Imaging of Ewing’s Sarcoma

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6 yo female presents with right clavicular mass and pain

- Area of mottled sclerosis and lucency in the distal half of the right clavicle, slightly expansive
- Lesion borders are ill-defined with associated periosteal reaction and cortical thinning
- Associated soft tissue mass, normal contour lost
- Lesion concerning for malignancy—MRI recommended
MRI

Suspicion of malignancy

-provides information on marrow and soft tissue involvement—spread through medullary better seen than on plain radiographs; can detect presence of skip lesions in bone

-must be obtained before biopsy because postoperative changes can confuse true extent of disease
Sagittal and axial T2 with fat suppression sequences show a soft tissue mass surrounding the right clavicle with associated edema.

There is communication of the soft tissue mass with the medullary cavity.

The soft tissue mass showed enhancement with gadolinium.
Differential Diagnosis

• Important things to consider when evaluating bony lesions:
  - age of patient
  - location, size
  - cortical destruction
  - associated soft tissue mass
Differential Diagnosis cont’d

- Osteomyelitis
- Osteosarcoma
- Ewing’s sarcoma
- Lymphoma
- Leukemia
- Metastatic neuroblastoma
- Langerhans cell histiocytosis

May have patterns of sclerosis

Can all have “moth-eaten” pattern and surrounding edema

Can have similar lytic pattern

PEAK AGES:
- 0-5: LCH, neuroblastoma, leukemia
- 5-10: Osteosarcoma
- 10-20: Ewing’s sarcoma
Biopsy (with CT guidance)
Our patient’s results showed
Ewing’s sarcoma

small round blue cells
Ewing’s Sarcoma

• Described in 1921 by Dr. James Ewing

• 2nd most common bone tumor in children

• Usually occurs in 2nd decade of life; rarely occurs after age 30

• Whites affected much more than other races

• Found mostly in flat and long bones (diaphysis)
Clinical Presentation

• Pain--usually intermittent at first, but can progress to intense pain

• Can present like osteomyelitis: Fever, anemia, leukocytosis, increased ESR or LDH

• Eventually a large mass may be palpable

• Less commonly, can present with pathological fracture
Plain Film: Typical Findings

- Ill-defined, destructive margins
- "moth-eaten" appearance (purple arrow)
- Overlying soft tissue mass
- Expanded cortex with displacement of periosteum (Codman’s triangle)
- "onion peel" appearance due to periosteal reaction (orange arrows)
MRI: Typical Findings

- 80% to 90% have soft tissue mass—best seen on T2-weighted/T1-weighted C+; heterogeneous contrast-enhancement

- Coronal or sagittal T1-weighted images can demonstrate intramedullary extent (arrows)
Now that we have seen a typical presentation of Ewing’s sarcoma, let’s review an atypical presentation.
Atypical Presentation: Pt #2

15 yo male, initially presented with fevers and hip pain.

Plain film of pelvis appears normal overall.
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Pt #2: Plain film

- Initial diagnosis of osteomyelitis made based on clinical presentation and findings on plain films and MRI

- Debridement, antibiotics—pt still had pain

- Plain film of right hip s/p debridement shows heterotopic bone along the right ilium--likely related to the debridement

- Another MRI done—findings consistent with osteomyelitis; a biopsy was non-specific
Pt #2: MRI #4

Another biopsy done after repeated failure to respond to antibiotics—Ewing’s sarcoma diagnosed.
Our Patient: Metastatic Workup

- Need to assess lung (most common site of metastases) with chest CT

- Our patient’s chest CT showed no evidence of metastatic disease; however, can visualize cortical destruction of clavicle (yellow arrow)
Metastatic Workup cont’d

Bone scintigraphy:

Whole body scan using Tc 99m-MDP

- technetium-99m (radioactive) is linked to methylene-diphosphonate (MDP) which is taken up by bone

- ‘hot spot’ occurs where tracer accumulates; denotes areas of ↑ physiological function (fracture, tumor)
Our Patient: Bone Scan

Area of uptake in right clavicle; no evidence of metastatic osseous disease
Treatment

• Chemotherapy
  - reduces local tumor volume
  - believed that majority of cases have subclinical metastatic disease at time of presentation

• Surgical resection of tumor

• Adjuvant radiation therapy if needed

• ~80% of limbs can be salvaged
Prognosis

• Unfavorable:

- presence of metastases (30% survival w/isolated lung mets, <20% w/bone mets)

- large size of primary tumor (>200ml)

- axial location vs. extremity

- male sex, age >12, anemia, ↑LDH, radiation therapy only for local control, poor chemo course
Treatment Evaluation

• MRI
  - necrotic intraosseous lesion with increased signal on T2
  - can have well-defined margin
  - however, changes in signal can reflect changes in bone marrow structure or nonspecific fibrosis → can make detecting residual tumor difficult

• Bone scan
PET

- Positron emission tomography (PET) with fluorodeoxyglucose (FDG)
  - most sensitive to detect changes in tumor metabolism following treatment
  - glucose analog is taken up and retained by tissues with high metabolic activity (brain, liver, most malignant tumors)
    (also a possible role for metastatic workup)

http://en.wikipedia.org/wiki/FDG-PET
Our Patient: Bone Scan, post-treatment

S/P chemo and resection; no evidence of uptake in previous area of neoplasm or osseus metastases
Imaging Algorithm

- Plain films (at least 2 planes)
- MRI for better characterization of extent and involvement
- Biopsy (CT guidance or open) for definitive diagnosis
- Chest CT and bone scan for evaluation of metastases
- MRI, bone scan, PET-FDG for treatment assessment
References


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