Jaccoud's Arthropy

Report of Seven Cases

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SUMMARY

Seven patients with severe rheumatic heart disease and joint deformities, in whom there were no signs of active arthritis, are described. They fulfill the criteria for the diagnosis of Jaccoud's arthropathy (or 'chronic post-rheumatic arthritis'). We postulate that hypermobility may be a predisposing factor in the development of the deformity. This rare condition should be distinguished from rheumatoid arthritis because of its relatively favourable prognosis.


It is generally accepted that rheumatic fever and rheumatoid arthritis are distinct entities. The former produces structural damage, predominantly in the heart, with characteristically transient joint problems, whereas the latter is usually responsible for irreversible articular damage and deformity in the absence of obvious cardiac disease. Occasionally, however, a patient presents with both chronic valvular heart disease and deforming peripheral arthropathy and, in such a case, 3 possibilities arise: (a) the patient may have rheumatoid arthritis with endocardial involvement as an extra-articular manifestation of the disease; (b) rheumatoid arthritis and rheumatic heart disease may occur coincidentally in the same patient; or (c) chronic joint damage as well as valvular heart disease may be produced by rheumatic fever.

Bywaters' and Weintraub and Zvaifler have shown clearly that patients with rheumatoid arthritis may develop rheumatoid valve damage or may have coincidental rheumatic heart disease. In addition, however, there is a small group of patients with severe rheumatic heart disease who have a deforming arthropathy which involves the hands, and sometimes the feet as well, and which appears to be distinct from rheumatoid arthritis. Bywaters has drawn attention to the fact that this condition of 'chronic post-rheumatic arthritis' was first described by Jaccoud in Paris in 1869, and his name is now generally used eponymously for this arthropathy. (The term 'arthropathy' is preferable to 'arthritis', since there is no active inflammation in this condition.)

Jaccoud's arthropathy is a rare condition. Between 1950 and 1975, 14 cases were reported in the English-language literature, and although some authors feel that it is a variant of rheumatoid arthritis, others believe that there is good evidence for its acceptance as a distinct entity.

Over a period of 4 years (1973 - 1976) we have seen 7 patients with rheumatic heart disease and hand deformities which are distinguishable from those of rheumatoid arthritis, and we wish to report them as further examples of Jaccoud's arthropathy.

CASE REPORTS

Patient 1

A 30-year-old man had been well until the age of 19, when he developed acute rheumatic fever. After recovery he continued to have attacks of intermittent joint pain. In 1967, at the age of 23, he was treated as an outpatient for severe joint pains of 2 months' duration. No objective evidence of joint changes was found and his heart was reported to be normal. Three years later, he was found to be in cardiac failure with mitral incompetence, tricuspid stenosis and tricuspid incompetence. In 1972 he was given a mitral valve prosthesis.

He was admitted to a medical ward at the beginning of June 1973 with fever, chest pain and intermittent joint pain. Physical examination revealed an ill, pale and jaundiced young man. There were no stigmata of chronic liver disease or infective endocarditis, and he was not in cardiac failure. There were signs of free aortic incompetence and a functioning Starr-Edwards mitral valve prosthesis. His lungs were clear. The liver and spleen were enlarged and palpable below the costal margin. The patient's neurological system was normal. Examination of the joints revealed deformities of the hands and feet, but no active arthritis. There was marked ulnar deviation and some palmar subluxation of the metacarpophalangeal joints of both hands. The joints were lax and hyperextensible and the deformity was correctable both passively and actively (Fig. 1). There was no deformity of the interphalangeal joints. The right hand was more severely involved than the left. There was no joint or tendon crepitus. In the feet, there was bilateral hallux valgus. All other joints were clinically normal. Laboratory studies showed evidence of an active haemolytic anaemia, which was later demonstrated to be thalassaemia minor. Tests for rheumatoid factor and antinuclear factor were negative. X-ray films of the patient's hands showed no evidence of joint destruction, and there was a questionable hook-like erosion at the metacarpal head of the left index finger.
The patient's acute illness eventually settled spontaneously, and since his discharge he has remained well. There is good control of his cardiac symptoms and he suffers only occasional minor episodes of joint pain.

Patient 2

A 39-year-old woman had suffered an attack of rheumatic fever at the age of 7, but had not received any treatment for it. Shortly thereafter she developed poliomyelitis, which affected her right arm, but after this the remainder of her childhood was said to have been unmarked by serious illness. At the age of 33 she was subjected to mitral valvotomy, at which time mild aortic incompetence and aortic stenosis were noted. She was readmitted in April 1974 for recurrence of her cardiac symptoms. There was no clear history of recurrent attacks of rheumatic fever, but for several years the patient had suffered from episodes of arthralgia in the knees and elbows and, more recently, in the hands as well. Shortly before her admission, she had had an attack of fairly severe migratory joint pains associated with the development of red patches on the skin around the affected joints. Her hand deformity appeared as the acute attack settled and at the time of her admission she was essentially free of joint pain. Physical examination revealed no signs of cardiac failure, but the murmurs of mitral incompetence, aortic incompetence and aortic stenosis were heard. Her right hand was atrophic and wasted from her childhood poliomyelitis. Her left hand showed mild swelling of the second and third metacarpophalangeal joints. There was moderate ulnar deviation of the fingers, which was correctable both passively and voluntarily. There was minimal tenderness of the affected joints. The interphalangeal joints were normal and there was no joint or tendon crepitus. There was a hallux valgus of the left foot, but the right foot was normal. All other joints appeared normal. An X-ray film of the left hand showed minimal ulnar deviation of the fingers with intact joint surfaces. Questionable hook lesions were discernible on the second and fifth metacarpal heads. Tests for rheumatoid and antinuclear factors were negative.

Patient 3

A woman aged 67 had had rheumatic fever at the ages of 16, 21 and 38 years. In each case she had experienced a fleeting polyarthritis and had been left with valvular heart disease. She had recently been treated with digoxin and a diuretic for atrial fibrillation and mild cardiac failure. Apart from the episodes of rheumatic fever, she had had no other joint disease, but she had noticed the gradual development of ulnar deviation of the fingers. She was first seen by us during an admission to hospital for treatment of a peripheral arterial embolism.

On examination, the typical changes of Jaccoud's arthropathy, i.e. flexion and ulnar deviation of the metacarpophalangeal joints of both hands, were found to be present. This deformity was not fixed, being easily correctable both actively and passively. There was also hyperextension of the proximal interphalangeal joints of all her fingers. There was no active synovitis in the joints. There were no rheumatoid nodules, but there was bilateral hallux

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Fig. 1. Patient 1 — left hand. Deformity of the thumb and metacarpophalangeal subluxation are evident. Ulnar deviation of the fingers has been completely corrected.

Fig. 2. Patient 3 — radiograph of hands showing well-preserved joint surfaces. Ulnar deviation evident in right little finger. Hook lesions visible in the original plates are not well shown.
valgus. The patient had the features of mitral stenosis and incompetence.

X-ray films of the chest showed a heart markedly enlarged in its transverse diameter with a mitralized contour. The lung fields were congested. X-ray films of the hands revealed features consistent with those of Jaccoud's arthropathy, apart from minimal osteoporosis around the small joints. There were some hook-like erosions at the metacarpophalangeal joints (Fig. 2). Tests for rheumatoid factor and lupus erythematosus cells were negative. Anti-streptolysin-O titre was less than 200 Todd units.

Patient 4

A 14-year-old boy presented during his second attack of rheumatic fever. This was characterized by a subacute arthritis that had been present for 4 months and involved his fingers, wrists and ankles. There was low-grade fever, early clubbing of the fingers and the signs of established aortic and mitral valve disease.

Examination of his hands showed a flexion deformity of the metacarpophalangeal joints of both hands, ulnar deviation of the fingers and hyperextension at the proximal interphalangeal joints.

Leucocytosis, elevated erythrocyte sedimentation rate and significant antistreptolysin-O titre were found. A β-haemolytic streptococcus was cultured from his throat. Tests for rheumatoid factor were negative and there were no radiological signs of erosions or destructive joint disease. The joint manifestations were voluntarily completely correctable.

Patient 5

A 40-year-old woman had had two attacks of rheumatic fever, which had started in adulthood. The first attack presented as a subacute polyarthritis with nodules in the tendo Achillis and was associated with an elevated antistreptolysin-O titre, a positive culture from the throat, fever, and leucocytosis. The patient was hospitalized for 4 weeks.

She presented for the second time 2 years later with a similar illness characterized by a sore throat, fever and malaise, followed by a polyarthritis. Clinical examination on this occasion revealed a subacute polyarthritis involving the metacarpophalangeal joints of both hands, wrists, knees and ankles. Several small nodules were palpable over the extensor tendons of the right hand. There were signs of established mitral valve disease. Leucocytosis, fever, elevated erythrocyte sedimentation rate and a raised antistreptolysin-O titre were present. Tests for rheumatoid factor were negative. X-ray examination showed some soft-tissue swelling, but no erosive arthritis. Biopsy of a tendon nodule showed features of a rheumatic fever nodule.

She was treated with soluble aspirin in full doses and, after protracted hospitalization, she was finally discharged after the resolution of the arthritis. Attempts were made to maintain penicillin prophylaxis, but she was lost to follow-up. Two years later, when seen again, she showed no active synovitis, but there was a mild flexion deformity of the metacarpophalangeal joints of both hands, ulnar deviation of the little fingers and hyperextension of the proximal interphalangeal joints of the ring and little fingers of both hands. The deformity was painless and voluntarily correctable. X-ray films showed no abnormality. Tests for rheumatoid factor were negative.

Patient 6

A woman aged 40 years presented with a 3-week history of an influenza-like illness followed by a polyarthritis which affected the ankles, the right knee, and the wrists, shoulders and fingers. Before this she had been in perfect health. On examination the woman was found to be ill, with acute arthritis involving the ankles, the right knee, the wrists and the metacarpophalangeal and proximal interphalangeal joints of both hands in an asymmetrical manner. Cardiac examination revealed the features of mixed mitral valve disease and aortic incompetence. There was a transient pericardial friction rub. The leucocyte count was elevated and the erythrocyte sedimentation rate was 100 mm/1st h. A β-haemolytic streptococcus was cultured from the throat. The antistreptolysin-O titre was elevated to 1 250 Todd units. Rheumatoid factor was negative.

After a period of 5 weeks the patient was discharged and was followed up as an outpatient. She was seen regularly and complained intermittently of joint pain, for which she had to take salicylates. Examination of her hands 18 months later revealed marked hyperextension of the proximal interphalangeal joints which was voluntarily correctable. There was ulnar deviation of both little fingers and a mild correctable flexion deformity at the metacarpophalangeal joints of both hands (Fig. 3). The features of established mixed mitral and aortic valve disease were again documented. Serological tests for rheumatoid factor were negative and X-ray films of her hands showed no abnormality.

Fig. 3. Patient 6 — deformity mainly involving little fingers. Ulnar deviation has been corrected.

Patient 7

A 67-year-old woman was referred to the arthritis clinic because of a supraspinatus tendinitis of the left
shoulder. She was uncommunicative and it was difficult to elicit an adequate history, but she had apparently only once before experienced any joint pain (in her hands, 4 years previously) and she denied ever having had acute rheumatic fever. She was, however, known to have suffered from rheumatic heart disease for at least 9 years, and in 1969 she had undergone a closed mitral valvotomy. On that occasion, the presence of 'chronic rheumatoid arthritis' was noted, but no comment was made about her joints. At the time of her referral, she was receiving medical therapy for cardiac failure and had the signs of mitral stenosis and incompetence and aortic incompetence.

Apart from her shoulder problem, the patient was found to have mild correctable ulnar deviation and slight subluxation of the metacarpophalangeal joints of both hands. More striking was the hyperextension of the proximal interphalangeal joints of all her fingers (Fig. 4). Hand closure (Fig. 5) and other hand function was excellent. There was no swelling or tenderness of her finger joints. In addition to her hand deformity, she was found to have hyperextension of the knees and the right elbow. Radiologically, the joint surfaces appeared normal. The erythrocyte sedimentation rate was normal and rheumatoid factor tests were negative, but the patient had an antinuclear factor titre of 100. A careful search revealed no evidence of collagen-vascular disease, and this finding was therefore considered to be of no significance.

Criteria for the diagnosis of Jaccoud's arthropathy have been proposed by Bywaters, with modifications by Zvaifler and Murphy and Staple and may be enumerated briefly as follows:

1. There is a history of recurrent, severe attacks of rheumatic fever.
2. Recovery is slow, with joint stiffness, and is followed by the insidious development of joint deformity.
3. Pathology reveals that the lesion appears to be fibrosis of the peri-articular tissues, fascia and tendons, and not synovitis.
4. The deformity consists of ulnar deviation with flexion and/or subluxation of the metacarpophalangeal joints. These deformities are characteristically correctable. There may be slight soft-tissue swelling and associated hyperextension of the proximal interphalangeal joints.
5. Tendon crepitus may be elicited.
6. The disease appears to be relatively inactive, with minimal symptoms, little evidence of synovitis, good hand function and a normal erythrocyte sedimentation rate.
7. Radiologically, there is little or no bone damage, which is often in striking contrast to the degree of deformity. In some cases, characteristic hook lesions may be seen on the radial and palmar aspect of the metacarpal heads.
8. Tests for rheumatoid factor should be negative.

The 7 cases reported in this paper fit in well with these criteria. All the patients but one had had attacks of severe rheumatic fever and all had serious cardiac lesions. In some there seems to have been slow resolution with joint stiffness after at least one attack. None of our patients was able to remember clearly the rate of onset of joint deformity, which suggests that it was probably insidious in each case. The joint deformities tallied closely with the classic description, both clinically and radiologically. In all cases, deformities were correctable, the disease appeared inactive, erythrocyte sedimentation rates were normal and tests for rheumatoid factors were negative.
Tendon crepitus was not elicited, but this does not seem to have been a feature of most of the reported cases in the literature. Bywaters mentions it in one of his cases and it has been reported on two other occasions. Interestingly, in one of these cases there was a feature which would tend to exclude a diagnosis of Jaccoud's arthropathy, namely a positive rheumatoid factor, while in the other there was the unusual feature of subcutaneous nodules of the rheumatoid type. We suggest that tendon crepitus be dropped from the list of diagnostic criteria.

Arcuiartic tissues were not available for histological examination in these patients, so we are not able to offer any comment on the pathological aspects of their diagnosis. The pathogenesis of this unusual sequel to rheumatic fever has not been clarified. The most comprehensive report which included pathological data was published by Bywaters, who considered the deformity to result from pericapsular and tendon fibrosis. Rheumatic fever predominantly affects the deeper layers of the synovium and capsule and it may be assumed that the evocative factor is the inflammatory episode in these sites. Small-joint arthritis is a documented feature of some cases of prolonged rheumatic fever and we have had the opportunity to see this inflammatory episode precede the deformity by 2 and 1½ years respectively in 2 patients (patients 5 and 6). The inflammatory episode in both these patients was non-specific and was attended by moderate joint thickening, stiffness, mild loss of function and a reaction to salicylates that was slower than expected in rheumatic fever. At the time of discharge, neither of these patients had any residual deformity or joint swelling. There are a few reports of patients who did not have preceding acute rheumatic fever or evidence of established valve disease. It may be argued that they represent disease of silent onset, analogous to the silent development of valvular heart disease, but this must be unusual in view of the frequent reference in the literature to the severity of the attacks of rheumatic fever preceding the deformity. The frequency with which rheumatic fever nodules are reported may be cited as evidence in favour of severe attacks. In the present series, only patient 7 had no history of acute rheumatic fever and this may be an example of silent onset of both heart and joint lesions.

Those who have previously written on the subject of Jaccoud's arthropathy have assumed that the development of the characteristic hypermobile deformity is simply the result of previous inflammatory episodes. We wish to postulate an additional factor. Joint hypermobility has been accepted as part of the rheumatic fever diathesis and, in one study, hypermobility of the metacarpophalangeal joints was demonstrated in subjects with rheumatic fever when compared with controls. It was proposed as another clue to the diagnosis of rheumatic fever. No prospective studies have been published on the development or outcome of rheumatic fever in subjects with hypermobile joints, but it is possible that hypermobility combined with an inflammatory reaction of suitable duration and intensity could give rise to the characteristics of Jaccoud's arthropathy. In this regard, the hypermobile knees and elbow noted in patient 7 may be of some significance.

Finally, are there any practical implications in making a diagnosis of Jaccoud's arthropathy? The answer, we believe, is 'yes', since, although there is joint deformity, there does not appear to be the threat of chronic pain and loss of function that would be implied by a diagnosis of rheumatoid arthritis. A striking feature of many of the cases in the literature is the inactivity and non-progressive nature of the disorder. In these patients the deformity had been present for periods of 12-38 years without pain, loss of function or joint surface damage. One of our patients, patient 7, had excellent, painless hand function 7 years after having been labelled as having 'chronic rheumatoid arthritis'. By contrast, when genuine rheumatoid arthritis develops in a patient with rheumatic heart disease, the joint disease progresses in the expected manner. Most patients with Jaccoud's arthropathy have severe valvular heart disease and it will undoubtedly be reassuring to them to know that they will not be further crippled by their joint disease.

REFERENCES