AMSER Case of the Month:
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25 y/o male with gradual onset of right shoulder swelling and discomfort

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Patient Presentation

• **HPI:** 25 y/o male who presented with 3 month history of gradual swelling of the right humerus and mild right shoulder discomfort. Patient denies a history of trauma

• **PMHx + PSHx:** Nothing relevant

• **Social:** Drinks alcohol. Denies smoking or recreational drug use

• **Physical Exam:** Tense skin over right upper arm with increased arm circumference compared to left. Full range of motion of both upper limbs with no neurologic deficits

• **Relevant labs:** Alk Phos – ↑1595 U/L (38 – 126 U/L)

What is the first imaging we should order?
### ACR Appropriateness Criteria

#### Variant 1: Atraumatic shoulder pain. Initial imaging.

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Appropriateness Category</th>
<th>Relative Radiation Level</th>
</tr>
</thead>
<tbody>
<tr>
<td>Radiography shoulder</td>
<td>Usually Appropriate</td>
<td></td>
</tr>
<tr>
<td>CT arthrography shoulder</td>
<td>Usually Not Appropriate</td>
<td>☂ ☂ ☂ ☂</td>
</tr>
<tr>
<td>CT shoulder with IV contrast</td>
<td>Usually Not Appropriate</td>
<td>☂ ☂ ☂</td>
</tr>
<tr>
<td>CT shoulder without and with IV contrast</td>
<td>Usually Not Appropriate</td>
<td>☂ ☂ ☂</td>
</tr>
<tr>
<td>CT shoulder without IV contrast</td>
<td>Usually Not Appropriate</td>
<td>☂ ☂ ☂</td>
</tr>
<tr>
<td>MR arthrography shoulder</td>
<td>Usually Not Appropriate</td>
<td>O</td>
</tr>
<tr>
<td>MRI shoulder without and with IV contrast</td>
<td>Usually Not Appropriate</td>
<td>O</td>
</tr>
<tr>
<td>MRI shoulder without IV contrast</td>
<td>Usually Not Appropriate</td>
<td>O</td>
</tr>
<tr>
<td>US shoulder</td>
<td>Usually Not Appropriate</td>
<td>O</td>
</tr>
<tr>
<td>X-ray arthrography shoulder</td>
<td>Usually Not Appropriate</td>
<td>☃</td>
</tr>
</tbody>
</table>

This imaging modality was ordered by the physician.
X-ray Findings

- Sunburst periosteal reaction with soft tissue mineralization
- Permeative process “Moth-eaten appearance”
- Wide zone of transition
- Medullary and cortical bone destruction
- X-ray Findings

Sunburst periosteal reaction with soft tissue mineralization
Differential Diagnosis

1. Malignant lesions
   - Osteosarcoma
   - Ewing sarcoma
   - Bone metastases from a separate primary malignancy

2. Benign lesions
   - Osteochondroma
   - Osteoid osteoma

3. Non-neoplastic processes
   - Osteomyelitis
   - Aneurysmal bone cyst
### Additional Imaging - ACR Appropriateness Criteria

**Variant 8:** Lesion on radiographs. Aggressive, suspicious for malignancy.

<table>
<thead>
<tr>
<th>Radiologic Procedure</th>
<th>Rating</th>
<th>Comments</th>
<th>RRL*</th>
</tr>
</thead>
<tbody>
<tr>
<td>MRI area of interest without and with IV contrast</td>
<td>9</td>
<td></td>
<td>O</td>
</tr>
<tr>
<td>MRI area of interest without IV contrast</td>
<td>8</td>
<td>This procedure is especially useful for areas with complex osseous anatomy.</td>
<td>O</td>
</tr>
<tr>
<td>CT area of interest without IV contrast</td>
<td>7</td>
<td>Perform this procedure if there is concern that the lesion represents multiple myeloma.</td>
<td>Varies</td>
</tr>
<tr>
<td>Te-99m bone scan whole body</td>
<td>6</td>
<td>This procedure is particularly helpful to look for multifocal disease.</td>
<td>⭐⭐⭐⭐</td>
</tr>
<tr>
<td>X-ray skeletal survey</td>
<td>5</td>
<td></td>
<td>⭐⭐⭐⭐</td>
</tr>
<tr>
<td>CT area of interest without and with IV contrast</td>
<td>5</td>
<td>Perform this procedure if MRI is contraindicated.</td>
<td>Varies</td>
</tr>
<tr>
<td>FDG-PET/CT whole body</td>
<td>5</td>
<td>This procedure is particularly helpful to look for multifocal disease.</td>
<td>⭐⭐⭐⭐⭐</td>
</tr>
<tr>
<td>CT area of interest with IV contrast</td>
<td>2</td>
<td></td>
<td>Varies</td>
</tr>
<tr>
<td>US area of interest</td>
<td>1</td>
<td></td>
<td>O</td>
</tr>
</tbody>
</table>

**Rating Scale:** 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate

*Relative Radiation Level*
Extensive lesion involving bone and soft tissue

Relative signal decrease due to medullary bone involvement
Involvement of cortical and medullary bone

Lesion measured to be 15 x 10 x 10 cm (H x W x L)

Heterogenous appearance due to overlapping areas of mineralization and soft tissue involvement
Technetium-99 MDP Bone Scan

- Intense radiotracer uptake in irregular pattern in right upper arm
- Normal radiotracer uptake in bladder
- Young patient with no uptake to suggest degenerative change

Final Diagnosis (after CT-guided biopsy)

*High grade osteosarcoma*
Case Discussion - Osteosarcoma

- 2nd most common form of primary bone tumour (20%) after multiple myeloma (30%)

- Primary form affects younger patients, often the metaphysis of long bones due to increased activity
  - Common sites = Femur (40%), tibia (16%), humerus (15%)

- Secondary form occurs in older patients and is more common in flat bones like the pelvis
  - Often sequelae of Paget’s disease or previous primary bone lesions treated with radiation

- Macrometastases demonstratable in 10-20% of cases at time of presentation with lungs being most common site

- Patients without overt metastases are presumed to have occult micrometastatic disease and are managed as such
Case Discussion - Management

• Slight variations based on subtype and staging but in general
  = CHEMOTHERAPY + SURGERY

• Neoadjuvant chemotherapy ➔ Re-staging ➔ Surgical resection ➔ Adjuvant chemotherapy
  - Adjuvant chemotherapy regimen will be decided based on response to neoadjuvant treatment

• Range of chemotherapy options
  - This patient ➔ Cisplatin, doxorubicin and MTX (with leucovorin rescue)
  - Consider pre-chemotherapy fertility consultation for appropriate patients

• Prognosis
  - 70% long-term survival but <20% in patients with a large metastatic burden
  - 35-40% long-term survival rate if limited pulmonary metastases
1. UpToDate – Osteosarcoma: Epidemiology, pathogenesis, clinical presentation, diagnosis and histology
   - Accessed July 2019

