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Deforming arthritis in systemic lupus erythematosus

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SUMMARY Of 45 patients in the McGill Lupus Registry 6 were found to have Jaccoud's deformities. One of these 6 and 4 others had fixed flexion contractures of the elbows. Those with Jaccoud's deformities were similar to the other patients with systemic lupus erythematosus in all respects except that those with Jaccoud's syndrome had had their disease and their arthritis significantly longer. It is concluded that Jaccoud's deformities are the result of longstanding arthritis and that elbow contractures occur via a different mechanism.

Articular symptoms are the commonest presenting feature and clinical manifestation of systemic lupus erythematosus (SLE).[1 2] The non-deforming and nonerosive nature of the arthritis is distinctive,[3] and it is a diagnostic criterion of the disease.[4] While deforming arthritis in SLE was noted in early descriptions,[5] there remained difficulty in clearly distinguishing SLE from rheumatoid arthritis until the titered antinuclear antibody test, the Farr assay for detecting antibodies to DNA, and serum complement determinations became available and aided separation of these 2 entities.[6] Since then deforming arthritis has been considered to be an uncommon feature of SLE, occurring in less than 5% of cases,[6–9] though a higher incidence has occasionally been found.[2 10 11] Bywaters noted the similarity of the hand deformities in SLE to the Jaccoud's deformities that could occasionally follow rheumatic fever.[6]

Of the initial 45 SLE patients seen at the McGill Lupus Registry 6 had Jaccoud's deformities and 5 elbow contractures. The clinical features and laboratory abnormalities of these 2 groups of SLE patients with a deforming arthritis have been compared to those of the remaining patients.

Patients and methods

The McGill Lupus Registry was started in 1977, and 41 of the 45 patients included in this report have been followed up at the Montreal General Hospital. They have come from the rheumatology, immunology, and nephrology services. Since inception of the registry data have been collected prospectively in accordance with the American Rheumatism Association (ARA) standard database.[12] At the first visit the date of disease onset was determined from the patients and their records as the date at which a systemic illness developed or an episode meeting an ARA criterion not explained by another illness occurred.

Sjögren's syndrome was defined as keratoconjunctivitis sicca with xerostomia that was confirmed by either lip biopsy or an abnormal salivary scan.

All laboratory tests were performed at the Montreal General Hospital. Antibodies to deoxyribonucleic acid (DNA) were measured by the Farr assay[13] and rheumatoid factor was assayed by the sheep cell agglutination test.[14–15] The third and fourth components of complement were measured by nephelometry.

Results

Six patients had Jaccoud's syndrome (group A), and the deformities are noted in Table 1. Five had fixed flexion contractures of the elbows (group B) with from 5 to 60° limitation of extension, and 1 of these had Jaccoud's deformities. Three patients had symptomatic radiologically confirmed aseptic necrosis and they were included in the remaining 35 patients (group C).

All but 3 of the 45 patients met 4 ARA criteria. One patient in group B had a positive antinuclear antibody, a butterfly rash, discoid lupus, Raynaud's phenomenon, and arthritis, though the last could not be included because bilateral elbow contractures were present. Two patients with 3 criteria were

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Table 1 *Abnormalities in patients with Jaccoud's deformities*

	Ulnar deviation	*Swan-neck deformities*	*Hyperextension thumb*	*Subluxation MCPs*	*Radiographic cysts*	*Radiographic hooks*
Case 1	+	+ 2,3,4,5	−	+	+	+
Case 2	+	R 2,3 L 3,4,5	L	−	−	−
Case 3	+	+ 3,4,5	+	−	−	−
Case 4	+	R 3,4,5 L 4,5	R	−	−	−
Case 5	+	R 3,4 L 4,5	−	−	−	−
Case 6	+	+ 2,3,4,5	−	−	+	..−

+ =present bilaterally; − =absent; L=present on left; R=present on right; numerals refer to digit involved with swan-neck deformity.McP= metacarpophalangeal joints.

included in group C. One had arthritis, alopecia, and pericarditis and the other arthritis, a butterfly rash, and leucopenia. Both had positive antinuclear antibody tests and raised DNA binding tests.

There was no difference in the female to male ratio, the number of ARA criteria met, or the age of disease onset within the 3 groups (Table 2). Arthritis without deformity was excluded as a criterion in groups A and B, though its inclusion would not have produced a statistically significant difference between the groups. The individual criteria were found in a similar frequency in groups A, B, and C, and although Raynaud's phenomenon was most common in group A this increased frequency was not significant. Sjögren's syndrome occurred in 1 patient in group A and 3 in group C. The only significant difference was that the duration of disease and the duration of arthritis was longer in group A than either group B or group C (Table 2, $P<0{\cdot}001$).

The frequency of positive LE cell preparations, raised DNA binding tests, and hypocomplementaemia did not differ between the groups. A positive rheumatoid factor was present most commonly in group B, though this did not differ significantly from the other 2 groups (Table 2).

Corticosteroids had been used in 66%, 40%, and 63% of patients in groups A, B, and C respectively.

Table 2 *Characteristics of subgroups of SLE*

	Group A	*Group B*	*Group C*
Number of patients	6	5	35
No. female	4	3	29
No. ARA criteria	5·7±1·4	4·8±1·5	5·4±1·8
Age at disease onset (yr)	33 ±19	35 ±15	33 ±14
Duration of disease (yr)	22 ±12*	7·0±4·8	8·3±6·5
Duration of arthritis (yr)	22 ±11*	6·8±4·6	7·2±6·3†
Raynaud's phenomenon	5	2	15
Rheumatoid factor positive	2	2	3

*$P<0{\cdot}001$ by *t* test. †No. with arthritis=32. Group A: those with Jaccoud's deformities. Group B: those with elbow contractures. Group C: remainder with SLE. One patient is common to both group A and group B.

The deformities were reducible in all patients with Jaccoud's syndrome. None of the patients in either group A or group B had functional impairment and none had erosive disease radiographically. Two patients with Jaccoud's syndrome had small cystic lesions in several metacarpal heads, and an ulnar hook-like lesion was seen in 1. It was not possible accurately to date the onset of the deformities in those with Jaccoud's syndrome, though 2 of the 3 patients with the longest history of polyarthritis did not have radiographic cystic changes or hooks.

Discussion

Six of our 45 (13%) SLE patients had deforming arthritis in the hands. While Russell *et al.* found Jaccoud's deformities in 7 of 30 (23%) lupus patients referred to a rheumatic disease unit[11] and another group reported ligamentous laxity in 50%, swan-neck deformities in 38%, and thumb hyperextension deformities in 30% of 50 SLE patients,[16] Jaccoud's syndrome has been found in from 2·4 to 7·0% of the largest series of SLE patients reported since 1970.[2 6–9] Jaccoud's syndrome appears to be a late manifestation of longstanding arthritis,[7 11 17] as our study confirms, and with longer survival of SLE patients the frequency of deforming arthritis of the hands can be expected to increase.[18]

Jaccoud's deformities consist most commonly of ulnar deviation, metacarpophalangeal subluxation, swan-neck deformities, hyperextension deformities of the interphalangeal joint of the thumb, and less frequently boutonnière deformities.[2 6–11] The deformities are usually reducible and only rarely disabling, though the hyperextension deformity of the thumb can lead to a decreased pinch.[7 16]

Erosions are uncommon and when present tend not to progress.[2 6 19] Bywaters has noted the frequent appearance of radiographic hook-like deformities at the metacarpal heads,[6] and while it has been suggested that they may be a late finding in Jaccoud's

syndrome[20] we did not find them in our patients with the most longstanding arthritis. They are not specific and may be found in rheumatoid arthritis, osteoarthritis, pseudogout, and gout, in addition to Jaccoud's syndrome.[6 21]

Most authors suggest that the deformities arise from a periarthritis involving the joint capsule, ligaments, and tendons and the resultant muscular imbalance.[6 7 21 22] This has been confirmed pathologically in some cases.[20–22] It follows that, when the deformities are disabling, surgical correction should be directed at correcting the muscular and ligamentous imbalance rather than at the joint, which is usually normal.[9 22]

Both Russell *et al.*[11] and Kramer *et al.*[17] noted a markedly increased frequency of Sjögren's syndrome in those with Jaccoud's deformities, though we could not confirm this.[11 17]. In fact, the presence of Jaccoud's deformities correlated only with the duration of disease and arthritis.

Fixed elbow contractures have not been noted in other series, and the finding of rheumatoid factor in the sera of 2 of the 5 suggests that it may occur via a different mechanism. Loss of elbow extension is one of the earliest signs of a true arthritis of the elbow,[23] and these patients may have developed their deformity from a true synovitis rather than the periarthritis presumed to be the cause of Jaccoud's deformities. Unlike group A those with elbow contractures had not had their disease or their arthritis longer than those in group C, lending further support to the concept that this deformity is distinct from the Jaccoud's syndrome.

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