66yo male with history of spinal surgery and recurrent numbness in both legs.


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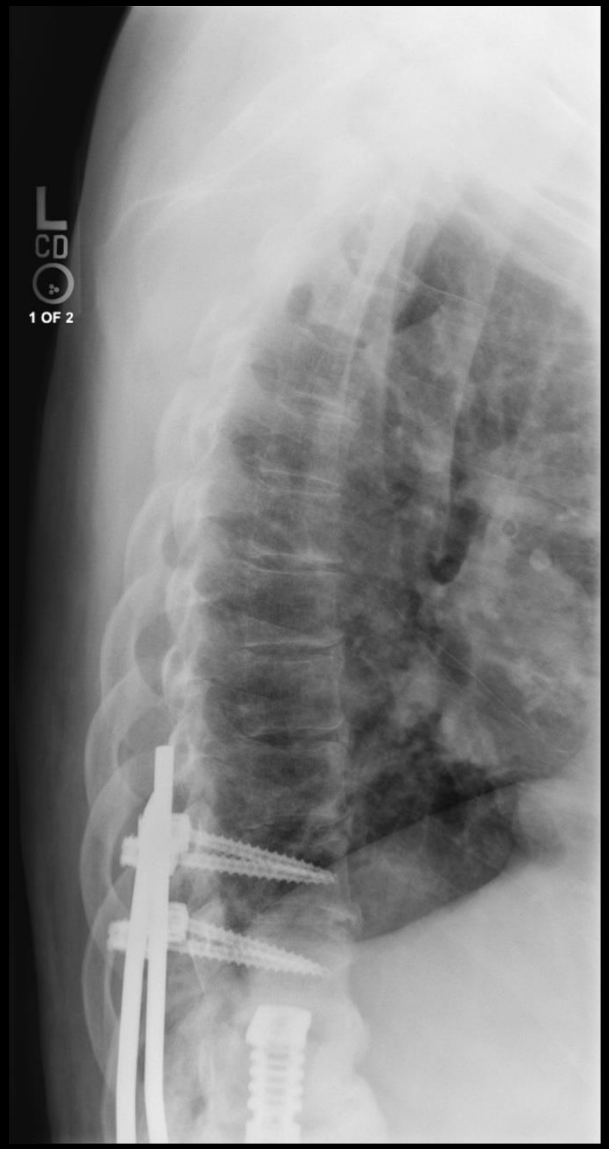
SUPINE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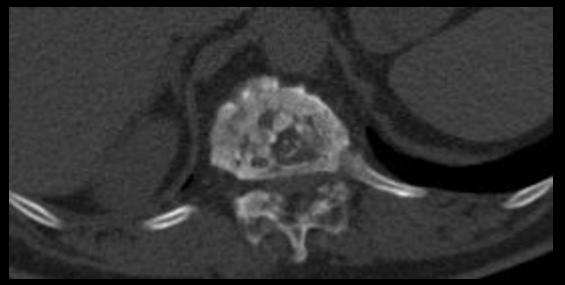


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STIR

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

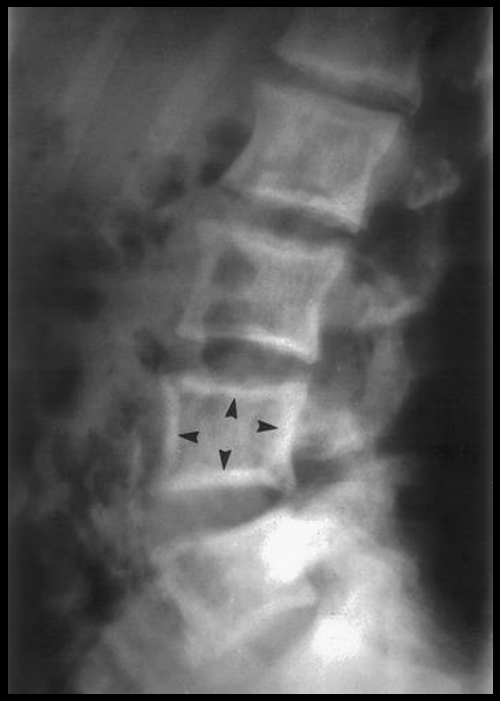
  Lumbar (58%; L3, L4) > Thoracic > Cervical

- 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

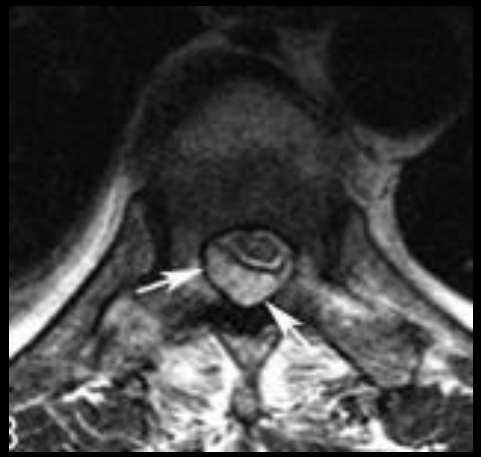
- Proposed mechanism:
  - Osteoclastic resorption of subchondral bone
  - Pagetic tissue replaces cartilage at vertebral endplates
  - Resorption of disc tissue by nonosseous pagetic tissue with subsequent ossification
  - Vertebral ankylosis (4.4% incidence)
- Complications:
  - Pain, fracture

Saifuddin et al. Clin Radiol. 2003

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



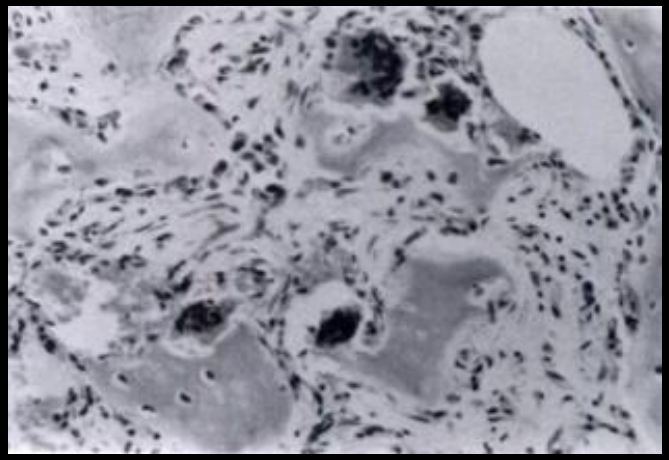
Oikonomou et al. Spine (Phila Pa 1976). 2007

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



Douglas et al. J Bone Joint Surg [Br] 1981.

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%)

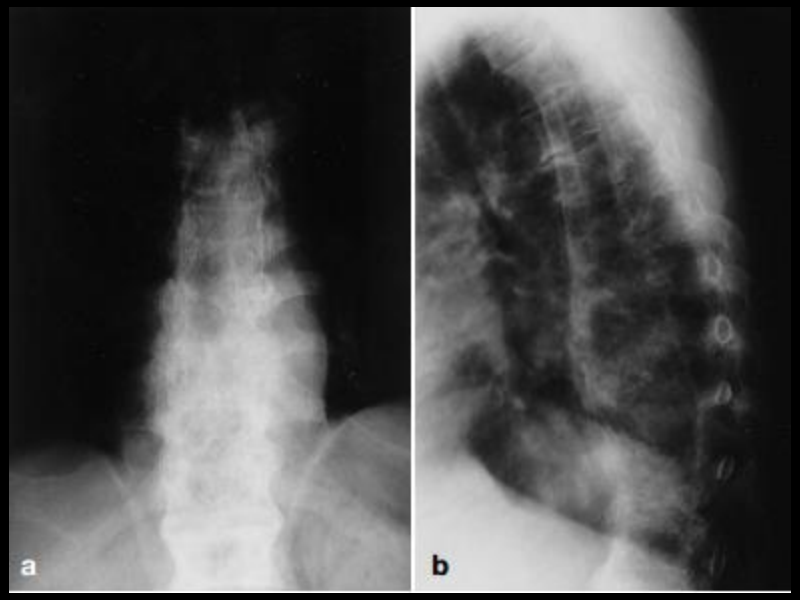
- Types:
  - Osteosarcoma (50-60%)
  - Fibrosarcoma
  - Chondrosarcoma
  - Malignant fibrohistiocytoma
  - Reticulosarcoma

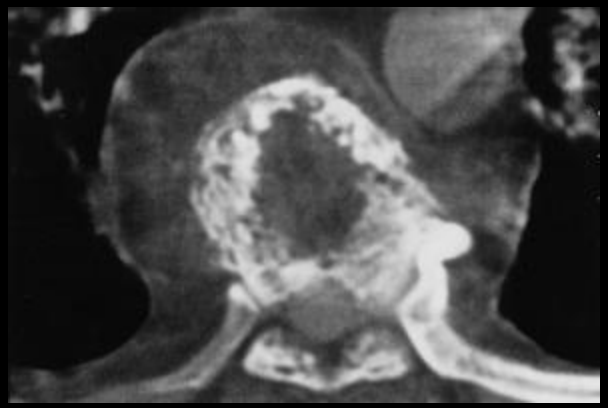
- Caution new lytic lesion in pagetic bone
- MRI excludes malignant degeneration if medullary fat SI preserved on T1WI

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





Relea et al Eur Radiol. 1999.

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

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