Annals of the Rheumatic Diseases, 1982, **41**, 147–152

The articular manifestations of progressive systemic sclerosis (scleroderma)

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SUMMARY The articular manifestations of progressive systemic sclerosis (PSS) were studied in 38 patients. Of these, 66% experienced joint pain and 61% had signs of joint inflammation. Limitation of joint movement was seen in 45%. Radiological abnormalities included periarticular osteoporosis (42%), joint space narrowing (34%), and erosions (40%). Erosive disease did not correlate with disease duration, presence of rheumatoid factor, antinuclear antibodies, distal tuft resorption, or the extent of the scleroderma skin changes. Calcinosis was seen more frequently in those patients with articular erosions (67%). Erosive osteoarthritis of the distal interphalangeal joints (7 patients) was associated with impaired finger flexion. Joint involvement in PSS occurs frequently and may resemble rheumatoid arthritis in the early stages but is less destructive. The occurrence of unrelated arthropathy, such as primary osteoarthritis, is not uncommon, and its differentiation from true PSS joint disease can be difficult.

Articular manifestations are common in progressive systemic sclerosis (scleroderma, PSS), and joint pain is a frequent presenting feature of this disease.[1] Joint symptoms have been noted in 12 to 66% of patients at the time of diagnosis[2-6] and in 24 to 97% of patients at some time during the course of their illness.[4 5 7] Histological evidence of synovitis has been found in up to 66% of synovial biopsies from patients with PSS,[6] but, clinically, arthralgias have been considered to occur more frequently than frank arthritis.[8]

Radiological changes in the joints of PSS subjects include periarticular osteoporosis, intra-articular calcification,[9] erosions, loss of joint space,[6 10-13] and rarely aseptic necrosis.[14 15] Despite previously published studies, however, the pattern of joint involvement in this disease has not been clearly established. The wide variation in the reported prevalence of both clinical and radiological features, as well as their correlation with other disease parameters, requires clarification. We report the results from a systematic study of the articular manifestations in 38 patients with PSS seen over a 6-month period.

Patients and methods

Thirty-eight patients attending the Scleroderma Clinic at the Wellesley Hospital, as part of a prospective study and fulfilling the preliminary diagnostic criteria for PSS[16] were evaluated by one author (M.B.). A history of joint manifestations was obtained in each case according to a standard protocol. All peripheral joints were examined for evidence of active inflammation, limitation of movement (20% or greater loss in flexion, extension, or total range), deformity, and tenosynovitis (including friction rubs). An actively inflamed joint was defined as being tender to palpation, painful on movement (stress pain), or exhibiting signs of an effusion. With the metacarpophalangeal and interphalangeal joints, however, it was necessary for stress pain to be accompanied by one other clinical sign of inflammation, such as soft tissue swelling or an increase in warmth.

Finger flexion was assessed by the estimation of 'fist', that is the ability to approximate the tip of the middle finger to the mid palmar crease. A full fist was arbitrarily assigned a grade of 4, 75% as grade 3, 50% as grade 2, 25% as grade 1, and grade 0 when no flexion was possible. Patients were assessed as having limited skin (scleroderma) involvement when the appropriate changes (thickening or atrophic and bound-down) were confined to the distal parts of the limbs (distal to mid forearm) and face, and diffuse when the trunk was involved in addition.

Standard anteroposterior roentgenograms of the hands and wrists were obtained for all patients and

Accepted for publication 7 May 1981.
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Table 1 *Clinical and serological features of 38 patients with progressive systemic sclerosis*

Sex, female:male	34:4
Mean age (range)	54·0 (16–76) years
Mean duration of disease (range)	7·3 (0·5–17) years
Number (%) with diffuse skin involvement	17 (45)
Number (%) with limited skin involvement	21 (55)
Number (%) with rheumatoid factor	25 (66)
Number (%) with antinuclear antibodies	22 (58)

read 'blindly' by 2 independent observers (M.B. and P.L.). Any discrepancies were resolved by concurrent interpretation. Joints with marked flexion deformities were omitted in the assessment of joint space narrowing. The serological status of the patients for rheumatoid factor and antinuclear antibodies (ANA) was determined by the presence (or absence) of a positive test at the time of this study or during the previous 2 years of follow-up. Rheumatoid factor titres were performed by the latex fixation method.[17] Titres equal to or greater than 1:160 were considered to be positive. The presence of ANA was determined by the indirect immunofluorescent technique using mouse kidney as substrate. Titres of ANA equal to or greater than 1:10 were considered to be significant. The disease duration was calculated from the time of diagnosis of PSS in each case. Statistical analyses were carried out with the Student's *t* test for unpaired variants and the chi-square test with Yates's correction when applicable.

Results

The clinical and serological features of the 38 patients at the time of the study are shown in Table 1.

Table 2 *The clinical history of joint involvement in 25 progressive systemic sclerosis patients with articular manifestations*

Nature of joint manifestations	*Number*	*(%)*
Mode of onset		
Acute	10	(40)
Insidious	15	(60)
Course		
Intermittent	11	(44)
Chronic remittent*	4	(16)
Slowly progressive	9	(36)
Rapidly progressive	1	(4)
Pattern		
Monoarticular	5	(20)
Oligoarticular	10	(40)
Polyarticular	10	(40)
Joint swelling	10	(40)
Morning stiffness of 30 minutes or greater	17	(68)

*A chronic remittent course is one with persistent symptoms but having periods of distinct remissions.

Twenty-five (66%) gave a history of joint pain occurring at some time during the course of their disease and 21 (55%) had joint pain at the time of the present evaluation. The onset of joint symptoms preceded the diagnosis by 1 year or more in 5 patients (20%), occurred during the year of diagnosis in 8 (32%), and developed after the diagnosis in 12 (48%).

The mode of onset, course of the joint symptoms, and pattern of articular involvement by history are seen in Table 2. Also shown are the numbers of patients with a history of joint swelling and morning stiffness of 30 minutes' duration or greater. In terms of morbidity 5 patients (20%) considered their joint symptoms to be an insignificant problem when compared to other aspects of their disease. Joint pain was considered to be of minor significance in 7 (28%), moderate in 8 (32%), and of considerable significance in 5 (20%). Treatment had included salicylates or other nonsteroidal anti-inflammatory drugs in 21, low-dose corticosteroids (less than 7·5 mg prednisone equivalent daily) in 8, and a course of gold in 1 patient.

Examination of peripheral joints revealed the presence of tenderness, stress pain, or effusions in 23 patients (61%). Painless effusions were seen in 2 cases (knees). Effusions, found in 10 patients, were small and confined to the knees, with the exception of 1 proximal interphalangeal (PIP) joint. In all instances there were no clinical features or radiological abnormalities to suggest other possible underlying causes for the effusion, such as osteoarthritis. Synovial thickening was not a clinical feature of the arthritis seen in these patients. The distribution of the clinically inflamed joints is shown in Table 3. The pattern was symmetrical and polyarticular in 14 (61%), oligoarticular in 5 (22%), and monoarticular in 4 (17%). Tendon friction rubs were noted in 12 (32%). Five patients giving a history of joint pain showed no clinical evidence of joint inflammation, but one of these had documented evidence of a pre-

Table 3 *Distribution of clinically inflamed joints and those with restricted range of movement in 23 progressive systemic sclerosis patients with articular manifestations*

Joints involved	*Number of patients*	
	Inflamed	*Restricted movement*
Metacarpophalangeal	12	—
Proximal interphalangeal	8	—
Distal interphalangeal	9	—
Wrist	10	10
Elbow	1	3
Shoulder	7	6
Hip	—	1
Knee	10	1
Ankle	2	7
Metatarsophalangeal	6	—

Table 4 *Radiological abnormalities of the hands and wrists in 38 patients with progressive systemic sclerosis*

Radiological abnormality	*Number patients (%)*
Osteoporosis	
Generalised	10 (26)
Periarticular	16 (42)
Joint space narrowing: site	13 (34)
Distal interphalangeal (DIP)	12
Thumb interphalangeal	2
First carpometacarpal (CMC)	4
Metacapophalangeal (MCP)	2
Radiocarpal	2
Trapezioscaphoid	1
Erosions: site	15 (40)
DIP	7
MCP	9
Thumb interphalangeal	2
PIP	3
First CMC	2
Inferior radioulnar	3
Radiocarpal	4
Terminal phalangeal tuft resorption	14 (37)
Resorption of distal ulna	3
Calcinosis	19 (50)

vious symmetrical polyarthritis. Since flexion contractures of fingers are frequently due to soft tissue involvement, these deformities were not individually recorded. Limitation of joint movement otherwise occurred in 17 patients (Table 3). Addition joint deformities included metacarpophalangeal (MCP) subluxation in 2 cases, ulnar deviation (1 case), and swan neck deformity (1 case).

The radiological abnormalities seen in the hands and wrists of the patients in this study are summarised in Table 4. Erosive changes were most frequently seen in the MCP joint (9 cases), usually in the form of isolated, small, discrete lesions at the periarticular margin. Similar changes, resembling those seen in early rheumatoid arthritis, were noted involving the PIP and distal interphalangeal joints (DIP) (Fig. 1). Fig. 2 shows an example of the erosive lesion seen involving the inferior radioulnar joint in 4 patients. In all of these cases there was an associated limitation in range of wrist movement, but 3 of the 4 had no signs of active joint inflammation at the time of this study.

Table 5 *Correlation between the presence of articular erosions, serological status, extent of scleroderma skin involvement, and other radiological features. Figures represent number and (%) of patients*

	Erosive disease n = 15	*Nonerosive disease n = 23*
Positive RA latex	11 (73)	14 (61)
Antinuclear antibodies present	9 (60	13 (57)
Limited skin involvement	8 (53)	13 (57)
Diffuse skin involvement	7 (47)	10 (43)
Terminal tuft absorption	6 (40)	8 (35)
Calcinosis	10 (67)	9 (39)

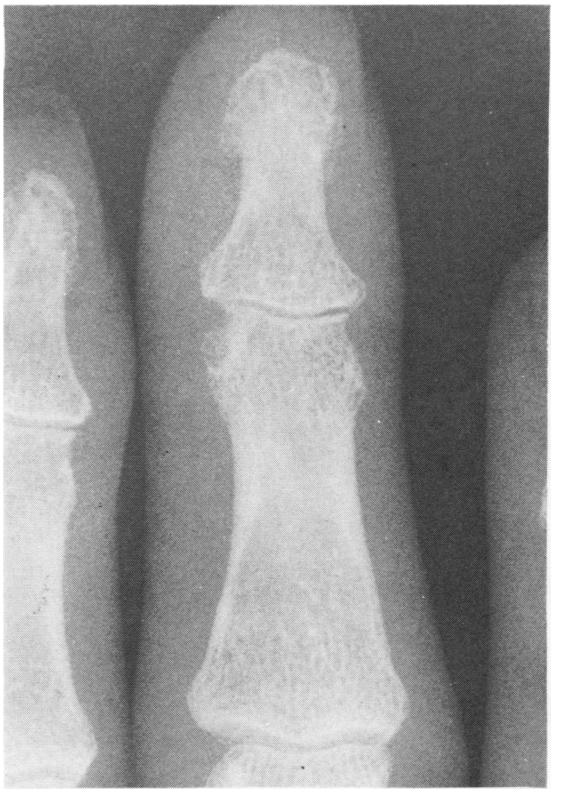

Fig. 1 *Discrete, marginal erosion of a distal interphalangeal joint in progressive systemic sclerosis.*

The 15 patients with articular erosions had a greater mean (±SD) duration of disease (7·6 ± 3·7 years) than those without erosive changes (5·4 ± 3·7 years), but the difference was not significant ($p>0·05$). Similarly, there was no significant difference in the mean (±SD) duration of disease between those with (7·4 ± 5·0 years) and without (5·7 ± 3·9 years) radiological evidence of joint space narrowing ($p>0·05$). No significant differences occurred between those with and without erosive disease with respect to the presence of rheumatoid factor, antinuclear antibodies, extent of scleroderma skin involvement, and occurrence of terminal tuft absorption (Table 5). The prevalence of calcinosis was higher in the group with erosive disease ($\chi^2 = 2·75$, $0·05<p<0·1$).

Twelve patients had joint space narrowing involving the DIP joint. In 5 the appearances were those of classical primary osteoarthritis, but the remainder (7 patients) resembled erosive osteoarthritis, which was severe in several cases, with fragmentation of the articular structures and deformity, producing a 'gull

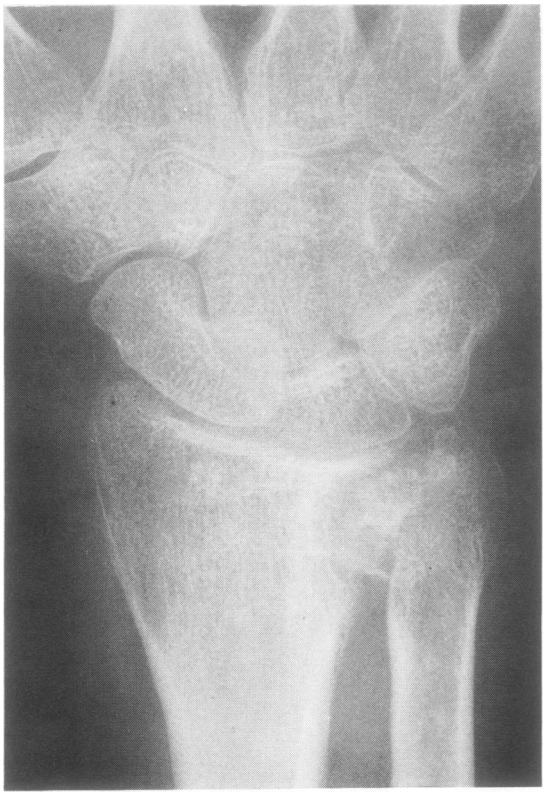

Fig. 2 *Erosive lesion of an inferior radioulnar joint in progressive systemic sclerosis.*

Table 6 *The 'fist' scores of progressive systemic sclerosis patients with and without erosive osteoarthritic changes in the distal interphalangeal joints*

'Fist' score	*Number of patients*	
	With erosive osteoarthritis	*Without erosive osteoarthritis*
3 or less	7	15
4	0	15

wing' appearance (Fig. 3). A similar lesion is shown in Fig. 4, showing the rapid progression which had occurred in the 16 months between the 2 radiographs. The mean age of the 7 patients with erosive osteoarthritis was 61·5 (±SD 7·2) years compared with 53·0 ± 15 years in those without this abnormality. The occurrence of the radiological changes in the DIP joints resembling erosive osteoarthritis was invariably associated with impaired finger flexion with a fist score of 3 or less (Table 6).

Discussion

The frequent occurrence of articular manifestations in PSS has been previously reported.[5,6] In the present series of 38 patients nearly two-thirds recall having experienced joint symptoms at some time during the course of their disease. The time of onset of the joint complaints varies widely but in the majority is early,

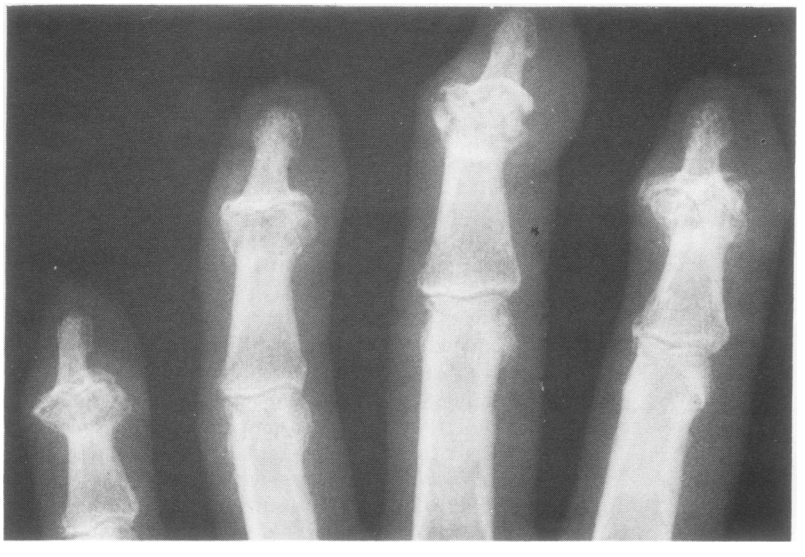

Fig. 3 *Joint abnormalities resembling erosive osteoarthritis in a patient with progressive systemic sclerosis.*

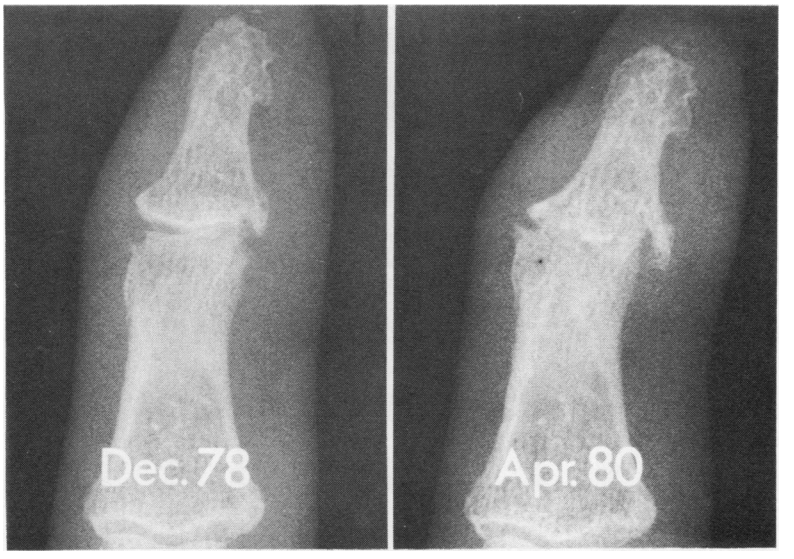

Fig. 4 *Progression of an erosive lesion seen in the distal interphalangeal joint over a period of 16 months.*

preceding or coinciding with diagnosis of the disease. With a symmetrical polyarticular pattern (40%) a misdiagnosis of rheumatoid arthritis could easily be made in the early stages, before the characteristic skin changes become evident.

Clinical evidence of joint inflammation was seen in 61% of the patients in our series. However, this was largely determined on the basis of joint tenderness and stress pain. Effusions tended to be small and were asymptomatic in 2 cases. Although 40% of those with articular involvement had a history of joint swelling, the absence of the latter in the current examination of these patients was a notable feature. Signs of active synovitis were seen more frequently by Rodnan,[6] but again did not appear to have been either prominent or persistent. The course of the joint manifestations in the present study was either intermittent or chronic remittent in the majority, and hence, unless a prospective study is carried out with frequent observations, any swelling or increase in warmth that might occur could have been missed. Synovial biopsies from scleroderma joints showed histological evidence of inflammation with lymphocytic and plasma cell infiltration, but late in the course of the disease there was evidence of increasingly severe synovial fibrosis.[14] The mean duration of disease at the time of the present study was 7·3 years, and therefore many could already have progressed to the fibrosed, noninflammatory, late stage of joint involvement.

Interestingly, in Rodnan's study[6] 6 of 9 patients in whom only synovial fibrosis was found did complain of joint pain or had tenderness or pain on joint motion. This suggests that, at least in the advanced stages of PSS, joint pain and tenderness need not reflect an underlying synovitis. It has also been noted that PSS synovium has little if any tendency to proliferate and form pannus[14 18] and is consistent with the clinical impression that the arthritis seen in this disorder is relatively benign.

Despite the apparent blandness of the arthritis in PSS at least 50% of those in the present study with articular involvement considered joint pain to be a significant manifestation of their illness. However, since many have flexion deformities of the fingers due to soft tissue contractures, the precise cause of the resulting disability is often difficult to define. Joint deformities such as MCP subluxation, ulnar deviation, and of the swan-neck variety, more typical of rheumatoid arthritis, were seen infrequently.

In the experience of Rodnan[6] erosive changes were rarely seen in PSS, the main roentgenographic findings being narrowing of the cartilage space and juxta-articular osteoporosis confined largely to the interphalangeal and carpal bones. That erosive changes do occur in PSS is now well established[10 12 13 19] and was seen in 40% of the present series. However, it is evident[11 19] that the destructive articular changes seen in this disease are frequently atypical of rheumatoid arthritis, in both their distribution and their radiological appearances. Single, small, discrete lesions are the general rule, and this reflects the relatively noninvasive nature of the synovium in PSS. We found no correlation between the occurrence of joint erosions and duration of disease, the presence of rheumatoid factor, antinuclear

antibodies, terminal tuft erosions, or extent of the skin involvement in the patients studied. The occurrence of calcinosis, however, was much more frequently seen in those having erosive disease.

The DIP joint was the most common site in this series for joint space narrowing. Many of these cases were associated with erosive changes and a radiological picture compatible with erosive osteoarthritis.[20 21] Since the majority of our patients were postmenopausal females, a group susceptible to developing erosive osteoarthritis, the occurrence of such an arthropathy unrelated to PSS could not be discounted. However, as has been the experience of Brower *et al.*,[19] the DIP lesions appear to be both deeper and more destructive than the usual picture of erosive osteoarthritis. A case is illustrated, indicating the rapid progression of this lesion over a period of 16 months. Pain was not a prominent feature associated with this DIP joint arthropathy, but impaired ability to make a fist due to soft tissue contractures was found to be a significant association and is likely to be an important aetiological factor. Soft tissue tightening, which could raise the intra-articular pressure and cause crush fractures of the articulating structures, has also been postulated as being responsible for the changes seen in primary osteoarthritis of the interphalangeal joints.[22] Lovell and Jayson[13] found no correlation between the presence of radiological evidence of articular manifestations and measurement of both grip strength and finger-palm flexion, but their analyses were not directed towards the involvement of any particular group of joints.

Selective involvement of the first carpometacarpal joint has been described in this disease.[12] Joint space narrowing was prominent in our 4 cases, but the marked erosive feature of the lesions described by Resnick *et al.*[12] was seen only in 2. With the exception of the latter characteristic, differentiation from primary osteoarthritis is difficult.

Murray Baron is a fellow and Peter Lee an associate of the Canadian Arthritis Society, to which we are grateful for continued support.

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