Generalized Musculoskeletal Disorders

Thomas Pope, MD, FACR
Vice Chairman
Radisphere National Radiology Group
Beachwood, Ohio and Westport, CT

Formerly: Professor of Radiology (Med Univ of SC, Wake Forest University, University of Virginia)
ARRS AFIP Fellow
Visiting Scientist, AFIP

Email: thomas.pope@radispheregroup.com
Lecture origin and analogies

• In 1997, Dr. Murphey asked me to cover “group of entities not well covered in the course”
• Analogies:
  – Sixth man lecture - NBA
  – Sixth woman lecture - WNBA
  – “Clean up” - Baseball
  – “Catfish” - Creek or river
  – “Bottom fish” - Salt water (ocean)
  – “Opossum” - Woods

“The ocean is the place where I rebalance my life and remember what is important!”
Learning objectives

- Describe entities not covered in course at that time
- Outline the imaging features of these diseases
- Introduce these entities so you can study them further if you desire ("lifelong learning")
- In interest of time, will skip some of word slides that are in the syllabus...study these at your leisure!
Topics

- Osteoporosis
- Osteogenesis imperfecta
- Neurofibromatosis
- Collagen vascular-like diseases
  - SLE
  - Scleroderma
  - Polymyositis/dermatomyositis
Terminology

• **Osteopenia** – “paucity of bone”
  – “Decreased calcification or density of bone; a descriptive term applicable to all skeletal systems in which such a condition is noted; CARRIES NO IMPLICATION ABOUT CAUSALITY”

• **Osteoporosis**
  – “Abnormal loss of bone tissue resulting in fragile porous bones attributable to a lack of calcium, most common in post-menopausal females”
  – Decreased bone mineral density
  – Normal in quality
  – Decreased in quantity

• **REMEMBER:**
  – 30-50% of cancellous bone must be missing to recognize on radiography
Types of osteopenia

- Localized
- Regional or segmental
- Generalized or diffuse
Localized osteopenia “test”

Brodie’s Abscess

Gout

Osteoarthrosis
Regional osteopenia

- Segmental area of decreased BMD
- Differential diagnosis:
  - Disuse (immobilization)
  - Chronic regional pain syndrome (CRPS) (RSD-Reflex sympathetic dystrophy)
  - Transient osteoporosis (bone marrow edema)
  - Regional migratory osteoporosis
Disuse/Immobilization osteopenia

- **Major causes**
  - Immobilization for traumatic injury
  - Motor paralysis
  - Inflammatory lesions of bones and joints
- **Changes apparent in 7-10 days**
- **Maximam at 2-3months**
- **Patterns: uniform, spotty, bands, cortical lamination or scalloping**
- **May appear very aggressive!!!**
Disuse osteopenia

76 yo s/p left hemispheric CVA
Reflex sympathetic dystrophy
(Chronic regional pain syndrome)

- Elderly
- Trivial trauma
- Pain, swelling, temperature changes
Reflex sympathetic dystrophy
(Chronic regional pain syndrome)
Reflex sympathetic dystrophy
(Chronic regional pain syndrome)
Regional osteoporosis

- Rapidly developing, self-limited, reversible diseases
- No identifiable inciting event
- Major types:
  - Transient osteoporosis (bone marrow edema) of the hip
  - Regional migratory osteoporosis
Transient regional osteoporosis (bone marrow edema) of the hip

- Women in the 1/3 trimester of pregnancy
- Middle aged males
- LE > UE
- Osteoporosis and BME
- Generally spontaneously resolves in 9-12 months
- Differential diagnosis: AVN, infection

B. C. Vande Berg, J. J. Malghem, F. E. Lecouvet, J. Jamart, and B. E. Maldague
Idiopathic Bone Marrow Edema Lesions of the Femoral Head: Predictive Value of MR Imaging Findings
Radiology, August 1, 1999; 212(2): 527 - 535.
Transient osteoporosis (bone marrow edema) of the hip
Regional migratory osteoporosis
Generalized osteopenia/osteoporosis

- Diffuse decreased bone mineral density (BMD)
- Differential diagnosis/causes:
  - Senile osteoporosis (most common by far!)
  - Medications (Steroids, heparin)
  - Systemic diseases (Deficiency states)
    - Scurvy
    - Malnutrition
    - Calcium deficiency
Senile osteoporosis

- Most commonly encountered metabolic disease
- Reduction in bone “quantity”
- Normal in “quality”
- F>M, 4:1 (equal incidence > 80 yo)
- Most pain source: compression fx’s and kyphosis
- PE - kyphosis, shortened stature, and spinal rigidity
Epidemiological data

- **Surgeon General Report, October, 2004**
  - Half population in US > 50 yo
    - Low bone mass and risk for fracture
    - 34 million with hip osteopenia
    - 1.5 million/year osteoporosis-related fx
- **Risk of fracture in Caucasians > 50 yo**
  - Females 40%, Males 13%
- **Hip fracture:**
  - Risk of mortality within 3 months is 4X greater than normal
  - 20% of fx victims die or wind up in nursing home within year after event
- **Annual cost:** $18 BILLION
Pathology of osteoporosis

Osteoporotic rib

Deficient trabeculae

Normal rib

Normal Bone vs Osteoporotic Bone
Osteoporosis

Life cycle

Fracture index
Osteoporosis measurement

- Conventional X-ray (radiogrammetry)
- Single photon absorptiometry (SPA)
- Dual photon absorptiometry (DPA)
- Quantitative CT (QCT)
- Dual energy X-ray absorptiometry (DEXA)
- Neutron activation analysis
Dual energy X-ray absorptiometry (DEXA)

- Dual energy X-ray source
- Measures relative tissue attenuation
- Easily performed with reproducible technique
- Least coefficient of variation (COV)
- Detects changes of 1-3%
- Expressed in gm/cm²
- Primary indication: Estrogen deficiency to determine therapy
Hologic (Delphi)
BMD terms

- **BMD measured in gm/cm²**
- **T-score:** Patient’s BMD compared to normative data (Normal = 25 yo women)
- **Z-score:** Patient’s BMD compared to her aged-matched controls
- **World health organization (WHO) uses T scores to classify a patient’s bone mineral status**
WHO classification scheme
BMD

- **STANDARD**: Mean BMD of 25 yo women
- **NORMAL**: T-score from the mean to 1 standard deviation below the mean (mean to -1SD)
- **OSTEOPENIA**: T-score from 1 to 2.5 SD below mean (-1SD and -2.5 SD)
- **OSTEOPOROSIS**: T-score below 2.5 SD below mean (> -2.5 SD)
  - Osteoporosis also established by presence of a non-traumatic vertebral compression fracture
Why measure BMD?

• Fracture risk doubles:
  – For each drop of 0.1 below T score mean
  – For each decade the patient is over 50 yo

• Ultimate goal:
  – Be able to calculate an “absolute fracture risk”
  – More holistic and understandable information for patients
  – “You have a 70% of developing a vertebral fracture”
    than “Your T-score is -2, etc …”

• http://www.shef.ac.uk/FRAX/ (website to calculate fracture risk)
DEXA
Lumbar spine printout
“Ward’s triangle” Where compression and tensile trabeculae cross...most lucent area of the femoral neck.
DEXA Wrist Printout
Louisville oral boards story break

“Homerun”

View from my favorite restaurant
Vickery’s on Shem Creek
Mount Pleasant, SC
Senile osteoporosis

**IMAGING:**
- Increased radiolucency on X-ray ("osteopenia")
- Cortical thinning
- Altered trabecular patterns

**COMPLICATIONS:**
- Acute fractures
  - Spine (L>T>C)
  - Distal radius (Colles)
  - Proximal femur
  - Humerus (neck)
  - Ankle (malleoli)
- Insufficiency fractures
Complications of osteoporosis
Fall with hip pain
MR – non-displaced fracture
Spinal osteoporosis

- Decreased bone density
- Accentuation of primary trabeculae
- Cortical thinning
- Alterations in vertebral morphology:
  - Endplate deformities (Schmorl’s nodes, cortical irregularities)
  - Wedged vertebrae
  - “Biconcave” – “fish” shape
  - Vertebrae plana (“pancake”/silver dollar)
Senile vertebral osteoporosis
“Codfish” vertebral bodies

**Kindly sent to me by a former AFIP student Dr. Timothy O’Connell, Oak Brook, Ill**
Types of stress injury***

- **“Fatigue” fracture**
  - Abnormal muscular stress
  - Bone of normal elastic resistance

- **“Insufficiency” fracture**
  - Normal or physiologic activity
  - Bone deficient in mineral or elastic resistance
  - May be mistaken for metastatic disease

***Imaging findings similar, age groups different
Insufficiency fracture

- Not uncommon in elderly
- May have aggressive imaging features
- May be mistaken for “tumor” or “infection”
- Often history can help in suggesting diagnosis
  - Pelvis: Elderly (osteoporsis) and h/o ext beam radiation
- You, as radiologist, likely to be most knowledgeable about this entity in your hospital
Risk factors
Insufficiency fractures

- Osteoporosis
- Metabolic disease
- Hyperparathyroidism
- Osteomalacia/Rickets
- Cushing’s disease
- Paget disease
- Diabetes mellitus

Condensation of cancellous bone
Perpendicular to the long axis
High risk patients

72 yo s/p pelvic surgery and irradiation
Note lack of significant STM
56 yo lady with back pain
Bilateral sacral insufficiency fractures on RBS

Rheumatoid on steroids
Husband had stroke 6 weeks ago
MBR on second floor

“Honda sign”
MR imaging of fatigue fracture**
23 yo female runner

**Imaging findings almost identical to those of insufficiency fracture!
Osteogenesis imperfecta

- Common inheritable disorder
- Disturbance in Type I collagen synthesis
- Four distinct types
  - Type I (blue sclera) mildest form
- 1/30,000 affected
- 20-50,000 in US
Type II OI
Osteogenesis imperfecta
Type I

Osteopenia
Bowing deformities
Exuberant callus formation
Osteogenesis imperfecta
Type I
Osteogenesis imperfecta
Type I
Neurofibromatosis (NF)  
(von Recklinghausen disease)

- Phakomatosis (tuberous sclerosis, Sturge-Weber and Von Hippel-Lindau)
- Defects in schwann cells, melanocytes, and endoneurial fibroblasts
- Can affect any organ in the body
- 1/3000 births  ~100,000 affected in US
- One of most common genetic disorders
- Mutation rate is 1/10,000 gametes/generation
- AD with variable gene expression (FH in 60%)
Neurofibromatosis

- **Clinical forms:**
  - NF-2 - Acoustic neuromas ~ 10% (covered in Neuro lectures)
  - NF-1 (von Recklinghausen’s) - Café-au-lait spots, neurofibromas, skeletal deformities ~85%

- **Clinical features:**
  - Disfigurement
  - Blindness
  - Deafness,
  - Dermal/brain/spinal tumors
  - Loss of limbs
  - Malignancies
  - Learning disabilities
Neurofibromatosis lesions

- Café-au-lait spots
- Molluscum fibrosum
- Focal gigantism
- Lisch nodules
NF skin nodules
Skeletal abnormalities (50% of patients)

- **Appendicular skeleton:**
  - Bowing deformities
  - Pseudoarthroses
  - “Whittling” of bone

- **Spine:**
  - Dural ectasia, vertebral scalloping, foraminal enlargement, pedicle erosion
  - Scoliosis (sharply angulated with < 6 segments)
  - Pencilling and spindling of the transverse processes
Cervical spine

Vertebral dysplasia
Kyphosis
Fusion
IV foramen enlargement
Shortened pedicles
Wedge shaped vertebral bodies
Thoracolumbar spine – NF1

Sharply angulated kyphoscoliosis
Lumbar spine – NF1

Posterior scalloping
Dural ectasia
Whittling of bone
Erosions
“Pseudoarthrosis”

Tibial bowing, lytic bone lesion with sclerotic marign, fibular pseudoarthrosis, and foot deformity in a 10-year old male with type 1 neurofibromatosis.
Pelvis

“Whittling” of bone
Meningoceles

- 2/3 of patients with NF1
- Most common presentation: Asymptomatic post mediastinal mass
- Protrusion of dura and arachnoid through IV foramen and posterior rib cage into the extrapleural thoracic cavity - nonca++
- Presence of ca++ excludes meningocele
Intrathoracic meningocele
Intrathoracic meningocele
Collagen Vascular Disease

Dr. Felson lecture on interstitial lung disease

Leiden, the Netherlands
Collagen vascular disease

- SLE
- Scleroderma
- Connective tissue disease
- Idiopathic inflammatory myopathies
  - Polymyositis
  - Dermatomyositis
  - Sporadic inclusion-body myositis
Systemic lupus erythematosus (SLE)

- "Lupus" - Latin for wolf
- Malar erythema
- Young (15-40 yo), Female 5:1, rare over the age of 45
- Positive ANA
- Fever, anorexia, weight loss, polyarthritis, skin rash
- Chronic unremitting disease with variable prognosis

Major types of SLE:
- Discoid-skin rash only, 20% of patients with SLE
- Systemic-chronic, inflammatory, multisystem disorder of the immune system
- Drug-induced-Chlorpromazine, hydralazine, isoniazid, methyldopa, procainamide
SLE

• **Typical findings:**
  – Symmetric nonerosive polyarthritis of small joints
  – Jaccoud-like arthropathy (joint deformities in absence of joint destruction)
  – Myositis

• **Complications:**
  – Osteonecrosis (mainly in the hip)
  – Bone infarctions
  – Tendon rupture
  – Septic arthritis
SLE

Major Differential Dx: Jacoud’s (Post-Streptococcal) arthropathy
AVN associated with SLE
Spontaneous rupture
(Extensor carpi ulnaris tendon)
Scleroderma

- Progressive systemic sclerosis
- “Disease that turns a person into stone”
- Pathology:
  - Increase collagen deposition
  - Perivascular mononuclear cell infiltration
  - Vascular abnormalities
- 90% are females

- 3rd to 5th decade
- Incidence:
  - 1/100,000 per year
- 65% MSK involvement at presentation
- Variable prognosis
- Death: Lung, heart and renal involvement
Scleroderma

Sclerosis of frenulum  Raynaud’s
Classic variants of scleroderma

- **Morphea (localized scleroderma)**
  - Localized cutaneous sclerosis (no internal organ involvement)
- **Secondary**
  - Exposure to PVC, bleomycin, solvents, etc
- **CRESTA**
  - Calcinosis
  - Raynaud’s phenomenon
  - Esophageal dysfunction
  - Sclerodactyly
  - Telangiectasia
  - Arthritis
  - Better prognosis
Calcinosis
Calcinosis
Raynaud’s phenomenon

- Often the first symptom of scleroderma
- Ischemia of fingers, toes and ears
- Numbness, tingling and burning pain
- Attacks precipitated by cold, vibration and emotional stimuli
Acroosteolysis
Esophageal dysfunction

- Fibrosis and atrophy of the smooth muscle
- Hypermobility, dysphasia, reflux esophagitis and strictures
Sclerodactyly

- Thickening of skin of hands and feet
- Dense collagen bundles
- Phases: Edematous, indurative and atrophic
- Skin appears smooth and is tightly bound
- Fingers narrow and taper distally
Telangiectasia

- Permanent dilatation of capillaries and venules
- **Location:**
  - Face
  - Lips
  - Tongue
  - Fingers
  - Mucosal surfaces of GI tract
Arthritis
Idiopathic inflammatory myopathy

- 2:1 female to male ratio
- 5 cases/million/year (incidence increasing)
- Polymyositis (muscle involvement)
  - Direct cytotoxic effect of CD8+ lymphocytes on muscle
- Dermatomyositis (muscle and skin involvement)
  - Complement-mediated (terminal attack complex) vascular inflammation
- Both occur in association with scleroderma and SLE
Idiopathic inflammatory myopathies
Association with malignancy

- **Dermatomyositis**
  - First peak at 9 yo, second at 40-70 yo
  - Proximal muscle weakness and pain - Quadriceps predominant
  - Associated malignancies - 20% of cases:
    - Lung, prostate, female pelvic organs, breast or GI tract
  - May precede detection of tumor by months to years

- **Polymyositis**
  - Less common than dermatomyositis
  - 40-60 yo
  - SubQ and skin edema with rash
  - Associated malignancies:
    - Lung, NHL
Histology

Necrotic and regenerating muscle fibers in perivascular regions

Perivascular inflammation
Imaging findings

- **ST thickening and edema with cal++ abnormalities**
- **Articular abnormalities**
  - Juxtaarticular osteoporosis (most common)
  - Erosions of multiple sites in hands
  - Flexion deformities (MCP)
    - “Swan neck” deformity
  - Radial subluxation or dislocation of IP of thumb (“floppy thumb”) = quite characteristic
Polymyositis
Dermatomyositis
Dermatomyositis
MR imaging

- **MR Imaging (targeted or whole body)**
  - Increased SI on T2W and STIR imaging correlates with most involved regions
  - Late stages:
    - Muscle atrophy
    - Fatty replacement
  - Useful for biopsy guidance for histologic confirmation

Dermatomyositis – 13yo
Summary

- **Osteoporosis**
  - Most common metabolic disease
  - Z-score and T-scores
  - Increased susceptibility to fractures
  - “Fish” vertebral bodies
- **Osteogenesis imperfecta**
  - Diagnosis of exclusion
  - Young patient with osteopenia out of proportion to age
  - Easily fractured and exuberant callous formation
• **Neurofibromatosis**
  – Pencilling, “pseudarthrosis,”, posterior scalloping, thoracic meningocele

• **Systemic lupus erythematosus**
  – Ulnar deviation without erosions (differential is Jacoud’s (post-streptococcal arthritis)

• **Scleroderma**
  – CRESTA
  – Acroosteolysis

• **Inflammatory muscle disease**
  – Dermatomyositis and polymyositis
  – Nonspecific findings (may resemble scleroderma)
  – Remember association with malignancies
Instructions to Hostesses
Air Canada (1949)

• Warn passengers against throwing cigarettes and cigars out the window
• Keep the clock wound up in the passenger cabin
• Carry a railroad timetable in case the plane is grounded
• Keep an eye on passengers when they go to the lavatory to be sure they don’t mistakenly go out the emergency exit!!!
The Future???

- The “status quo” of radiology will not remain
- Imaging’s “golden age” may be over with changes in health care financing/delivery
- But radiology is STILL the best specialty in medicine, bar none—you made a great choice
- The “only constant” for the future will be change
- When there is change, there is a chance for improvement
- Be ready for it and react to it positively
- But regardless, being an imager should remain one of, if not the most rewarding and fulfilling jobs to do…..
Hope you have learned something…

And I know you will enjoy the rest of the AIRP course and its faculty!!

Always, remember that life is short, enjoy it, and think of these two phrases/sayings to help you through your future days:

“It is your work in life that is the ultimate seduction.” Pablo Picasso

“Don’t worry, be happy!!!” Caribbean phrase
Thanks for your attention! Enjoy the rest of the course and the beautiful city of DC!

Come to visit Charleston and the beautiful Lowcountry of SC!!