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Nineteen-Year Radiographic Follow-up of Untreated Paget’s Disease of Bone

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Radiographic and histologic findings in Paget’s disease of bone have been previously well described, and characteristic features of each stage have been determined. Long-term radiographic follow-up of untreated patients is rarely reported. We had the opportunity to observe the radiographic evolution of untreated Paget’s disease of the tibia. Sequential radiographic studies spanning a 19-year period have allowed us the opportunity to study the natural course of this disease. (Henry Ford Hosp Med J 1986;34:127-9)

Case Report

A 63-year-old man was initially seen at Henry Ford Hospital in 1963 because of increasing shin pain which had been present for several months. At that time a diagnosis of Paget’s disease was made, and a brief trial of aspirin therapy was instituted. The patient’s anterior tibial pain persisted and worsened with prolonged use of the leg. The patient experienced periods both of exacerbation and remission of symptoms, but over the last two decades no further therapy was deemed necessary by him.

In August 1985, calcitonin therapy was instituted after several months of right ankle pain. The patient reported separate instances of trauma to the right leg caused by falls in 1968, 1974, and 1980. He also described “sensitivity to kneeling,” with persistent pain in the knees and occasional brief “locking” for the past several years. He has a past history of peripheral trauma to the right leg caused by falls in 1968, 1974, and 1980. He also described “sensitivity to kneeling,” with persistent pain in the knees and occasional brief “locking” for the past several years. He has a past medical history of gout and essential hypertension. Physical examination revealed moderate deformity of the right leg and slight warmth over the tibia. Laboratory investigation revealed an alkaline phosphatase of 1.033 IU/L; past measurements had ranged from 200 to 300 IU/L.

Radiographic evaluation was obtained intermittently between 1966 and 1985. In 1966, roentgenograms of the right tibia revealed a well-defined radiolucency involving the anterior cortex in the proximal diaphyseal region, with a characteristic V-shaped distal margin. No sclerosis, cortical thickening, periosteal reaction, or soft tissue swelling was evident. The lytic region was seen to spare the proximal end of the tibia (Fig 1A).

In 1972, the lesion had progressed distally to the mid-diaphysis, with minimal proximal extension. At this time, the border between normal and pagetoid bone was less well-defined than in 1966. Smudging and coarsening of the trabecular pattern within the region of abnormality were evident, along with irregular periosteal cortical thickening (Fig 1B).

In 1980, further progression to the distal tibial diaphyseal region was observed. Radiographs revealed marked undulent cortical thickening throughout the tibial diaphysis, with regions of lysis and sclerosis and disorganization of the trabecular pattern. Ill-defined sclerosis was seen in formerly lytic zones proximally, with predominantly lytic involvement of the more recently affected distal tibia (Fig 1C).

On radiographic evaluation in 1985, further distal progression of the changes were evident. Cortical thickening and trabecular coarsening were combined with poorly marginated patches of sclerosis. A large bony spur superior to the tibial tubercle also demonstrated findings of Paget’s disease. The changes appeared to extend to both the proximal and distal articular surfaces of the tibia (Fig 1D).

Radionuclide bone scan demonstrated intense increased uptake throughout the entire length of the right tibia (Fig 2). Increased uptake was also evident in the left hemipelvis, proximal left femur, and right humerus. Classic radiographic changes of Paget’s disease were seen in these latter locations, with evidence for previous pathologic fracture through the distal right humerus.

Discussion

Osteitis deformans was first described by Sir James Paget in 1877 (1). This bone disease generally has its onset in mid-adult life and increases in incidence with age, although cases have occurred in young adults (2). The pathologic changes are commonly divided into three phases: initial, active, and inactive (3,4). The early phase is characterized roentgenographically by bone resorption, appearing classically as a radiolucent area with a V-shaped distal margin involving both cortical and cancellous bone. This initial phase corresponds to the so-called “flame” or “blade of grass” appearance, with a sharply defined oblique border between normal and abnormal bone (4). The term osteoporosis circumscripta is used to describe the appearance of this early phase in flat bones such as the calvarium. Intense increased uptake is usually seen in active Paget’s disease on radionuclide bone scan, although areas of photopenia have been postulated as possible precursors of active Paget’s disease (5).

The findings on the initial radiographs in our patient demonstrate the characteristic appearance of the lytic phase of the disease. At this point, biopsy usually shows intense osteoclastic

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Fig 1—Lateral radiographs of the tibia spanning a 19-year period. (A) 1966: Note the characteristic radiolucent area with a sharply defined oblique inferior margin (arrows). (B) 1972: Progression of resorption distally, with less well-defined border between normal and pagetoid bone. (C) 1980: Further distal progression of lysis, with disorganization of the trabecular pattern and marked periosteal new bone (arrows). (D) 1985: Extension to involve the entire tibia. Note the increased size and anterior bowing of the tibia.

Fig 2—Isotope bone scan (1985). Markedly increased uptake throughout the entire length of the right tibia, indicating active disease.
Remineralization is also seen, with gross bowing deformities resulting from the extensive remodeling and weakening of bone.

Pathologically, the new pagetoid bone develops in an irregular pattern in contrast to the organized longitudinal osteon orientation of normal bone. Hypervascularity is prominent within the bone marrow, with loose fibrotic changes (4). A wide zone of spongy periosteal bone, which has been likened to pumice, may form around the original cortex (9). Similarly, endosteal new bone may also cause cortical thickening, although this is less frequently recognized (3,4). Ultimately, the remodeling, with constant interplay between resorption and new bone formation, results in the classic histologic mosaic pattern. At the most recent evaluation, the disease in our patient was still active, as evidenced by symptoms and the findings on isotope bone scan.

The inactive phase is reached after many years, with reduction of abnormal osteoblastic and osteoclastic activity. Radiographically, the changes of inactive Paget's disease may be difficult to differentiate from advanced active disease (4); hence, the value of the isotope bone scan. Deformities of an affected bone may lead to malalignment of joints with secondary osteoarthritic changes. Basilar invagination of the skull, spinal stenosis, and stress fracture, usually in the later stages, may occur. Sarcomatous degeneration is a well-documented late complication (10).

Drugs commonly used to treat Paget's disease, such as calcitonin and mithramycin, appear to produce remission by causing an inhibition of excessive osteoblastic and osteoclastic cellular activity (11-14). Following the recent exacerbation of symptoms at the distal right tibia, calcitonin therapy was initiated in our patient.

Summary

Although the radiographic and pathologic changes in the three commonly recognized phases of Paget's disease of bone have been well described, they are rarely observed over a long period of time in untreated patients. Our report of a patient, who was followed clinically and roentgenographically for 19 years, has allowed us the unusual opportunity to study the natural progression of this disease.

References