Imaging of Chondrosarcoma

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History of Present Illness

• 34-year-old woman presents with one-year history of "tightness" in her medial right proximal thigh. She describes deep aching soreness in the medial proximal right thigh.

• No history of trauma, penetrating injury, numbness or tingling in the right leg.

• Fail to improve with physical/occupational therapy and time.
Patient’s History

- PMH: none
- Past surgical history: Fibroadenoma excision from right breast
- Ob/gyn: Two children delivered by spontaneous vaginal delivery. Youngest is 3 years old
- Medication: Multivitamin
- Family history: Hodgkin’s lymphoma
Physical Exam

• Right thigh with slightly more girth than contralateral side. Firm, fixed, nontender mass in the medial aspect of right proximal thigh on palpation. Mass measures at least 10cm in diameter in all dimensions. Range of motion is supple without evidence of limitation.

• Right foot is warm with good capillary refill. Normal pulses, sensation. 5/5 foot dorsiflexion, foot plantar flexion, great toe extension, knee flexion, knee extension, hip flexion.

Next step: Radiograph of the right femur……
First, let’s review the approach to evaluation of bone lesions……

- Location (cortex vs. medulla, epiphysis/metaphysis/diaphysis)
- Underlying bone abnormality
- Single vs. Multifocal
- Margin
  - Well-define, rim of sclerosis = Less aggressive
  - moth-eaten, permeative = More aggressive
- Cortical expansion or destruction
- Soft tissue mass
Approach to evaluation of bone lesions......

- Periosteal reaction (Solid, lamellated, Sunburst, Codman’s triangle, “onion-skinning,” spiculated “hair-on-end”)

- Mineralized matrix
  - Cloud-like, amorphous = osteoid
  - Rings and arcs, lobulated = chondroid
Our patient’s radiograph of the right femur

**LOCATION**

- Medial aspect of the proximal right femoral diaphysis, inferior to the level of lesser trochanter.
- Precise localization difficult: Along the surface or adjacent soft tissue
- ? Broad-based or pedunculated
Magnified views of the radiograph of the right femur

Matrix:
- Rings and arcs, lobulated, implying cartilaginous matrix
- Cloud-like, implying osteoid

Cortex:
- Saucerization of the medial femoral diaphyseal cortex

PACS, BIDMC

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Radiograph of the Right Femur: Impression

- Large mass with probable cartilaginous matrix located medial to the proximal right femur just inferior to the level of the lesser trochanter
- Precise localization of this mass is difficult from the radiographs. The lesion seems to be located along the surface of the right femur or less likely in the adjacent soft tissue
- Saucerization of the medial aspect of the proximal femoral diaphyseal cortex
- Further characterization of the lesion with CT or MRI is advised
Differential Diagnosis Based on Radiograph

- Malignant transformation of an osteochondroma
- Surface-based chondrosarcoma
- Surface-based osteosarcoma (Parosteal, Periosteal) with chondroblastic components
- Simple osteochondroma
Coronal and axial images on T1 show large mass in the right proximal medial thigh.

- Isointense to muscle on T1 sequence
- Base of the lesion demonstrates low signal
**MRI of the Right Femur: STIR**

- Coronal and axial images on STIR sequence shows large multilobulated mass
- Markedly increased signal on STIR, isointense to muscle on T1, and “rings-and-arcs” on radiograph suggest cartilaginous matrix
- Mass arises from the medial cortex of the femur 1 cm below the lesser trochanter, with medial displacement of the rectus femoris, adductors, and sartorius
- No connection with the medullary cavity, bone marrow edema in the adjacent femur
- Base of the lesion again demonstrates low signal, consistent with dystrophic calcifications
MRI of the Right Femur: Post-contrast

- The mass demonstrates heterogeneous lace-like patchy enhancement post-contrast, excluding the possibility of a cystic lesion.
- Again demonstrates lobulated lesion with adjacent bone marrow edema.
Patient’s MRI of the Right Femur: Impression

- Large mass arising from the medial cortex of the proximal femur with displacement of the adjacent soft tissue structures
- The lesion demonstrates cartilaginous components
- No involvement of the adjacent neurovascular structures
- There is no connection with native medullary cavity of the femur, excluding malignant transformation of osteochondroma from the differential diagnosis
- Differential diagnosis: Surface chondrosarcoma, parosteal or periosteal osteosarcoma with chondroblastic component
The patient underwent CT-Guided biopsy of the mass
Bone window demonstrates calcified area of the lesion

Soft tissue window demonstrates soft tissue component of the lesion subtle on the bone window
Pathology result shows fragments of benign cartilage …..
Discordance between the image findings and pathology: What is the next step?

1. Treat like benign lesion: Preservation of femur, piecemeal-type excision

2. Treat like low-grade chondrosarcoma: Resect proximal femur

3. Second biopsy (calcified and cartilaginous portions) attempting to decrease sampling error
After extensive discussions amongst the patient, orthopedics, radiologists, and pathologists, the patient decided to undergo another biopsy……
CT-Guided Biopsy #2: Pathology

Area of hypercellularity

Focal chondrocyte atypia

* While this tumor may represent a benign cartilaginous or osteocartilaginous neoplasm, the presence of hypercellular cartilage raises the possibility of a low-grade chondrosarcoma. There is no evidence of osteosarcoma in this sample.
The patient also underwent tumor work-up with contrasted CT of the chest and bone scan for possible metastatic lesions……
CT Chest with Contrast: Impression

- Multiple 2-3 mm noncalcified pulmonary nodules. If there is a proven primary malignancy, then follow-up should be obtained in three months to exclude small foci of metastatic disease.
- Sclerotic lesion in the L1 vertebral body. Correlation with bone scan is recommended.
Bone Scan

Abnormal uptake corresponding to the known large mass centered in the medial proximal right femur and thigh. No evidence for distant osseous lesions.
Clinical Decision

- Although the pathology is not sufficient to reach the diagnosis of a chondrosarcoma, the hypercellularity and chondrocyte atypia, in conjunction with the image finding, suggest the possibility of a chondrosarcoma.

- The patient will be treated as having a low-grade chondrosarcoma with en bloc resection of the proximal femur.
Chondrosarcoma

- Malignant tumor of cartilaginous origin
- Primary – arising de novo
- Secondary – malignant transformation of benign cartilaginous neoplasm (enchondroma or osteochondroma)
- Primary chondrosarcoma constitute 20% - 27% of all primary malignant osseous neoplasm
- Primary: age 20 – 60 years old. Most tumors arise in patients > 40
- Male > Female (1.5-2 to 1)

Chondrosarcoma

• Different types include: conventional intramedullary (most common), clear cell, juxtacortical, myxoid, mesenchymal, extraskeletal, and dedifferentiated

• Common Sites: Pelvic bone, femur, humerus, ribs, scapula, sternum, spine

• Proximal metaphysis more common than diaphysis in long bone

• Hands and feet involvement usually due to secondary malignant transformation in multiple enchondromatosis syndrome
Chondrosarcoma
Clinical Presentation

• Dull pain for months
• Pain may be worse at night
• Focal swelling
• If tumor close to joint, effusion and limited movement
• Pathologic fractures
• Average duration of symptoms prior to presentation is 1-2 years
Chondrosarcoma Imaging

• Grow with lobular type architecture
• Hyaline cartilage nodules with high water content (increase signal on T2 and STIR)
• Radiograph - peripheral enchondral ossification with “ring-and-arc” appearance
• Aggressive features include irregular calcifications, endosteal scalloping, and soft-tissue extension
• CT - detects matrix mineralization, guide biopsy, pulmonary metastasis
• MR - shows high water content as low attenuation and very high signal intensity with T2-weighting, soft tissue extension
Because our patient has juxtacortical chondrosarcoma, which accounts for only 4% of chondrosarcomas, let’s look at a companion patient with conventional intramedullary chondrosarcoma……
Companion Patient #1: Conventional Intramedullary Chondrosarcoma

Radiograph shows lobulated, “ring-and-arc” appearance of chondroid matrix mineralization
Companion Patient #1: Conventional Intramedullary Chondrosarcoma

CT demonstrates low-attenuation soft-tissue mass and chondroid matrix calcification with cortical destruction.
Companion Patient #2: Atypical Location
Chondrosarcoma of the Posterior Rib: Radiograph

52-year-old nonsmoker presents with a month of cough and fatigue

5 cm soft tissue mass projecting posteriorly at the level of T7 and T8. This mass projects to the right of midline on the PA film. This mass does not have the typical “ring-and-arc” appearance.
Companion Patient #2: Atypical Location
Chondrosarcoma of the Posterior Rib: CT

- Mass with calcifications in the right lower hemithorax posteriorly involving the right posterior 8th rib
- Expansile, lytic components
Companion Patient #2: Atypical Location
Chondrosarcoma of the Posterior Rib: MRI

- Irregular areas of low signal on both T1 and T2-weighted images
- Mass enhances heterogeneously on post-contrast images
# Surgical Staging System for Musculoskeletal Tumors

<table>
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<tr>
<th>Stage</th>
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<td>3</td>
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<td>I B</td>
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<tr>
<td>II A</td>
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<tr>
<td>III</td>
<td>Low or high grade; Intra or extracompartmental</td>
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</tbody>
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Histologic Grade

• Low-grade = Grade 1 and 2: locally aggressive tumor with limited capacity to metastasize to distant organs

• High-grade = grade 3: high cellularity, marked cellular pleomorphism, high N/C ratio, frequent mitoses, significant metastatic potential
Treatment of Chondrosarcoma

1. Intralesional curettage, adjunct chemical or thermal ablation, and cementation or bone grafting of the defect

2. Wide excision with structural graft or metal reconstruction

3. Chondrosarcoma respond poorly to chemo and radiation therapy. Radiation therapy may be used in high grade tumors.
Prognosis

• Low-grade: Local recurrence = 9.5% with adequate initial resection
• High-grade: Local recurrence = 47%
• Metastasis: Lung, regional lymph nodes, liver
  10% - 50% for grade 2
  50% - 71% for grade 3
• Juxtacortical: Local recurrence and metastatic disease have been reported, but the prevalence is low, even in high-grade lesions
References

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