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The Other Arthritides

Roentgenologic Features of Osteoarthritis, Erosive Osteoarthritis, Ankylosing Spondylitis, Psoriatic Arthritis, Reiter's Disease, Multicentric Reticulohistiocytosis, and Progressive Systemic Sclerosis

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OSTEOARTHRITIS

Osteoarthritis is a disorder of movable (diarthrodial) joints characterized by deterioration and abrasion of articular cartilage and by formation of new bone at joint surfaces and margins. One form of osteoarthritis, primary generalized osteoarthritis, is characterized by involvement of multiple joints, with a familial incidence and a particular susceptibility of females (10 to 1). Heberden's nodes of the distal interphalangeal joints and Bouchard's nodes of the proximal interphalangeal joints of the hand are often present. The early stages of the disease are often accompanied by an inflammatory response of the synovium and adjacent soft tissues.^{10, 11} Another form of osteoarthritis, secondary osteoarthritis, features joint degeneration secondary to articular cartilage damage from trauma, metabolic disorders, prior inflammation, acquired or congenital malformations, or abnormal stress.

Since the anatomic changes of osteoarthritis are generally irreversible, their increasing severity and prevalence with advancing age are not surprising. In primary osteoarthritis with Heberden's nodes, seen mainly in women, spontaneous pain is experienced at an early stage, and is worse at night and after use of the

extremity.⁹ In time, as the cartilage and ligaments become ossified, the pain subsides. The excessive ossification results in an enlarged joint with a limited range of motion, but only moderate functional disability. The distribution of joint involvement, in order of decreasing frequency, is another characteristic feature: distal interphalangeal, first carpometacarpal, acromioclavicular, first metatarsophalangeal, first tarsometatarsal, trapezioscapoid, knee joints, and apophyseal joints of the spine.

An erosive form of osteoarthritis has been described in which destructive changes of the proximal and distal interphalangeal joints are characteristic.¹⁸ The onset of arthritic symptoms in middle aged women is usually abrupt, with associated development of Heberden's nodes. Many features are similar to those of primary generalized osteoarthritis. The erosions tend to be limited to the interphalangeal and first carpometacarpal joints of the hand.

In primary generalized osteoarthritis, marginal osteophytes are produced by cartilaginous metaplasia at the attachment of the capsule, tendons, or ligaments. Endochondral ossification follows, which extends into the supporting soft tissue structures. In addition, characteristic extracapsular osteochondromas form in the soft tissues adjacent to Heberden's or Bouchard's

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nodes, or adjacent to the arthritic changes of the first carpometacarpal and trapezioscapoid joints.

Roentgenographic Features

The formation of osteophytes and narrowing of cartilage are common to both forms of osteoarthritis. In secondary osteoarthritis, the earliest changes may be narrowing of cartilage in regions of stress and weightbearing combined with formation of osteophytes along nonweightbearing surfaces.^{7, 25} In the hip (Fig. 1), the nonweightbearing surfaces include the inferomedial portion of the femoral head, which is opposed by the acetabular fossa covered by connective tissue and synovium. The acetabular margins and fossa also form new bone, producing an incongruity of the joint surfaces that leads to lateral subluxation of the femoral head.¹⁹ New bone formed along the femoral neck, particularly its medial margin, causes a broadening of the bone, a process that has been called "buttressing." As narrowing of the cartilage progresses, the underlying bone is subjected to additional stress. The subchondral bone gradually becomes flattened and sclerotic as infarctions and ischemic necrosis lead to the formation of new bone along the nonviable trabeculae. Subchondral cyst-like cavities (geodes) form in the vicinity of greatest stress adjacent to the joint and within the zone of

sclerosis.²⁰ The cavities may become large and involve opposing bony surfaces. The cysts form as synovial fluid is driven by hydrostatic pressure through defects in the degenerated articular cartilage and into the underlying subchondral bone, gradually undermining it, as a waterfall gradually wears down the rocks at its base. The fluid within the geode is gradually replaced by connective tissue.

In primary generalized osteoarthritis (Figs. 2 and 3), well developed Heberden's nodes of the distal interphalangeal joints may cause flexion deformities and lateral deviation of the distal phalanges. The subchondral bony margins are irregular, and the thickness of the cartilage is diminished. Bony spurs originate from the adjacent surfaces of the distal and middle phalanges. Round or oval fragments of bone are frequently found along the medial, lateral, and occasionally volar surface of the soft tissues adjacent to the joint but outside its capsule. The fingers most commonly involved by Heberden's nodes are the index and middle fingers. Involvement of the proximal interphalangeal joints usually follows that of the distal interphalangeal joints. Osteoporosis is not a prominent feature, and is usually no more than would be expected for the age and sex of the patient. Involvement of the first carpometacarpal joint is common. As the cartilage narrows, the subchondral bone becomes sclerotic, and osteophytes produce incongruity and lateral

Figure 1. Secondary osteoarthritis of hip of 86-year-old man. Complete degeneration of articular cartilage has occurred superiorly, in association with subchondral sclerosis and formation of large geode in femoral head. Mature new bone along inferomedial margin of femoral head and adjacent acetabulum is associated with lateral migration of femoral head.

Figure 2. Primary generalized osteoarthritis affecting interphalangeal joint of hand of woman, age 74. Disorder is characterized by narrowing of articular cartilage, absence of erosions and presence of bony spurs projecting posteriorly from distal (Heberden's nodes) and proximal (Bouchard's nodes) interphalangeal joints.

subluxation of the base of the first metacarpal. This may be accompanied by severe pain and an adduction deformity. The trapezioscapoid joint is somewhat less frequently affected, and then, usually bilaterally and symmetrically.

Many of the radiographic features of primary generalized osteoarthritis are seen in erosive osteoarthritis (Fig. 4), including Heberden's and Bouchard's nodes and involvement of the first carpometacarpal and trapezioscapoid joints. The osteophytic changes nearly always precede the bony erosions. The erosions accompany irregular narrowing of the cartilage and

tend to involve the central surface of the joints most severely, with marginal erosions being uncommon. Bony fusion of the interphalangeal joints occurs in 12 to 15 per cent of patients with erosive osteoarthritis, and may be confused with the sequela of psoriatic arthritis.^{15, 24} Psoriatic erosions occur earliest and often predominate along the margins of the joints, resemble "mouse ears," and tend to persist, while the erosions of erosive osteoarthritis occur earliest and often predominate along the central surfaces of the joints, resemble "gull wings," and tend to heal.

ANKYLOSING SPONDYLITIS

Ankylosing spondylitis is a progressive, inflammatory, usually self-limited disease of the spine, involving the sacroiliac joints and, less commonly, the appendicular skeleton. The disease predominates in males (4 to 1). It is usually observed in the third decade of life, but it may begin in childhood. In males, prostatitis frequently accompanies the spondyloarthropathy. Most patients with ankylosing spondylitis possess the histocompatibility antigen HLA B27. Histologic tests for rheumatoid and lupus erythematosus factors are characteristically negative. The initial symptoms are usually low backache and stiffness, and discomfort in the thighs and buttocks. Ankylosis of the lumbar, thoracic, and cervical spine generally progresses for 10 to 20 years before severe rigidity of the back and kyphosis of the cervicothoracic spine su-

Figure 3. Primary generalized osteoarthritis involving first carpometacarpal and trapezioscapoid joints of 72-year-old woman. Space occupied by articular cartilage has all but disappeared. Sclerotic subchondral bone and prominent para-articular ossicles are characteristic.

Figure 4. Erosive osteoarthritis in woman, age 62. The distal interphalangeal joint of little finger manifests central erosion in association with posteriorly projecting osteophyte (Heberden's node). The proximal interphalangeal joint of same finger is strikingly narrowed. The distal interphalangeal joint of ring finger has undergone bony fusion, a change that occurs in 15 per cent of patients with erosive osteoarthritis.

pervene. The disease may subside spontaneously at any stage.²⁶

In contrast to the synovial lesions of rheumatoid arthritis, those of ankylosing spondylitis are less destructive. However, involvement of striking severity in the capsules, tendons, and ligaments ultimately leads to bone formation and ankylosis, in some instances with intact joints.¹ Histologic examination of tissue from various skeletal parts during active erosion of bone reveals a subacute inflammation of the soft tissues and bone, followed by replacement of the joint structures and intervertebral disks by granulation tissue. Eventually the granulation tissue becomes fibrotic, and metaplastic cartilage and bone form. In the spine, the inflammation is present in the areolar tissue and peripheral fibers of the annulus fibrosus at

their attachments to the upper and lower corners of contiguous vertebral bodies. Destruction of these corners produces the typical anterior "squaring" of the vertebral body. The ossification extends through the outer layers of the annulus fibrosus, but usually does not involve the paraspinous ligaments. In the diarthrodial joints, villous hyperplasia of the synovium is accompanied by inflammation and pannus formation, just as in rheumatoid arthritis. Interestingly, in the apophyseal, sternomanubrial, sacroiliac, and sometimes other joints, capsular ossification and bony ankylosis may take place over intact articular cartilage. Thus, the capsular ossification appears to be the primary event, and may then be followed by endochondral ossification and conversion of articular cartilage to bone as a result of disuse.

Radiographic Features

The sacroiliac joints are almost always affected early in the disease (Fig. 5). Initially the subchondral compact bone on the iliac side of the joint loses its sharp margin and becomes blurred. The sacral side of the sacroiliac joint tends to be involved later. Irregular erosions and erosive-like lesions develop, producing a ragged border and creating the illusion of joint widening, an appearance that results from irregular progression of endochondral ossification, with replacement of the dense subchondral bone plate by porous trabecular bone. The adjacent bone simultaneously increases in density because of appositional deposition of lamellar bone and the formation of woven bone at the capsular attachment. A similar appearance ultimately develops on the sacral side of the joint. Once the process becomes quiescent with joint fusion, the sclerotic bone assumes a normal density. Ankylosis of the joint capsules occurs first in thin sheets anteriorly and posteriorly and is the primary lesion. The disease is nearly always symmetrical in the two sacroiliac joints. The true sacroiliac joint comprises only the lower part of the gap between the sacrum and ilium. In many instances the sacroiliac ligamentous areas dorsal and cephalad to the true joint undergo ossification, as do the ligaments between the fifth lumbar transverse process and the sacrum and ilium.

Lesions similar to those in the sacroiliac joints develop, usually later, at other sites around the pelvis, adjacent to attachments of ligaments or tendons: the iliac crests, ischial tuberosities and spines, symphysis pubis, and greater and lesser trochanters. Similar abnormalities are seen sub-

Figure 5. Sacroiliitis in man, age 30, with 8-year history of ankylosing spondylitis. Bilaterally symmetrical involvement with adjacent fluffy sclerosis is characteristic.

sequently around the shoulder at the adjacent surfaces of the acromioclavicular joint, the coracoid process, and greater tuberosity of the humerus. Other sites include the sternal angle, sternoclavicular joints, costovertebral and costotransverse joints (Fig. 6), transverse and posterior spinous processes, and posterior and plantar surfaces of the calcaneus. The lesions tend to be bilaterally symmetrical, and consist of local superficial erosion of the cortical surface accompanied by underlying sclerosis and overlying soft tissue swelling.

The earliest and most common lesions of the spine develop at the thoracolumbar region and, next, at the lumbosacral articulation. If progressive, the disease usually ascends the spine to involve the cervical spine last. Since the inflammatory reaction probably begins in the outer fibers of the annulus fibrosus of the disk and in the adjacent areolar tissue, the earliest bony lesions are demonstrated at the upper and lower corners of the vertebral bodies, best seen

on lateral radiographs. Initially, the corners of the vertebral bodies become superficially eroded, and small triangular areas of sclerosis become visible in the subjacent bone (Fig. 7). The erosions, when more extensive, convert the original concave anterior surface of the vertebral body to a straight, and ultimately a convex, surface (Fig. 8). Usually, the lower corner of one vertebral body and the upper corner of the vertebral body below it are affected simultaneously. The inflammatory process also extends into the annulus fibrosus, often producing a narrowing of the disk, and may extend to the adjacent vertebral endplates. With repair, bone often forms within the outer margin of the annulus and unites the two adjacent vertebrae (Fig. 9). Initially, this syndesmosphyte is thin with fuzzy contours, but eventually, additional sharply demarcated bone forms around its outer surface. This formation of bone produces bulging beyond the margin of the vertebral body that, when extensive, has

Figure 6. Ankylosing spondylitis in 32-year-old woman. The costotransverse joints have undergone bilaterally symmetrical bony fusion.

Figure 7. Ankylosing spondylitis (same patient as in Fig. 6). Anterior "squaring" of vertebral bodies has progressed to corner erosions.

Figure 8. Man, age 24, with ankylosing spondylitis since childhood. Thoracolumbar vertebral erosions have progressed from anterior "squaring" to the formation of convex anterior surfaces.

Figure 9. "Burnt-out" ankylosing spondylitis in 66-year-old man. Syndesmophytes represent ossification of outer fibers of annulus fibrosus. In ankylosing spondylitis, they are typically delicate or thin and attach to corners (margins) of vertebral bodies. The apophyseal joints are fused.

been termed a "bamboo spine" (Fig. 10). The anterior and lateral surfaces are predominantly involved. In the cervical spine the odontoid may be subluxated as in rheumatoid arthritis; but unlike rheumatoid arthritis, the cervical lesions are not usually associated with instability and subluxations of the lower five vertebrae. If the onset of ankylosing spondylitis is before age 20, apophyseal lesions prevail over those of the vertebral bodies; if the onset is after age 25, ankylosis is primarily of the vertebral bodies and less often of the apophyseal joints.

Localized back pain of recent origin in patients with ankylosing spondylitis may represent extensive destruction of the cartilage endplates, and occurs almost exclusively in spines with extensive ankylosis and in regions where an insufficiency fracture of the lamina, ossified disk, ligaments, or other supporting structures has occurred.²⁹ Occasionally, the destruction involves hypermobile, unankylosed segments of an otherwise severely ankylosed spine. Whatever the cause, the lesions appear radiographically as a destructive focus surrounded by sclerotic margins.

In about 60 per cent of patients, the joints of the appendicular skeleton are involved, in the

following order of decreasing frequency: acromioclavicular joints, knees, hips, shoulders, and tarsal joints.²² Less often involved are the remaining joints of the feet, hands, and wrists, in which the disease is usually mild, sometimes with only joint effusion and slight or no osteoporosis. The lesions of the hip and knee joints are often mild, with slight narrowing of the articular cartilage and minimal or no bone erosion. Osteoporosis during the acute phase may be followed by bony ankylosis of the joint capsule, although the articular cartilage appears to be of normal or slightly diminished width (Fig. 11). The hip lesions differ from those of rheumatoid arthritis in that they are less severe, usually do not produce protrusion, and may be accompanied by bony ankylosis. Another distinguishing feature is the presence in ankylosing spondylitis of a characteristic osteophytic skirt along the margin of the articular cartilage of the femoral head.

PSORIATIC ARTHRITIS

Psoriatic arthritis combines many roentgenographic features of rheumatoid arthritis, in which synovial inflammation predominates, and

Figure 10. Ankylosing spondylitis (same patient as in Fig. 9). The "bamboo spine" is characterized by generalized syndesmophytes, ligamentous ossifications, and fusion of apophyseal and sacroiliac joints.

Figure 11. Long-standing, quiescent ankylosing spondylitis in man, age 58. The capsules of hip joints have ossified, resulting in bilateral fusions. The sacroiliac joints and spine are completely fused.

ankylosing spondylitis, in which ligamentous inflammation predominates. Thus, although the early manifestations of psoriatic arthritis may be restricted to synovium lined joints, in at least one quarter of all patients, the inflammatory process eventually involves both synovium and ligaments. In cases that progress to severe deformity of the hands and feet, inflammation at sites of tendinous and ligamentous attach-

ments results in bone resorption or erosion with accompanying exuberant proliferation of new bone at the sacroiliac, acromioclavicular, and manubriosternal joints, as well as at the plantar surface of the calcaneus, the ischial tuberosities, and the femoral trochanters. The vertebral bodies may also be involved at their sites of ligamentous attachment.

The clinical manifestations comprise a triad

Figure 12. Psoriatic arthritis in man, age 43, characterized by distal interphalangeal marginal and surface erosions accompanied by striking fluffy periosteal new bone and soft-tissue swelling.

consisting of 1) psoriasis of the skin, scalp, and/or nails, 2) a negative rheumatoid factor, and 3) an erosive polyarthritis with a tendency to predominate at the distal interphalangeal joints, often bilaterally asymmetric, and which may precede the cutaneous manifestations by years.

Roentgenographic Features

In psoriatic arthritis, as in all inflammatory arthritides, joint swelling heralds the earliest erosions. In psoriatic arthritis and rheumatoid arthritis, these occur at the joint margins, where the cartilage is least effective in protecting the underlying bone from the ravages of the synovial pannus. Unlike the erosions of rheumatoid arthritis, which are often well defined and bilaterally symmetrical in location and severity, those of psoriatic arthritis usually have ill defined or fuzzy margins, are often bilaterally asymmetrical, are characteristically associated with florid proliferation of periosteal new bone (Fig. 12), and are less often accompanied by osteoporosis.²⁷

In the hands and wrists the "classical pattern" of bilaterally asymmetric erosive polyarthritis of the distal interphalangeal joints^{27, 28} (Fig. 12) is only one of several characteristic patterns. Other patterns are oligoarticular; "sausage" digit due to tenosynovitis (Fig. 13); rheumatoid arthritis-like, with metacarpophalangeal predominance; unilateral polyarticular, which may involve only a single ray (Fig. 14); arthritis mutilans; and fixed hand deformity. In Martel's series, the distribution of erosions was often unilateral, being polyarticular in 25 per cent and involving a single ray in another 21 per cent of patients (Fig. 14).¹⁵ Concomitant nail changes are usually present and vary from mild pitting, usually imperceptible in radiographs, to severe thickening and deformity. Erosions are sometimes limited to one or two joints in each hand (oligoarticular), and tend to be bilaterally asymmetrical and slowly progressive. Although the metacarpophalangeal joints undergo the greatest deformity, generalized ulnar deviation of the proximal phalanges, a characteristic and often early feature of rheumatoid arthritis, is not characteristic of psoriatic arthritis. Subluxations may occur in patients with longstanding psoriatic arthritis, but the direction tends to be haphazard, some directed one way, some another.

Eventually, the subchondral bone may undergo such severe destruction that the loss of bone length in combination with subluxation gives rise to arthritis mutilans, a condition that

Figure 13. Psoriatic arthritis in woman, age 37, manifested by tenosynovitis and producing "sausage" digit of index finger. The process also features fluffy periosteal new bone of proximal phalanx.

may also occur as the end result of rheumatoid arthritis, Reiter's disease, and multicentric reticulohistiocytosis. The characteristic radiographic feature of arthritis mutilans is the "pencil-in-cup" deformity; the "pencil point" is represented by the distal end of a metacarpal (or metatarsal in the foot), while the "cup" is represented by the eroded articular surface of the apposing phalanx. Adjacent phalanges may be involved in the same manner (Fig. 15). Although the radiographic appearance suggests that the changes derive from friction between unprotected apposing bone surfaces, pathologic analysis reveals that the bone ends are embedded in fibrous connective tissue.²³

Bony ankylosis of the interphalangeal joints is seen in 12 to 15 per cent of patients with psoriatic arthritis and may be a striking feature

Figure 14. Psoriatic arthritis in man, age 37. Single-ray pattern. The metacarpophalangeal and interphalangeal joints of thumb have undergone bony fusion. Extensive bony erosive and proliferative changes affect sesamoid bone adjacent to metacarpal head. The other joints in hand were unaffected.

that affects many joints simultaneously (Fig. 16). Resorption of bone along the dorsal and lateral aspects of the tips and shafts of the distal phalanges occurs in about 5 per cent of patients with psoriatic arthritis,²⁸ even less frequently in Reiter's disease,¹⁴ and may be related to inflammation beneath the nails (Fig. 17). The resorptive "whittling" may at times be revealed only in a lateral radiographic projection.

Just as the distal interphalangeal joints are usually the earliest to be affected in the hand, the interphalangeal joint of the great toe is a favored site of early involvement in the foot. An inflamed plantar aponeurosis may result in an exuberant calcaneal plantar spur. Similarly, a generalized tenosynovitis may lead to a fluffy periosteal reaction along the surfaces of other

tarsal bones. In patients with chronic, progressive arthritis, arthritis mutilans may become a significant complication. As in rheumatoid arthritis, the proximal phalanges may manifest a fibular drift, a change which may also occur in Reiter's disease.

The hips, knees, ankles, shoulders, and elbows may be involved in a bilaterally asymmetric fashion. Soft tissue swelling is succeeded by marginal erosions and, ultimately, surface erosions. The subchondral bone tends to undergo destruction in a uniform manner over the entire articular surface. As in the hands and feet, the erosions appear ill defined, and are frequently associated with exuberant subperiosteal new bone. Osteoporosis may be mild or absent.

Although patients with psoriatic arthritis may exhibit sacroiliitis without other spinal manifestations, the latter rarely occur in the absence of sacroiliitis. In a series of 21 patients with psoriatic spondylitis, radiographic evidence of sacroiliitis was seen in every case.¹² The erosions were bilaterally symmetrical in two thirds of

Figure 15. Psoriatic arthritis in 36-year-old woman. Arthritis mutilans of foot, characterized by "pencil-in-cup" deformities of interphalangeal joint of great toe and distal interphalangeal joint of second toe, whose proximal interphalangeal joint has fused.

Figure 16. Psoriatic arthritis (same patient as in Fig. 15). Bony ankylosis affects proximal interphalangeal joints of index and middle fingers. Erosions and adjacent fluffy periosteal new bone, present at multiple joints, are most severe at first and fifth metacarpophalangeal joints.

Figure 17. Psoriatic arthritis, characterized by "whittled" terminal tuft of thumb of 25-year-old woman. The overlying nail is displaced outward.

patients. Fusion was relatively infrequent, being found in only three of the patients. Since the sacroiliac resorptive and erosive changes result primarily as a consequence of inflammation of adjacent ligaments, they may be seen anywhere along the joint margins, and are not necessarily confined to the synovial, or lower portion, of the joint. The erosions are usually accompanied by fluffy sclerosis, the result of endosteal and periosteal proliferation of new bone. As in ankylosing spondylitis, it is not unusual for the erosions to be manifest on the iliac side of the joint before they can be detected on the sacral side (Fig. 18).

Psoriatic involvement of the spine is most often observed in association with severe cutaneous and arthritic lesions, and is characterized by the presence of coarse, asymmetrical syndesmophytes (Fig. 19), paravertebral ossifica-

Figure 18. Psoriatic arthritis in 21-year-old man. The erosions of sacroiliitis are usually initially manifested on iliac side of joint, as shown in this oblique view, and are bilaterally symmetrical in two thirds of cases.

Figure 19. Mature, long-standing anterior syndesmophyte at C2-3 level in man, age 40, with psoriatic arthritis. The ossification is characteristically coarse and nonmarginal (i.e., it attaches to C3 body beyond its anterosuperior corner or margin).

tion (Fig. 20),⁵ atlantoaxial subluxation, and relative sparing of the apophyseal joints. As in ankylosing spondylitis, erosions may arise anywhere along the surface of the vertebral body. They may simply heal or become closely associated with a mass of fluffy new bone. This new bone appears at a site of erosion or in the adjacent soft tissue, and often organizes into a dense bony ridge or syndesmophyte that extends around and through the disk annulus. The syndesmophytes of psoriatic arthritis are characteristically large and coarse (Fig. 19), tend to be asymmetrical and to skip areas (Fig. 20), and are most common in the middle and lower cervical spine and in the thoracolumbar and upper lumbar spine. They are often non-marginal, that is, they attach to the vertebral bodies beyond their corners. When the erosions heal, the normal anterior endplate prominences of the vertebral body may fail to reconstitute. The result is an apparent "squaring" of the anterior surface, sometimes accompanied by sclerosis at the corners of the "square." Vertebral squaring is much less common in psoriatic than in ankylosing spondylitis.

In contrast to syndesmophytes, osteophytes reflect a degenerative process of proliferation of bone at sites of ligamentous insertion, and are

Figure 20. Progression of paravertebral ossification over duration of 32 months in man, age 35, with psoriatic arthritis. Bone first appeared in paravertebral soft tissue, gradually matured, and became incorporated into annulus of disks as asymmetrical syndesmophytes.

common phenomena of aging. Osteophytes arise from the vertebral endplates, extend in a horizontal direction, may be irregular in contour but, in the absence of vertebral instability, are not fluffy. They may or may not be associated with narrowing of the disk. The annulus of the disk is not incorporated into the osteophytes. In practice, however, the distinction between syndesmophytes and osteophytes is often difficult. The presence of erosions, underlying sclerosis, and bridges of fluffy new bone are clues to the diagnosis of syndesmophytes.

Atlantoaxial subluxation is common in patients with psoriatic spondylitis. In one series it was present in 45 per cent of 20 patients who had roentgenograms of the cervical spine.¹² It should be remembered that atlantoaxial subluxation may not be detectable unless the lateral radiograph of the neck is obtained during *flexion*.

REITER'S DISEASE

Reiter's disease, like ankylosing spondylitis, manifests a strong association with the histocompatibility antigen HLA B27. Like psoriatic arthritis, it differs from ankylosing spondylitis in its inconstant involvement of the spine and its greater involvement of the peripheral joints. It is not always easy to distinguish between Reiter's disease and psoriatic arthritis, radiologically or pathologically. One difference is that

Reiter's disease tends toward selective involvement of the lower limbs, particularly the feet, with relative sparing of the hands and wrists, while psoriatic arthritis tends to affect the joints of the upper limbs as extensively as those of the lower. In Reiter's disease, the interphalangeal joint most prominently involved is that of the great toe, while in psoriatic arthritis there is, by contrast, a predilection for the distal interphalangeal joints of the other digits of the hands and feet as well as the interphalangeal joint of the great toe. In Reiter's disease there is a much stronger tendency for involvement of the knee.

Many similarities exist between Reiter's disease and psoriatic arthritis: osteoporosis is usually minimal or absent; periosteal new bone is often fluffy and irregular, and especially common at bony prominences (Fig. 21);¹⁶ sacroiliitis is frequently seen in both conditions, but is more common and less symmetrical in Reiter's disease. In both disorders, syndesmophytes are coarser and less symmetrical than in ankylosing spondylitis and tend to attach to adjacent vertebral bodies beyond their corners; hence, the term nonmarginal syndesmophytes.¹⁴

MULTICENTRIC RETICULOHISTIOCYTOSIS

The onset of this rare but dreaded disorder may occur at any time between adolescence and senescence. The histologic hallmark is the

Figure 21. Exuberant periosteal new bone along plantar and posterior surfaces of calcaneus of 35-year-old man with Reiter's disease.

presence in the skin, subcutaneous tissue, and synovium of multinucleate, PAS-positive, lipid-laden histiocytes that sometimes resemble foreign body type giant cells.⁶ Polyarthritides usually precedes the nodular cutaneous eruptions by an average of 3 years, a fact that emphasizes the importance of early roentgenologic recognition. Although it tends to wax and wane, in one third of cases the disorder undergoes rapid evolution into an incapacitating and deforming arthritis mutilans.⁴ The interphalangeal joints are the predominant sites of involvement in the hand. Cutaneous and subcutaneous nodules predominate in the vicinity of the ears, bridge of the nose, scalp, dorsum of the hands, and nail beds. Finally, after several years, the arthritis and nodules may become quiescent. At the end stage of the disease, the patient is frequently left with crippling deformities of the hands and a leonine facies.²

Roentgenographic Features

Multinodularity of the skin, subcutaneous tissue, and tendon sheaths may be reflected roentgenographically in uncalcified soft tissue masses. Bilaterally symmetrical marginal erosions rapidly progress to surface erosions, and predominate in the interphalangeal joints of the hands and feet. The erosions are exquisitely well circumscribed, and strikingly symmetrical (Fig. 22).⁶ Eventually all of the synovium lined joints become involved. Atlantoaxial subluxation may be present at an early stage.^{6, 14} The sharp margins of the erosions, mildness of ac-

companying osteoporosis, and absence of periosteal new bone reflect the mildness of the inflammatory response and resultant lack of repair by the underlying bone. Nevertheless, the end result is often arthritis mutilans.

PROGRESSIVE SYSTEMIC SCLEROSIS (DIFFUSE SCLERODERMA)

This generalized disorder of connective tissue is characterized by inflammatory, fibrotic, and degenerative changes, accompanied by vascular lesions in the skin, synovium, esophagus, stomach, intestinal tract, heart, lung, and kidney. The initial complaint is usually Raynaud's phenomenon, swelling of the distal parts of the extremities or gradual thickening and tightening of the skin of the fingers. Dystrophic calcification of soft tissues, present most often in women, usually occurs in the fingertips but may develop over bony prominences throughout the body.

The so-called CREST syndrome consists of calcinosis, Raynaud's phenomenon, esophageal hypomotility, sclerodactyly, and telangiectasis, and may remain confined to the skin for long periods before involving other systems. Large synovial effusions may appear in large joints, associated with tenderness, redness, or warmth.²¹ Later the fingers, knees, and elbows undergo flexion contractures. Esophageal dysfunction develops in 90 per cent of patients. Hypomotility and stasis in saccules or pseudodiverticula allow overgrowth of bacteria in the

Figure 22. Multicentric reticulohistiocytosis in man, age 31, with 7-year history of disease. Erosions are universal, exquisitely bilaterally symmetrical and well-circumscribed, and accompanied by disproportionately mild osteoporosis.

upper part of the small intestine, which interferes with normal absorption of fat as a result of deconjugation of bile salts. The lungs undergo functional aberrations consisting of restrictive insufficiency. Pulmonary fibrosis leads to pulmonary hypertension. Cardiac symptoms result from myocardial fibrosis and pulmonary artery hypertension. Cardiac arrhythmias are often present. Renal insufficiency is a major cause of death, and sometimes results from a highly malignant arterial hypertension of the renal arteries and arterioles. Inflammation of the joints is characterized by infiltration of lymphocytes and plasma cells within the synovium, changes that resemble early rheumatoid arthritis. Later, intense fibrosis of the synovium may encompass and obliterate the vascular structures.

Roentgenographic Features

Thickening of the soft tissues of the fingers with relative sparing of the terminal portions may be the initial manifestations of the disease. Later in the course of the disease the soft tissues become thin, and the subcutaneous fat is no longer apparent since it is replaced by fibrous tissue. Moreover, the transverse folds over the interphalangeal joints, usually seen in well exposed radiographs of normal joints, disappear

with tightening of the skin. Flexion deformities of the fingers and other joints gradually develop. Atrophy and ulceration of the pulp and lateral soft tissues of the finger tips may be associated with calcinosis circumscripta (Fig. 23) that tends to predominate on the volar

Figure 23. Progressive systemic sclerosis, characterized by calcinosis circumscripta and resorption of underlying terminal tufts.

Figure 24. Progressive systemic sclerosis in 23-year-old woman, featuring profound, bilaterally symmetrical resorption of posterior aspects of fourth through tenth ribs. Cardiomegaly is also present.

Figure 25. Progressive systemic sclerosis in woman, age 42, characterized by soft-tissue atrophy, flexion contractures of interphalangeal joints, and two changes similar to those of rheumatoid arthritis: marginal erosions of metacarpophalangeal joints, and ulnar deviation of proximal phalanges.

surface. Resorption of terminal tufts usually begins on the volar surface in contrast to psoriatic arthritis and Reiter's syndrome, in which resorption occurs along the dorsal and lateral surfaces adjacent to involved and thickened nails. The resorption is progressive, and various portions of the distal, middle, and proximal phalanges may be destroyed. Other prominences of bone may be resorbed, such as the medial surfaces of the bases of the fifth metacarpals, outer margins of the radius and ulna, distal clavicle, acromion process, mandibular condyle, and the superior margins of the posterior aspects of the midthoracic ribs. Rib resorption is often bilaterally symmetrical, predominates along the superior surfaces, and in rare cases may be profound (Fig. 24).⁸

Although marginal erosions are less common than in rheumatoid arthritis, they may develop in the metacarpophalangeal and interphalangeal joints (Fig. 25). In one review, of 55 patients with progressive systemic sclerosis, 12 showed radiographic evidence of inflammatory arthritis that ranged from isolated to generalized joint destruction.³

SUMMARY

Osteoarthritis may be divided into primary generalized and secondary forms. Primary generalized osteoarthritis is characterized by narrowing of cartilage, marginal osteophytes, and absence of erosions. The most common sites of involvement are the distal interphalangeal joints of the fingers and the first carpometacarpal joint. Secondary osteoarthritis also results in narrowing of cartilage in the absence of erosions, but in regions of mechanical stress.

Erosive osteoarthritis affects predominantly the proximal and distal interphalangeal joints, and evolves into bony fusion in 12 to 15 per cent of cases, about the same percentage of interphalangeal bony fusion that occurs in psoriatic arthritis.

Ankylosing spondylitis predominates in the axial skeleton where it eventually leads to fusion of the vertebrae and sacroiliac joints.

Psoriatic arthritis combines many features of rheumatoid arthritis, in which synovial inflammation predominates, and ankylosing spondylitis, in which ligamentous inflammation predominates. The hands and feet are involved to an equal extent, and in 20 per cent of patients the disorder also involves the sacroiliac joints and spine.

Reiter's disease, like psoriatic arthritis, differs from ankylosing spondylitis in its inconstant

involvement of the spine and greater involvement of peripheral joints. Reiter's disease differs from psoriatic arthritis in its predominant involvement of the lower limbs, particularly the feet, with relative sparing of the hands and wrists.

Multicentric reticulohistiocytosis is a rare disorder in which polyarthritis usually precedes the onset of nodular cutaneous eruptions, a fact that emphasizes the importance of early roentgenologic recognition. The interphalangeal joints are the predominant sites of involvement in the hands, but eventually all of the synovium lined joints become affected, with arthritis mutilans the end result in one third of cases. The erosions are strikingly symmetrical and well circumscribed, and accompanying osteoporosis is disproportionately mild.

Progressive systemic sclerosis is characterized by atrophy and dystrophic calcifications in the soft tissues, ultimately leading to joint deformities and resorption of the terminal tufts of the phalanges. Resorption of bone occurs at other sites as well, and marginal erosions may develop in the metacarpophalangeal and interphalangeal joints of the hands.

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