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Radiologic Spectrum of Paget Disease of Bone and Its Complications with Pathologic Correlation[1]

LEARNING OBJECTIVES FOR TEST 6

After reading this article and taking the test, the reader will be able to:

■ Recognize the radiologic spectrum of Paget disease of bone and its complications.

■ Describe the pathologic basis of the radiologic features of Paget disease of bone and its complications.

■ Identify the radiologic manifestations that allow distinction of sarcomatous transformation in Paget disease of bone.

Stacy E. Smith, MD • Mark D. Murphey, MD • Kambiz Motamedi, MD Michael E. Mulligan, MD • Charles S. Resnik, MD • Francis H. Gannon, MD

Paget disease of bone is a common disorder affecting approximately 3%–4% of the population over 40 years of age. The pathologic abnormality in Paget disease is excessive and abnormal remodeling of bone. Three pathologic phases have been described: the lytic phase (incipient-active), in which osteoclasts predominate; the mixed phase (active), in which osteoblasts cause repair superimposed on the resorption; and the blastic phase (late-inactive) in which osteoblasts predominate. Radiographic appearance of Paget disease reflects these pathologic changes and is often characteristic. Initially, there is osteolysis, particularly affecting the skull (osteoporosis circumscripta) and subchondral long bones, with subsequent development of trabecular and cortical thickening and enlargement of bone in the mixed phase of the disease. Finally, areas of sclerosis may develop in the blastic phase. Frequent sites of involvement include the skull (25%–65% of cases), spine (30%–75%), pelvis (30%–75%), and proximal long bones (25%–30%). Bone scintigraphy typically demonstrates marked increased uptake of radionuclide in all phases of Paget disease. Computed tomography and magnetic resonance imaging often show changes similar to those seen radiographically in noncomplicated Paget disease with maintenance of yellow marrow. Complications of Paget disease include the effects of osseous weakening (deformity and fracture), arthritis, neurologic symptoms, and neoplastic involvement. Sarcomatous transformation is the most feared complication, occurring in approximately 1% of cases, and is seen on images as focal bone destruction extending through the cortex with an associated soft-tissue mass. Recognition of the radiologic spectrum of the appearances of Paget disease usually allows prospective diagnosis and differentiation of its associated complications, which helps guide therapy and improve patient management.

Introduction

Paget disease of bone, also known as osteitis deformans, was first described in a small group of patients in 1877 by Sir James Paget (1). These patients had an "odd overgrowth" of their heads and extremities, which often resulted in bowing of the long bones and increased propensity to develop subsequent fractures (1). Because Paget disease is extremely common, radiologists are frequently confronted with its many manifestations, both in asymptomatic (noncomplicated) and symptomatic (complicated) patients. In the asymptomatic patient in whom Paget disease was incidentally detected, it is important not to confuse its imaging appearance with those of other diseases. Complications of Paget disease include the effects of osseous weakening (deformity and fracture), arthritis, neurologic symptoms, and neoplastic transformation or involvement (2–8).

This article illustrates the spectrum of radiologic manifestations of Paget disease, both uncomplicated and complicated. Although we emphasize its radiographic appearance, a multimodality approach including bone scintigraphy, computed tomography (CT), and magnetic resonance (MR) imaging is used. The pathologic basis of these imaging manifestations is also emphasized.

Demographics and Causes

Demographic distribution of Paget disease is particularly high in Great Britain, site of the first reported cases (1–5,9). Interestingly, lands settled by the British such as Australia, New Zealand, and the United States share a common prevalence of Paget disease. The disease is also common in continental Europe but rare in Asia and Africa. Although there is variation among studies, there is an overall mild male predilection in Paget disease and age of onset is slightly younger in men. Paget disease is extraordinarily common, affecting 3%–4% of the population over 40 years of age and up to 10%–11% after 80 years of age (2–7). Although Paget disease is unusual in people younger than 40 years of age (only about 4% of patients with the disease are in this younger age group), its characteristic imaging features are maintained and thus allow an accurate diagnosis (4).

The cause of Paget disease remains uncertain. Although still controversial, a probable viral origin has been proposed because intranuclear inclusion bodies (resembling those of a paramyxovirus variety) are found in the osteoclasts in histologic specimens of Paget disease (8,10–12). Giant osteoclasts composed of numerous nuclei characterize the active phase of Paget disease (13). These osteoclasts contain organized groups of microcylinders in the nuclei and cytoplasm. Both findings help support the viral origin theory, because both enormous osteoclasts and

inclusion bodies are also found in viral infections such as measles (caused by a paramyxovirus), an entity found in some patients with Paget disease.

A review of Paget disease cases in Japan noted that the disease was unknown among the population until the first case report in 1921 (14). Since that time, the number of cases in Japan has increased. This raises the question of whether it is a true increase in cases, secondary to a slow virus infection, or only an increase in detection, secondary to an increased number of imaging studies that reveal unsuspected disease in asymptomatic patients. Ashkenazi Jews have a higher prevalence of Paget disease, with an associated increased frequency of the serum marker HLA-DR2, a finding that suggests a possible genetic origin (3,4,15–17). Other purported causes include connective tissue disease, autoimmune disorder, vascular disease, metabolic disorder related to parathormone, or a true neoplastic process (4).

Pathologic Characteristics

Paget disease is characterized by excessive and abnormal remodeling of bone, with both active and quiescent phases (18,19). Three phases have classically been described as discrete and distinctive, although in reality they represent a continuum: the lytic phase (incipient-active), in which osteoclasts predominate (Fig 1a); the mixed phase (active), in which osteoblasts begin to appear superimposed on osteoclastic activity and eventually predominate; and finally, the blastic phase (late-inactive), in which osteoblastic activity gradually declines (Fig 1b).

Because there is often widespread osseous involvement and because individual sites progress at variable rates, Paget disease of differing phases may be seen in the same patient. The end result is a thickened, disorganized trabecular pattern of bone, referred to as a "mosaic" or "jigsaw" pattern (1,4,19) (Fig 1c). Cement lines along the coarsened and enlarged trabeculae are characteristically seen; these lines represent osseous resorption and bone formation (Fig 1b, 1c). The trabecular areas of thickening usually lack the interconnection seen in normal bone and thus are weakened and often referred to as "pumice" bone (Fig 1d). The cortex is also thickened and is the area of most active bone turnover and repair. These areas of increased bone resorption and formation also reveal hypervascularity with small caliber vessels (20,21).

Bone marrow changes are noted throughout the disease process. Fibrovascular tissue replaces the normal yellow marrow in more active disease, particularly in the lytic phase and less extensively in the early mixed phase (Fig 1a). A return to diffuse yellow marrow gradually occurs in the late mixed phase (Fig 1b). This process often ultimately results in an actual increase in marrow fat deposition (compared with normal yellow mar-
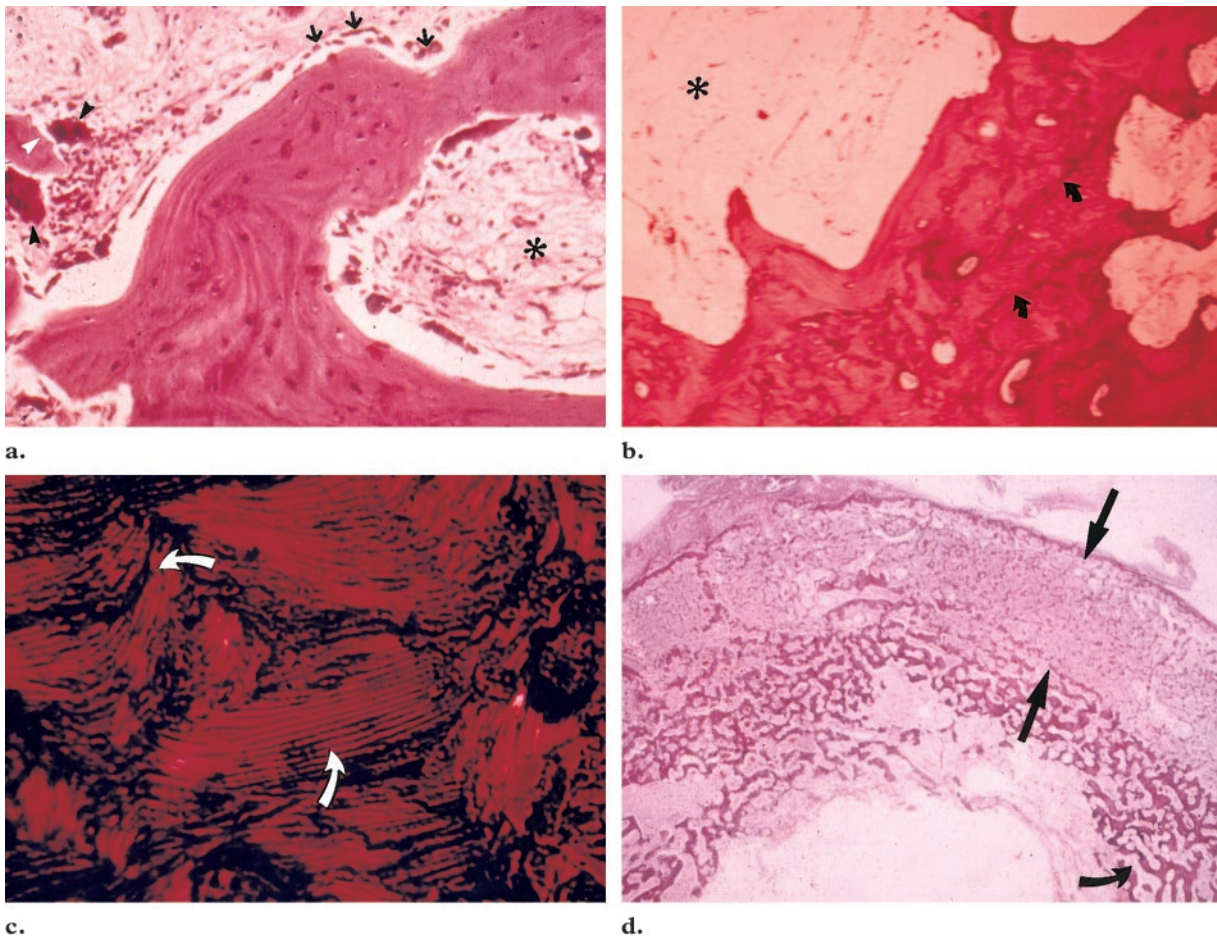
Figure 1.   Typical histologic appearance of Paget disease. **(a)** Lytic to mixed active phase. Photomicrograph (original magnification, ×150; hematoxylin-eosin [H-E] stain) reveals multiple osteoclastic giant cells (black arrowheads) and osteoblasts (arrows) causing bone resorption (white arrowhead) and formation. The marrow cavity is replaced by fibrovascular tissue (∗). **(b)** Blastic quiescent phase. Photomicrograph (original magnification ×175; H-E stain) shows cement lines (arrows) and that the marrow cavity has returned to fatty tissue (∗). **(c)** Mixed phase. Photomicrograph (original magnification ×200; H-E stain under polarized light) shows multiple cement lines and the disorganized or mosaic pattern in cortical bone (arrows). **(d)** Photograph of an axially sectioned, whole-mounted tibia (H-E stain) shows thickening of the cortex with weak bone (pumice) lacking interconnection (between large straight arrows), compared with more normal underlying cortex (curved arrow).

row component), which we refer to as atrophic marrow, during the final inactive phase (4,22).

Distribution of Disease

Paget disease is predominantly located in the axial skeleton, with the most commonly affected sites being the pelvis (30%–75% of cases), spine (30%–75%), and skull (25%–65%) (3,4,6,23–30). Proximal long bones are also frequently involved, with the femur affected in 25%–35% of cases (2–5). Less commonly affected sites include the shoulder girdle and forearm (humerus, 31% of cases; scapula, 24%; and clavicle, 11%) (2–5). Involvement of other sites, including the ribs, fibula, bones of the hands and feet, calcaneus, patella, and tibial tubercle, is infrequent (2–5). Paget disease of the long bones typically begins in a subchondral location, with an advancing wedge of lucency estimated to progress at a rate of 1 mm per month (4).

Monostotic disease (10%–35% of cases) is more often seen in the axial skeleton, although any site can be the sole region of involvement (23–32). Polyostotic disease (65%–90%) is more frequent than monostotic disease, tends to have right-sided predominance, and usually involves lower extremities (3,4). Pelvic involvement is more often asymmetric than symmetric, and appendicular involvement is frequently unilateral (25).

Clinical Findings

Twenty percent of patients with Paget disease are asymptomatic initially (2–5,32). Skeletal symptoms include localized pain, tenderness, increased warmth (related to lesion hypervascularity), increased bone size, bowing deformities, kyphosis of the spine, and decreased range of motion. Neuromuscular symptoms can result from bone enlargement that encroaches on neural foramina or canals and leads to mechanical compression of neurogenic structures (particularly the cranial

Figure 2.   Lytic phase of Paget disease of the skull in different patients with osteoporosis circumscripta. **(a)** Lateral radiograph of a 50-year-old man shows a well-defined area of lysis in the frontal and occipital regions (arrowheads). **(b)** Lateral radiograph of a 60-year-old woman with osteoporosis reveals frontal and occipital areas of osteolysis (∗) that are more difficult to detect in this clinical setting. **(c)** Bone scan of the same patient as in **b** reveals intense uptake of radionuclide in the frontal and occipital areas, as well as in the face. **(d)** Axial CT scan of a 55-year-old man with mild expansion of the head reveals a lytic lesion with sharp borders (arrows) in the frontal bone. **(e)** Photograph of a whole-mounted, longitudinal section of the calvaria (H-E stain) shows calvarial expansion and extensive fibrovascular tissue in the diploic space (∗).
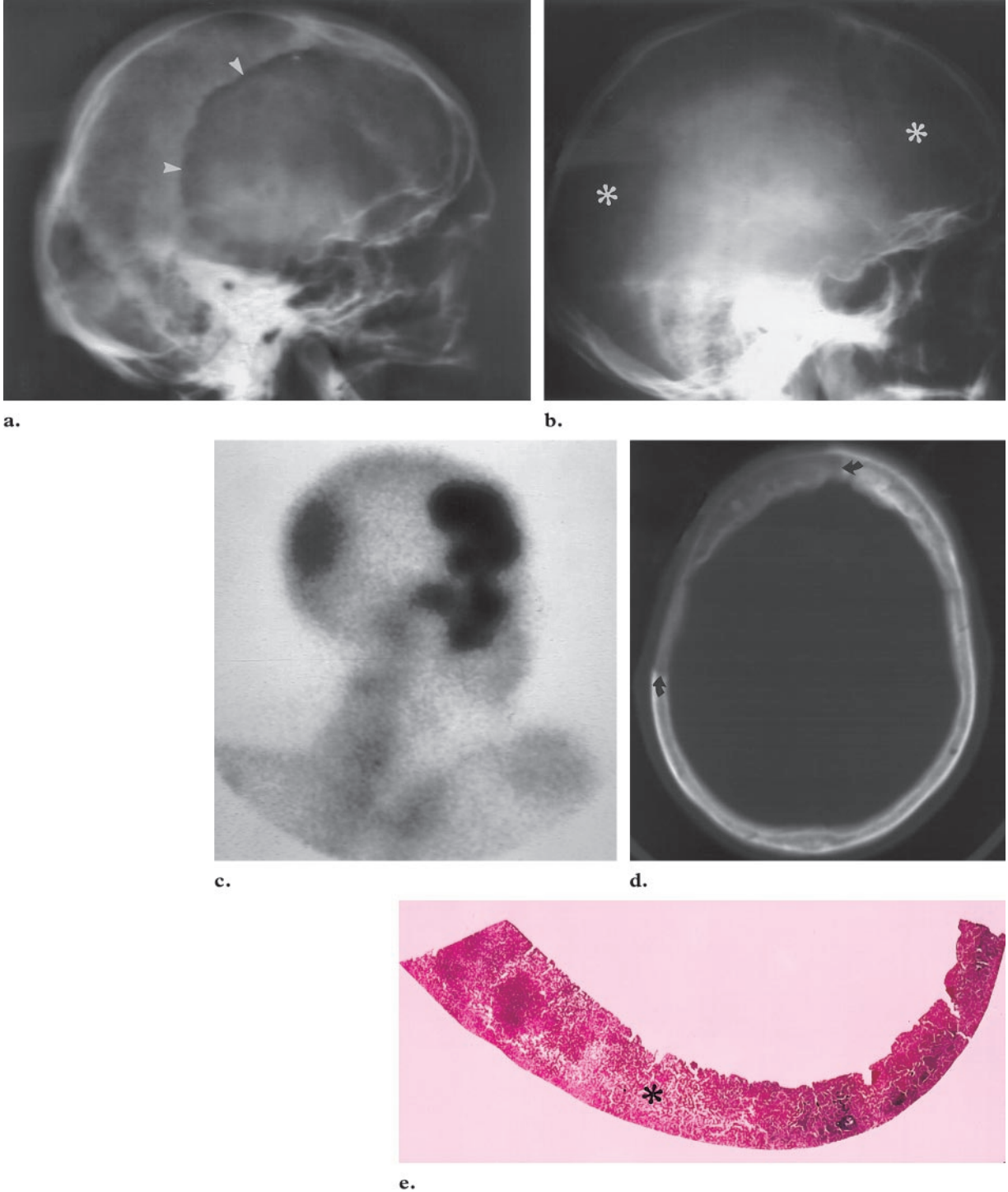

a.

b.

c.

d.

e.

Figure 3.   Lytic phase of Paget disease ("blade-of-grass" appearance) of the appendicular skeleton in different patients. **(a)** Anteroposterior radiograph of the knee in a 74-year-old man shows a large area of lysis beginning in subchondral bone with a sharp inferior margin (arrowheads). **(b)** Lateral radiograph of the upper tibia in a 41-year-old man shows a lytic lesion in the tibial tubercle, with blade-of-grass appearance superiorly and inferiorly (arrows). **(c)** Photograph of a sagittally sectioned, whole-mounted specimen (H-E stain) shows the intracortical, nonsubchondral location of the Paget disease (∗) and the sharp superior margin (arrow).

a.   b.   c.
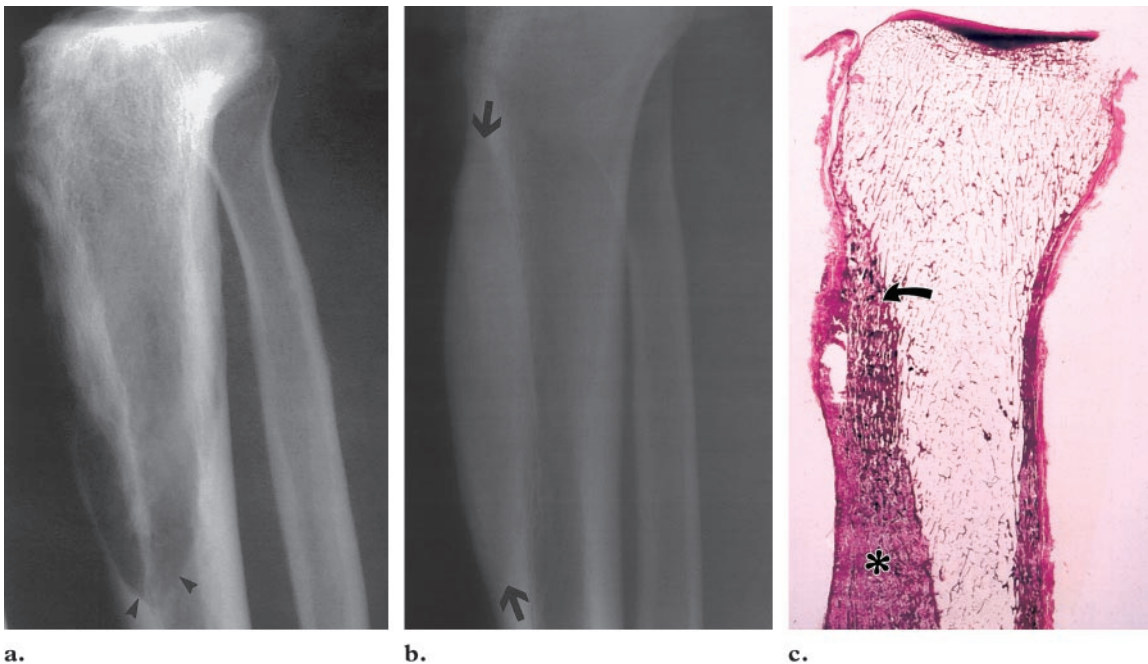
nerves), causing deafness, visual abnormalities, weakness, paralysis, and incontinence. Cardiovascular symptoms include high-output congestive heart failure, arterial calcification, and a rare report of Hashimoto thyroiditis (4,33). Recent studies have suggested a higher prevalence of aortic stenosis, heart block, and bundle branch block in severe cases of Paget disease (4,7).

Underlying osteoclastic and osteoblastic changes in bone are reflected in the patient's serum and urine laboratory values (4,5). Patients often have an elevated serum level of alkaline phosphatase (related to increased rate of bone formation), particularly during more reparative disease phases (mixed and blastic). Increased serum and urine levels of hydroxyproline are seen in the lytic phase (related to increased rate of bone resorption) and are an accurate marker of resorptive activity, even in monostotic disease. Serum levels of calcium and phosphate are usually normal, but 10% of patients may develop secondary hyperparathyroidism owing to hypercalcemia related to the aggressive bone remodeling (3,4).

## Imaging of Non-complicated Paget Disease

Radiography
Radiography is the mainstay of diagnosis in noncomplicated Paget disease. Most pathognomonic findings are easily depicted on radiographs alone and reflect the pathologic abnormalities.

Lytic Phase.—The early phase of Paget disease is characterized by osteolysis on radiographs, an appearance that reflects the unopposed osteoclastic activity seen pathologically. In the skull, osteolysis is frequently seen as well-defined, often large areas of radiolucency most commonly affecting the frontal and occipital bones; these areas are referred to as osteoporosis circumscripta or osteolysis circumscripta (24,34) (Fig 2). Both inner and outer calvarial tables are involved, with the former usually more extensively affected. This pattern is in contradistinction to that of fibrous dysplasia, which usually affects the outer table more prominently (35). There is a notable absence of peripheral sclerosis surrounding the calvarial osteolysis secondary to the lack of significant osteoblastic activity. These areas of lysis are usually easily identified on radiographs, although it may be more difficult in patients with osteoporosis and correlative bone scans are helpful (Fig 2b, 2c).

In the long bones, osteolysis begins as a subchondral area of lucency. The advancing wedge of osteolysis often demonstrates a characteristic sharp radiolucent margin without sclerosis likened to a blade of grass or flame (36) (Fig 3). In

Figure 4.   Mixed phase of Paget disease of the pelvis in different patients. **(a)** Anteroposterior radiograph of an 81-year-old woman shows extensive involvement by Paget disease with areas of cortical (iliopectineal and ilioischial lines, arrows) and trabecular (arrowheads) thickening throughout the pelvis and coxa varus deformity in the right hip. **(b)** Photograph of a coronally sectioned, whole-mounted femoral specimen (H-E stain) shows cortical (arrows) and trabecular (arrowheads) thickening, fibrovascular marrow (∗), and coxa varus deformity.

a.

b.
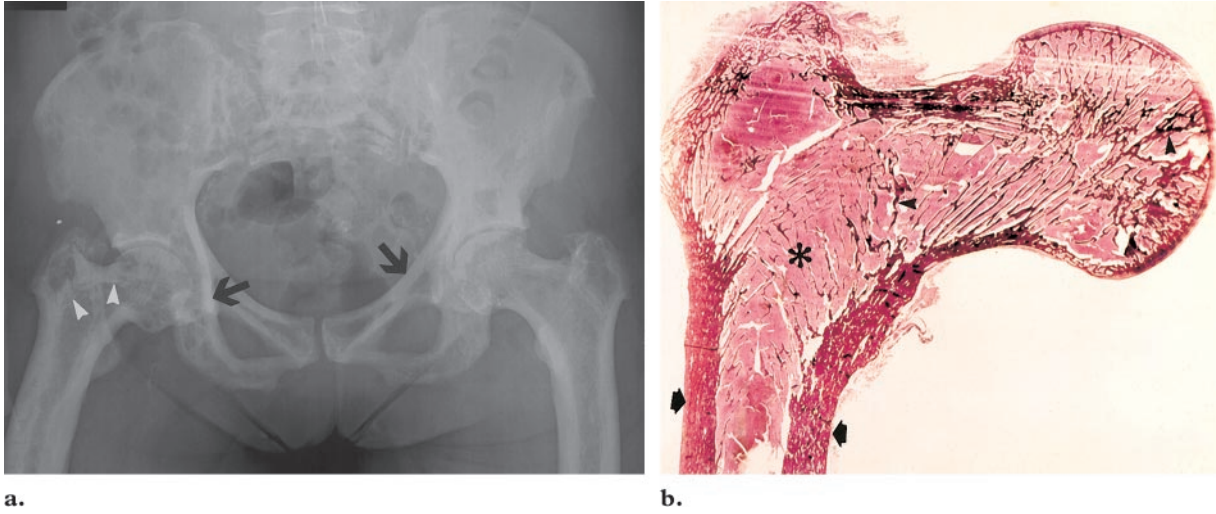
rare cases, the disease is isolated to the diaphysis, most commonly in the tibia, rather than subchondral bone, which can cause diagnostic confusion (29,37) (Fig 3b, 3c). However, in our experience, even these unusual cases often maintain the typical sharp margins that allow accurate diagnosis and differentiation from neoplastic disease (Fig 3b, 3c).

Early coarsening and prominence of the trabecular pattern of bone can be seen radiographically in the later stages of the incipient or active lytic phase, emphasizing the continuum with some overlap between phases.

Mixed Phase.—The vast majority of cases of Paget disease seen by radiologists are in the mixed phase. The characteristic manifestations seen radiographically are coarsening and thickening of the trabecular pattern and cortex. These findings reflect the underlying pathologic changes of osteoblastic repair and are usually pathognomonic on radiographs, particularly in long bones of either the upper or lower extremities (Fig 4). The trabecular thickening occurs primarily along the lines of stress, although disorganized areas are also seen (Fig 4a).

Paget disease of the pelvis usually manifests with cortical thickening and sclerosis of the iliopectineal and ischiopubic lines (5) (Fig 4). The iliac wing may also be involved. These findings are often asymmetric and more commonly seen on the right side. These manifestations are also often associated with enlargement of the pubic rami and ischium (2,3).

Paget disease of the spine frequently manifests with cortical thickening encasing the vertebral margins, which gives rise to the "picture frame" appearance on radiographs in mixed phase disease (4) (Fig 5). The osteoblastic activity is seen along all four margins of the vertebral body cortices, unlike the rugger jersey vertebrae in renal osteodystrophy, which only involves the superior and inferior endplates. Coarsening of the spinal trabeculae occurs, predominantly in a vertical direction. The vertical trabecular thickening pattern in Paget disease is coarser than the more delicate pattern seen in hemangiomas with which it can be confused. In addition, the condensation of trabeculae at the endplates coexisting with the picture frame appearance is not seen in spinal hemangiomas (Fig 5). Flattening or squaring of the normal concavity of the anterior margin of the vertebral body can be seen on the lateral spinal radiographs.

Blastic Phase.—Paget disease in the mixed phase may progress at a variable rate and extent to the blastic phase. In the long bones and pelvis, areas of sclerosis may develop and can be extensive, obliterating areas of previous trabecular thickening. Bone enlargement is particularly common in the blastic phase of Paget disease.

Figure 5.   Paget disease of the spine in different patients. **(a)** Anteroposterior radiograph of the lumbar spine in a 45-year-old man shows subtle vertical trabecular thickening (white arrowheads) and early picture frame appearance with condensation of trabeculae about the superior and inferior endplates (black arrowheads). **(b)** Lateral radiograph of the lumbar spine in a 54-year-old woman shows cortical thickening (picture frame appearance) about the entire vertebral body at all levels (arrowheads). **(c)** Sagittal CT reformatted image in a 50-year-old man shows similar trabecular thickening (arrowheads) with extension into the posterior vertebral elements (∗). **(d)** Photograph of a coronally sectioned, whole-mounted specimen (H-E stain) shows cortical thickening encasing the vertebral body at two levels (arrows) as the cause of the radiologic findings as opposed to the diffuse bone formation at the most superior level (∗), which could manifest as an ivory vertebral body (see Fig 7).
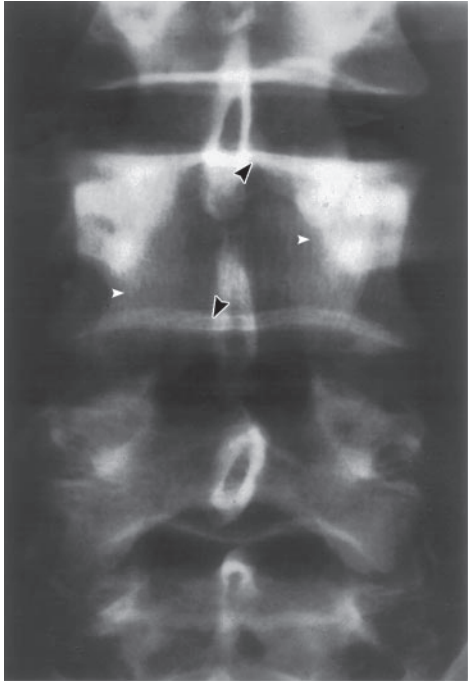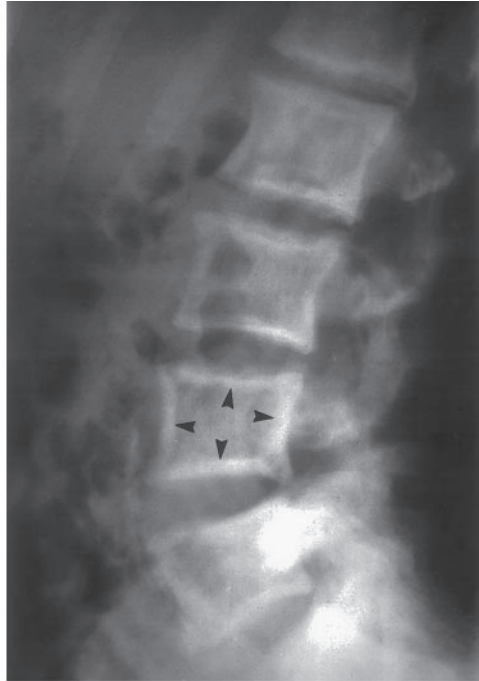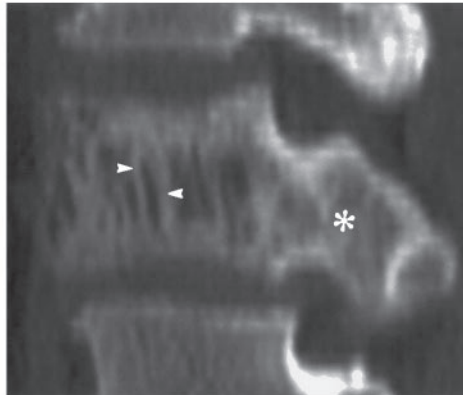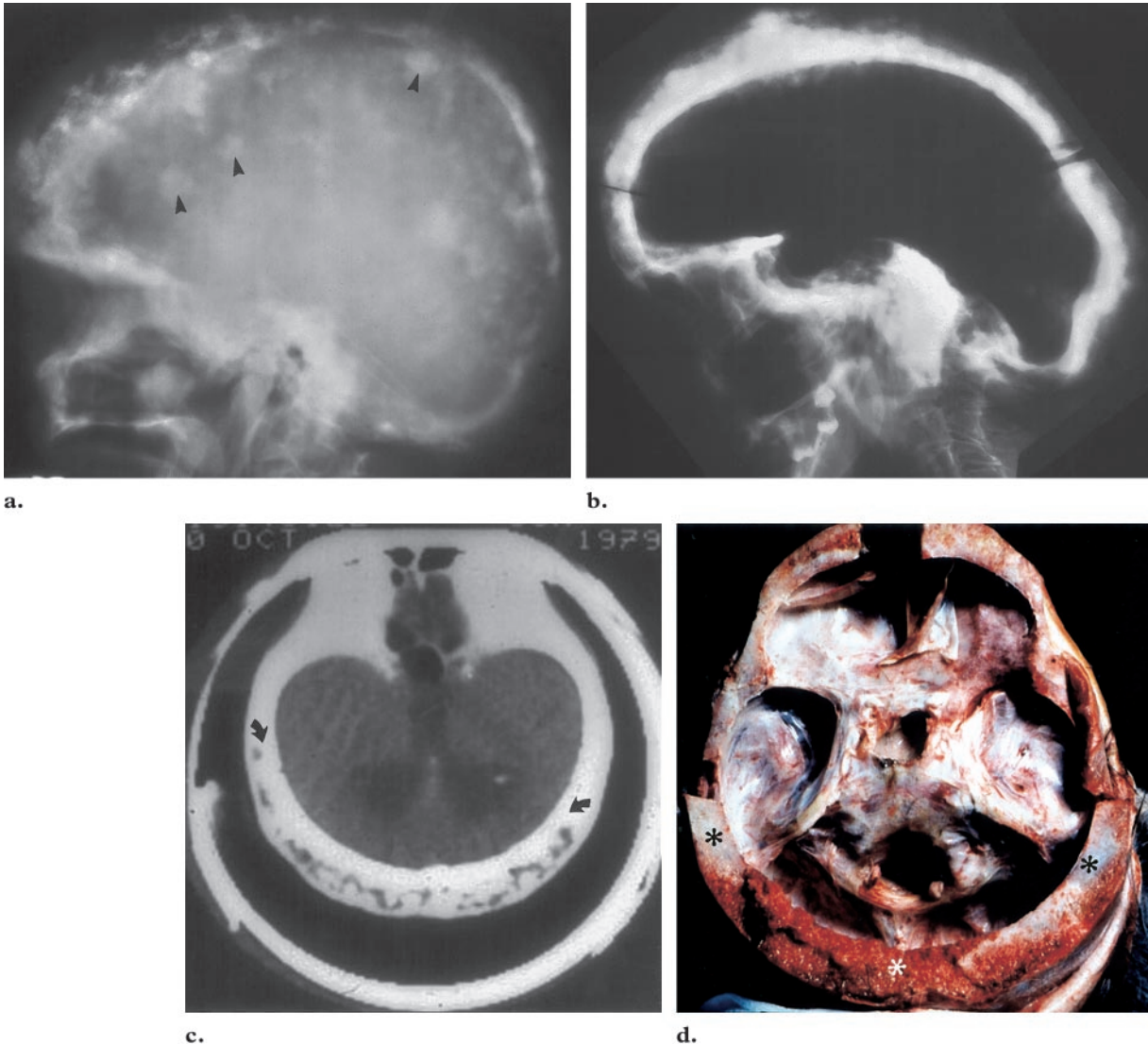
Figure 6.   Blastic phase of Paget disease involving the calvaria. **(a)** Lateral radiograph of the skull in a 56-year-old man shows diffuse calvarial thickening including the calvaria and maxillary sinus region, with several areas of focal sclerosis ("cotton wool" appearance) (arrowheads). **(b)** Lateral radiograph of a skull specimen from an 85-year-old woman reveals diffuse sclerosis and calvarial thickening with platybasia. **(c)** Axial CT scan (same case as **b**) also reveals the posterior thickening and mixed lysis and sclerosis (area between arrows). **(d)** Photograph of the sectioned calvaria viewed from above at autopsy (same case as **b** and **c**) also shows widening of the diploic space (white ∗) and sclerosis anteriorly (black ∗).

In the skull, the regions of sclerosis may become the predominant manifestation of Paget disease, with osteoblastic areas crossing sutures (Fig 6). This pattern may result in marked thickening of the diploic space, particularly the inner calvarial table, and has been referred to as a "tam-o'-shanter" skull, with resultant marked enlargement of the calvaria (Fig 6b). Initially, these areas of sclerosis may be circular and occur in previous areas of osteoporosis circumscripta (2–4). This pattern often creates focal areas of opacity that has been called the "cotton wool" appearance at radiography (2–4).

The blastic phase may cause the vertebral body to become diffusely sclerotic, creating an ivory vertebral body (38) (Fig 7). The posterior vertebral elements may also be affected. Bone enlargement of either the vertebral body alone or including the posterior elements is common and often aids in distinguishing Paget disease from other diseases. Spinal involvement may affect only one vertebral level, multiple levels, or even all the vertebral segments.

a.   b.

Figure 7. Ivory vertebral body caused by Paget disease in a 50-year-old asymptomatic man. **(a)** Anteroposterior radiograph shows diffuse sclerosis in the T10 vertebral body. **(b)** Posterior bone scan reveals intense radionuclide uptake at the involved vertebral level (see Fig 5d for histologic specimen of an ivory vertebral body).
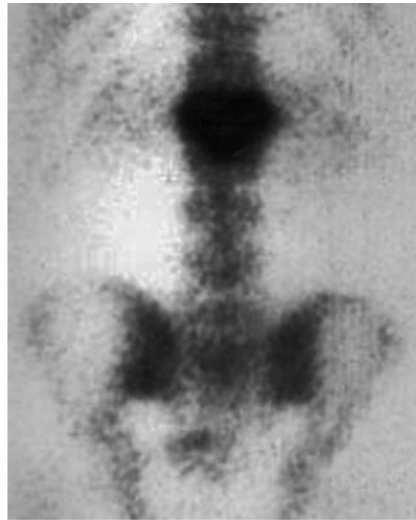
Figure 8. Anterior bone scan of the pelvis and proximal femora shows intense elongated areas of increased radionuclide uptake in the left hemipelvis and right femur, areas affected by Paget disease. Lateral bowing of the affected femur is seen.

The differential diagnosis of local vertebral body sclerosis includes osteoblastic metastatic disease (particularly from breast and prostate carcinoma), lymphoma, chordoma, and in rare cases unusual infection such as tuberculosis. Diffuse vertebral sclerosis has an extensive differential diagnosis, including osteoblastic metastatic disease, sickle cell disease, myelofibrosis, fluorosis, mastocytosis, and renal osteodystrophy.

Bone Scintigraphy

Bone scintigraphy (including blood flow, blood pool, and static images) classically demonstrates increased radionuclide uptake in the region of abnormal bone in all three phases of Paget disease (39–45) (Figs 2c, 7b, 8). It is a sensitive but not specific examination for detection of hyperemia and osteoblastic activity seen in Paget disease (40,41). The area of abnormal radionuclide uptake is usually elongated, reflecting the distribution of Paget disease, rather than the characteristic circular abnormality identified with metastatic disease or myeloma (Fig 8). Because scintigraphy is more sensitive to changes in vascularity, the hypervascular nature of Paget disease is often demonstrated as marked increased radionuclide uptake, which may be detected even before the typical radiographic lucency is evident (3). Scin-

tigraphy is most valuable in identifying the polyostotic distribution of the disease (42).

In our experience, there is a broader spectrum of radionuclide uptake in abnormal pagetic bone than has been reported, since not all cases are as "hot" as has been classically described. This concept is supported in studies by Lavender et al (44) and Khairi et al (45). In these studies, results from both bone scans and radiographs were abnormal in 56%–86%, were abnormal only on bone scans in 2%–23%, and were abnormal only on radiographs in 11%–20% of sites of pagetic involvement. It is likely that more quiescent late phase disease may show normal radionuclide activity yet appear abnormal on radiographs. Indeed, in the study of Khairi et al, all the patients with normal-appearing bone scans and abnormal radiographic findings were asymptomatic, whereas 73% of those with abnormal bone scan results and normal-appearing radiographs were symptomatic representing more physiologically active disease (45). It is important to recognize this spectrum of scintigraphic and radiographic disparity that can occur in Paget disease so as not to cause diagnostic confusion. The various complications associated with Paget disease cannot typically be differentiated with bone scintigraphy.

Figure 9.   Noncomplicated Paget disease found incidentally in an asymptomatic 58-year-old man. Axial CT scan of the pelvis (obtained for other reasons) shows cortical (large arrows) and trabecular (small arrows) thickening of the left iliac bone with maintained yellow marrow (∗).
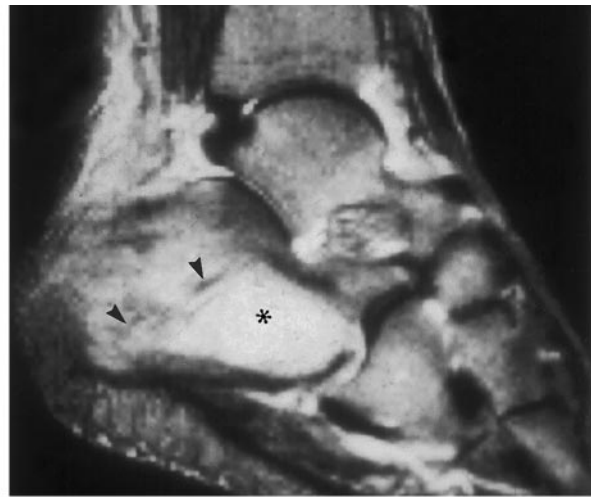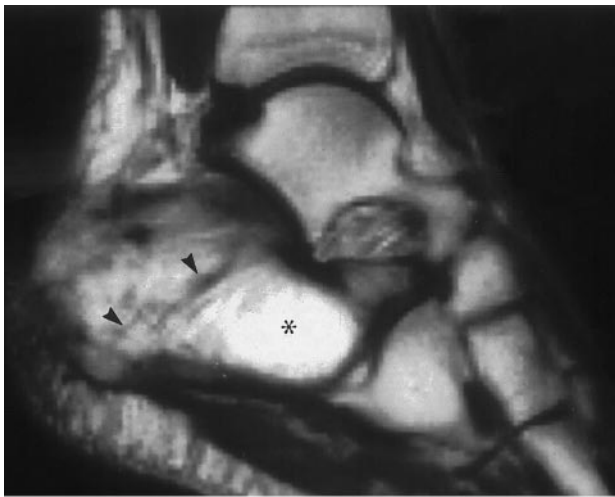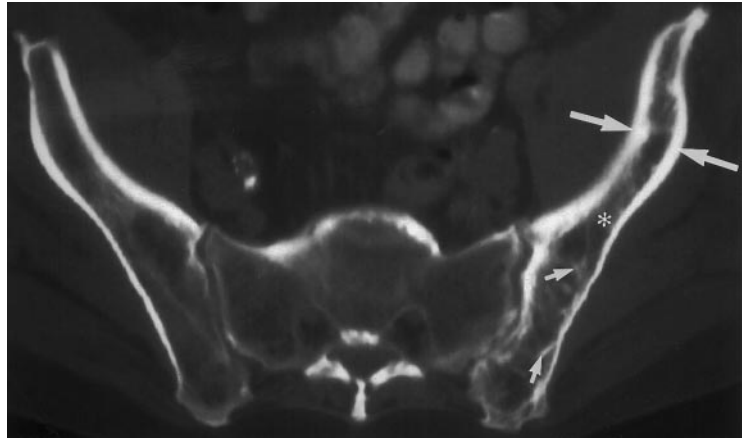
a.   b.

Figure 10.   Noncomplicated Paget disease of the calcaneus in a 58-year-old asymptomatic man. Sagittal T1-weighted (repetition time msec/echo time msec = 500/20) **(a)** and T2-weighted (2,000/80) **(b)** MR images show cortical and trabecular thickening (arrowheads) with maintained yellow marrow (∗). In fact, there is more fat in the affected calcaneal marrow, compared with that in the noninvolved talus.

CT and MR Imaging

Although Paget disease is usually apparent on radiographs, the disease may be discovered incidentally on CT or MR images obtained for other reasons (21,46–56). Familiarity with and the ability to recognize the typical appearance of this disease on CT or MR images is important so as to prevent misdiagnosis (particularly of metastatic disease in this age group of patients).

Findings at CT are largely identical to the radiographic findings of Paget disease previously discussed (49). Areas of lysis show loss of normal trabeculae (50). Cortical and trabecular thickening are also well demonstrated at CT (35) (Fig 9). The disorganized pattern of trabecular thickening seen pathologically is better demonstrated on CT scans than on radiographs. Areas of sclerosis may be seen in the blastic phase of the disease. The marrow space in Paget disease often reveals fat

attenuation. Noncomplicated Paget disease shows no evidence of cortical destruction or any soft-tissue mass.

Bone enlargement as well as trabecular and cortical thickening (which has low signal intensity with all MR pulse sequences) may be seen on CT and MR images; however, this pathognomonic appearance is easier to appreciate on radiographs. The MR imaging appearance of the remainder of the marrow in Paget disease is variable and depends on the disease phase and, more important, on the histologic composition of the marrow space as previously discussed (21). Three intrinsic patterns of the marrow space in noncomplicated Paget disease are noted on MR images. In the vast majority of these cases, the yellow marrow signal intensity is maintained regardless of pulse sequence, reflecting the fact that most disease is in the mixed phase and is relatively long-standing (51) (Figs 10, 11). In fact, in many

Figure 11.   Noncomplicated Paget disease of the distal femur in a 57-year-old woman. **(a)** Anteroposterior radiograph of the knee shows typical cortical and trabecular thickening of the entire distal femur to subchondral bone. **(b)** Coronal T1-weighted MR image (600/20) shows identical changes with maintained yellow marrow (∗) between thickened trabeculae. **(c)** Coronal fat-suppressed short-inversion-time inversion-recovery MR image (2,000/30/100) reveals mild increased signal intensity in the marrow resulting from small fibrovascular elements. **(d)** Photograph of a coronally sectioned, whole-mounted specimen (H-E stain) shows cortical and trabecular thickening (arrowheads) and yellow marrow centrally, with small components of fibrovascular tissue (∗).
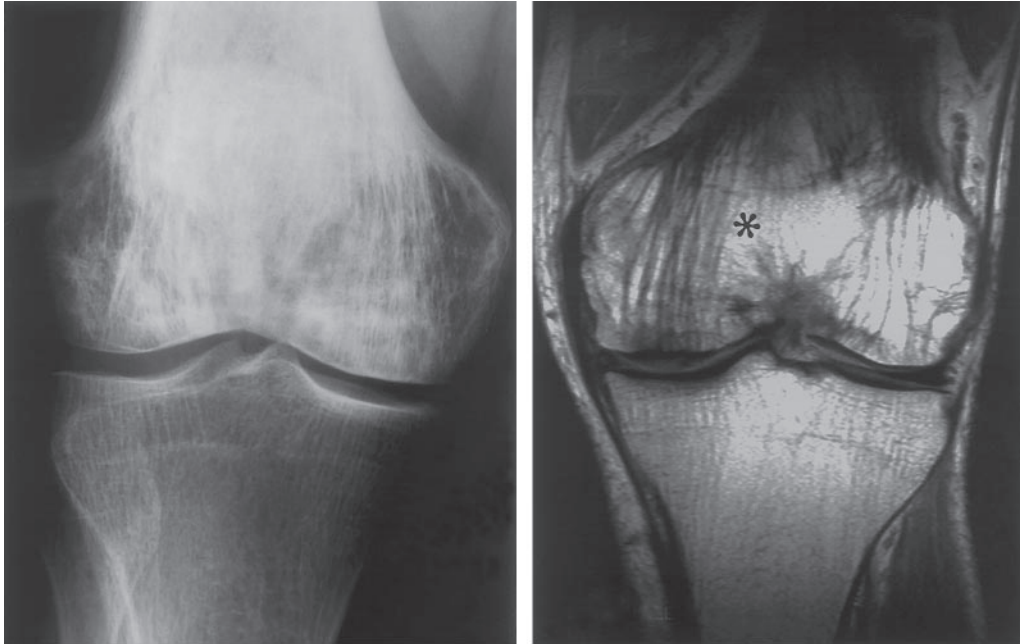
a.

b.
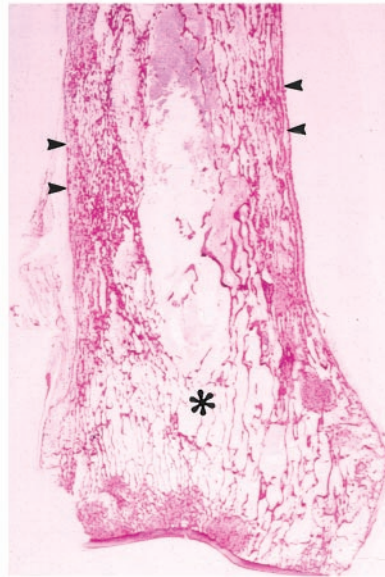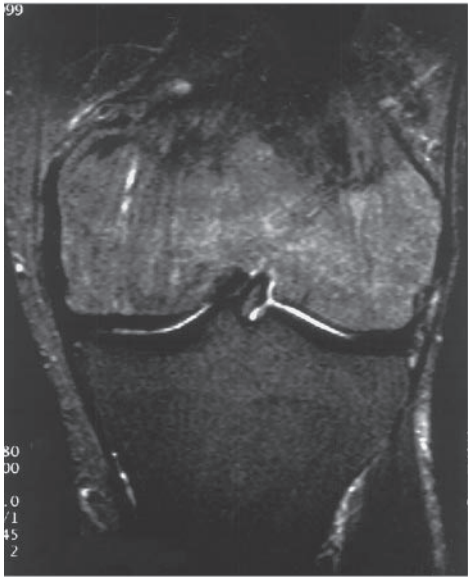
c.

d.

Figure 12.   Noncomplicated active Paget disease of the left distal tibia in an asymptomatic 45-year-old man with fibrovascular marrow. **(a)** Anteroposterior radiograph of the ankle shows typical cortical and trabecular thickening from Paget disease. **(b)** Coronal T1-weighted MR image (500/14) reveals patchy marrow replacement in the left distal tibia, although small foci of maintained yellow marrow are seen (arrows). **(c)** Axial fat-suppressed T1-weighted MR image (600/15) obtained after intravenous injection of gadolinium reveals patchy speckled enhancement in the marrow space (∗) and particularly prominent intracortical enhancement (arrowheads). **(d)** Axial fat-suppressed T2-weighted (5,333/84) MR image shows heterogeneous speckled high signal intensity in the marrow (∗) without cortical destruction or soft-tissue mass.
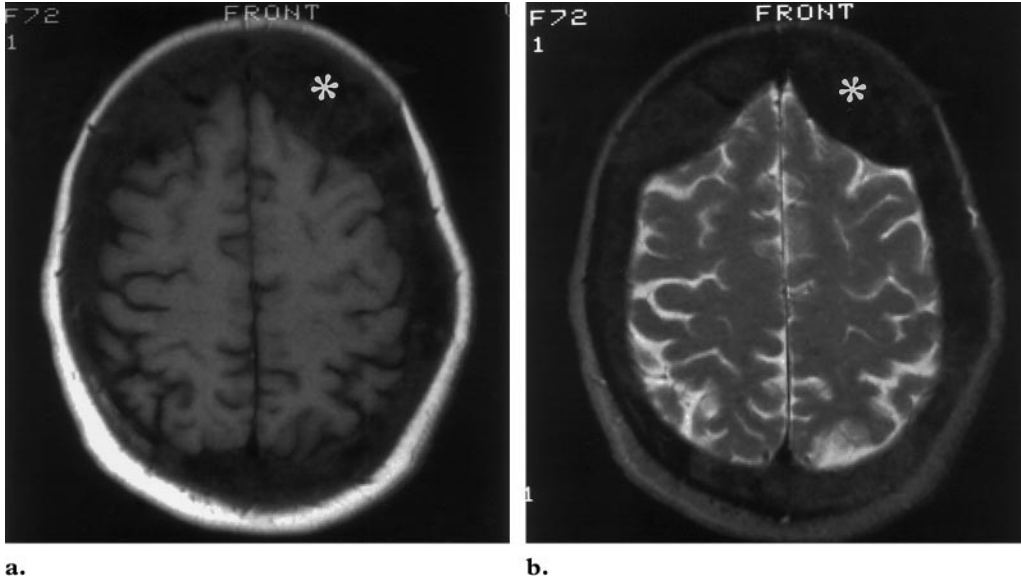

a.

b.

c.

d.

Figure 13.   Noncomplicated Paget disease of the skull in a 72-year-old woman. Axial T1-weighted (600/20) **(a)** and T2-weighted (4,000/75) **(b)** MR images of the head show diffuse calvarial thickening that remains low signal intensity with both pulse sequences (∗). Radiograph (not shown) revealed diffuse sclerosis.

a.   b.

cases, the marrow space of pagetic bone actually has more fat than found in the uninvolved bone, a finding that represents the atrophic marrow seen pathologically (Fig 10). The volume of the medullary canal can be decreased secondary to encroachment by cortical thickening.

The second MR imaging pattern is seen in the lytic to early mixed active phase, in which the marrow space has heterogeneous signal intensity with both T1- and T2-weighted sequences (Fig 12). On T1-weighted MR images, the marrow has decreased signal intensity, generally similar to that of muscle; however, it typically contains small to extensive foci of intermixed, normal and maintained yellow marrow. This feature is important because it excludes malignant transformation since no masslike marrow replacement is seen (48,55). On T2-weighted MR images, the marrow has heterogeneous high signal intensity, which is accentuated with water-sensitive pulse sequences (22,48,55). We believe this appearance corresponds to the fibrovascular marrow replacement seen pathologically in these more active phases of Paget disease. We refer to this pattern as the "speckled" appearance on MR images (Fig 12). Other investigators have suggested that dilated vascular channels in Paget disease may be responsible for the high-signal-intensity changes on T2-weighted images secondary to their slow flow state. In our experience, however, we have not recognized a serpentine appearance on MR images or large vascular structures pathologically.

The final MR imaging pattern is seen in the late blastic inactive phase, in which the marrow space has low signal intensity representing sclerosis regardless of pulse sequence (Fig 13).

The increased blood flow seen pathologically in Paget disease is reflected as increased enhancement, compared with the appearance of normal marrow, after intravenous administration of gadolinium contrast material. In our experience, the most prominent enhancement is often noted in more active disease and in the intracortical component, since there is more metabolic activity at this site. Intramedullary enhancement after gadolinium injection also often has a "speckled" pattern (Fig 12c).

Nonneoplastic Complications of Paget Disease

Common nonneoplastic complications include osseous weakening (resulting in deformity, bowing, and fractures), arthritis, and neurologic abnormalities (57–65). Miscellaneous entities such as osteomyelitis, extramedullary hematopoiesis, recrudescence of the lytic phase of the disease, and transference of Paget disease to a new osseous site following surgical placement of bone graft material harvested from pagetic areas are also rare, reported nonneoplastic complications (4,66,68).
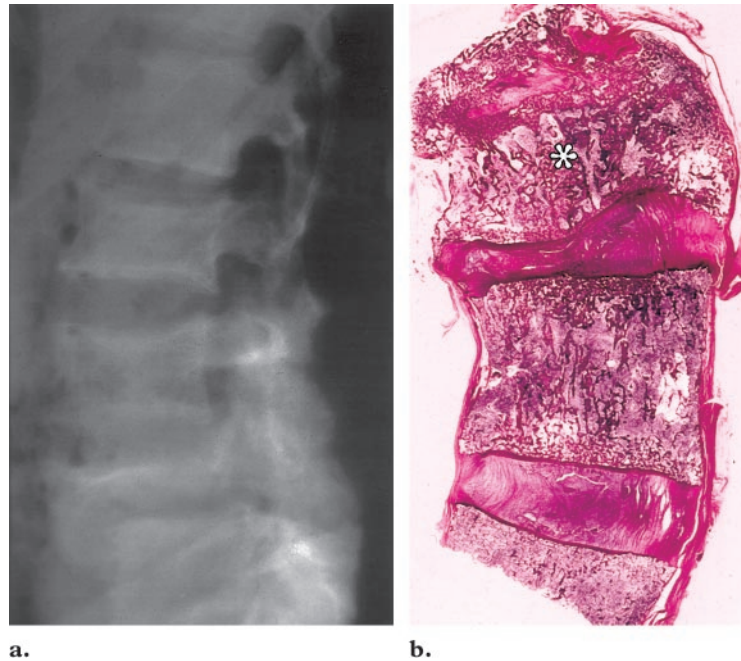
Figure 14. Paget disease of the spine with associated fracture in a 50-year-old woman. **(a)** Lateral radiograph of the lumbar spine shows the typical picture frame appearance of Paget disease with severe compression (vertebrae plana) at L4. **(b)** Photograph of a coronally sectioned, whole-mounted specimen (H-E stain) of the lumbar spine in a different patient with Paget disease shows compression fracture (∗) and scoliosis.

a.   b.

Osseous Weakening

The combination of progressive osteoclastic and osteoblastic activity gives rise to the dichotomy of osseous enlargement but weakening of the bone. Sequelae of this osseous weakening are the most common complications of Paget disease. Bowing of the appendicular skeleton is seen both clinically and radiologically. Anterior or lateral bowing of the tibiae or femora is particularly frequent (Figs 4, 8). Bowing in the spine may result in scoliosis, and in the pelvis, it may lead to protrusio acetabuli (particularly with pagetic involvement on both sides of the joint) (Fig 14).

Fractures are the most common complication of Paget disease and may cause symptoms that require orthopedic intervention (4,24,60,64) (Figs 14, 15). Initially, single or multiple, small, linear, cortical, lucent regions representing incomplete fractures may develop on the convex surface of the long bones (unlike the Looser zones in osteomalacia that occur on the concave side of bones). The radiolucent areas in Paget disease contain immature callus pathologically, as opposed to the nonmineralized osteoid seen in osteomalacia. These fractures may progress and become complete and are often referred to as "banana" fractures owing to their horizontal nature. Common sites of fractures associated with Paget disease include the femur, tibia, humerus, pelvis, and spine (2–4,24,60,64) (Figs 14, 15). Femoral fractures are the most common and frequently occur proximally in the subtrochanteric region (Fig 15). Fractures usually affect patients with longstanding disease and are more frequent in women (4).

Fractures associated with Paget disease have a higher prevalence of nonunion. Some authors have suggested that biopsy of these fractures should be performed to exclude secondary malignancy associated with Paget disease (4). However, we believe that true pathologic fractures have a much more aggressive radiologic appearance and that lesions with only linear lucent areas result from fracture alone and do not require biopsy.

Arthritis

Secondary osteoarthritis is common in longstanding Paget disease secondary to altered biomechanics across abnormal bone resulting from osseous enlargement and bowing (62,63,69). Hip and knee involvement are most common, although any joint with adjacent pagetic bone may develop this complication. The appearance of secondary osteoarthritis in Paget disease may be atypical because of the altered mechanics and alignment (Fig 16). This altered appearance of osteoarthritis associated with Paget disease most commonly affects the hip with axial narrowing related to preexisting protrusio acetabuli, rather than the typical superolateral narrowing pattern (Fig 16). This variation in the pattern of osteoarthritis of the hip in Paget disease must be recognized on radiographs so that an inflammatory arthritis cause is not suggested.

Monosodium urate deposition (gout), calcium pyrophosphate dihydrate crystal deposition disease, and rheumatoid arthritis have been associated with Paget disease (65,69). The latter two are still considered controversial and have only a very weak association at best (4,65,69). The association with gout is related to the high cell turn-
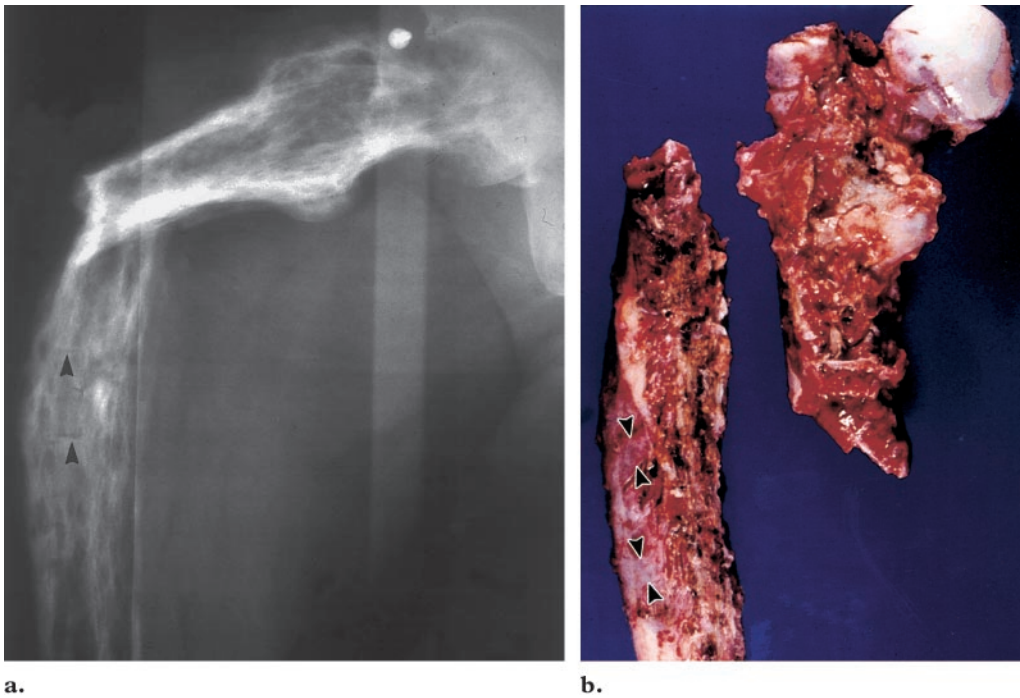
a.   **b.**

Figure 15.   Paget disease with both incomplete and complete fractures of the femur in a 70-year-old woman. **(a)** Anteroposterior radiograph of the femur shows pathognomonic changes of Paget disease: cortical and trabecular thickening extending to subchondral bone and subtrochanteric complete fracture. Additional incomplete fractures (arrowheads) are seen along the lateral convex femoral surface with callus formation. **(b)** Photograph of a partially sectioned gross specimen reveals the complete displaced fracture and the incomplete fractures with callus (between arrowheads).

Figure 16.   Paget disease of the hip with osteoarthritis. Anteroposterior radiograph of the left hemipelvis shows extensive Paget disease with osteoarthritis involving the hip. The axial pattern of narrowing of the hip (arrowheads) is unusual and is likely caused by the underlying osseous weakening and mild protrusio acetabuli.

over state in Paget disease resulting in hyperuricemia reported in up to 40% of patients in one study (69). However, the number of patients with gout and Paget disease is low, and the typical radiologic appearance is not modified by this association.

Neurologic Abnormalities

In patients with Paget disease of the spine or skull, several neurologic complications have been noted. The major complications in the spinal column include spinal stenosis and foraminal en-

croachment (secondary to osseous expansion of the vertebral bodies), fractures of the spine, basilar invagination, and spinal cord hypoxia (57–59,61). Calvarial enlargement can result in clinical complications related to cranial nerve compression, including impairment of smell, absence of pupillary reflexes, blindness, ptosis, facial pain, tinnitus, dysphagia, trigeminal neuralgia, sensory loss, hearing loss, and extraocular muscle palsies (7,61).

In the later stages of Paget disease, enlargement of involved vertebral bodies or posterior vertebral elements (either at single or multiple levels, with or without accelerated degenerative disease) can cause encroachment on the spinal canal or foraminal recesses, with resultant neurologic compression and vertebral pain. Although CT is best for assessing the overall enlargement of

Figure 17.   Transport of Paget disease from iliac crest bone graft donor site to engrafted proximal tibial area in a 64-year-old man. **(a)** Anteroposterior radiograph of the knee shows a depressed lateral tibial plateau fracture (arrow) sustained in a motor vehicle accident. **(b)** Postoperative anteroposterior radiograph shows bone graft and elevation of the depressed fragment fixed by a single lag screw. **(c, d)** Follow-up anteroposterior radiographs obtained 2 **(c)** and 3 **(d)** years later show progressive development of characteristic Paget disease with cortical (large arrow) and trabecular (small arrows) thickening and a sharply defined, blade-of-grass lucent area (arrowheads). **(e)** Radiograph of the pelvis reveals extensive Paget disease throughout the pelvis and proximal femora, including the bone graft donor site (∗) in the left iliac crest. No radiographs of the pelvis were obtained before surgery.

bone and degree of stenosis, MR imaging is more sensitive for evaluating the degree of spinal cord and nerve root encroachment (48,49,59). Boutin et al (48) noted that although Paget disease is diagnosed more economically with radiography, MR imaging is well suited for demonstrating the presence and extent of not only neoplastic but also nonneoplastic complications. The width of the vertebral canal is narrowest in the upper thoracic region, making it the most vulnerable site for spinal stenosis. Intermittent neurogenic claudication can occur in patients with involvement of the lumbar spine and compression of the cauda equina.

Spinal stenosis associated with Paget disease can result from bone expansion alone or in combination with fracture. Fractures of the involved vertebral column can result in fragment displacement and hemorrhage, which compromise the spinal canal or lateral recess, as well as associated kyphosis or scoliosis. Fractures of the spine in Paget disease can also cause severe diffuse loss of vertebral height leading to vertebrae plana.

a.   b.   c.

Figure 18.   Recurrent lytic phase of Paget disease after tibial tubercle avulsion with fixation and non-weight-bearing in a 75-year-old man. **(a)** Lateral radiograph of the lower leg shows an avulsion of the patellar tendon attachment at the tibial tubercle (black arrow). Changes of Paget disease, with trabecular and cortical thickening (white arrow), are subtle. **(b, c)** Follow-up anteroposterior radiographs obtained 2 weeks **(b)** and 2 months **(c)** after screw fixation, bracing, and non-weight-bearing reveal progressive osteolysis (white arrowheads) with apparent focal destruction of the medial cortex (open arrow). Changes of Paget disease with trabecular (black arrowheads) and cortical (curved arrow) thickening and sharp inferior margin of lysis (solid straight arrow) are now more apparent. **(d)** Photograph of the coronal gross specimen obtained at amputation (performed because of misdiagnosis of sarcomatous transformation) shows hemorrhagic marrow space (∗) without a mass or cortical destruction. (Case courtesy of Arthur H. Newberg, MD, New England Baptist Hospital, Boston, Mass.)

d.

Basilar invagination in which the vertebral column prolapses into the skull base with secondary stenosis at the level of the foramen magnum is an acquired abnormality reported in up to 30% of patients with Paget disease of the skull (4,57) (Fig 6). It is reported to occur more frequently in women and in those with more severe disease. Syringomyelia, obstructive hydrocephalus, and brain stem compression have also been reported in association with Paget disease (58). Although CT may be useful for assessment of cortical and trabecular detail, MR imaging is best suited for this evaluation, given its excellent contrast resolution and multiplanar technique that uses direct sagittal imaging of the cervical occipital junction.

An additional interesting potential cause of neurologic symptoms in patients with longstanding Paget disease of the spine is cord hypoxia.

The purported theory is that hyperemic pagetic bone with increased blood flow steals from the blood supply meant for the central spinal cord, causing hypoxia. As with other thecal sac disease, MR imaging can be useful for assessing signal-intensity changes or degree of enlargement of the spinal cord.

Miscellaneous

Secondary osteomyelitis, extramedullary hematopoiesis, and transfer of pagetic bone to initially uninvolved bone following bone graft surgery are also reported in rare cases (Fig 17). A recrudescence of the lytic phase has also been described, simulating malignancy, particularly after trauma or following restricted weight-bearing for treatment of secondary fractures in Paget disease (68) (Fig 18).

RadioGraphics

Figure 19.   Sarcomatous transformation of Paget disease in the tibia to malignant fibrous histiocytoma in a 75-year-old woman. **(a)** Lateral radiograph of the tibia shows typical changes of Paget disease, with cortical and trabecular thickening involving the entire bone and anterior bowing. Focal destruction of the anterior cortex is seen superiorly, with a soft-tissue mass (arrowhead). **(b)** T1-weighted (500/20) MR image shows focal marrow replacement at the proximal tibial site of malignant transformation with cortical destruction and associated soft-tissue mass (∗). Yellow marrow signal intensity is seen throughout the remainder of the tibia. **(c)** T1-weighted (500/20) MR image obtained with gadolinium shows marked enhancement. T2-weighted MR image (not shown) revealed similar high signal intensity in the sarcoma. **(d)** Photograph of the sagittally sectioned gross specimen reveals the margins of the soft-tissue component of the sarcoma (∗). *f* = femur, *t* = tibia.
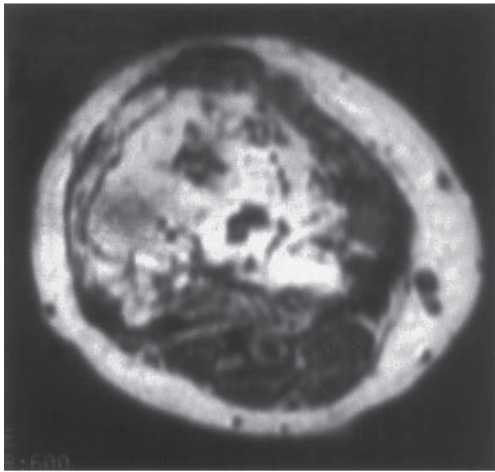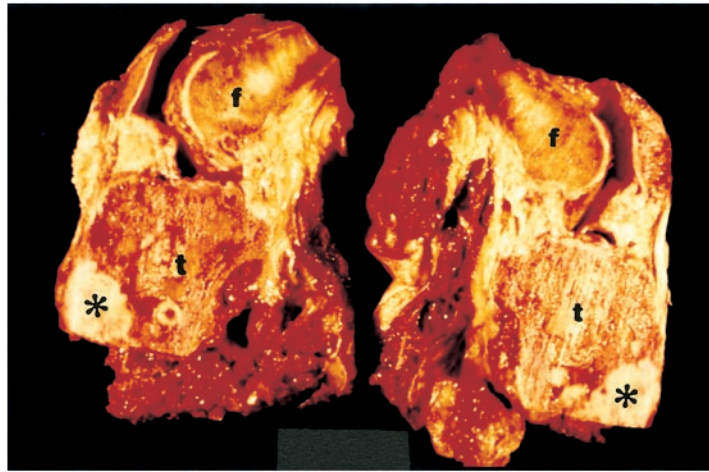
Neoplastic Complications of Paget Disease

Neoplastic complications associated with Paget disease are rare but include sarcomatous transformation (to osteosarcoma, malignant fibrous histiocytoma/fibrosarcoma, chondrosarcoma), other tumors such as giant cell tumors (benign and malignant) and superimposed metastatic disease, multiple myeloma, leukemia, and lymphoma (70–102) (Figs 19–21).

Neoplastic complications of Paget disease are relatively rare. Sarcomatous degeneration has been estimated to occur in 1% of patients with longstanding disease, although others believe it is far less common (4,73,74,83). Patients with severe polyostotic disease have an increased risk of
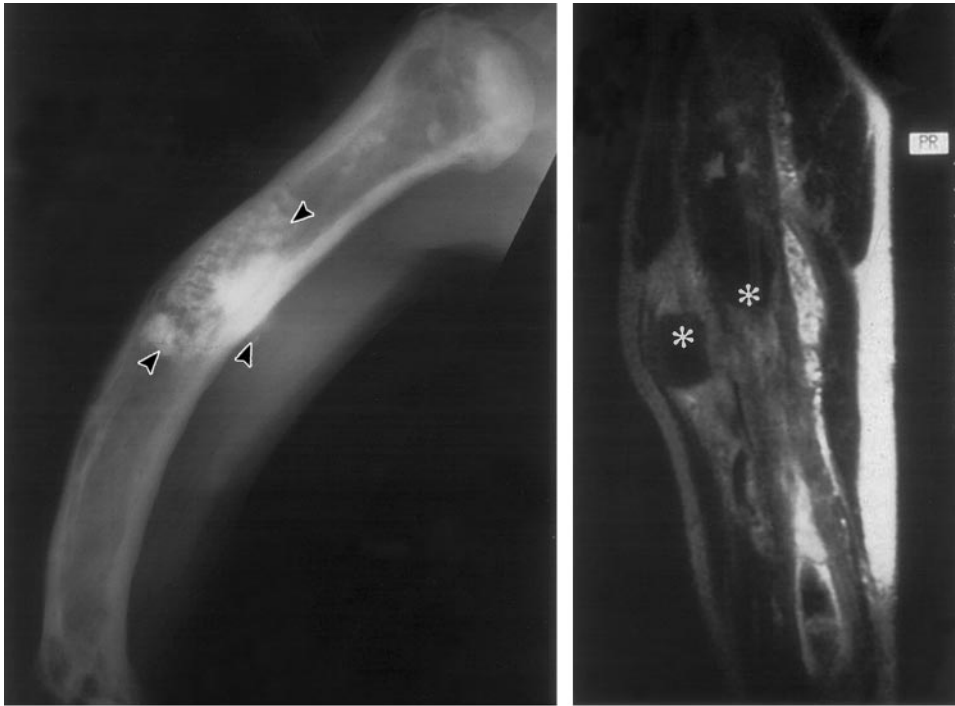
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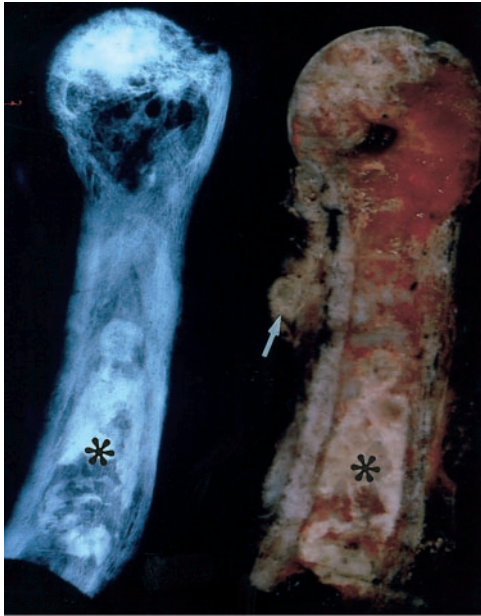
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Figure 20. Sarcomatous transformation to osteosarcoma of Paget disease of the humerus in a 77-year-old woman. **(a)** Anteroposterior radiograph of the humerus shows typical cortical and trabecular thickening from Paget disease involving the entire humerus and lateral bowing. Area of medullary sclerosis with mineralization in the soft tissue is also seen (arrowheads). **(b)** Sagittal T2-weighted (3,500/105) MR image reveals focal marrow replacement and associated soft-tissue mass (∗). **(c)** Anteroposterior radiograph and photograph of the coronally sectioned gross specimen show the osteosarcoma (∗) in the humeral midshaft and adjacent soft tissues (arrow).

sarcomatous transformation of up to 5%–10% of cases (2–7). Men are more commonly affected (2:1 ratio), and patients are usually between 55 and 80 years of age (2–7). New focal pain and swelling are the most common clinical symptoms that may portend this dire sequela, followed by pathologic fracture. The most frequently affected locations include bones about the hip, pelvis, and shoulder. The proximal humerus has a much higher prevalence than expected in proportion to the frequency distribution of uncomplicated Paget disease. In contrast, the skull and spine have a much lower prevalence than expected from the distribution of disease (4,7).

Osteosarcoma (50%–60% of cases) followed by malignant fibrous histiocytoma/fibrosarcoma

Figure 21. GCT of the face associated with Paget disease.
(a) Axial CT scan shows characteristic thickening and sclerosis of the diploic space (arrows) caused by Paget disease. Expansile mass involving the maxillary sinus (∗) represents the associated GCT. **(b, c)** Coronal T1-weighted (500/20) **(b)** and T2-weighted (2,500/80) **(c)** MR images obtained after intravenous injection of gadolinium also reveal Paget disease with calvarial thickening and maintained yellow marrow signal intensity (arrowheads). The maxillary GCT shows focal expansion, intermediate signal intensity with both pulse sequences, and mild diffuse enhancement (∗).
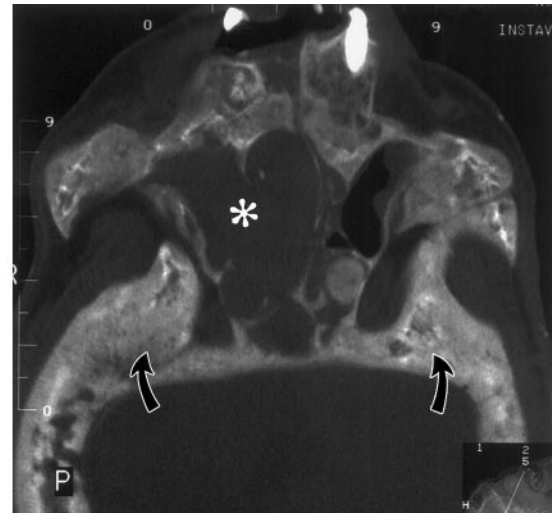
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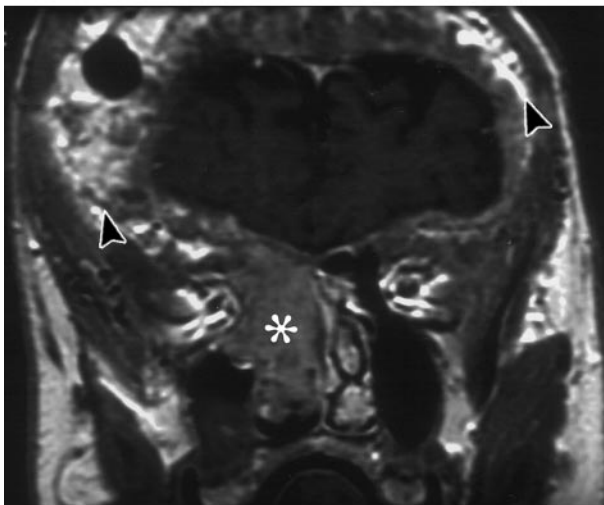
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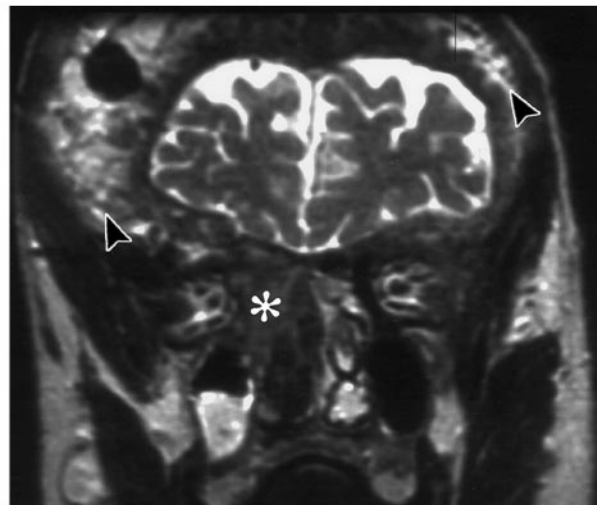
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(20%–25%), and chondrosarcoma (10%) are the most common histologic lesions associated with sarcomatous transformation (4,73,83). Lymphoma and angiosarcoma have also been reported in association with Paget disease but are very rare, representing only 1%–3% of reported cases (71,89,100). Prognosis for patients with sarcomatous transformation is poor, with over 90% of patients dying within 3 years of diagnosis despite the use of neoadjuvant chemotherapy and aggressive surgical resection (84). This poor prognosis reflects the high degree of anaplasia seen pathologically in these secondary malignancies and accounts for why we do not believe distinction of the histologic type of sarcoma is important from a clinical or treatment perspective. Metastatic disease is frequent and most commonly affects the lung.

Radiographic hallmarks of sarcomatous degeneration include aggressive osseous lysis, cortical destruction, and the presence of a soft-tissue mass (2–7,24,70–102) (Figs 19, 20). The majority of malignancies associated with Paget disease (including osteosarcoma, which is usually blastic when not associated with Paget disease) are largely lytic, an appearance that reflects the high degree of anaplasia in sarcomatous transformation, although matrix mineralization is occasionally prominent (Fig 20). Periosteal reaction, an important radiographic indicator of bone sarcoma in younger patients, is often not present in these sarcomatous lesions in older patients with Paget disease because the rapidity of bone destruction does not allow for significant osseous repair (2–7). On bone scans, sarcomatous transformation is suggested by photopenic foci in areas of increased activity from Paget disease, findings that also reflect rapid bone destruction. Distinction of sarcomatous transformation from manifestations of lytic components of Paget disease alone can be
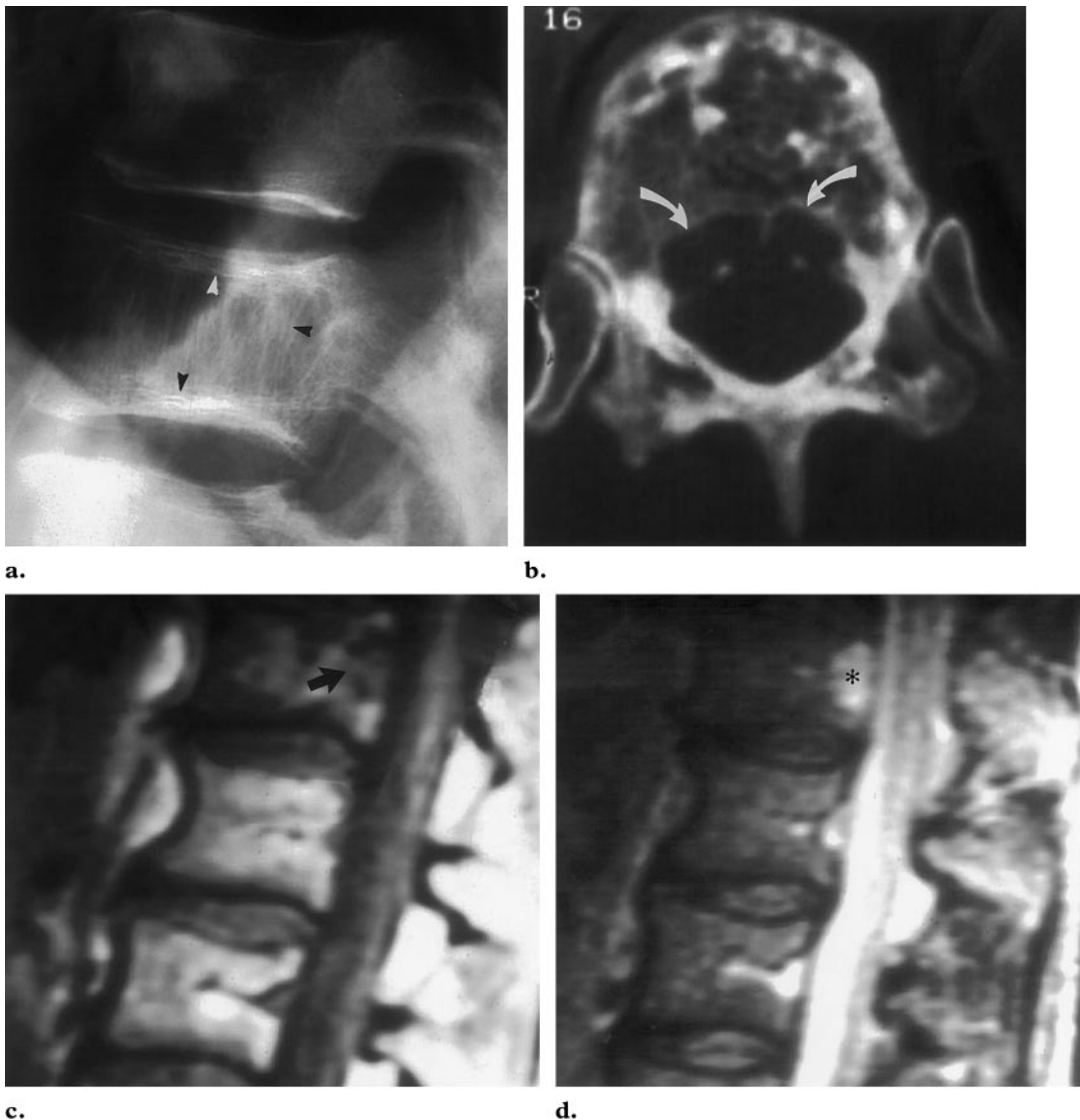
RadioGraphics

a.

b.

c.

d.

Figure 22.   Mixed lytic and blastic Paget disease of the spine simulating malignant transformation in a 60-year-old man. **(a)** Lateral radiograph of the thoracic spine shows typical picture frame appearance of Paget disease with cortical and trabecular thickening (arrowheads). **(b)** Axial CT scan reveals similar changes with a focal area of lysis posteriorly (arrows) that could be caused by lysis from Paget disease or malignant transformation. **(c)** Sagittal T1-weighted (600/25) MR image shows patchy areas of maintained yellow marrow signal intensity in the area of concern posteriorly (arrow), a finding that excludes malignant transformation. **(d)** Sagittal T2-weighted (2,500/90) MR image reveals high signal intensity in the lytic focus posteriorly (∗), but there is no epidural soft-tissue mass. This area remained unchanged for 5 years and represents a lytic component of Paget disease.

difficult (Fig 22). Comparison with previous radiographs is often helpful to detect new areas of aggressive bone lysis from sarcomatous transformation, particularly in a patient with a previously stable radiographic appearance of Paget disease. However, cases in which malignant transformation is a diagnostic concern require CT or MR imaging for further evaluation. As with other neoplastic processes, MR imaging is usually superior to CT owing to its improved contrast resolution and multiplanar capabilities. Malignant transformation is typically quite apparent on either CT or MR images. Masslike marrow replacement and cortical destruction are easily seen, and associated soft-tissue masses are almost invariably present and are often quite large and infiltrative (Figs 19, 20). The intrinsic imaging characteristics of sarcomatous transformation are typically nonspecific, and the attenuation of affected areas is similar to that of muscle on CT scans. Subtle areas of matrix mineralization are most easily detected with CT. Sarcomatous transformation shows intermediate signal intensity on T1-weighted images, high signal intensity on T2-weighted

images, and enhancement following intravenous administration of contrast material. Central necrosis is commonly seen on both CT and MR images. MR imaging also enables accurate lesion staging and can be helpful to direct biopsy away from necrotic or hemorrhagic regions.

Giant cell tumor (GCT) is another neoplasm that is associated with Paget disease in rare cases and can be solitary or multiple (up to 27 lesions in one patient) (76–79,87,95). Conventional GCTs (ie, those unrelated to Paget disease) are approximately 50–100 times more common than GCTs associated with Paget disease (76–79). GCTs involving the skull or facial bones (unlike the distribution of conventional GCTs, which most commonly occur at the ends of long bones) are almost invariably associated with Paget disease (Fig 21). The age of patients with GCT associated with Paget disease is much higher (range, 32–85 years; mean, 61 years) than the typical age of patients with conventional GCT (range, 20–50 years) (75–79,87). There is a male predilection (1.6:1 ratio) (75,76). Although GCT is most commonly associated with the polyostotic form of Paget disease (91% of cases), rare reports of GCT in monostotic Paget disease (9% of cases) are noted (75,76). The patients generally have a longstanding diagnosis of Paget disease (average, 12 years; range, 1–30 years) before the development of the GCT (76–79).

Several researchers have noted an interesting demographic similarity in patients with multifocal GCT and Paget disease (75,76,87). These patients were often born and raised in northern New Jersey and had ancestry that could be traced back to the Avellino, Italy area (a small village near Naples), a history that suggests the diseases have a familial relationship. Solitary and multifocal GCTs associated with Paget disease often have an indolent clinical behavior. These lesions only rarely lead to patient demise and generally do not have metastatic potential, with the exception of one recent report by Leonard et al in which a patient with a malignant GCT of the cranium associated with monostotic Paget disease died after developing pulmonary metastatic disease (88). This more indolent clinical behavior has treatment implications that differ from those of conventional GCT in that a good response has been seen with systemic steroids alone in some patients. Long-term clinical and imaging evaluation is necessary to identify new lesions (may occur over long intervals) and any aggressive features that may alter therapy.

Radiologically, solitary GCT associated with Paget disease is a lytic lesion typically without periosteal reaction and usually without an associated soft-tissue mass (75,76). Evidence of underlying Paget disease is always apparent usually both radiographically and pathologically, although in rare cases it is seen only at histologic evaluation. Interestingly, in patients with multiple GCTs and Paget disease, an associated soft-tissue mass is more frequent (76). Similar to the CT and MR imaging appearance of sarcomatous transformation, CT or MR imaging of GCT demonstrates masslike marrow replacement, which allows it to be distinguished from an abnormality that represents only the lytic phase of Paget disease (55,77). Although in our experience the lesion margins of GCT are often better defined than those seen in sarcomatous transformation and they may suggest GCT, it is usually not possible to adequately differentiate a GCT from sarcomatous transformation at imaging evaluation and biopsy is required. In our experience, the solid portions of GCT associated with Paget disease usually have heterogeneous low to intermediate signal intensity on T1-weighted and T2-weighted MR images, secondary to their high cellularity and fibrous content (Fig 21). Cystic and hemorrhagic regions may also be present. Contrast-enhanced T1-weighted MR images of GCT associated with Paget disease show a diffuse enhancement of the solid areas of the tumor.

Other neoplastic conditions that may coexist with Paget disease include metastatic disease, lymphoma, leukemia, or multiple myeloma, although this association may be fortuitous (2–7,71,72,89,97). The differentiation of these neoplastic conditions from sarcomatous transformation is difficult unless they involve bone not affected by the Paget disease. The prevalence of metastatic disease in bone affected by Paget disease is controversial (4,72,97). Some investigators have found that metastatic deposits more often involve bone affected by Paget disease, and they speculate that the hematogenous spread is more likely to occur in this hyperemic bone. Other researchers disagree and observe that metastatic disease is more frequent to affect bone not involved by Paget disease. Clinical history is often helpful if the patient has a known history of a primary tumor. The presence of multiple lesions (lytic or blastic) in sites not affected by Paget disease suggests metastases or myelomatous, leukematous, or lymphomatous involvement rather than sarcomatous transformation.

In rare cases, a soft-tissue mass arising from pagetic bone can be detected that is not neoplastic, either malignant or benign (70,103–105).

RadioGraphics

Such a pseudomass is believed to represent the lifting of the periosteal membrane by active Paget disease in subjacent bone. In the report by Tins et al of two patients with pseudosarcoma in Paget disease, neither case showed aggressive bone lysis on radiographs and MR images revealed maintained areas of yellow marrow without replacement in both cases (70). These features provide convincing evidence against the true diagnosis of sarcomatous transformation.

Treatment

Multiple agents have been used to treat Paget disease including calcitonin, biphosphonates, mithramycin, and gallium nitrate (2–7,106–111). The goal of therapy is the control, reduction, and alleviation of pain, rather than the return to normal bone. These medications all act through the inhibition of bone resorption and are quite successful in relieving pain. Oral biphosphonates are currently the most frequently used medication for Paget disease and have been employed since 1971, when etidronate was first proposed for clinical use (109–111). Etidronate decreases both bone resorption and formation in patients with uncomplicated Paget disease (109–111). However, its effect on osseous resorption precedes bone formation, and it ultimately inhibits normal skeletal mineralization, leading to a clinical and histologic picture of focal osteomalacia. Complications such as an increased risk of fractures including vertebral collapse have been reported in long-term use of etidronate, and long-term use may require radiologic evaluation (109–111). Calcitonin was one of the earliest medications employed in the treatment of Paget disease, and it has been used alone or in combination with biphosphonates (106–111). Calcitonin directly inhibits the osteoclastic activity of bone resorption and typically rapidly relieves pain within several weeks of the initiation of therapy. Mithramycin is a cytotoxic antibiotic that is typically used only in recalcitrant cases (4). Although mithramycin is effective in relieving pain, its toxicity severely restricts its use.

The treatment of Paget disease may improve the radiographic appearance, although these changes are usually quite subtle and often difficult to perceive, even with previous images for comparison (4,106–111). This is most apparent with calcitonin and mithramycin and in the lytic phase of the disease with ensuing sclerosis. Additional radiographic improvements associated with treatment include a return to normal bone size, shape, and cortical thickness and distinctness. In the past, bone scintigraphy was employed to document the positive effects of therapy, which was defined as progressive reduction in the degree of radionuclide uptake during treatment. However, this method of evaluation is no longer used because of its high cost, compared with the relatively inexpensive follow-up of laboratory (alkaline phosphatase and hydroxyproline) or clinical (pain) parameters.

Summary

Paget disease of bone represents a common disorder affecting 3%–4% of the population over the age of 40 years. Paget disease demonstrates a wide spectrum of radiologic patterns and appearances related to the phase of disease seen pathologically. Radiography is sufficient for diagnosis in the majority of cases and typically reveals pathognomonic changes in the lytic phase of Paget disease, with sharply defined areas of osteolysis in the skull or beginning in subchondral bone of the appendicular skeleton; in the mixed phase, with characteristic trabecular and cortical thickening and bone enlargement; and in the blastic phase, with areas of sclerosis. These manifestations correlate to the overall degree of remodeling of bone, which reflects the underlying extent of osteoclastic versus osteoblastic activity seen histologically. It is important to recognize uncomplicated Paget disease at CT or MR imaging because of the frequency with which these examinations are performed and so as not to cause diagnostic confusion. Bone enlargement as well as trabecular and cortical thickening are also seen on CT and MR images, with the additional frequent finding of maintained yellow marrow in noncomplicated Paget disease. Complications of Paget disease include the effects of osseous weakening (deformity and fracture), arthritis, neurologic symptoms, and neoplastic transformation or involvement. CT and MR imaging have proved particularly useful in differentiating the most feared complication of Paget disease, sarcomatous transformation, which occurs in approximately 1% of these patients. Sarcomatous transformation is heralded by findings of masslike replacement of the marrow space, cortical destruction, and an associated soft-tissue mass on CT or MR images. Recognition of the radiologic spectrum of the appearances of Paget disease usually allows prospective diagnosis and differentiation of its associated complications, helping to guide therapy and improve patient management.

RadioGraphics

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