The Spectrum of Cartilaginous Tumors

EP

- 34y/o F with “chronic LBP,” underwent appendectomy 2 weeks prior to presentation. Abdominal CT identified lytic sacral lesion. Endorses diffuse/aching sacral pain, denies B/B complaints, perineal numbness.
- PMH: type 1 diabetes
- PSH: L5-S1 discectomy for HNP (2000)
- Meds: insulin
- All: vancomycin
- Soc Hx: Kindergarten teacher, 1 son.
- PE: TTP at L sacrum, no ST mass. NVI.
86yo F, with chronic “L knee pain” presents with atraumatic onset of pathologic fracture L distal femur.
PMH: p. vera, stroke, hypertension, GERD
PSH: ORIF L hip fracture ~5y prior, appy, hysterectomy
Meds: hydroxyurea
All: labetolol
Soc Hx: Lives independently. Performs all ADLs, occasionally uses walker/cane.
PE: TTP at fracture site, mild swelling/ecchymosis, NVI
JC
- 61 y M, 9 mo painless, slowly growing anterior chest mass
- PMH: Charcot Marie Tooth
- PSH: Foot
- Meds: lisinopril, vitamins
- NKDA
- SocHx: Manages stockroom, enjoys golfing
- PE: Painless firm prominence over sternum. Normal BUE.

JG
- 52 y M, very active, farm equipment mechanic with 1 y worsening right hip pain.
- PMH: neg, but does not see a PCP
- PSH: T&A
- Meds/All: none
- Soc Hx: +tobacco
- Fam Hx: neg
- PE: antalgic gait, irritable hip
• 32 y/o healthy F sustained a R shoulder injury doing yoga 6 weeks prior to presentation. Imaging identified R scapular lesion for which she was referred. Shoulder pain completely resolved at our appointment. No prior shoulder pain/limitation.
• PMH/PSH/meds/All: none
• Soc Hx: Desk job, enjoys volleyball, staying active
• PE: Prominence at R inferomedial scapula, nontender. Full/symmetric ROM. RUE ~2cm shorter than LUE.
### Spectrum

**Benign → → → → → → → → → → → Malignant**

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Benign Cartilage Forming Tumors

- Enchondroma

Benign Cartilage Forming Tumors

- Periosteal chondroma

Benign Cartilage Forming Tumors

- Chondromyxoid Fibroma
Benign Cartilage Forming Tumors

- Chondroblastoma

Benign Cartilage Forming Tumors

- Osteochondroma

Spectrum

Benign → → → → → → → → → Malignant

Enchondroma
Periosteal chondroma
Osteochondroma
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Chondromyxoid Fibroma
Synovial (Osteo)Chondromatosis

Chondrosarcoma
Conventional IM
Secondary/Transform
Periosteal
Clear Cell
Mesenchymal
Dedifferentiated
Chondrosarcoma

- Most common bone sarcoma after 20 years old, usually in Px's 50y or older.
- Primary complaint is a painful mass.
- Most common locations in order = pelvis, proximal femur, shoulder girdle.

CHONDROSARCOMA

- Cartilage forming malignant cells without osteoid formation
- Primary – central conventional CS
  - Grade I/II/III 10y survival: 83%/64%/29%
- Secondary
  - Secondary chondrosarcs can arise from enchondroma, osteochondroma (~1%/lesion/year), Px's c MHE, enchondromatosis (Ollier's DZ ~25% and Maffucci Syn ~100%).
- Rare Variants
  - Mesenchymal/Dedifferentiated/Clear Cell/Periosteal

Chondrosarcoma

- Neither radiology or histopathology can always predict biologic behavior.
- Location in skeleton MATTERS
  - Finger & toe lesions: aggressive rads/histo, less so clinically
  - Axial lesions: behave more aggressively
- Exostotic (secondary peripheral or periosteal) are more likely lower grade
- Increase in size or pain may indicate malignant transformation
- Change over time (radiological/clinical)
CHONDROSARCOMA - Histology

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<th>Grade</th>
<th>Cytology</th>
<th>Matrix</th>
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<td>I (50-60%)</td>
<td>Similar to enchondroma, slightly increased cellularity</td>
<td>Chondroid, Rare, focal necrosis</td>
</tr>
<tr>
<td>II (40-50%)</td>
<td>Increased cellularity, irregular nuclei</td>
<td>Chondroid to myxoid, focal necrosis</td>
</tr>
<tr>
<td>III (5-10%)</td>
<td>Very cellular, pleomorphism, atypia, spindling, mitosis</td>
<td>Prominent myxoid stroma, extensive necrosis</td>
</tr>
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</table>

*Presence of tumor entrapment of host bone lamellae - most important indication of malignancy*

CHONDROSARCOMA - Radiology

- Large (mean size=9.5cm).
- 75% mineralized (rings, arcs, stippled calcifications)
- 84% cortical abnormalities (endosteal scalloping over 50% of cortex, bone destruction, expansion, ST mass)

Radiology

- **High grade:** Destructive, symmetric bone expansion, predominant lysis w mineralization, soft tissue mass
- **Low grade:** intraosseous, no cortical destruction vs endosteal scalloping

1. Characteristics to distinguish low grade CS from benign enchondromas?
   - Cold bone scan = benign
   - FDG-PET: grade II/III higher uptake than grade I
   - MRI: soft tissue mass (HGCS) and entrapped fat within tumor (LGCS)
   - DCE MRI: 100 % sensitivity, 63.3 % specificity and 93.4 % accuracy for LGCS but 36.7% FP with enchondromas
2. Effective use of imaging studies to plan biopsy site?
Spectrum

Benign → Enchondroma → Low grade Chondrosarcoma → Chondrosarcoma → Malignant

Enchondroma or Low Grade Chondrosarcoma?

Chondrosarcoma

Low grade Chondrosarcoma

Conventional IM
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Periosteal
Clear Cell
Mesenchymal
Dedifferentiated

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Osteochondroma
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Chondrosarcoma
SecondaryTransform
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Dedifferentiated Chondrosarcoma

- Bimorphic pattern - low grade chondrosarcoma with sharply juxtaposed high grade sarcoma.
- 10% CS transform into more anaplastic, high grade lesions
- Tx= wide resection, chemo for those who can tolerate it.
- 5y survival: 0-18%

Cartilage Tumor Syndromes

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<tr>
<th>Ollier’s Disease</th>
<th>Maffucci’s Disease</th>
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<tr>
<td>Multiple enchondromas</td>
<td>Multiple enchondromas + soft tissue hemangiomas</td>
</tr>
<tr>
<td>Non hereditary</td>
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</tr>
<tr>
<td>1/100,000</td>
<td>100% malignancy risk (usually visceral)</td>
</tr>
<tr>
<td>Assymmetric distribution</td>
<td>Enchondroma: ~56%</td>
</tr>
<tr>
<td>Highly variable presentation</td>
<td></td>
</tr>
<tr>
<td>Skeletal deformity, LLD</td>
<td></td>
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<td>~30% malignancy risk</td>
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Enchondromatosis

Metachondromatosis
Spondyloenchondrodysplasia
Dysspondyloenchondromatosis
Genochondromatosis
Cheirospondyloenchondromatosis

Ollier's Disease

Maffucci's Disease
CHONDROSARCOMA - Treatment?

• SURGICAL DISEASE
  ○ Chemo and radiation DO NOT WORK

• Treatment
  ○ GRADE & LOCATION
    ▪ Low grade: curettage and grafting
    ▪ High grade: wide resection

Treatment Issues

• Acral tumors: can curettage even if not low grade
• Axial Skeleton/Flat Bones (Pelvis, Scapula)
  ○ Wide excision for all grades
• Low grade CS in long bones
  ○ Intralesional Excision and adjuvant therapy vs. Wide Excision
• Intermediate to High grade
  ○ Wide resection, limb salvage
  ○ Inadequate margins: 33-69% LR rate

Prognosis

Metastasis (10y survival)
  Grade I: 0% (77-89%)
  Grade II: 10-33% (53-59%)
  Grade III: 70% (36-38%)
Thank you!