Cartilage Forming Tumors

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Cartilage Forming Tumors

- Osteochondroma
- Enchondroma
- Chondroblastoma
- Chondrosarcoma
Osteochondroma

- Most common benign skeletal tumor
  - 20-50% of all benign bone tumors
- Most frequent in 1st and 2nd decade of life
- Male : female = 1.5 : 1
- Most often in juxta-epiphyseal / metaphaseal area of long bones (distal femur, proximal tibia)
  - 40% around the knee (also shoulder, hip)
Osteochondroma

- Bony exostosis with cartilage cap
- Typically grow away from physis
- Growth ceases after maturity
- Growth after maturity indicates malignant transformation
- < 1% risk of malignant transformation for solitary lesions
Clinical Presentation

- Painless bony mass
- Can be painful if ......
  - Mechanical irritation (nerves, vessels, muscles, tendons, bones)
  - Fracture
  - Bursa formation
Radiographic Appearance

- Diagnostic
- Sessile or stalk-like (exostosis)
- Metaphyseal bone may be expanded and remodeled
- Cartilage cap may be calcified
- Base of lesion contiguous with cortex of bone
X-Ray
Differential Diagnosis

- Juxtacortical myositis ossificans
- Periosteal chondroma
- Surface (parosteal) osteosarcoma
- Soft tissue osteosarcoma
- Chondrosarcoma arising in an osteochondroma
CT

- Determine if marrow and cortices of lesion are continuous with bone

MRI

- Proximity to other structures
- Cartilage cap
Gross: irregular bony mass with bluish-grey cartilage cap

Microscopic: cartilage cap disorganized, covered with thin layer of periosteum maturing into trabecular bone

Medullary canal is continuous with bone
Histology

Cartilage

Bone
Treatment

- **Asymptomatic**
  - No treatment

- **Painful**
  - Excise (watch proximity to growth plate)

- **Change in cartilage cap**
  - Biopsy

- **Very large lesions**
  - Follow regularly to detect malignant transformation early (low-grade chondrosarcoma or osteosarcoma)
Hereditary multiple osteochondromatosis
- Autosomal dominant
- Causes skeletal deformities
- Symptoms in 1st decade of life
- ??? 25-30% risk of malignant transformation
- Sessile more likely to transform than exostosis
Multiple Hereditary Exostosis
Enchondroma

- Solitary, benign, intramedullary cartilage forming tumor
- Peak incidence in 3rd decade
- Male : female = 1 : 1
- Common in short tubular bones of hands and feet
  - Most common primary tumor in the hand, usually in the diaphysis
Clinical Presentation

- Hand > Foot lesions more commonly active
  - Cortical bone erosion resulting in pain, bony mass, or pathologic fracture
  - Expansile lesions may cause palpable bony mass
- Found incidentally in long bone
  - < 1% risk of malignant transformation
Radiographic Appearance

- **Long bones**
  - Metadiaphysis
  - Most common in femur and humerus

- **Tubular bones**
  - Diaphysis

- **Matrix** – “popcorn”, “comma shaped”, “stippled” calcification
Enchondroma
Features strongly associated with malignant transformation

- Large size, lytic component, significant thinning of adjacent cortex
- Progressive destruction of chondroid matrix by an expanding, nonmineralized component
- Enlarging lesion associated with pain
- Soft tissue mass
Differential Diagnosis

- What else has calcified matrix?
  - Chondrosarcoma
  - Fibrous dysplasia
  - Medullary bone infarct

Calcification in periphery
Other Imaging

- **Bone Scan**
  - Always hot
  - Multiple lesions suggests Ollier’s disease

- **CT**
  - Useful for evaluating calcifications / matrix mineralization
  - Useful for evaluating endosteal scalloping / erosion and lucency within lesion (enchondroma vs chondrosarcoma)

- **MRI**
  - Not definitive for benign vs malignant unless soft tissue mass present
Bone Scan
Histology / Pathology

- Lobules of hypocellular hyaline cartilage
- Chondrocytes may cluster and some lacunae may be binucleate
- May see thin rim of bone surrounding lobules
Histology
Treatment

- Latent lesions
  - Plain x-rays with F/U x-rays showing lack of progression (3 months, 6 months, 1 year)

- Biopsy when
  - Lesion changes over time
  - Persistent pain without other cause
  - Worrisome features
Special Note

- **Ollier’s Disease**
  - Multiple enchondromatosis
  - Non-heritable

- **Maffucci’s Syndrome**
  - Multiple enchondromas and hemangiomas of soft tissue

- Usually one side of body affected
- 30% risk of malignant transformation (Maffucci’s higher)
Chondroblastoma

- Rare, benign tumor derived from chondroblasts (5% of benign bone tumors)
- Epiphysis of long bones (also apophyseal)
- Most common sites
  - Femur, humerus, tibia
Chondroblastoma

- Male : female = 3 : 2
- Mean age: skeletally immature
- May have behaviour not normally associated with benign tumors (pulmonary metastases, local bone / soft tissue invasion)
Clinical Presentation

- Pain near a joint without history of trauma
- Tumor can induce a secondary synovitis
- Patient may have a joint effusion
- Pathological fracture rare
Radiographic Appearance

- Lytic
- Well-defined margins
- Scalloping or erosion of cortical bone may be present
- Fine calcifications (punctate, rings)
Differential Diagnosis

- Clear cell chondrosarcoma
- Infection
- Eosinophilic Granuloma
- Degenerative Geode

Geode = A hollow or partly hollow, hard, globular body, usually from 2.5 to 30 cm or more in diameter found in certain limestone beds and, rarely, in some shale beds, characterized by a thin and sometimes incomplete outermost layer of chalcedony, by a cavity that is partly filled with a lining of inward-projecting crystals deposited from solution on the cavity walls, and by evidence of growth by expansion in the cavities of fossils or along fracture surfaces of the shells.
Other Imaging

**CT**
- Useful for defining relationship of lesion to the joint, integrity of cortex, intralesional calcifications

**MRI**
- Peritumoral edema
Histology / Pathology

- Uniform, polygonal, closely packed cells
- Scant chondroid matrix may be superimposed by a pericellular deposit of calcification (“chicken wire”)
- Giant cells often present
Chicken Wire Calcification
Treatment

- Biopsy + curettage + bone graft
- Excise all pulmonary nodules
- Recurrence rate: 10 – 30%
Chondrosarcoma

- Malignant tumor that produces cartilage matrix

Primary chondrosarcoma
- Very uncommon, arises centrally in bone, found in children

Secondary chondrosarcoma
- Arises from benign cartilage defects (osteochondroma - surface, enchondroma - intramedullary)
Chondrosarcoma

- Occurs in 5th or 6th decade of life
- Male : female = 1.5 : 1
- Location: femur, humerus, ribs, pelvis
- Higher risk to occur in patients with Ollier’s Disease and Maffucci’s Syndrome (3rd or 4th decade of life)
Clinical Presentation

- Pain
- Enlarging mass

Abnormal!!
Radiographic Appearance

- Partially lytic lesion
- Scalloping of inner cortex with periosteal reaction
- May have extension into soft tissue
- Matrix: punctate or stippled calcification
Differential Diagnosis

- Enchondroma
- Osteochondroma
- Osteosarcoma (cartilaginous subtype)
- Chordoma (can calcify)
Other Imaging

CT
- Useful for defining integrity of cortex and distribution of calcification

MRI
- Useful for surgical planning
- Useful in evaluating possible malignant transformation of osteochondromas (size of cartilage cap > 2 cm)
CT

Oink!!
Histology / Pathology

- Multiple cells per lacunae, binucleated cells, nuclear pleomorphism

Grades 1 (low) – 3 (high)

- Low grade: close in appearance to enchondromas and osteochondromas, occasional binucleated cells
- High grade: increased cellularity, atypia, and mitoses
Chondrosarcoma
Low Grade Chondrosarcoma
Histology

Medium Grade

High Grade
Treatment

- **Wide surgical excision**
- Classically **does not** respond to chemotherapy or radiation
- Excellent survival rate for low-grade chondrosarcomas
- Overall 50% survival rate (high-grade much poorer prognosis)
- Recurrence rates associated with adequacy of resection
Clear Cell Chondrosarcoma

- Malignant cartilage tumor
- Rare, low-grade tumor with better prognosis than other chondrosarcomas
- Radiographically similar to chondroblastoma but occurs in adult population
Dedifferentiated Chondrosarcoma

- Most malignant form of chondrosarcoma
- Mix of low-grade and high-grade spindle cell sarcoma (can no longer identify that spindle cells have cartilage origin)
- **5-year survival: 10%**

Special thanks to the Chinese!!
Osteochondroma

Enchondroma

Chondroblastoma

Chondrosarcoma
Thanks!!