Osteosarcoma: Two Different Presentations

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Our Patient: Clinical History

- 25 year old male from El Salvador
- Currently presenting with
  - Pain in right thigh on walking and in daily activities
  - Restricted range of motion of the right knee
- Two month history of right thigh pain and swelling
- Associated with
  - Fever and night sweat
  - Night pain
  - General Malaise
  - 15 pound weight loss over past 2 months
- Not relieved by oxycodone and ibuprofen
Our Patient: Physical Findings

- Significantly increased size to his right thigh with a large palpable mass around the distal femur
  - Firm
  - Tender
  - Extends both medially and laterally
- Right thigh is warm compared to the contralateral side
- Small effusion in right knee
- Significant pain at the extremes of flexion and extension of right knee
- Motor power on right lower limb 5/5
- No popliteal or groin lymphadenopathy on the right
- Neurovascularly intact
Osteosarcoma:
Classical Clinical History

• Age from 5 to 30 years, most commonly between 10 to 20 years
• Dull, aching pain
  – For weeks or months
  – Can become more severe suddenly
• Night pain
• Swelling
• Rarely: Fever, night sweats, weight loss
Osteosarcoma:
Classical Physical Findings

• Local tenderness
• Swelling, mass, deformity
• Restricted range of movement
• Uncommonly: Pathologic fracture
• Rarely: Lymphadenopathy
Osteosarcoma: Menu of Tests

- Radiography
- Magnetic Resonance Imaging
- Computed Tomography
- Bone Scintigraphy
Osteosarcoma: Radiographic Findings

• Originate in the metaphyseal region of bone
• Commonly mixed sclerotic and lytic lesion, rarely purely sclerotic or lytic lesion
  – Sclerosis: Small, irregular, cloud-like densities due to osteoid formation
  – Lytic: ill-defined edges or multiple cortical holes due to destruction of bony cortex
• Aggressive periosteal reaction
  – Codman’s triangle
  – Multilaminated/ spiculated/ sunburst reactions
• Soft tissue extension and formation of soft tissue mass
  – Mineralized osteoid within soft tissue adjacent to bone
  – Extensions near joints may be mistaken for effusion
• Pathologic fractures can also be identified
• Useful in diagnosis of osteosarcoma, but often underestimates the extent of the tumor, both within and outside the bone
Our Patient:
Radiograph of Right Femur
At time of presentation

- Soft tissue mass
- Increased periosteal reaction
- Soft tissue swelling
- NO fracture
Our Patient: Radiograph of Right Femur 2 months after initial presentation

- Mineralized soft tissue osteoid
- Codman’s triangle
- Sun burst periosteal reaction
- Cortical destruction
Our Patient:
Radiograph of Right Femur
3 months after initial presentation

- Articular extension
- Sun burst periosteal reaction
- Mineralized soft tissue osteoid

AP radiograph
BIDMC PACS

Lateral radiograph
BIDMC PACS
Our Patient: Radiographic Differential Diagnoses

• Malignancy
  – Osteosarcoma
  – Ewing's sarcoma
  – Chondrosarcoma
  – Fibrosarcoma
  – Osteoblastoma
  – Giant cell tumor

• Infection
  – Osteomyelitis

• Others
  – Aneurysmal bone cyst
  – Fibrous dysplasia
Osteosarcoma: Magnetic Resonance Imaging

- Determine intraosseous and extraosseous extent of tumor
- Detect soft tissue mass
- Detect skip lesion
  - More likely to have distant metastasis and shorter periods of disease-free survival
- Accurate local staging of osteosarcoma
  - Assess involvement of muscle compartments
  - Avoid contamination of previously uninvolved compartments during biopsy
- Determine relationship of tumor to neurovascular bundle, to estimate resectability
- Modality of choice because of excellent bone marrow and soft tissue contrast and multiplanar capabilities
- Insensitive to small amounts of calcium - appear hypointense in all sequences
Osteosarcoma: MRI Findings

- Most accurate sequence for determining the longitudinal extent of disease is the T1-weighted sequence
- Intramedullary disease best shown by T1-weighted sequences
- Subtle extra-compartmental disease best shown by T2-weighted sequences
- Tumour appears
  - Low to intermediate signal on T1-weighted sequences
  - Hyperintense on T2-weighted sequences
Our Patient: MRI Right Femur
At time of presentation

- Large heterogeneous mass enhancing heterogenously
- Extensive soft tissue component
- Central areas of low signal intensity – necrosis
- Peripheral areas of high/intermediate signal intensity - intratumoral hemorrhage
- Displaces femoral neurovascular bundles and sciatic nerve
Our Patient: MRI Right Femur
At time of presentation

- High signal intensity extends from level of tensor fascia lata to gastrocnemius
Our Patient: MRI Right Femur
2 months after initial presentation

- Large non-enhancing areas consistent with tumor
- Diffuse mottling of the femoral diaphyseal cortex
- Cortical breakthrough in femoral condyles with intra-articular extension
Our Patient: MRI Right Femur
2 months after initial presentation

- Increased intramuscular edema
- Increase in size and number of locules with fluid-fluid levels – hemorrhage
- Moderate joint effusion
- NO pathological fracture
Osteosarcoma: Bone Scintigraphy

• Osteosarcomas typically show increased uptake of radioisotope technetium-99

• Bone scans can help determine
  – Polyostotic multifocal involvement
  – Intraosseous extent of tumor; skip lesions
  – Metastatic disease
    • Bone
    • Lung

• Entire axial and appendicular skeleton is imaged
Our Patient: Bone Scintigraphy
At time of presentation

- Marked uptake in right distal femur
- Mild proximal tibial uptake – reactive changes
- Uptake in central chest – lung metastases
Osteosarcoma: Chest Radiograph

- Detect pulmonary metastasis
Our Patient: Chest Radiograph
At time of presentation

- Retro-sternal opacity
Osteosarcoma: Computed Tomography

• Less sensitive than MRI in local evaluation of the tumor
• CT may be helpful locally when extensive necrosis and surrounding edema are present
• Most accurate modality for staging of pulmonary metastases
• Used in post-surgical surveillance
Our Patient: CT Thorax
At time of presentation

- Multiple calcified and non-calcified pulmonary nodules
- Largest nodule partially calcified, measuring 2 x 2 cm
Our Patient: CT Thorax
At time of presentation

- Multiple calcified and non-calcified pulmonary nodules
- Largest nodule partially calcified, measuring 2 x 2 cm
Osteosarcoma: Epidemiology

- Most common bone sarcoma/ primary malignant tumor of bone, excluding plasma cell myeloma
- Third most common malignancy in children and adolescents
- About 900 new cases diagnosed in the United States every year
  - About 400 of these are in patients younger than 20 years old
  - About 80 percent of these tumors are non-metastatic at time of presentation
- Peak incidence in persons aged 10-20 years
- Male to female ratio is 3:2
Osteosarcoma: Pathology

• Occurs most commonly in the metaphyses of long tubular bones
  – Distal femur
  – Proximal tibia
  – Proximal humerus
  – Any other bone

• Commonly extends from the metaphysis into the adjacent diaphysis or epiphysis, with uncommon intra-articular extension

• Systemic metastasis
  – Lungs
  – Bone
Osteosarcoma: Staging

- Two systems
  - Enneking System
  - ACJC System
Osteosarcoma: Staging Enneking System

• Stage 1: Low-grade tumor
  – 1A: Intra-compartmental
  – 1B: Extra-compartmental

• Stage 2: High-grade tumor
  – 2A: Intra-compartmental
  – 2B: Extra-compartmental

• Stage 3: Either grade with metastase
Osteosarcoma: Staging
ACJC

- Stage 1: G1 – G2
  - 1A: T1, N0, M0
  - 1B: T2 – T3, N0, M0

- Stage 2: G3 – G4
  - 2A: T1, N0, M0
  - 2B: T2, N0, M0

- Stage 3: T3, N0, M0, G3 – G4

- Stage 4: any T, any G
  - 4A: N0, M1a
  - 4B: N1, any M OR any N, M1b
Osteosarcoma: Treatment & Prognosis

- Primary treatment is surgical
  - Limb salvage is undertaken whenever possible
- Pre-surgical chemotherapy is vital in the treatment
- Improve limb salvage and survival rates
- Reduce number of pulmonary metastases or delay their appearance, aiding surgical removal
- Preoperative radiologic or biopsy assessment of the response of the tumor to chemotherapy allows an estimate of prognosis
- 5-year survival rate for patients with localized osteosarcoma is between 60% to 80%
- 5-year survival rate for patients with metastatic disease is about 15% to 30%
  - Survival rate is closer to 40% if the cancer has spread only to the lungs or if all of the tumors (including metastases) can be removed surgically
Our Patient: Outcome

• Treated with pre-surgical chemotherapy
• Underwent radical resection of osteosarcoma of the right femur and proximal tibia
• Final pathology revealed high-grade osteosarcoma with extra-compartmental involvement is
• Tumor showed approximately 20% necrosis secondary to treatment effect
• Progression of metastatic disease with innumerable pulmonary nodules and masses increasing in both size and number
• Local recurrence of disease
• Expired within 6 months of initial presentation
Now that we have seen a patient with classical presentation of osteosarcoma, let’s see a patient with an unusual presentation.
Companion Patient: Clinical Presentation

• 12 year old female
• Unsteady gait for past two months
• Progressive bilateral lower limb weakness
• Numbness in bilateral lower limbs
• Acute bowel incontinence
Companion Patient: Radiograph of Spine

- Predominantly osteoblastic lesion
Companion Patient: MRI of Spine

- Low signal intensity on T1-weighted sequence
- Low signal intensity on T2-weighted sequence
- Abnormally enhancing (low T1, high T2) soft tissue compresses cord
Companion Patient: Bone Scintigraphy

- Uptake in thoracic spine
Osteosarcoma of the Spine: Statistics

- 3.6% to 14.5% of primary spinal tumours
- 1.7% to 4% of all osteosarcomas
- Most commonly involved spinal regions:
  - Lumbar (32.3%), sacral (20.7%), thoracic (33.3%), cervical (13.6%)
- Majority of tumours involve the body alone (21%) or the body and posterior elements (79%)
- Involvement of 2 levels in 17%
- Spinal canal invasion in 84%
- Metastases to the lungs, pleura and other bones, liver and abdominal viscera
2 Different Presentations of Osteosarcoma: A Comparison

• Different presentation
  – Pain & swelling versus cord symptoms
• Different location of lesion
  – Distal femur versus thoracic spine
• Different radiographic findings
• Similar age group (10 – 30 years old)
Summary

• Osteosarcoma is a common malignancy
  – in the young
  – of the bone

• Although it has a classical presentation, it can also present atypically through the involvement of other bony sites

• A healthy index of suspicion would be helpful

• Early prompt treatment may improve outcome
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References