Radiologic Manifestations of Bone Disease in Sickle Cell Anemia

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Presentation Outline

• Meet patient #1
• MSK imaging modalities
• Brief review of Sickle cell anemia and its clinical manifestations
• Radiologic imaging of the bone complications of SCA
Meet Patient #1

- 44-year old African-American male with hx of SCA and associated chronic pain syndrome
- Presents with complaint of left knee pain
- Reports increased pain while walking and occasionally at rest
- Has had to “dip” into MSContin for additional analgesia
Bone Imaging modalities

• Plain radiograph:
  – initial radiologic evaluation of the majority of suspected musculoskeletal abnormalities
  – Relatively specific in differentiating potential etiologies of lesion b/c of proven ability to characterize features of the lesion
  – May not detect certain processes until they are far progressed
Bone imaging modalities

- **CT**
  - Can detect lesions not seen on routine radiographs or when plain film evaluation is difficult due to nature of lesion or anatomic site
  - best technique in assessment of matrix mineralization, cortical detail, and detection of the cystic and fatty lesions
  - Can identify subtle bony abnormalities (occult fractures) and their relationship to adjacent organs
  - May fail to demonstrate early vascular and marrow abnormalities
Bone imaging modalities

- **MRI**
  - Method of choice for imaging the soft tissues of MSK system
  - Sensitive to changes in bone marrow (i.e. subtle marrow edema, marrow infarcts)
  - Can detect and stage primary bone tumors and occult bony metastases
  - Difficulty visualizing fine bone detail or small calcifications
Bone imaging modalities

• Bone radionuclide scan
  – Sensitive, but nonspecific for measuring bone formation and perfusion
  – Detects lesions not otherwise seen on radiograph
  – Effective for imaging bone marrow and identifying areas of red marrow replacement
  – Effective screen for detecting multiple lesions
  – Lacks sufficient spatial resolution to detail extent and anatomic involvement of disease
Bone imaging modalities

• Ultrasound
  – High frequency transducers allow markedly improved image resolution
  – Can detect joint effusions, subperiosteal fluid, soft tissue masses, congenital hip dysplasias, etc.
  – Ineffective in visualizing bone detail
Pt #1: AP films of left knee

Ill-defined sclerotic densities
Pt #1: Lateral oblique of knee

Ill-defined sclerotic densities
Differential dx for multiple sclerotic densities

- Bone infarct
- Chronic osteomyelitis
- Osteoblastic metastases
- Enchondromatosis
- Bone islands
Pt #1: Left knee MR – T2 sagittal

heterogeneous bone marrow
Pt #1: Left tibia MR – T2 axial

- Diffusely heterogeneous bone marrow
- Sclerotic bone
Pt #1: Left knee MR - coronal

- Sclerotic bone
- Heterogeneous bone marrow

BIDMC PACS
Differential dx of bone infarct*

- Idiopathic
- Occlusive vascular disease
- Sickle cell anemia
- Fat embolism
- Infection, osteomyelitis

*diff dx for diametaphyseal ischemia
Quick Review of SCA

- Most common hemoglobin variant in the world (HbS)
- Autosomal recessive
- ~8% of African Americans are carriers
- High frequency of HbS gene attributed to “balanced polymorphism”
- Clinical severity can vary among those afflicted with SCA
Pathogenesis of SCA

- Mutation endows deoxy HbS new ability to polymerize distorting the RBC contours

= HbS

Nucleation → Growth → Alignment
Pathophysiology of SCA

• In addition to polymerization,
  – Alteration in RBC membrane and cytoskeleton
  – Increased RBC adherence to vascular endothelium
  – Disordered RBC volume control
  – The above factors act in concert to contribute to the vasooclusive episodes and hemolysis seen in SCA

http://carnegieinstitution.org/first_light_case/horn/lessons/sickle.html
Clinical manifestations of SCA

- Symptoms typically begin after 6 mo. of age
- Heterogeneity in clinical and hematologic severity
- Hematologic:
  - chronic anemia
  - acute severe anemia (splenic sequestration, aplastic crisis, hyperhemolytic crisis)
  - acute vassocclusive episodes
- Gastrointestinal: pigmented gallstones
- Neurologic:
  - Transient ischemic attacks
  - infarctive stroke
  - intracerebral hemorrhage
  - retinopathy
Clinical Manifestations of SCA

• Cardiac: acute myocardial infarction and high output failure

• Pulmonary: acute chest syndrome (incl. pneumonia and infarction due to in situ thrombosis)

• Renal:
  – papillary necrosis
  – focal glomerulosclerosis

• Infectious: bacteremia, osteomyelitis, meningitis
Bone complications in SCA

• Among the most common manifestations of SCA

• Due to one or both of the following factors:
  – Bone marrow hyperplasia secondary to chronic hemolysis
  – Vaso-occlusion leading to ischemia and infarction
Marrow hyperplasia

- widespread expansion of hematopoietic bone marrow throughout the body

- Commonly seen in chronic hemolytic anemias

- fatty marrow

- hematopoietic red marrow
Skull – marrow hyperplasia


Skull – marrow hyperplasia

- Occurs secondary to thinning of the outer table
- Not frequently seen in SCA, but more common in β-thalassemia

Spinal complications of SCA

- Vertebral sclerosis due to many small infarcts beginning early in life
- Central cupping of the end-plate (aka H-shaped vertebrae)
- Vertebral collapse
H-shaped vertebrae
Vertebral blood supply

Hands and Feet

• Dactylitis also known as hand-foot syndrome

• Most often occurs between 6 and 18 months of age

• Usually the first ischemic manifestation of SCA

• May involve one bone or multiple bones

Dactylitis

- soft tissue swelling of index finger
- periostitis along 3rd through 5th metacarpals

Diff dx includes:
- Pyogenic osteomyelitis (esp. Salmonella)
- Tuberculosis

Generalized Osteosclerosis

- secondary to multiple widespread tiny infarcts
- differential dx includes osteoblastic mets
Long bone circulation

- The nutrient artery supplies the medullary cavity and inner cortex

- Multiple periosteal vessels supply the outer cortex

- The metaphyseal and epiphyseal regions are well-perfused by their own network of vessels

Shoulder: bone infarct

- metadiaphyseal
- medullary infarct
Lower extremity infarcts-bone scan

- Targets bone with Tc-99m methylene diphosphonate

- Immediately after infarction, scan demonstrates decreased uptake

- With reactive bone formation, there is increased uptake

- May be difficult to distinguish infarct from osteomyelitis

*BrighamRADS website*
Pt #1 revisited: Plain film of hips - AP

sclerosis of femoral head

sclerosis of femoral head

BIDMC PACS
Companion pt: coronal Hip MR

low signal intensity in femoral head

flattened femoral head

BIDMC PACS
Femoral head anatomy

- vascular supply to femoral head is an end-organ system with poor development of collaterals
- femoral head more vulnerable to episodes of vaso-occlusion

http://home.pacific.net.au/~rossjones/avn.htm
Complications of bone infarction

• Fractures
  – Resulting from full thickness cortical necrosis

• Osteomyelitis
  – Necrotic bone marrow fertile site for 2º infection
  – Most common offender is Salmonella

• Growth disturbances
  – Usually secondary to deficiency in the nutrient artery circulation to the central region of the metaphyseal side of the growth plate
Radiographic progression of osteomyelitis

Initial radiograph

7 days later

5 weeks later

Summary

• Bone abnormalities in SCA result secondary to marrow hyperplasia and episodes of ischemia leading to infarction

• Multiple modalities can be used in conjunction with one another to image these bony changes

• Notably, MRI is most sensitive to the detection of bone marrow changes which are commonly seen in SCA
References

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