Paget's disease: A historical review and imaging with plain radiographs

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Learning objectives

To recall the history of Paget's disease. To describe its pathophysiological basis. To review the commonest types of the disease’s bone lesions on plain radiographs.

Background

Osteitis deformans was first described by Sir James Paget in 1876. Different chapters of the presentation call to mind the disease's history and main theories on its pathophysiological basis. Radiographs are still a very useful modality for the initial diagnosis and follow up of the disease.

Imaging findings OR Procedure details

Imaging diagnosis still relies basically on plain radiographs of the skull, spine, pelvis and long extremity bones. Different types of imaging findings are reviewed: skull osteoporosis circumscripta, thickening of diploic space, cottonwool effect, generalised lytic/sclerotic mixed lesions, cortical thickness increase, V-shaped transition zone, bowed femur and tibia, elongated long bones, pelvic trabecular coarsening, indistinct thickened iliopectineal line, hip joint space narrowing, protrusio acetabuli, vertebral body width increase-collapse and picture frame appearance, dental lesions etc.

Discussion

Paget's disease of the bone was first described by Sir James Paget (1814-1899), a British surgeon and pathologist, who had also two more diseases named after him: Paget's disease of the breast and extramammary Paget's disease. He named the bone disease "osteitis deformans" in 1876, suspecting that its pathophysiological nature is inflammatory.

It is a chronic bone condition characterised by disorder of the normal bone remodelling process [1]. It is the second most common bone disorder, osteoporosis being the first [2]. In bones affected by Paget's disease, osteous remodelling is disturbed and not synchronised. As a result, the bone that is formed is abnormal, enlarged, not as dense, brittle, and prone to breakage [3]. It's estimated that 1% of adults in the U.S. have Paget's disease. Paget's disease is rarely diagnosed in people younger than 40 years of age.
Men are more commonly affected than women [1, 4, 5]. The disease is characterised by enlarged and deformed bones and occurs mainly in the axial skeleton.

The cause of the disease is still not known entirely, but genetics are certain to be involved and viral factors are also suspected [2]. Pathogenesis includes 3 stages: Osteoclastic, mixed osteoclastic/osteoblastic and exhaustive (burnt out). The characteristic mechanism is excessive osteoclastic bone resorption with increased and disorganised bone formation. Resulting osteolysis is followed by a compensatory increase in bone formation induced by osteoblasts recruited to the area. This is associated with accelerated deposition of lamellar bone in a disorganised fashion. This intense cellular activity produces a chaotic picture of trabecular bone ("mosaic" pattern), rather than the normal linear lamellar pattern [1].

Frequent sites of involvement include the skull (25%-65% of cases), spine (30%-75%), pelvis (30%-75%), and proximal long bones (25%-30%) [6]. Paget's disease of the bones is usually asymptomatic. Polyostotic involvement and activity are common at the time of diagnosis [7]. Symptoms depend on the bones affected and the severity of the disease. Pain is the commonest symptom, originating from any bone involved and expanding to adjacent areas. Enlarged bones can pinch adjacent nerves, causing tingling and numbness. Bowing of the legs can occur. Hip or knee involvement can lead to limping, as well as pain and stiffness. Headache, loss of vision and hearing can occur when bones of the skull are affected. Vascular steal syndrome of the skull and vertebrae can cause drowsiness or paralysis. Teeth involvement causes hypercementosis.

Complications of Paget's disease include the effects of osseous weakening (deformity and fracture), arthritis, neurologic symptoms and neoplastic involvement [8, 6].

On plain radiographs, Paget's disease lesions show signs of thick trabeculae and cortex, as well as cyst-like areas caused by fat or blood filled cavities, necrosis and degeneration [9]. Skull pathology is expressed by diploic widening, lytic areas (osteoporosis circumscripta), mixed lytic/blastic lesions in thickened calvarium (cotton wool lesions), skull base sclerosis, basilar invagination, maxillar deossification and sclerosis. Long bone involvement usually occurs at the ends of the bones, including V-shaped lysis of the diaphysis and curvature of the femur and tibia, commonly complicated by fractures. In the spine, the enlarged vertebral body with lucent internal part and coarse trabeculae produces the "picture frame" appearance, while ivory vertebra is caused by increased blastic process. Ligaments of the spine, soft tissues and intravertebral disks may ossify. However, vertebral involvement is better evaluated by CT or MR. Both CT and MR are recommended in spine complications (pathological fractures, radicular or cord compression syndromes, malignant degeneration) [10, 11]. In the pelvis, there are
thickened trabeculae in the sacrum and ilium, thickened iliopectineal line and acetabular protrusion.

**Images for this section:**

![Skull radiograph shows cotton wool lesions.](image)

**Fig. 1:** Skull radiograph shows cotton wool lesions.
Fig. 2: Diploic widening is evident on skull radiograph.
Fig. 3: Picture frame appearance of lower thoracic and first lumbar vertebrae.
**Fig. 4:** Thickened iliopectineal line and sclerotic lesions of the left pelvic bones.
Fig. 5: Thickened iliopectineal line and mixed (sclerotic/lytic) lesions of the left pelvic bones.
Fig. 6: Generalised increased density produces the ivory vertebra appearance.
**Fig. 7:** Trabecular thickening and increased cortical thickness is seen in the right femoral bone.
Fig. 8: Mixed lesions of the pelvis and left femur.
Fig. 9: Bilateral sclerotic pelvic lesions.
Conclusion

Paget's disease is common in everyday clinical practice. Plain radiographs are the first imaging examination to be performed. Often no further imaging is needed. Knowledge of the different types of bone lesions is mandatory for all radiologists.

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References


