66yo male with history of spinal surgery and recurrent numbness in both legs.
Paget’s Disease

- Disordered bone remodeling due to overactive osteoclasts and osteoblasts

- Etiology unclear
  - Viral: intranuclear inclusion bodies in osteoclasts resembling measles, viral antigens, IL-6 upregulation
  - Familial: autosomal dominant (18q, 5q)
  - Benign neoplasm of mesenchymal cells
  - Zoonosis
Paget’s Disease of the Spine

- Poly-ostotic or Mono-ostotic

- Spine is the second most common site affected, after pelvis
  - Lumbar (58%; L3, L4) > Thoracic > Cervical

- Variable incidence of back pain in patients with Paget’s disease (PD)
  - 1/3 of patients with PD have symptomatic spinal canal stenosis
  - Pain attributed to PD in 12-24% of patients
  - Most often in thoracic spine (caliber of cord relative to spinal canal)
  - Remainder of cases attributed to degenerative changes, etc.
Osseous expansion of vertebral body and neural arch

- Picture frame vertebra
  - Condensation of bone in the periphery of the vertebral body with rarefaction internally
  - Squared vertebral body

- Other appearances
  - Ivory vertebrae
  - Discrete lytic lesion

- Complications:
  - *Compressive myelopathy* from central canal stenosis
  - Compression fracture

Smith et al. Radiographics. 2002
Pagetic facet arthropathy

- Abnormal remodeling leading to facet joint overgrowth and incongruity
- Destruction of articular cartilage
- Facet joint ankylosis

- Complication:
  - *Nerve root compression* from neural foraminal stenosis
Intervertebral disc invasion by pagetic tissue

• Proposed mechanism:
  – Osteoclastic resorption of subchondral bone
  – Pagetic tissue replaces cartilage at vertebral endplates
  – Resorption of disc tissue by nonosseous pagetic tissue with subsequent ossification
  – Vertebral ankylosis (4.4% incidence)

• Complications:
  – Pain, fracture

Ossification of epidural fat and ligamentum flavum at level of bone involvement

- Hypothesized to be due to hyperemic blood flow to pagetic bone

- Complication
  - *Cord compression or cauda equina*
  - Paraparesis

Arterial Steal Phenomenon

- Pagetic bone is hypervascular
  - contains 6x more vessels per cross-sectional area than normal bone
  - Shunts blood away from cord

- Pain may be localized to a different level than that suggested by anatomic findings.

- Note, Impairment of blood supply to the cord may also occur from mechanical compression by pagetic bone overgrowth

Other complications

- Platybasia
- Syringomyelia
- Vertebral body fracture
  (from PD or bisphosphonate)
- Subluxation
- Spontaneous hemorrhage
Sarcomatous Transformation of Paget’s Disease

• Rare complication (0.7% of PD overall, even less common in spine)
  – Most common sites: hip, pelvis, shoulder
• More common in polyostotic PD
• Swelling, worsening and persistent pain with rapid deterioration
• Poor prognosis (3-year mortality rate > 90%)

• Types:
  – Osteosarcoma (50-60%)
  – Fibrosarcoma
  – Chondrosarcoma
  – Malignant fibrohistiocytoma
  – Reticulosarcoma

• Caution new lytic lesion in pagetic bone
• MRI excludes malignant degeneration if medullary fat SI preserved on T1WI
Extramedullary hematopoiesis

• Anemia is rare in PD due to compensatory hematopoietic marrow proliferation in appendicular skeleton

• Thought to be from proliferation of extruded hematopoietic marrow following pathologic fractures

Paget’s Disease and DISH

- Incidence of DISH in PD: 12 – 30%

- *Morales et al.* found a statistically significant higher incidence of DISH in patients with PD compared with a control population.
  - Partly attributable to gender

- *Marcelli et al.* found higher incidence of ankylosis in the thoracic spine, in men, and an association with DISH.
  - Proposed mechanism of pagetic process infiltrating bridging osteophytes
Paget’s Disease and Ankylosing Spondylitis

- Rarer association than DISH

- Proposed mechanism of PD spreading via the path of bridging syndesmophytes

McKean et al. BJR case reports 2016.
Treatment

- Analgesic medications
- Bisphosphates
- Calcitonin
- Mithramycin
- Gallium nitrate
- Ipriflavone

- Surgical decompression of spinal stenosis
References

13. Statdx