

# RADIOLOGICAL CONFERENCE

## Clinical History:

Incidental radiographic finding (**Figure 1**) in the pelvis of a 43-year-old Caucasian man. He does not have any significant medical history.

**Figure 1: Frontal radiograph of the pelvis**



Answer  
on  
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## What is the diagnosis?

- a) Paget's disease
- b) Myelosclerosis
- c) Fluorosis
- d) Metastases from prostatic carcinoma
- e) Melorheostosis

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## Answer:

a) Paget's disease

## Radiological findings

Radiograph of the pelvis (**Figure 2**) shows increased density and expansion of the bones in the right hemipelvis. There is coarsening of the trabecular pattern and cortical thickening, with narrowing of the medullary space. Features are typical of those of Paget's disease.

## Discussion

### Paget's disease

Paget's disease, also known as osteitis deformans, is a disease of unknown origin. In this condition,

normal bone is reabsorbed and replaced by abnormally coarsened trabecular bone which is excessively remodelled in bizarre fashion. It is rare in Asia but is commonly encountered in Europe, Australia and the United States. The disease usually manifests during middle age, only occasionally occurring before the age of forty years. It is more prevalent among males. Pain is the most common presenting symptom, although about 20% of the patients are asymptomatic at the time of initial diagnosis. Biochemically, the serum calcium and phosphorus levels are normal, with a strikingly high level of alkaline phosphatase.

Paget's disease has a predilection for bones of the axial skeleton and the proximal femur. The common sites of involvement are the pelvis, lumbar spine, femur and skull. The sequential stages of this disease are the active (or osteolytic), mixed (combined osteolytic and osteosclerotic), and inactive (quiescent or osteosclerotic) phases, though there may be overlap of these stages. The radiographic hallmarks of Paget's disease are coarsening of trabeculae, bony expansion, and modelling deformity.

**Figure 2:** Same radiograph as **Figure 1** with the addition of arrows. There is asymmetrical involvement of the right hemipelvis by sclerosis, coarse trabeculation, bony expansion (curved arrows) and cortical thickening (small arrows). Characteristic iliopectineal line thickening (open arrowheads) is seen



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The manifestations in the skull range from osteoporosis circumscripta in the active phase to 'cotton wool' and diffuse sclerosis in the quiescent stage. Bone softening can result in basilar invagination. The changes in the long bones start with a subarticular 'flame-shaped' lucent lesion, likened to a 'blade of grass'. Bowing deformities and incremental transversely-orientated fractures may result from weight bearing and abnormal mechanical stress. In the spine, vertebral body expansion, cortical thickening and trabecular coarsening can produce an 'ivory' vertebral body appearance. Spinal cord and nerve root compression results from the pressure of distorted pagetoid bone or pathological fracture.

The changes in the pelvis are most often seen during the mixed or inactive stages. There is asymmetrical expansion and coarsened trabeculation of the pelvic ring. These changes are frequently accompanied by characteristic thickening of the iliopectineal line. Mechanical stress in pagetoid bone can lead to protrusio acetabuli and premature osteoarthritis. In the ilium, there may be multiple curvilinear lines running parallel to the iliac crest. Besides fractures and deformity from bone softening, another important complication of Paget's disease is sarcomatous change. This occurs in 1% of patients, being as high as 5-10% if there is widespread bony involvement. The femur, pelvis and humerus are the sites most commonly involved. In our patient, the radiographic features in the pelvis are typical of those of Paget's disease.

## Myelosclerosis

Myelosclerosis is one of the chronic myeloproliferative disorders. This group of diseases is characterized by self-perpetuating haematopoietic cell proliferation. The body responds by production of fibrotic tissue and endosteal new bone. Clinically, the patient is anaemic, and may present with weakness, dyspnoea and abdominal pain. Bleeding due to abnormal platelet function and joint pain from secondary gout may occur. Typical radiographic appearances consist of widespread diffuse increase in bone density, endosteal thickening,

and patchy lucencies due to persistence of fibrous tissue. There may be associated hepatosplenomegaly caused by extramedullary haematopoiesis. None of these features were present in our patient.

## Fluorosis

Fluorosis is caused by fluorine poisoning or excessive intake of fluorine. The incidence of fluorine poisoning is now exceedingly low, occurring mainly from agricultural and industrial (e.g. chemical, mine and steel plants) sources. Fluorosis usually results from chronic intake in certain geographical regions which have large amounts of fluorine in the drinking water. It can also be caused by long-term therapy with sodium fluoride for osteoporosis. Affected patients are usually asymptomatic although children may have severe stiffness and pain. Radiographically, there is diffuse skeletal sclerosis, with cortical thickening and periosteal reaction. These changes are most pronounced in the axial skeleton. There is often ossification of ligaments, musculotendinous attachments to bone and interosseous membranes. Paget's disease, with typical features of increased trabeculation and bone expansion, is radiographically distinct from fluorosis.

## Metastases from prostatic carcinoma

Osteosclerotic metastases are less frequently encountered compared to lytic metastases. They are most commonly due to prostatic carcinoma, and are also found in breast carcinoma and carcinoid. Radiographically, the presence of multiple focal areas of sclerosis within the medullary cavity of the skeleton poses no problems with regard to the correct diagnosis. A careful search should be made for cortical destruction or a lytic component, which would support the diagnosis of a metastatic lesion. However, osteosclerotic metastases may manifest diffusely, superficially resembling Paget's disease. The distinction can be made in most cases from careful radiographic evaluation for features of Paget's disease and from the clinical history of a known primary neoplasm.

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## Melorheostosis

Melorheostosis is a rare non-hereditary progressive disease of unknown aetiology. The hallmark of this disease is the presence of peripherally located cortical hyperostosis in one bone or a series of bones. Melorheostosis usually manifests early in childhood, causing pain, peri-articular swelling and limitation of motion. It may be associated with skin and soft tissue abnormalities. Bony involvement is usually asymmetrical, extra-axial and most often, unilateral. Densely sclerotic cortical bone involving one side of a long bone and crossing a joint to the most adjacent long bone has been likened to the "guttering of a candle" or

"burning candle wax" appearance. Bone masses may protrude into adjacent articulations, appearing as osteochondromas. None of these clinical or radiologic features were present in our patient. ■

## Further reading

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