JACCOUD'S ARTHRITIS

A CASE REPORT

BY

EITHNE BEAUSANG, EUGENE V. BARNETT, AND SIDNEY GOLDSTEIN

From the Department of Medicine of the Rochester General Hospital and the Department of Medicine of the University of Rochester School of Medicine and Dentistry, N.Y., U.S.A.

A woman admitted for cardiac catheterization was found to have chronic arthritis of both hands. She gave a history of many recurrent attacks of rheumatic fever with gradual development of her joint deformity. On further study, these changes were found to consist of marked ulnar deviation of the fingers at the metacarpophalangeal joints and hyperextension of the proximal interphalangeal joints of the 2nd, 3rd, 4th, and 5th digits unaccompanied by clinical evidence of bone destruction. Serological tests for rheumatoid factors were negative and the patient had a normal erythrocyte sedimentation rate. Her catheterization data showed severe aortic stenosis. The above results conform to the criteria outlined by Jaccoud (1869) as necessary for the diagnosis of chronic post rheumatic fever arthritis.

The relationship of chronic deforming arthritis and valvular heart disease is complex and has been the source of a large number of investigations both clinical and pathological. It is now apparent that rheumatic fever is a late non-suppurative complication of Group A streptococcal infection manifested clinically by many symptoms (Master, 1944; Massell and Jones, 1944; Hubble, 1943), the most typical of which is migratory polyarthritis. Permanent joint deformity is not one of the criteria used in the diagnosis of rheumatic fever (Duckett Jones, 1944).

Rheumatoid arthritis may also present as an acute febrile polyarthritis but is not usually associated with streptococcal infection. A distinction between the two varieties of arthritis may be made

on the basis of clinical history and the results of certain laboratory tests, the most important of which are the serological tests for rheumatoid factors which are negative in patients with rheumatic fever (Rose, Ragan, Pearce, and Lipman, 1948). However, occasional cases may still present difficulty in differentiation. Between 20 and 50 per cent. of patients who suffer from rheumatic fever eventually develop chronic heart disease (Coombs, 1924; Arnsø, Brøchner-Mortensen, and Hastrup, 1951) with one or more valves involved. To add to the complexity of the problem, it is now known that, in 12 to 50 per cent. of autopsied cases of rheumatoid arthritis, valvular lesions indistinguishable from those of rheumatic heart disease are present (Ragan and Snyder, 1955; Kellgren and Lawrence, 1956). In ankylosing spondylitis also (Graham and Smythe, 1958), a disease similar to rheumatoid arthritis but usually distinguishable from it, there is frequently a dilatation of the root of the ascending aorta and aortic insufficiency.

The above facts would presuppose a possible relationship between cardiac and articular disease which is further suggested by the fact that rheumatic fever itself may produce chronic joint deformity. Jaccoud (1869) gave the first detailed description of chronic arthritis after repeated attacks of rheumatic fever. Sporadic reports of similar cases have appeared in the literature (Bywaters, 1950; Thomas, 1955; Zvaifler, 1962; Twigg, 1963), but Jaccoud's arthritis has not been considered a separate clinical entity by many (Hollander, 1960; American Rheumatism Association, 1964). The clinical features of the disease are those of marked ulnar deviation of the fingers at the metacarpophalangeal joint and hyperextension of the proximal interphalangeal joints of the 2nd, 3rd, 4th, and 5th digits unaccompanied by clinical evidence of bone destruction. This paper will present one case which fulfils the criteria for the diagnosis of this rare form of arthritis.

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†Senior Investigator, Arthritis Foundation, present address: Department of Medicine, Center for the Health Sciences, University of California, Los Angeles, California.

Reprint requests to: Rochester General Hospital, 1425 Portland Avenue, Rochester, New York, 14621. U.S.A.

Case Report

A 50-year-old white female restaurant hostess was in good health until the age of 11 years, at which time she had rheumatic fever characterized by arthritis, fever, and chorea. During the next 3 years at 6-monthly intervals she suffered from recurrent episodes of arthritis involving the ankles and wrists and the small joints of the hands. At the age of 16 she had a serious recurrence with a sore throat and fever and later swelling and erythema of the right ankle. At this time strict rest in bed at home was prescribed. She was treated with salicylates and some red liquid heart medication and was told that she had a heart murmur and definite heart involvement. Over the next 9 months she gradually recovered, but states that during her teens she had decreased exercise tolerance and that she noted, with the passage of time, gradual disfigurement of her hands with progressive ulnar deviation and flexor deformation most noticeable in the 3rd, 4th, and 5th metacarpophalangeal joints bilaterally. At the age of 30 she became pregnant but a therapeutic abortion and a bilateral tubal ligation were performed, because of severe shortness of breath. At age 35 she was admitted to the Rochester General Hospital because of shortness of breath and paraesthesia. Digitalization was attempted but had to be discontinued because of nausea and vomiting. She denied any joint pain, swelling, or morning stiffness. She did well over the next 10 years and noticed *one-flight* dyspnoea on exertion but no orthopnoea. At age 50 she was again admitted to the Rochester General

Hospital again because of *one-flight* dyspnoea and recurrent anterior chest pain on exertion. She denied any haemoptysis, black-out spells, fever, or night sweats.

Examination.—She was a small, slightly obese female who appeared chronically ill. Blood pressure was 160/110/80 in both arms with the patient in a supine position. Jugular venous pulse was normal in height and contour. Pulse was 80 and regular and respiration rate 16. Head, ears, eyes, nose, and throat were unremarkable. Chest was clear to percussion and auscultation.

Cardiac examination showed a point of maximum impulse palpable only in the left lateral decubitus position. Heart size was normal on percussion. On auscultation S2 was louder than S1. There was a grade 3 systolic murmur at the apex but heard loudest at the aortic area with radiation into the carotid vessels. The carotid pulse demonstrated a systolic thrill. A grade 2/6 decrescendo diastolic murmur was noted along the left sternal border.

Liver, spleen, and kidneys were not palpable.

Joint examination disclosed striking changes in the hands, more marked on the right side. At rest the hands were held in a position of ulnar deviation most prominent at the right 3rd, 4th, and 5th metacarpophalangeal joints (Fig. 1).

These joints were enlarged but without warmth or tenderness and there was a reducible subluxation of the

Fig. 1.—Hands at rest. The chalk outline of the fingers was drawn with the ulnar deviation passively reduced and both hands pressed firmly on the table to prevent ulnar deviation.

Fig. 2.—Hands with ulnar deviations passively reduced by the examiner and both hands pressed firmly on the table to prevent ulnar deviation.

right 4th and 5th metacarpophalangeal joints (Fig. 2, opposite).

The proximal interphalangeal joints showed a mild hyper-extension deformity (Fig. 3).

The distal interphalangeal joints of the 2nd and 3rd

digits on the right and the 3rd and 4th digits on the left showed enlargement and radial deviation. The resting deformities at the metacarpophalangeal joints could be voluntarily corrected in the left hand but only incompletely on the right (Fig. 2). There was a lateral dis-

Fig. 3.—Hands showing enlargement of metacarpophalangeal joints, hyper-extension at proximal interphalangeal joints of 2nd, 3rd, and 4th fingers bilaterally, and flexion deformity at distal interphalangeal joints of 2nd and 3rd fingers of the right hand and the 3rd and 4th digits of the left hand.

placement of the extensor tendons at rest which were seen to lie in the ridges between and to the ulnar side of the metacarpophalangeal joints. These returned to normal alignment when the deformity was corrected. No tendon nodules or Dupuytren's contracture were found.

Urine analysis was within normal limits. Haemoglobin 12.5 per cent. and hematocrit 35 per cent. Erythrocyte sedimentation rate was repeatedly normal. Serum protein electrophoresis normal. Wassermann reaction for syphilis negative. Repeated antistreptolysin-O titres within normal limits. Antinuclear factor and L.E. preparations negative. Bentonite flocculation tests for rheumatoid factors all normal. Electrocardiogram interpreted as showing nonspecific ST and T wave abnormalities.

Chest x ray (Fig. 4) showed no evidence of pulmonary venous engorgement, but some slight left ventricular enlargement was noted with calcification in the region of the aortic valve. A supravulvar angiogram demonstrated thickening and rigidity of the aortic valve cusps and a mild degree of aortic insufficiency. Cardiac catheterization revealed a cardiac index of 2.05 L/min.M². and the absence of a gradient across the aortic valve.

Fig. 4.—Posterior anterior chest x ray, showing left ventricular hypertrophy.

X ray of the hands showed a minimal but definite ulnar deviation of the proximal phalanges at the metacarpophalangeal joints most marked in the 3rd, 4th, and 5th joints (Figs 5 and 6, opposite). There was considerable flexion deformity of the first metacarpophalangeal joint on the right apparently due to old trauma (Fig. 6). Slight distension of the joint capsules of the metacarpophalangeal joints was noted. The radial aspect of the metacarpal head of the left index finger had an indentation similar to the hook-like erosions described at this location by Bywaters (1950) in Jaccoud's arthritis.

Discussion

Jaccoud (1869) first described the chronic arthritis appearing after frequent and severe attacks of rheu-

matic fever which he named chronic fibrous rheumatism. His patient was a 19-year-old youth who initially suffered an attack of rheumatic fever in the course of which he developed aortic stenosis and insufficiency. After two additional attacks, deformities of the hands and feet made their appearance. These changes later became permanent and were characterized by marked ulnar deviation of the fingers at the metacarpophalangeal joints and hyperextension of the proximal interphalangeal joints of the 2nd, 3rd, and 4th digits with no evidence of bone destruction. Keil (1938), in a summary on rheumatic fever nodules, noted that they may affect the flexor tendons and give rise to shortening. He also pointed out that in cases of acute rheumatic fever contractures occur similar to those of Dupuytren, being due to involvement of the aponeurosis of the palmaris longis and differing from the true Dupuytren's contractures by not showing adherence to the underlying structures. Jaccoud had reported that the chief changes were found in the joint capsules which became distended. It was stated that, because of this distension, the tendons could slip to the ulnar side of the metacarpophalangeal heads and consequently pull the phalanges obliquely in the ulnar direction. The fifth finger would be the one most severely involved because of the absence of a barrier to the ulnar deflection. Unlike rheumatoid arthritis, the gradual development of the deformity of the hands occurs without symptoms, with little evidence of active synovitis, and with the maintenance of functional capacity.

Bywaters (1950) suggests a list of features which characterized Jaccoud's arthritis:

- (1) A history of severe rheumatic fever with repeated and prolonged attacks.
- (2) Recovery delayed and associated with stiffness in the metacarpophalangeal joints which later results in the appearance of joint deformity.
- (3) The deformity appears to be due to periarticular fascial and tendon fibrosis rather than to synovitis.
- (4) The deformity consists of flexion at the metacarpophalangeal joint with some associated periarticular soft tissue swelling and ulnar deviation most marked in the fifth finger.
- (5) Associated hyperextension at the proximal interphalangeal joints.
- (6) Joint disease is usually inactive with little or no symptoms and good functional capacity.
- (7) "Radiologically, the earliest bone change is erosion of the metacarpal head on the palmar and radial part of their circumference in an anteroposterior projection producing a hoof like erosion."

Fig. 5.—X ray of left hand, showing increased soft tissue densities at the metacarpophalangeal joints. The encircled indentation possibly represents an old erosion of the hand of the second metacarpal bone.

Fig. 6.—X ray of right hand, showing old traumatic injury of first metacarpophalangeal joint. There are increased soft tissue densities at all metacarpophalangeal joints, with ulnar deviation and erosion with loss of joint space and radial deviation at the distal interphalangeal joint of the middle finger.

The erythrocyte sedimentation rate is normal. Radiological changes may be minimal or absent and the rheumatoid factor cannot be demonstrated in the patient's serum. The cardiovascular abnormalities are often very prominent unlike rheumatoid heart disease in which the articular symptoms overshadow the cardiac abnormalities. In Jaccoud's arthritis with rheumatic heart disease, the joint deformities rarely cause any discomfort. There is no associated anaemia and the erythrocyte sedimentation rate is normal. The serological tests for rheumatoid factors are negative.

In the case reported here the moderate severity of the heart disease, the normal erythrocyte sedimentation rate, the absence of the rheumatoid factor, and the presence of the characteristic joint deformities both clinically and radiologically would all lead to a diagnosis of Jaccoud's arthritis. There was no evidence of any other joint involvement either clinically or radiologically. This patient had no history of subcutaneous nodules. A case was recently reported by Ruderman and Abruzzo (1966) which fulfilled the criteria for chronic post-rheumatic fever arthritis (Jaccoud's) in which subcutaneous nodules, histologically resembling rheumatoid nodules, were found. Bywaters and Zvaifler (1966) felt that the presence of such a nodule did not exclude the diagnosis of Jaccoud's arthritis.

The alternative diagnosis of osteo-arthritis combined with rheumatic disease was considered but excluded because of the age at onset of the joint deformities in this patient, the absence of osteophytes or bone destruction, and the presence of joint capsular enlargement with hyperextensible joints. In rheumatoid arthritis with rheumatoid heart disease, it is noted that the joints are severely and widely involved with evidence of active synovitis. The rheumatoid factor is nearly always present in the serum. There may be an anaemia consistent with the activity of the rheumatoid state. The

murmurs of aortic insufficiency or mitral valvular disease may be present, but the patients are rarely incapacitated by their valvular disease (American Rheumatism Association, 1964; Weintraub and Zvaifler, 1963). In ankylosing spondylitis with aortitis, the hands are not deformed but there is ankylosis of the spine and involvement of sacroiliac and the large diarthrodial joints. Patients with rheumatic heart disease and osteo-arthritis of the hands frequently have Heberden's nodes, osteophyte formation, and painful crepitant joints. Painless joint deformities with reducible subluxations at the metacarpophalangeal joints would not be expected in osteo-arthritis.

The present case of painless joint deformities and reducible subluxations at metacarpophalangeal joints, with apparent rheumatic aortic valvulitis and insufficiency resembles the original case of Jaccoud's arthritis and fulfils criteria for this disease.

Summary

The combination of valvular heart disease with chronic deforming arthritis may be due to several different diseases, for example:

- (1) Rheumatoid arthritis and rheumatoid heart disease.
- (2) Rheumatoid arthritis developing in a patient with rheumatic heart disease.
- (3) Ankylosing spondylitis with aortic insufficiency.
- (4) Osteo-arthritis in a patient with rheumatic heart disease.
- (5) Rheumatic heart disease with Jaccoud's arthritis.

The purpose of this paper is to present a case of the last-named entity and to discuss the criteria for its diagnosis.

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L'arthrite de Jaccoud

RÉSUMÉ

L'association de la maladie valvulaire du coeur et de l'arthrite chronique déformante peut être due à plusieurs maladies différentes, telles que:

- (1) polyarthrite chronique évolutive et la maladie rhumatoïde du coeur;
- (2) polyarthrite chronique évolutive survenant chez un malade atteint de maladie rhumatismale du coeur;
- (3) spondylarthrite ankylosante avec insuffisance aortique;
- (4) ostéoarthrose chez un malade atteint de maladie rhumatismale du coeur;
- (5) maladie rhumatismale du coeur avec arthrite de Jaccoud.

Cet article a pour but la présentation d'un cas de cette dernière entité et la discussion de ses critères diagnostiques.

La artritis de Jaccoud

SUMARIO

La asociación de la enfermedad valvular del corazón y de la artritis crónica deformante se puede deber a varias enfermedades diferentes, tales como:

- (1) poliartritis crónica evolutiva y enfermedad reumatoide del corazón;
- (2) poliartritis crónica evolutiva ocurriendo en un enfermo con carditis reumática;
- (3) espondilartritis anquilosante con insuficiencia aórtica;
- (4) osteoartrosis en un enfermo con carditis reumática;
- (5) carditis reumática con artritis de Jaccoud.

Se ha escrito este artículo con el propósito de presentar un caso de esta última enfermedad y de discutir sus criterios diagnósticos.