Deforming arthropathy or lupus and rhupus hands in systemic lupus erythematosus

R M van Vugt, R H W M Derksen, L Kater, J W J Bijlsma

Abstract

Objective—Although deforming arthropathy in systemic lupus erythematosus (SLE) is characterised by a number of manifestations, definitive criteria for the different forms have not yet been established. To define deforming arthropathy and its different types a study was undertaken of 176 SLE patients.

Methods—Using as criterion any deviation from any of the metacarpus finger axes 17 patients (16 women, one man) were identified with clinical deforming arthropathy. These patients were evaluated according to a standardised protocol that covered all known characteristics of deforming arthropathy. By means of “Jaccoud’s arthropathy index” three different forms were identified.

Results—Three patients had an erosive form of deforming arthropathy (or rhupus hand) such as those seen in frank rheumatoid arthritis (RA), eight patients were identified as having Jaccoud’s arthropathy (or lupus hand), and the remaining six patients had mild deforming arthropathy. Jaccoud’s arthropathy is characterised by severe deformation of the hands (ulnar deviation, swan neck deformities, and Z deformity of the thumb) and feet with multiple non-erosive subluxations, mild aching and little or no evidence of synovitis. All patients, but one, fulfilled just four criteria of the ACR classification and joint symptoms were always found to precede the diagnosis of SLE. Furthermore a remarkable association of Jaccoud’s arthropathy with fetal loss, thrombosis—both venous and arterial—and the presence of antiphospholipid antibodies was found.

Conclusions—These data suggest that Jaccoud’s arthropathy represents a subset of SLE. Subdivision of deforming arthropathy into several clinical forms can facilitate the clinical management of this disorder.


Involvement of the joints in systemic lupus erythematosus (SLE) (lupus arthropathy) is one of the earliest and most common manifestations of this multisystemic disease. The degree of involvement may range from minor arthralgia to severe deforming arthritis. Pain and stiffness are more common than objective abnormalities and the synovitis of SLE is generally transient, migratory, and reversible. Occasionally it may take a more chronic course, leading to joint deformity that affects predominantly the finger joints, the wrists, and the knees. In the extreme case the hand deformities form a typical picture of ulnar deviation and subluxations that together resemble that of rheumatoid arthritis (RA).

Bywaters was the first to point out the similarity between the deforming arthritis of SLE and that reported in 1869 by Jaccoud for recurrent rheumatic fever. Although Jaccoud’s arthropathy has been described together with a variety of disorders, no definitive criteria for diagnosis have been published so far.

Other forms of deforming arthropathy have not yet been described in detail and the question is whether the erosive form represents the coexistence of SLE and RA. The purpose of this study was to describe deforming arthropathy, identify the different forms and correlate these forms with the various clinical and laboratory features of the disease.

Methods

In accordance with Alarcon-Segovia et al we considered any deviation, assessed with an angle goniometer correctable or not, from any of the metacarpus finger axes as deforming arthropathy in SLE. Using this criterion we identified 17 patients with clinical deforming arthropathy of the hands in our total group of 176 patients (160 female and 16 male) who fulfilled the ACR revised criteria for the classification of SLE. The study group consisted of 16 women and one man with a mean age at onset of disease of 20.8 years (range 13–40) and a mean disease duration of 15.6 years (range 4–35).

These patients were evaluated according to a protocol that covered all known characteristics of deforming arthropathy. Physical examination included a detailed standardised examination of the hands and feet. The following items were evaluated in each case: signs of arthritis, ulnar deviation of fingers, metacarpo-phalangeal subluxation, swan neck deformities of the fingers, Z deformity of the thumb, boutonnière deformities of the thumb and fingers, ulnar drift and ulnar deviation of the hand. Other forms of deforming arthropathy have not yet been described in detail and the question is whether the erosive form represents the coexistence of SLE and RA. The purpose of this study was to describe deforming arthropathy, identify the different forms and correlate these forms with the various clinical and laboratory features of the disease. Other forms of deforming arthropathy have not yet been described in detail and the question is whether the erosive form represents the coexistence of SLE and RA.

Conclusions—These data suggest that Jaccoud’s arthropathy represents a subset of SLE. Subdivision of deforming arthropathy into several clinical forms can facilitate the clinical management of this disorder.

Deformities, grip strength (assessed with a Martin vigorimeter and calculated as the mean of three measurements), hallux valgus and hammer toes.

Patients were asked about Raynaud’s phenomenon, pain, and functional impairment of the hands and problems with the feet (pain or deformities necessitating semi-orthopaedic shoes or surgery). These items were scored as “positive” or “negative”.

Previous history with special attention to the interval between first joint symptoms and time of diagnosis, the number of ACR criteria for SLE, fetal loss, and venous and arterial thrombosis were obtained from medical records. Radiographs of hands and feet were reviewed according to a standard data form that included such details as (sub)luxation, joint space narrowing, metacarpal hook formation, and erosive changes.

The following serological studies were performed in all cases; antinuclear antibody (indirect immunofluorescence on Hep-2 cells), anti-dsDNA (Farr assay), anti-cardiolipin (enzyme immunoassay), lupus anticoagulant (LAC), anti-ENA (counter immunoelectrophoresis), and rheumatoid factor (Rose-Waaler test).

On the basis of these data a subdivision was made between evident erosive and the non-erosive forms of deforming arthropathy (fig 1). The patients of the non-erosive group were then assessed for the presence of definite Jaccoud’s arthropathy, using a “Jaccoud’s arthropathy index”, which is dependent upon the different clinical symptoms and the severity of the deformities (table 1). The remaining patients were classified as having mild deforming arthropathy.

Because of the small number of patients in the different groups Fisher’s exact test was used for statistical analysis.

Results

Patients with deforming arthropathy could be separated into three groups. The first group consisted of three patients with an erosive form

<table>
<thead>
<tr>
<th>Patient characteristics</th>
<th>Jaccoud’s arthropathy (n=8)</th>
<th>Erosive arthropathy (n=3)</th>
<th>Mild deforming arthropathy (n=6)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at onset (range)</td>
<td>23.3 (17–39)</td>
<td>19.6 (14–24)</td>
<td>23.8 (19–25)</td>
</tr>
<tr>
<td>Disease duration (range)</td>
<td>23.7 (11–39)</td>
<td>10.6 (4–18)</td>
<td>11.6 (8–17)</td>
</tr>
<tr>
<td>Functional impairment</td>
<td>3/8</td>
<td>2/3</td>
<td>3/6</td>
</tr>
<tr>
<td>Pain</td>
<td>1/8</td>
<td>2/3</td>
<td>4/6</td>
</tr>
<tr>
<td>Foot involved</td>
<td>8/8</td>
<td>1/3</td>
<td>0</td>
</tr>
<tr>
<td>Hypermobility</td>
<td>1/8</td>
<td>0</td>
<td>1/6</td>
</tr>
<tr>
<td>Synovitis wrist</td>
<td>0</td>
<td>3/3</td>
<td>0</td>
</tr>
<tr>
<td>Synovitis fingers</td>
<td>0</td>
<td>1/3</td>
<td>4/6</td>
</tr>
<tr>
<td>Pain at pressure</td>
<td>0</td>
<td>3/3</td>
<td>2/6</td>
</tr>
<tr>
<td>Swan neck deformity</td>
<td>3/8</td>
<td>1/3</td>
<td>3/6</td>
</tr>
<tr>
<td>1–4 fingers</td>
<td>5/8</td>
<td>1/3</td>
<td>0</td>
</tr>
<tr>
<td>5–8 fingers</td>
<td>7/8</td>
<td>2/3</td>
<td>0</td>
</tr>
<tr>
<td>Boutonniere deformity</td>
<td>2/8</td>
<td>1/3</td>
<td>0</td>
</tr>
<tr>
<td>Z deformity one or both thumbs</td>
<td>8/8</td>
<td>2/3</td>
<td>2/6</td>
</tr>
<tr>
<td>Mean grip strength (0–120 kpa)</td>
<td>31</td>
<td>33</td>
<td>44</td>
</tr>
<tr>
<td>Hallux valgus</td>
<td>4/8</td>
<td>1/3</td>
<td>1/6</td>
</tr>
<tr>
<td>Metatarsal subluxation</td>
<td>3/8</td>
<td>1/3</td>
<td>0</td>
</tr>
<tr>
<td>Hammer toes</td>
<td>5/8</td>
<td>0</td>
<td>1/6</td>
</tr>
</tbody>
</table>

Figure 1 Flow diagram by which the diagnosis of the three different forms of deforming arthropathy is reached.

Figure 2 Radiograph of erosive arthropathy (rhapus hand) showing joint space narrowing and rheumatoid-like bone erosions in the intercarpal and radiocarpal joints. Mild erosive damage of some MCP joints.

Table 2 Comparison of characteristics of the three types of deforming arthropathy

Figure 3 (A) Characteristic joint deviation with MCP subluxation, ulnar deviation, swan neck deformity of the fingers, and Z deformity of both thumbs in Jaccoud’s arthropathy of the hands (lupus hand). (B) Radiograph of same hands showing hook formation (arrows) and alignment disorders without noticeable erosions.
deforming arthropathy

**Table 3** Comparison of ACR criteria in the total SLE population and the three types of deforming arthropathy

<table>
<thead>
<tr>
<th>ACR criteria SLE</th>
<th>Jaccoud's arthropathy (n=8)</th>
<th>Erosive arthropathy (n=3)</th>
<th>Mild deforming arthropathy (n=6)</th>
<th>Non-deforming arthropathy (n=159)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Malar rash</td>
<td>2/8</td>
<td>2/3</td>
<td>5/6</td>
<td>99/159</td>
</tr>
<tr>
<td>Discoid rash</td>
<td>1/8</td>
<td>1/3</td>
<td>3/6</td>
<td>33/159</td>
</tr>
<tr>
<td>Photo-sensitivity</td>
<td>1/8</td>
<td>2/3</td>
<td>4/6</td>
<td>74/159</td>
</tr>
<tr>
<td>Oral ulcers</td>
<td>0</td>
<td>1/3</td>
<td>3/6</td>
<td>32/159</td>
</tr>
<tr>
<td>Arthritis</td>
<td>8/8</td>
<td>3/3</td>
<td>6/6</td>
<td>128/159</td>
</tr>
<tr>
<td>Serositis</td>
<td>0</td>
<td>1/3</td>
<td>3/6</td>
<td>49/159</td>
</tr>
<tr>
<td>Renal disorder</td>
<td>1/8</td>
<td>2/3</td>
<td>2/6</td>
<td>90/159</td>
</tr>
<tr>
<td>Neuropsychological disorder</td>
<td>1/8</td>
<td>0</td>
<td>1/6</td>
<td>15/159</td>
</tr>
<tr>
<td>Haematological disease</td>
<td>6/8</td>
<td>2/3</td>
<td>2/6</td>
<td>123/159</td>
</tr>
<tr>
<td>Anti-nuclear antibody</td>
<td>8/8</td>
<td>3/3</td>
<td>6/6</td>
<td>156/159</td>
</tr>
<tr>
<td>Immunological disorder</td>
<td>5/8</td>
<td>3/3</td>
<td>6/6</td>
<td>143/159</td>
</tr>
</tbody>
</table>

*ENA was only performed in 135 of 159 patients.
†Four of eight and 66 of 159 patients had been pregnant.

**Table 4** Comparison of laboratory data and thrombotic history of the three types of deforming arthropathy and non-deforming arthropathy

<table>
<thead>
<tr>
<th>Laboratory data/thrombotic history</th>
<th>Jaccoud's arthropathy (n=8)</th>
<th>Erosive arthropathy (n=3)</th>
<th>Mild deforming arthropathy (n=6)</th>
<th>Non-deforming arthropathy (n=159)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Livedo reticularis</td>
<td>4/8</td>
<td>0</td>
<td>0</td>
<td>47/159</td>
</tr>
<tr>
<td>Raynaud’s phenomenon</td>
<td>8/8</td>
<td>3/3</td>
<td>0</td>
<td>81/159</td>
</tr>
<tr>
<td>Arterial/venous thrombosis</td>
<td>5/8</td>
<td>0</td>
<td>1/6</td>
<td>38/159</td>
</tr>
<tr>
<td>Fetal loss</td>
<td>4/4†</td>
<td>0</td>
<td>0</td>
<td>25/159</td>
</tr>
<tr>
<td>Rose-Waaler test</td>
<td>0</td>
<td>3/3</td>
<td>1/6</td>
<td>not known</td>
</tr>
<tr>
<td>LAC</td>
<td>6/8</td>
<td>0</td>
<td>1/6</td>
<td>99/159</td>
</tr>
<tr>
<td>Anticardiolipin IgG</td>
<td>7/8</td>
<td>1/3</td>
<td>2/6</td>
<td>73/159</td>
</tr>
<tr>
<td>Anticardiolipin IgM</td>
<td>4/8</td>
<td>0</td>
<td>1/6</td>
<td>49/159</td>
</tr>
<tr>
<td>Anti-U1RNP</td>
<td>0</td>
<td>2/3</td>
<td>0</td>
<td>19/159*</td>
</tr>
<tr>
<td>Anti-Sm</td>
<td>1/8</td>
<td>0</td>
<td>2/6</td>
<td>26/159*</td>
</tr>
<tr>
<td>Anti-SSA</td>
<td>1/6</td>
<td>0</td>
<td>1/6</td>
<td>36/159*</td>
</tr>
<tr>
<td>Anti-SSB</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>8/159*</td>
</tr>
</tbody>
</table>

All patients, but one, of the Jaccoud’s arthropathy group fulfilled just four criteria of the ACR classification (table 3) and joint symptoms were always found to precede the diagnosis of SLE (mean 13 months, range 6–36 months). Five of them had a history of arterial or venous thrombosis and four of them had had a fetal loss. Renal disorder was the only ACR criterion significantly lower (p=0.02) and fetal loss significantly higher (four of four and 25 of 66 patients who had been pregnant respectively, p=0.022) in patients with Jaccoud’s arthropathy group compared with patients without deforming arthropathy.

Table 4 gives the laboratory profiles and the history of thrombosis. Seven of the eight patients with Jaccoud’s arthropathy were positive for anticardiolipin (IgG positive in seven cases and IgM positive in four). In six of the seven patients positive for IgG anticardiolipin LAC was also present. In the other two groups only three patients were positive for IgG, one of them also for IgM anticardiolipin. The prevalence of LAC and IgG anticardiolipin was significantly higher in patients with Jaccoud’s arthropathy compared with patients without deforming arthropathy (seven of eight patients and 73 of 159 patients, p=0.028). All patients with joint erosions were positive for rheumatoid factor. In the other two groups radiological changes were minimal (table 5).

Irregularity of bone (quite unlike that in RA) was seen in five of the eight patients with Jaccoud’s arthropathy. Two of them with marked subluxation showed hook formation of the MCP joints. Four patients showed scapholunate dissociation (fig 4). All patients with Jaccoud’s arthropathy had some kind of deformity of the feet (hallux valgus, hammer toes and/or subluxation of metatarsophalangeal joints).

**Discussion**

Bywaters was the first to point out the similarity between the deforming arthropathy of SLE and that originally described in 1869 by Jaccoud for recurrent rheumatic fever; he reported an incidence of five per cent. The clinical picture of Jaccoud’s syndrome is characterised by chronic non-erosive deformities (subluxation of the MCP joints, ulnar deviation, which is correctable in the early stages, hyperextension at the proximal interphalangeal joints) that superficially resemble...
Deforming arthropathy or lupus and rhupus hands in SLE

Deforming arthropathy or lupus and rhupus hands in SLE

543

Alarcon-Segovia

approach in their study.7 Alarcon-Segovia

index to ensure a standard

cal relation between the extensor carpi ulnaris

resulting from subluxation. The close anatomi-
capsule and the altered mechanical forces

rub caused by overlying inflamed tendons or

ect of synovitis. The irregularity

muscles acting across the hand rather than the

combination with the compressive forces of the

periarticular fibrosis or synovial vasculitis) in

straint (resulting from subsequent capsular

consequence of the loss of ligamentous con-
in Jaccoud’s arthropathy seems to be the

underlying mechanism. The deformity seen

both the synovial membrane and the capsule as

recurrent low grade inflammatory activity in

tered patients with scant and asymmetric

atients also included patients with scant and asymmetric

joints. When we used Alarcon-

Segovia’s criterion to define patients with deforming arthropathy, subdivided this group into those with an evident erosive and those with a non-erosive form and then applied “Jac-
could’s arthropathy index” to the non-erosive

group, we identified three forms of deforming arthropathy (Jaccoud’s arthropathy, erosive arthropathy, and mild Arthritic arthropathy).

Arthritis associated with SLE is usually non-
erosive, but in a small subset of patients an ero-
sive disease resembling RA develops. We iden-
tified erosive arthropathy of the hands (rhubus

hand) in three patients (1%), which is similar to the 1–2% reported by Dubois.2 Whether these patients represent a subset of SLE arthri-
tis or the true coexistence of the two diseases is not clear, but there probably is a real overlap. The serological overlap between RA and SLE is well known with up to 20 per cent of RA patients exhibiting positive antinuclear anti-
bodies (ANA). However, patients with con-
comitance of RA and SLE are rare, as there are only a small number of well documented cases in literature.1

Whereas in RA loss of bone and joint stabil-
ity are secondary to hypertrophic synovitis, Jaccoud’s arthropathy seems to involve mainly ligaments and periarticular soft tissue. Based on histological findings, such as mild but typi-
cal fibrous synovitis with little or no round cell infiltration and microvascular changes, Bywaters and others4 reported a prolonged or recurrent low grade inflammatory activity in both the synovial membrane and the capsule as the underlying mechanism. The deformity seen in Jaccoud’s arthropathy seems to be the consequence of the loss of ligamentous con-
straint (resulting from subsequent capsular periarticular fibrosis or synovial vasculitis) in combination with the compressive forces of the muscles acting across the hand rather than the destructive effect of synovitis. The irregularity of bone, which is quite unlike the erosions seen in RA, is thought to be attributable to friction rub caused by overlying inflamed tendons or capsule and the altered mechanical forces resulting from subluxation. The close anatomi-
cal relation between the extensor carpi ulnaris
tendon (as part of the ulnocarpal complex) and

the ulnar styloid may explain the high preva-

cence of erosive damage to this bone mentioned in the studies of Alarcon-Segovia1 and Reilly.5

flammation of the tendon probably plays a

major part in the pathogenesis of sudden
tendon rupture, which is uncommon in SLE

but has been described in several case reports.8

Another characteristic of deforming ar-

thropathy observed and used as criterion by

Alarcon-Segovia was non-erosive carpal collapse.5 In RA hypertrophic synovitis is

responsible for disruption of the ligamentous

sling within the wrist, producing the joint

changes that lead to carpal collapse and

subluxation of the joints.5 In four patients with

Jaccoud’s arthropathy we found scapholunate
dissociation and rotation of the scaphoid itself

that had to be the consequence of weakening or

slackening of the (radio)-carpal ligaments. It is

interesting that Bywaters4 had already noted

the gross subluxation of the wrist and radiographs printed in several publications10 11 showed evident, scapholunate luxation or ulnar translocation of the wrist that was not de-
scribed. For patients with Jaccoud’s arthropa-

thy the estimated prevalence of articular hyper-
mobility lies between 7 and 50 per cent, depend-

ing upon the method of evaluation used.12 Although hypermobility in the development of Jaccoud’s arthropathy seems plausible we found no association.

Jaccoud’s arthropathy seems to be a general-
ised pathological capsular and periarticular

condition involving all joints. Although most

reports have focused on the hands, Morley et

al5 reported three cases of deforming arthropa-

thy of the feet in SLE and coined the term “lupus foot”. In accordance with Mizutani and

Reilly6 we also found a significant association

with deformities of the feet (hallux valgus and/or subluxation of metatarsophalangeal joints) in patients with Jaccoud’s arthropathy.

Given the wide variety of clinical features

associated with SLE, there have been many

attempts to identify subsets of patients for

whom a given antibody specificity can be iden-
tified with deforming arthropathy. Several

associations, such as the presence of antibodies

to U1 RNP,5 RA 33,11 SS-A/Ro and

SS-B/La,12 have been reported previously. We

noted that the patients in our group with

Jaccoud’s arthropathy represent a subset of

SLE. Although all of these patients had clinical

features or serological abnormalities suggestive

of SLE (mainly arthritis, ANA+, leuco-

trombocytopenia, rash) it took several years to

satisfy just four of the ACR classification crite-

ria for the diagnosis of SLE. Previously these

patients were considered as having a lupus-like

syndrome. Furthermore in our study there

seems to be a remarkable association with fetal

loss, thrombosis—both venous and arterial—

and the presence of antiphospholipid antibod-

ies. Our data suggest a striking coexistence of

Jaccoud’s and the antiphospholipid syndrome6

although we cannot explain the pathogenetic

link. It is possible that small vessel vasculopathy plays a part in the genesis of the periarticular

fibrosis. Some evidence for this hypothesis

could be found in “fibrin like material
obliterating small vessel lumens” described in certain synovial biopsy specimens. Bywaters already reported a correlation with mitral stenosis and Libman-Sacks endocarditis, both said to be associated with antiphospholipid syndrome. Sturgess et al and Palazzo et al described patients with hypocomplementaemic urticarial vasculitis syndrome and the combination of Jaccoud’s syndrome, valvulopathy and vasculopathy of the small cutaneous vessels.

The group with mild deforming arthropathy did not differ in any respect from the SLE population without deforming arthropathy.

In conclusion, allowing for the limitations of a definition of deforming arthropathy, we have presented some clinical guidelines for subdividing deforming arthropathy into several clinical forms that are relevant in clinical practice for early diagnosis and the differential diagnosis of SLE (for instance, distinguishing it from RA).

Patients with Jaccoud’s arthropathy were characterised by a slow evolution into classifiable SLE, distinct radiological features, association with foot involvement and association with the presence of lupus anticoagulant and antinuclear antibodies. We believe that these guidelines will facilitate the clinical management of deforming arthropathy.

In conclusion we have presented some guidelines that facilitate the division of lupus related deforming arthropathy, into distinct subgroups with differing clinical and serological characteristics. Patients with Jaccoud’s arthropathy are notable for the slow evolution of their disease into classifiable SLE, unique radiological features, foot involvement and the association with lupus anticoagulant and antiphospholipid antibodies, while those with a “mild deforming arthropathy” do not seem to differ in any respect from SLE patients without arthropathy. We believe that defining lupus arthropathy in the manner described will assist in clinical management, from the time of diagnosis, for example, in distinguishing RA from SLE, through treatment decisions based on likely progression.

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