Paget's disease is rare in the Chinese. We present a patient who had a rare monostotic Paget's disease involving middle diaphysis, distal metaphysis and epiphysis of left femur. Characteristic radiological findings include cortical thickening, coarse trabeculae, osseous expansion and enlargement, and bowing deformity of bone. The patient also had osteo-arthritis of the knee, which can be a complication of Paget's disease.

Key words: Paget's disease, Osteitis deformans

Paget's disease is a common disorder of middle-aged and elderly patients in Europe and America, but very rare in Asia. We report a rare monostotic Paget's disease in a Chinese patient, who had characteristic radiological findings and complication of secondary degenerative osteo-arthritis.

CASE REPORT

A 67-year-old woman visited our hospital presenting with left knee pain and deformity for about 2 years. Varus deformity of left knee joint and limited range of flexion to 45° were noted on physical examination. Radiographs of left knee (Figures 1, 2) showed typical radiological findings of Paget's disease, including cortical thickening, coarse trabeculation, bony expansion and bowing deformity from mid-diaphysis of left femur to femoral condyle and associated with osteoarthritis. Serum biochemistry and CBC were normal except increased alkaline phosphatase (455 IU/L). Radionuclide scintigraphy (Figure 3) showed increased radiotracer uptake at left femur (from mid-diaphysis to distal epiphysis) and the first lumbar vertebral (L1) body, but thoraco-lumbar spine radiographs showed osteoporosis with compression fracture of L1 without evidence of Paget's disease involving L1. MRI findings (Figures 4, 5, 6) included cortical thickening with foci of increased signal intensity on intermediate-weighted (PD) image and T2-weighted (T2W) image, coarse trabeculation, bowing deformity and enlargement of bone, and osteo-arthritis of left knee. The signal characteristics in the medullary cavity had resembled either normal fatty bone marrow or focal fatty replacement. The patient received total knee replacement and calcitonin therapy. Histopathological diagnosis (Figure 7) was compatible with late phase of Paget's disease with the findings including irregular bony trabeculae of various sizes and shapes embedded in fibrovascular stroma, and typical mosaic pattern of bony trabeculae and cement lines.

Reprint requests to: Dr. Chi-Chen Hou
Department of Diagnostic Radiology, Chi-Mei Foundation Hospital.
No. 901, Chung Hwa Road, Yung Kang 710, Tainan, Taiwan, R.O.C.
Paget’s disease of bone (osteitis deformans) first described by Sir James Paget in 1877 is a condition of unknown etiology that affects approximately 3% of people over 40 years old in the United Kingdom, Australia, and New Zealand [1, 2]. However, it is rare in Asia, Africa, and Scandinavia; and it is extremely rare among the Chinese.

Paget’s disease predominates in middle-aged and elderly individuals. It affects two times more often in men than in women, and frequently asymptomatic, and often diagnosed incidentally on radiographs. Clinical symptoms and signs vary with the distribution and extent of the disease. It is a slowly progressive disorder characterized by initial osteoclastic activity of bony resorption, then excessive and abnormal remodeling of bone, leading to enlargement and deformity of bone. It passes through active, intermediate and quiescent stages. Paget's disease is a sporadic disease with no clear genetic transmission. The etiology is uncertain, but several factors have been suggested to play a role, including environmental factors, nutritional deficiencies, and viral infections.

**Figure 1.** (a,b) Two views of left knee show typical radiological findings of Paget’s disease, including cortical thickening, coarse trabeculation, osseous enlargement and expansion with a cyst-like osteolytic lesion (arrow-head) in lateral femoral condyle, and later MRI confirm this osteolytic lesion to be a focal fatty replacement or fatty bone marrow; it should not be mistaken as neoplasm.

**Figure 2.** a. Anteroposterior and b. Lateral view of left femur X-ray show lateral and anterior bowing deformity of left femur in contrast to frequent lateral bowing deformity of Pagetic femur.

**Figure 3.** Tc-99m bone scan in blood pool phase show increased radiotracer uptake at left femur (from mid-diaphysis to distal epiphysis) and L1 body, but more markedly increased radionuclide activity at left femoral shaft than L1 body.
cent phase, leading to a combination of osseous resorption and apposition that produce a distinctive radiological and pathological appearance in which has cortical thickening, osseous enlargement and expansion, and bowing deformity of long bones, and disorganized & coarsened trabeculae (mosaic pattern) (Figures 1, 2).

Although Paget’s disease is polyostotic in most cases, it may be initially or totally monostotic in 10-35% of cases [2]. As noted by Groh [2], monostotic Paget’s disease appears to predominate in axial skeleton (particularly in lumbar spine), although any portion of the skeleton may present with the only site of involvement as our case. Monostotic Paget’s disease can be a distinct entity. Groh [4] presented nine cases of monostotic paget’s disease under observation long enough to be convinced that Paget’s disease can occur in a single bone only and never become generalized.

The musculo-skeletal complications of Paget’s disease include osseous deformity, fracture, neoplasms, neurologic deficits, and degenerative joint disease. The most serious and deadly complication of Paget’s disease is sarcomatous degeneration [2]. Unlike the findings in primary degenerative disease, the majority of secondary degenerative change by Paget’s disease has a radiographic pattern characterized by uniform narrowing of the articular cartilage and joint space, and minimal hypertrophic changes [6]. The pathogenesis of osteo-arthritis changes associated with osteitis deformans is possibly hypothesized that hypervascularity and rapid bone turnover in Pagetic subchondral bone may lead to accelerated and disturbed endochondral ossification at the expense of articular cartilage which is eroded from beneath, and

Figure 4. Sagittal plane of left femur on PD (1850/40) show high signal intensity within medullary canal, either represent fatty bone marrow or focal fatty replacement, and can exclude secondary sarcomatous transformation of Paget’s disease.

Figure 5. Axial plane through meta-diaphysis of left femur on a. PD (1850/40) show linear and patchy increased signal intensity (arrow-head) within thickened cortex, and enhanced after Gadolinium b. T1WFS (500/13). This finding is postulated to be caused by resorption and remodeling of cortical bone, with the introduction of cellular marrow element.
loss of cartilaginous inner calcified zone, resulting in thinning and irregularity of remaining cartilage. Secondary bowing deformity of Pagetic bone produce further abnormal stress of joint biomechanics, and accelerate secondary degenerative joint change of Paget's disease [6].

In our case, the dominant signal intensities of bone marrow are similar to that of fat, which had a 100% negative predictive value in excluding Pagetic sarcoma or other tumors [7]. And this MRI finding can explain the pseudoneoplastic osteolytic cystic lesion of femoral condyle shown on plain radiographs as fatty signal intensity on all pulse sequences, representing focal fatty replacement. MRI is indicated when Pagetic bone has osteolytic foci possibly representing with the advancing radiolucency in initial Paget's disease, or pseudo-neoplastic osteolytic foci and focal fatty replacement in osteosclerotic phase of Paget's disease, or giant cell tumor and other sarcoma in complicated Paget's disease. Linear and patchy increased signal intensity within thickened cortex was noted on PD and T2W. The lesions were enhanced after intravenous Gadolinium administration (Figure 5). This finding is probably due to resorption and remodeling of cortical bone with the introduction of cellular marrow element [8]. Other MR findings include osseous expansion and enlargement, bowing deformity of left femur, coarse trabeculae across stress lines (Figure 6), secondary osteoarthrosis of left knee joint, and without cortical destruction or extra-osseous soft tissue mass. In view of plain radiographs and MRI and bone scan, our case is a monostotic Paget's disease of osteosclerotic phase with focal fatty replacement (pseudo-neoplastic cystic lesion in femoral condyle).

In summary, radiographic features of Paget's disease are characteristic and diagnostic. Scintigraphy can detect the extent and activity of Paget's disease. CT and MRI can help to evaluate the complications of Paget's disease.

REFERENCES
單一骨頭Paget’s Disease：病例報告

侯其禎¹ 羅際衛² 張晉民¹ 曾慶誠³

奇美醫院 放射診斷科¹ 骨科² 病理科³

Paget’s disease非常少見於亞洲，尤其是中國人和日本人，較常見於英格蘭、澳洲和紐西蘭。它常是多發性骨頭疾病，常侵犯骨盆骨頭、腰椎、頭蓋骨、股骨、腸骨，甚至廣泛侵犯全身骨頭。單一骨頭Paget疾病約占全部Paget疾病的10-35%。我們提出一個非常少見的單一骨頭Paget疾病，左頸的股骨包括骨幹，遠端的骨端都被Paget疾病浸潤，而且X光的表現相當的典型：如皮質變厚，骨小樑變粗，骨頭彎曲變形和變大。病人有二度退化性關節炎，是Paget疾病的併發症。核子醫學掃瞄檢查是否有其它骨頭被侵犯，核磁共振檢查是否有肉瘤的變化，它是Paget疾病最嚴重的併發症。

關鍵詞：骨頭、變形骨炎

Monostotic paget’s disease

121