AMSER Case of the Month

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56-year-old male presents with acute lower right leg discomfort

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Patient Presentation

<u>HPI</u>: 56-year-old male, with a history significant for prior right leg fracture treated with ORIF, presents with acute lower right leg discomfort near a prior external fixator incision, with no new trauma.

PMH: Cardiomyopathy, hypertension, and type-2 diabetes mellitus

<u>Medications:</u> Empagliflozin (JARDIANCE), Metformin (FORTAMET), Valsartan-Sacubitril (ENTRESTO), Carvedilol (COREG), Digoxin (LANOXIN)

Allergies: None

Vitals: BP: 121/72 mmHg, HR: 106, RR: 20, SpO2%: 99%, Temp: 97.7°F

<u>Physical Exam:</u> Asymmetric enlargement and swelling of the right lower leg. Right lower leg tenderness to palpation.



Patient Presentation

Pertinent Laboratory Findings

Complete Blood Count (CBC) with Differential

Normal RBC (5.42 x 10⁶/uL)

Normal WBC (8.76 x 103/uL)

Normal Hemoglobin (16.6 g/dL)

Normal Platelets (167 x 10³/uL)

↓ MCH (30.6 pg)

↑ MPV (13.0 fL)

Basic Metabolic Panel (BMP)

Normal Sodium (137 mmol/L)

Normal Potassium (4.8 mmol/L)

↑ Glucose (165 mg/dL)

↓ CO2 (21 mmol/L)

C-Reactive Protein

↑ CRP (0.94 mg/dL)

Alkaline Phosphatase

↑ 201 U/L



What Imaging Should We Order?



ACR Appropriateness Criteria

Revised 2024

American College of Radiology ACR Appropriateness Criteria® Stress (Fatigue-Insufficiency) Fracture Including Sacrum Excluding Other Vertebrae

<u>Variant 1:</u> Adult. Suspect stress fracture, excluding vertebrae. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
Radiography area of interest	Usually Appropriate	Varies
Bone scan whole body with SPECT or SPECT/CT area of interest	May Be Appropriate (Disagreement)	222
US area of interest	Usually Not Appropriate	0
MRI area of interest without and with IV contrast	Usually Not Appropriate	0
MRI area of interest without IV contrast	Usually Not Appropriate	0
CT area of interest with IV contrast	Usually Not Appropriate	Varies
CT area of interest without and with IV contrast	Usually Not Appropriate	Varies
CT area of interest without IV contrast	Usually Not Appropriate	Varies



Findings (unlabeled)







Findings (labeled)

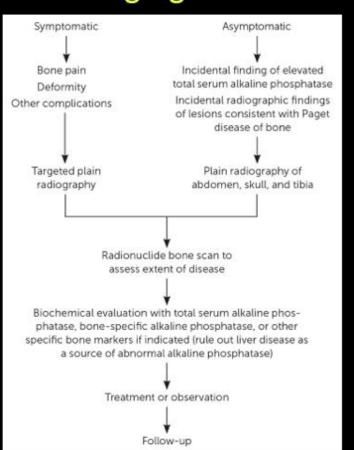
Cortical and trabecular thickening within the tibia. Transverse insufficiency fracture of the distal tibial diaphysis with surrounding callous formation. An anteriorly displaced fibular fracture traverses the proximal diaphysis.





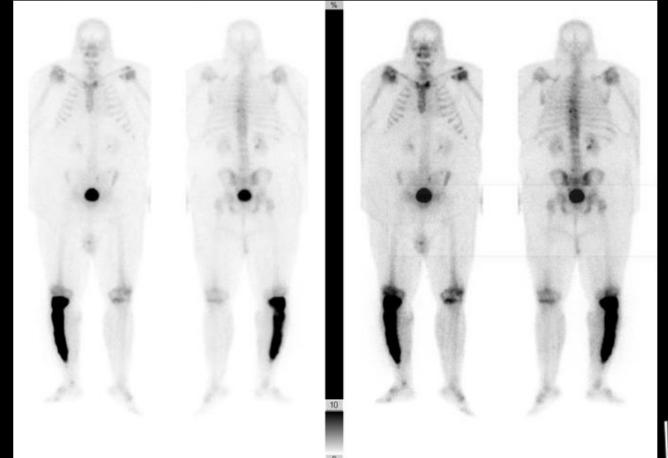


What imaging to order next?





Findings (unlabeled)

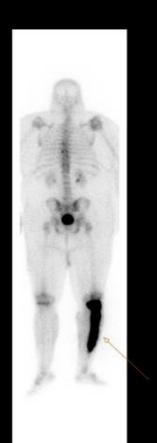




Findings (labeled)

Anterior and posterior bone scintigraphy images show avid radiotracer uptake within the right tibia. Radiotracer uptake within the bilateral shoulders, sternoclavicular joints, knees and spine is consistent with degenerative joint disease.







Differential Diagnosis

- 1. Paget's Disease
- 2. Fibrous dysplasia
- 3. Osteoporosis
- 4. Osteomalacia
- 5. Underlying bone malignancy



Final Diagnosis:

Paget's Disease



History of Paget's Disease:

- Paget's Disease was first detailed by Sir James Paget in a manuscript in 1877²
- He described it as osteitis deformans, suspecting an underlying inflammatory etiology^{2,3}
- Paget's Disease is a chronic, progressive skeletal growth disorder featuring abnormal bone growth^{4,5}
 - This can lead to skeletal deformities and pain



History:

Paget's Disease refers to a skeletal
 growth disorder due to excessive
 bone resorption followed by abnormal
 bone formation^{4,6}

Clinical Presentation:

Most patients present as asymptomatic.

However, a multitude of features can develop:

- Bone pain (most common)
- Diffuse joint stiffness

- Back, hip, or neck pain
- Fractures
- · Abnormally enlarged skull
- Bowed limbs or scoliosis
- Deafness
- Migraines
- Cranial nerve palsies
- Congestive heart failure
- Osteosarcoma (rare complication)



Epidemiology:

- Second most common metabolic bone disorder in older patients after osteoporosis
- Worldwide prevalence of 1.5% to 8.3%, affecting 1-3 million individuals in the US⁴
- Most commonly seen in Caucasians of Northern European origin⁴
- More commonly seen in individuals 50 years of age and older, with males and females affected equally
- Most commonly involves the pelvis, skull, spine, femur, and tibia⁸
- 1% have osteosarcomatous transformation.



Pathophysiology:

- Unknown cause but suspected interplay of genetic and environmental factors⁸
- Occurs in 3 stages
 - Lytic stage, featuring localized excess of osteoclastic activity leading to an initial lesion^{3,7}
 - Hybrid lytic/blastic stage, featuring increased abnormal osteoblastic bone formation with concurrent osteoclastic resorption
 - Sclerotic stage, featuring osteoblastic woven bone formation with declining osteoblastic and osteoclastic activity, with little bone formation or resorption present

Etiology:

- Risk factors include⁸:
 - Age (uncommon under 40)
 - Family History
 - Anglo-Saxon descent



Diagnostic Imaging:

- Radiography
 - Initial diagnosis
 - More specific than radionucleotide bone scan in making a diagnosis⁷
- Radionuclide bone scan (scintigraphy)
 - Most sensitive test in evaluation of extent of lesions⁷
 - During the osteoclastic lytic phase of disease, may underestimate disease activity⁷
- CT
 - Evaluate regions with articular abnormalities, and neurologic complications⁷
 - Better visualization of bone and posterior fossa⁷
- MRI
 - Evaluate regions with articular abnormalities and neurologic complications⁷
 - Better detailing of the brain, spinal cord, cauda equina, and soft tissues⁷
 - Pagetic sarcomas better evaluated⁷



Treatment:

- Treatment options for Paget's Disease consist of the following:
 - Bisphosphonates
 - First-line treatment⁴
 - Calcitonin
 - Second-line treatment, for patients unable to take or tolerate bisphosphonate therapy⁵
 - Denosumab
 - In the case of bisphosphonate contraindication or intolerance⁴
 - Calcium and Vitamin D supplements
 - NSAIDS for secondary osteoarthirite pain⁷



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