clinical Data:

- Very rare; Nonhereditary
- <u>Multiple Enchondromas</u>
- <u>Cavernous Hemangiomas</u> 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 softtissue

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#### Plain X-Ray of Hand: Maffucci Syndrome





#### Plain X-Ray of Hand: Maffucci Syndrome





## 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

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• Knee area 35% of cases

# **Radiographic Presentation**

### • Plain X-rays:

- Projects from bone with narrow (pedunculated) to broad (sessile) stalk
- <u>Corticomedullary continuity</u>: 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,

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#### Plain X-ray: Osteochondroma of Proximal Tibia



### **Proximal Fibula Osteochondroma: Cortical-Medullary Continuity; Ring and Arc Calcifications**



#### 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



# Specimen







### Plain Xray/MRI: Distal Femur Sessile Osteochondroma



### 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 w/ 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



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**Medullary Bone** 

#### **Cartilage Cap**

#### Microscopic Pathology: Osteochondroma

#### Junction of Cap with Medullary Bone: Similar to Growth Plate



## 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

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• 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
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#### 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 Episphysealis 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 <u>immature cartilage cell</u> (chondroblast) proliferation
  - Cells resemble chondrocytes/chondroblasts
  - Marked predilection for arising from the epiphysis
- Usually occurs in skeletally immature patients
- <1% of osseous neoplasms</p>
- Benign Aggressive with high propensity for local recurrence

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• 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:

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- 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**

- <u>Geographic</u> /<u>defined/well circumscribed</u>
   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

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# CT Scan of Proximal Humerus Chondroblastoma



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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



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#### CT Scan: Expansile Chondroblastoma of Proximal Tibia: Subtle Calcifications within Tumor



### MRI T1 Weighted Image Proximal Tibia Chondroblastoma Intermediate Signal on T1 Weighted Image



#### MRI T2 Weighted Image Chondroblastoma of Proximal Tibia

- 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

Low Signal Areas on T2

## 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



### **CT Scan: Chondroblastoma of Talus**



### **MRI T1: Chondroblastoma of Talus**



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### **CT Scan: Distal Femur Chondroblastoma**





# MRI T1 and T2 Weighted Images Chondroblastoma of Distal Femur





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# **Microscopic Pathology**

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

Tightly Packed Cells Dark, Thick Cell Membrane Bean Shaped Nuclei Abundant Cytoplasm No Pleomorphism No Atypical Mitotic Figures





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#### **Chicken Wire Pattern of Calcification**

#### The calcium is deposited along the cell membranes and perimeter of the cells in a linear manner



#### Von Kossa Stain for Calcium



S 100 Protein Immunostain is <u>Positive</u> in Chondroblastoma Cartilage stains positive for S-100 Wittig Orthopedic Oncossy

### Treatment

 Intralesional curettage resection and bone grafting is the most common treatment.
 High risk of local recurrence after curettage alone



# Chondromyxofibroma CMF



# General Information Chondromyxofibroma

- 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.

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## **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
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# Radiology of Chondromyxoid Fibroma (CMF)

### <u>Imaging:</u>

- Geographic, well circumscribed lesion
- Eccentric metaphyseal location
- <u>Rare matrix calcification</u> 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 Wittig Orthopedic Oncol

#### Plain X-ray: Chondromyxofibroma of Right Posterior Ilium



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#### Plain X-Ray: CMF of Posterior Ilium: Geographic, Expansile Lesion



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# 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



## 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**



**Expanding** Cortex

rp 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



### **CT: CMF of Femoral Neck**



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### Plain X-ray: Chondromyxofibroma of **Proximal Phalanx of Toe**

Detected

in CMF)

Sclerotic Medullary Border **Expansile Lesion** Well Circumscribed Geographic **No Mineralization** (Mineralization is not always detected Wittig Orthopedic Oncors

## CT Scan (sagittal): CMF of Proximal Phalanx of Big Toe



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# MRI T2: CMF of Proximal Phalanx of Big Toe (High Signal)



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### Plain X-Rays of CMF of Proximal Right Tibia



### **CT Scan of CMF of Proximal Tibia** Calcifications within Lesion are Demonstrated



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### 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**





#### **Fibrous Area**

#### CMF Pathology

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High Power of Chondromyxoid Area with Stellate Types of Cells Stellate Cells have Long Cytoplasmic Processes Wittig Orthopedic Oncodes

### Treatment

- Intralesional curettage and bone grafting
- Recurs 30%-60% of the time after intralesional 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 intralesional 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
- Calcifcations in a "Ring and Arc" manner and/or stippled calcifications



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## 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



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# **Biological Behavior**

- No metastasis
- No malignant change
- Exceedingly rare recurrence
- Non aggressive



## **Treatment & Prognosis**

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

# **Thank You!**

