

## Pagetic disease of the femur

Published on 14.01.2002

**DOI:** 10.1594/EURORAD/CASE.1352

**ISSN:** 1563-4086

**Section:** Musculoskeletal system

Case Type: Clinical Cases

**Authors:** A. Loshkajian

**Patient:** 70 years, male

### Clinical History:

Femur fracture occurring on a pathological bone.

### Imaging Findings:

The patient complained of intense pain in the left leg after a fall. An X-ray of the left femur was performed. The radiogram revealed a transverse incomplete cortical fracture of the distal femur. The other features seen are a bowing deformity of the bone, a coarse demineralisation with widening of the bone and thickened cortex. There is also a thickening of the trabecular pattern.

The most likely diagnosis is a stress fracture occurring in a pagetic long bone.

### Discussion:

Paget's disease (PD) is a relatively common disease occurring in approximately 3-4% of people over 40 years of age. Most patients are mildly symptomatic or asymptomatic. No clear aetiology is known. Pathologically, the disease occurs in three phases. Initially, bone is resorbed secondary to increased osteoclastic activity. Osteoblasts then form abnormal new bone. The primitive woven bone production is disorganised, contains increased vascularity and connective tissue, and does not contain Haversian systems. These two phases are repeated until the osteoclastic activity subsides. The final, or inactive phase is reached when there is a cessation of osteoblastic activity. The disease usually involves more than one bone although 10-35% of patients have monostotic disease.

The three distinct radiographic phases (lytic, mixed and sclerotic) correspond to the pathological phases. In the lytic phase, most commonly recognised in the skull as osteoporosis circumscripta and in long bones, a purely lucent defect is noted. As the disease progresses, through the mixed, to the inactive sclerotic phase, new pagetoid bone enlarges the bone predominantly through cortical apposition. Coarse and thickened trabeculae are also noted and in the skull a "cotton wool" appearance may be seen. In the inactive or sclerotic phase, a diffuse increase in bone density as well as bone enlargement is noted.

The long bone most frequently involved in Paget's disease is the femur, but the clavicles, the scapula, and the fibula may also be involved. The sternum, patella and the bones of the hands and feet are rarely involved.

In the long bones, the initial osteolytic lesion usually starts in the subarticular region of the epiphysis and extends toward the diaphysis with a well-demarcated V-shaped zone ("blade of grass") separating the normal and abnormal bone. With progression of the disease, the radiograms reveal:

- characteristic bowing of the long bones especially the femur and tibia and deformities such as the "shepherd's crook" of the proximal femur and anterior tibial bowing
- a thickening of the trabecular pattern which leads to distortion of the normal architecture
- a widening of the bone with a thickened cortex
- pathological fractures of the transverse type, most commonly in the femur or the tibia
- incomplete cortical stress fractures healing at the periosteal and endosteal surfaces, leaving a residual midcortical lesion

Most difficulty in diagnosis will occur in the mixed phase, when patchy areas of sclerosis are present with minimal or no bone enlargement. The main differential diagnoses are metastasis to the bone; fibrous dysplasia, if the facial bones are involved; myelofibrosis; fluorosis; renal osteodystrophy; and haemolytic anemias. The clinical context and biological and haematological tests lead usually to the correct diagnosis of Paget's disease.

**Differential Diagnosis List:** Pagetic disease of the femur

**Final Diagnosis:** Pagetic disease of the femur

#### **References:**

Guyer PB, Clough PW.

Paget's diseases of bone: some observations on the relation of the skeletal distribution to pathogenesis.

Clin Radiol. 1978 Jul;29(4):421-6. (PMID: [679616](#))

Mirra JM, Brien EW, Tehranzadeh J.

Paget's disease of bone: review with emphasis on radiologic features, Part II.

Skeletal Radiol. 1995 Apr;24(3):173-84. (PMID: [7610409](#))

Griffiths HJ.

Radiology of Paget's disease.

Curr Opin Radiol. 1992;4(6):124-8. (PMID: [1292546](#))

**Figure 1**

a



**Description:** X-ray demonstrating a transverse fracture of the femur (blue arrow). The bone is bowed (green arrows) with coarsened and thickened trabeculae. **Origin:**

**Figure 2**

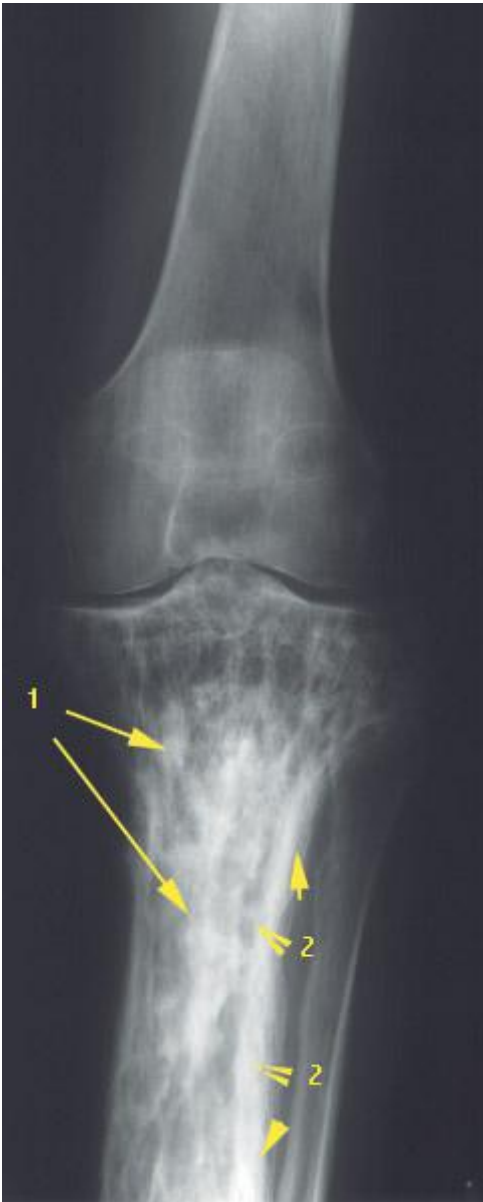
**a**



**Description:** Well-demarcated V-shaped zone (black arrows) separating the normal and abnormal bone. This finding is highly suggestive of pagetic disease. The cortex is thin (arrows) corresponding to the lytic phase of the disease. **Origin:**

**Figure 3**

a



**Description:** There is a typical pagetic aspect of the tibia with thickened cortex, bone enlargement and coarsening of the bone trabeculae (arrows). **Origin:**