Chronic post-rheumatic fever (Jaccoud's) arthropathy

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This paper describes two patients with Jaccoud's arthropathy.

Case reports

Case 1,* a married woman, a former nurse, aged 66 years, was admitted to the West London Hospital on November 12, 1967, after a syncopal attack provoked by climbing a long flight of stairs. On regaining consciousness she had had a small haematemesis. There was no history of dyspepsia and she had not taken any drugs.

She had suffered from a severe and prolonged attack of rheumatic fever 40 years before, for which she had received treatment in hospital for 7 months, and she had developed a second attack of rheumatic fever the following year. In both attacks the polyarthritis involved the small joints of the hands. She subsequently remained in good health, though aware of a painless ulnar deformity of the fingers of both hands. In recent years she had twice fallen on outstretched hands, injuring both wrists. In 1957 she developed exertional dyspnoea and orthopnoea and in April, 1967, she first experienced angina of effort.

EXAMINATION

The cardiovascular system showed pulsus bisferiens and sinus rhythm. Jugular venous pressure was normal and there was no dyspnoea or ankle oedema. The left ventricle was enlarged and there was a loud aortic systolic murmur with a thrill and a soft early diastolic murmur. The blood pressure was 130/80 mm. Hg.

The hands (Fig. 1) showed a reversible ulnar deviation of the fingers; this could be actively corrected at will by the patient. Both first metacarpophalangeal joints were subluxed. There was no synovial thickening of the hand joints but the ulnar styloid processes were prominent. The other joints were normal and there were no subcutaneous nodules.

INVESTIGATIONS

Haemoglobin 11 g. per cent.; white cells 5,300/mm.³ with a normal differential count. Erythrocyte sedimentation rate (Westergren) 20 mm./1st hr. Sheep cell agglutination, latex-fixation, and anti-nuclear factor tests were all negative, and the antistreptolysin-O titre was 100 Todd units. An electrocardiogram revealed left ventricular preponderance.

FIG. 1 Case 1. Ulnar deviation of fingers and subluxation of first metacarpophalangeal joints.

Chest x ray showed cardiomegaly due to left ventricular enlargement and pulmonary venous congestion. Lateral films showed calcification in the aortic valve.

X rays of the hands (Fig. 2) confirmed the ulnar drift of the fingers with bilateral subluxation of the first metacarpophalangeal joints. In addition 'hook' lesions were seen on the radial aspect of the fifth left metacarpal head (Fig. 3) and on the first right metacarpal head (Fig. 4). Degenerative changes were present in the wrist joints (Fig. 2), but it is difficult to know how much they were due to the previous injuries. No rheumatoid erosions were seen in either the hands or the feet. The sacroiliac joints were radiologically normal. A barium meal examination revealed no abnormality.

A diagnosis was made of chronic rheumatic endocarditis with aortic stenosis and regurgitation, and left ventricular failure; and chronic post-rheumatic fever (Jaccoud's) arthropathy. No cause for the haematemesis was found. With appropriate treatment the left ventricular failure remitted and a subsequent chest x ray showed a reduction in heart size. She returned home 4 weeks after admission with an improved exercise tolerance and subsequent progress was uneventful until March, 1968, when she suddenly died at home. No autopsy was carried out.

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* Case 1 was presented at a meeting of the Royal Society of Medicine (Grahame, 1968).

FIG. 2 Case 1. Antero-posterior radiograph of hands.

FIG. 3 Case 1. Enlargement of a portion of Fig. 3 to show 'hook' lesion on radial aspects of left 5th metacarpal head.

FIG. 4 Case 1. Oblique view of right first metacarpal head showing probable 'hook' lesion.

Case 2, a 65-year-old managing director of an engineering firm, was admitted to the West London Hospital on March 6, 1969, having suffered from malaise and intermittent pyrexia for 2 weeks. This had not responded to tetracycline but was improving on penicillin and Septrin, when a further pyrexial episode precipitated admission.

Between 1920 and 1931 (aged 17 to 28 years) he had suffered four illnesses diagnosed as acute rheumatic fever. Joint involvement occurred only in the second episode, at the age of 18, when the joints of the hands and fingers became inflamed for a few months. No other joints were involved. After the pain and swelling had settled, he noticed ulnar deviation of the fingers and this subsequently remained unchanged. He had no further pain in these joints and disability was minimal. Cardiac involvement of unspecified nature had been diagnosed dur-

ing the second attack, but this gave rise to no trouble until 1959 (when he was aged 55), when manipulation under anaesthetic was performed on his right knee because of a torn cartilage. Atrial fibrillation developed and persisted subsequently.

EXAMINATION

He was thin and afebrile by the time of admission. The cardiovascular system showed atrial fibrillation and a collapsing pulse. There were signs of left and right ventricular enlargement. The blood pressure was 170/60 mm. Hg. There was an aortic systolic murmur accompanied by a thrill and an early aortic diastolic murmur conducted down the left sternal border. A pansystolic

apical murmur was heard radiating to the axilla. An opening snap and short mid-diastolic murmur localized to the apex were audible. The second heart sound in the pulmonary area was accentuated.

The jugular venous pressure was not raised, no basal rales were present, and there was no oedema. A non-tender liver was enlarged to 3 cm. below the costal margin. There were no splinter haemorrhages, splenomegaly, or clubbing.

The hands (Fig. 5) showed bilateral ulnar deviation without synovial thickening. Subluxation of the metacarpophalangeal joints together with ulnar deviation (Fig. 6) prevented active extension of the fingers, but almost complete passive extension was possible. Proximal and distal interphalangeal joints were normal apart from hyperextensibility of the left second proximal interphalangeal joint. Other joints were normal and no subcutaneous nodules were present.

FIG. 5 Case 2. Ulnar deviation of fingers and subluxation of metacarpophalangeal joints.

INVESTIGATIONS

Haemoglobin 12.7 g. per cent.; white cells 3.900/mm.³ with a normal differential count. Erythrocyte sedimentation rate (Westergren) 14 mm./1st hr. Blood cultures were sterile.

An electrocardiogram confirmed atrial fibrillation and left ventricular hypertrophy. Chest x ray showed cardiac enlargement, extensive nodular opacities in both lung fields, consolidation of the right middle lobe, and pleural reactions in both costophrenic angles.

The sheep cell agglutination, latex-fixation, and anti-nuclear factor tests were all negative.

X rays of the hand (Fig. 7) showed ulnar deviation with subluxation of metacarpophalangeal joints. There were no erosions and no definite hook lesions present. The sacroiliac joints were normal. X rays of the feet showed bilateral hallux valgus but no other abnormality.

FIG. 6 Case 2. Subluxation of 5th metacarpophalangeal joint with lateral displacement of extensor tendon.

FIG. 7 Case 2. Antero-posterior radiograph of hand.

A diagnosis was made of resolving right middle lobe pneumonia complicating chronic rheumatic carditis with atrial fibrillation; and chronic post-rheumatic fever (Jaccoud's) arthropathy.

Treatment with digoxin and diuretics was continued. He remained apyrexial and a follow-up chest x ray showed clearing of the right middle zone and left costophrenic angle shadowing. The heart shadow was definitely enlarged but had diminished slightly. Mottling of the lung fields and right pleural reaction persisted. Pericardial calcification was now seen. (These residual abnormalities had been seen in an x ray of the chest in 1960.)

He was discharged as asymptomatic on March 14, 1969, and prophylactic oral penicillin was added to his regime. He has remained well and at work.

Discussion

It is just over a century since Jaccoud (1869) described a case of chronic ulnar deformity in the hands of a patient who had previously suffered no fewer than six attacks of rheumatic fever.

The first report of this condition in the British literature is probably that of Garrod (1890), who illustrated the hand of a patient showing ulnar deviation of an extreme type. This patient had suffered from a number of attacks of rheumatic fever.

It took a further 60 years before the confusion surrounding the relationship between valvular heart disease and chronic arthritis cleared with the re-emergence of chronic post-rheumatic fever arthritis (Jaccoud's arthritis) as a clinical entity (Bywaters, 1950). This classic study reviewed the necropsy findings in 27 cases of 'rheumatoid-like' arthritis. Amongst those cases showing a coincidental heart valve lesion, three types of association were recognized: rheumatoid arthritis developing in a patient suffering from unrelated rheumatic heart disease; rheumatoid granulomata involving the heart valves;

and Jaccoud's arthritis. A pathognomic 'hook' lesion seen on the radial aspect of the affected metacarpal head on x ray was described in the last condition. (To these three types of association might be added that of aortic incompetence occurring with ankylosing spondylitis (Ansell, Bywaters, and Doniach, 1958) and Reiter's disease (Csonka, Litchfield, Oates, and Willcox, 1961)).

Despite the increasing awareness of this condition, there have only been a handful of reports in the literature of further cases, all emanating from the United States—Short, Bauer, and Reynolds (1957) 1 case; Engleman (1960) 1 case; Zvaifler (1962 and 1966) 4 cases; Twigg and Smith (1963) 2 cases; Ruderman and Abruzzo (1967) 1 case; Burda and Sanders (1967) 1 case; Beausang, Barnett, and Goldstein (1967) 1 case.

The diagnosis of Jaccoud's arthropathy in the present study is based on the findings of a painless reversible ulnar deformity of the hands of insidious onset in the wake of recurrent acute rheumatic polyarthritis involving the small joints of the hands, coupled (in the first case) with the finding of hook lesions on x ray. It is of interest that in this patient the wrist joints were also involved, although the trauma which the patient had previously sustained may have contributed. Rheumatoid arthritis is highly improbable because of the absence of symptoms in the affected joints and the absence of a raised erythrocyte sedimentation rate, subcutaneous rheumatoid nodules, erosions on x ray, and rheumatoid serum factor.

With the declining prevalence of rheumatic fever it is likely that this already uncommon articular complication of rheumatic fever will become rarer still. Nevertheless, the importance of its recognition lies in distinguishing it from rheumatoid arthritis with which it has so often been confused in the past.

ANSELL, B. M., BYWATERS, E. G. L., AND DONIACH, I. (1958) *Brit. Heart J.*, **20**, 507 (The aortic lesion of ankylosing spondylitis).

BEAUSANG, E., BARNETT, E. V., AND GOLDSTEIN, S. (1967) *Ann. rheum. Dis.*, **26**, 239 (Jaccoud's arthritis).

BURDA, C. D., AND SANDERS, C. V. (1967) *Arch. intern. Med.*, **120**, 712 (Chronic postrheumatic fever (Jaccoud's) arthritis).

BYWATERS, E. G. L. (1950) *Brit. Heart J.*, **12**, 101 (The relation between heart and joint disease including 'rheumatoid heart disease' and chronic post rheumatic arthritis (type Jaccoud)).

CSONKA, G. W., LITCHFIELD, J. W., OATES, J. K., AND WILLCOX, R. R. (1961) *Brit. med. J.*, **1**, 243 (Cardiac Lesions in Reiter's disease).

ENGLEMAN, E. P. (1960) *Arch. Interamer. Rheum.*, **3**, 616 (Questionnaire).

GARROD, A. E. (1890) 'A Treatise on Rheumatism and Rheumatoid Arthritis'. Griffin, London.

GRAHAME, R. (for J. T. Scott) (1968) *Proc. roy. Soc. Med.*, **61**, 1109 (Jaccoud's arthropathy).

JACCOUD, S. (1869) 'Leçons de clinique médicale faites à l'Hôpital de la Charité', 2nd ed. Delahaye, Paris.

RUDERMAN, J. E., AND ABRUZZO, J. L. (1966) *Arthr. and Rheum.*, **9**, 640 (Arthritis Rounds: Chronic postrheumatic fever arthritis (Jaccoud's): Report of a case with subcutaneous nodules).

SHORT, C. L., BAUER, W., AND REYNOLDS, W. E. (1957) 'Rheumatoid Arthritis; A Definition of the Disease and a Clinical Description Based on a Numerical Study of 293 Patients and Controls', pp. 22-23. Harvard University Press, Cambridge, Mass.

TWIGG, H. L., AND SMITH, B. F. (1963) *Radiology*, **80**, 417 (Jaccoud's arthritis).

ZVAIFLER, N. J. (1962) *N. Engl. J. Med.*, **267**, 10 (Chronic post rheumatic fever (Jaccoud's) arthritis).

— (1966) *Arthr. and Rheum.*, **9**, 646 (Arthritis Rounds: Discussion).