CHRONIC POST-RHEUMATIC FEVER ARTHRITIS (JACCOUD'S) — REPORT OF 2 CASES

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SYNOPSIS

Two cases of chronic post-rheumatic fever arthritis (Jaccoud's) are presented. The clinical, biochemical and radiological features are described.

INTRODUCTION

Joint involvement in rheumatic fever is familiarly known as a transient, migratory polyarthritis occurring during the acute episode or exacerbation of rheumatic fever, seldom leaving permanent residual joint deformity. Jaccoud in 1869 was the first to describe chronic arthritis in a young adult with recurrent rheumatic fever and rheumatic valvular disease. Although rheumatoid arthritis and ankylosing spondylitis can produce valvular heart disease (usually aortic incompetence), these diseases are distinctly different from rheumatic fever—clinically, radiologically and serologically.

Valvular heart disease and chronic joint deformity can be related in several ways:

(1) Rheumatoid arthritis occurring in a person with pre-existing rheumatic heart disease.

(2) Rheumatoid arthritis with rheumatoid heart disease.

(3) Ankylosing spondylitis with peripheral arthritis and aortitis.

(4) Rheumatic heart disease with chronic post-rheumatic fever arthritis.

These two cases are being presented because of their rarity. Only 9 cases have been reported in the English literature so far—Bywaters (1950), Ruderman (1966), Sandler (1968), Zvaflier (1962).

CASE REPORTS

Case 1

L.M.L., a 46 year old Chinese fisherman who has severe mitral stenosis (restenosis) and moderate aortic incompetence, developed rheumatic fever in 1953 (age 26) with arthritis involving his hands and wrists. He remembered having had 5 attacks of rheumatic arthritis involving his hands. In June 1963, he was admitted for the first time because of exertional dyspnoea. Ulnar deviation of his fingers were noted but was thought to be due to co-existing rheumatoid arthritis. Physical examination then revealed a thin, frail man with severe isolated mitral stenosis.

Cardiac catheterisation done in 1964 revealed a pulmonary artery pressure of 60/38 mm. Hg., and a pulmonary arterial wedge pressure of 27 mm. Hg. (mean). The Erythrocyte Sedimentation Rate was 9 mm./hour Westergren. The Rheumatoid Arthritis factor was negative. At surgery the mitral orifice was found to be less than 1 cm. in diameter. The valve edges were calcified and a large calcareous mass was found on the free edge of the anterior cusp. The valve orifice was dilated to 3 cm. There was good symptomatic improvement after operation, but from 1969 his effort tolerance progressively deteriorated. He was readmitted several times for congestive cardiac failure and haemoptysis.

Examination in 1973 revealed evidence of critical mitral stenosis and moderate aortic regurgitation. On repeat cardiac catheterisation the pulmonary artery pressure was found to be 60/40 mm. Hg. Aortic angiogram confirmed moderate aortic incompetence. There was ulnar deviation of the fingers, most marked in the 4th and 5th fingers and slight flexion deformity of the metacarpophalangeal joints (Fig. 1). There was a full range of movements of his fingers. The ulnar deviation could be completely corrected, voluntarily, by placing his palm on a flat surface (Fig. 2). He was unaware of his joint deformity until he was told about it and he never experienced pain or stiffness in his fingers apart from the earlier episodes of rheumatic arthritis.

The haemoglobin was 15 gm.%, Erythrocyte Sedimentation Rate 2 mm./hour (Westergren), Lupus Erythematosus cells and Rheumatic Arthritis factor were repeatedly negative. The electrocardiogram showed atrial fibrillation and right

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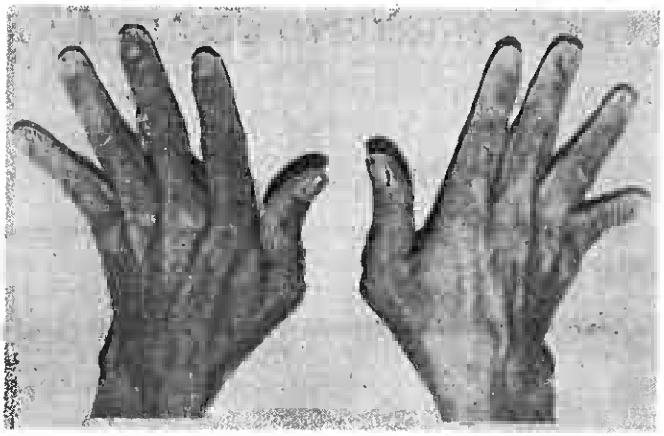

Fig. 1. Case 1. There is ulnar deviation of the thumb and fingers, more marked in the 4th and 5th fingers. The extensor tendons lie in the ridges on the ulnar side of the corresponding fingers.

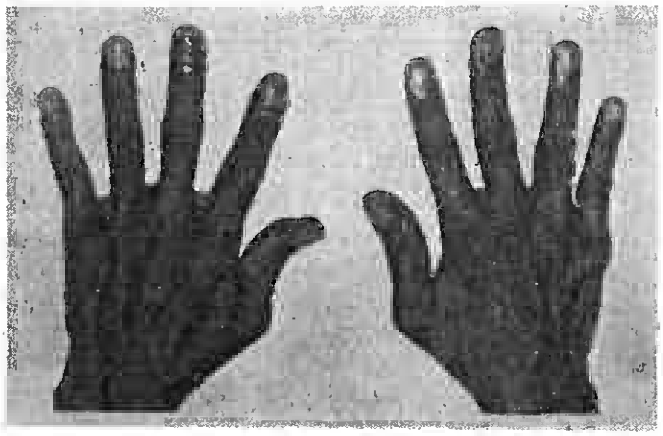

Fig. 2. Case 1. Ulnar deviation of the fingers has been voluntarily corrected.

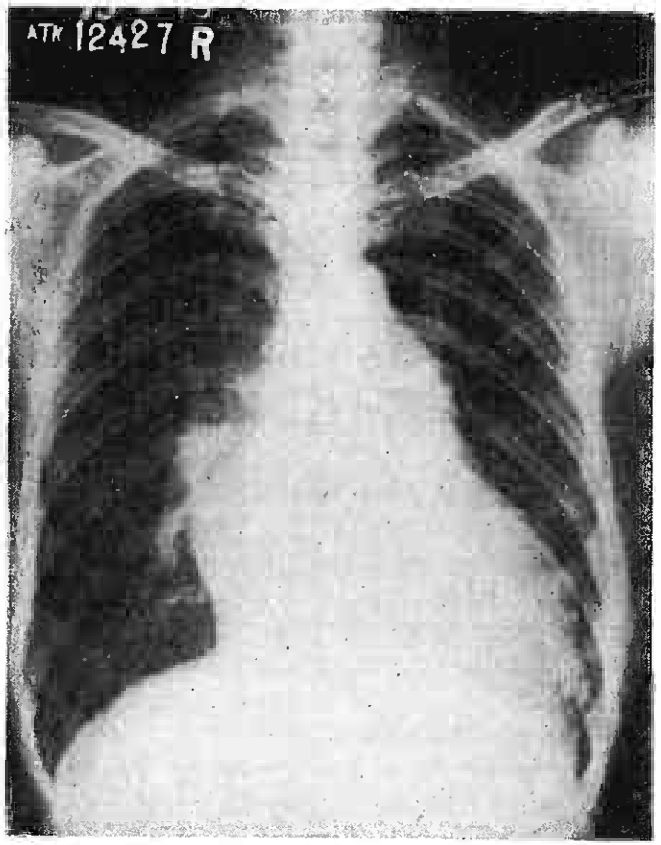

Fig. 3. The chest X-ray of Case 1 shows cardiomegaly, left atrial enlargement, pulmonary haemosiderosis, and lung ossification.

ventricular hypertrophy while the chest X-ray revealed a large heart, with aneurysmal dilatation of the left atrium, pulmonary haemosiderosis and lung ossification (Fig. 3). X-rays of both hands showed normal bone density, normal joint spaces, ulnar deviation of the phalanges (Fig. 4) and subluxation of the distal phalanx of the thumb.

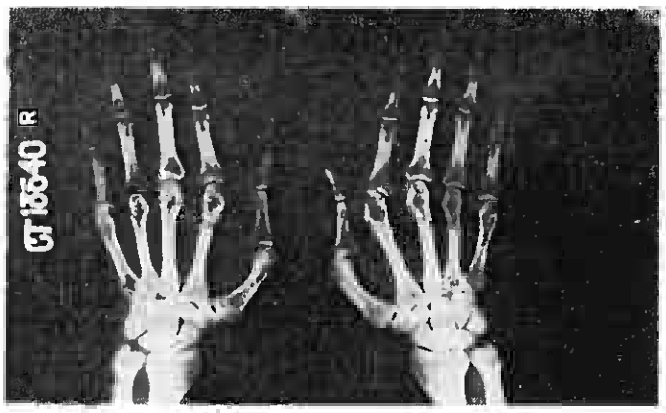

Fig. 4. Case 1. X-ray of both hands show ulnar deviation of the fingers, normal joint spaces and normal bone density.

Case 2

L.C.S., a 45 year old Chinese male was first admitted for haemetemesis and epigastric pain in 1961. He was found to have severe mitral incompetence and mitral stenosis, and moderate aortic incompetence and atrial fibrillation. Barium meal confirmed the presence of a duodenal ulcer. Ulnar deviation of his fingers was noted then, but was thought to be due to co-existing rheumatoid arthritis. He never had pain or stiffness in his hands. He had an episode of cerebral embolism in 1969 and an episode of congestive cardiac failure in June 1972.

Latest examination revealed ulnar deviation of his fingers and flexion deformity of the metacarpophalangeal joints which could only be partially corrected voluntarily (Figs. 5 and 6). There

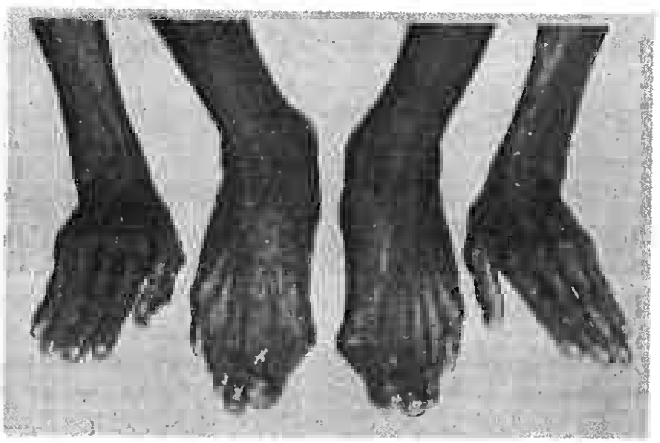

Fig. 5. Case 2. There is flexion deformity and soft tissue swelling of the metacarpophalangeal joints and ulnar deviation of the fingers. There is hallux valgus of both feet.

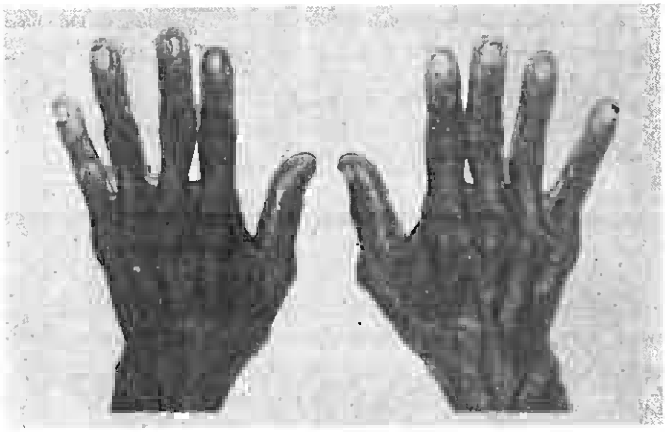

Fig. 6. Case 2. The flexion deformity of the metacarpophalangeal joint is only partially corrected voluntarily. Soft tissue swelling of the metacarpophalangeal joint is clearly seen.

was thickening of the periarticular tissue of the metacarpophalangeal joints and crepitus could be elicited from them.

When the fingers were held in the resting position, the extensor tendons were seen to be in the ridges on the ulnar side of the corresponding metacarpophalangeal joints (Fig. 5).

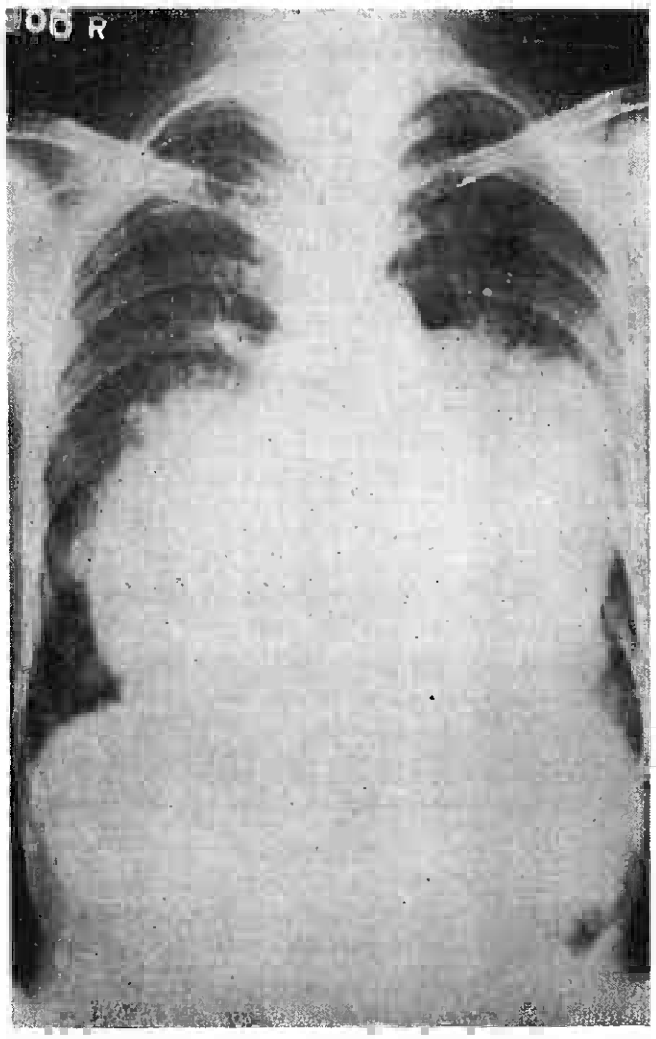

Fig. 7. The chest X-ray of Case 2 shows aneurysmal dilatation of the left atrium and pulmonary venous hypertension.

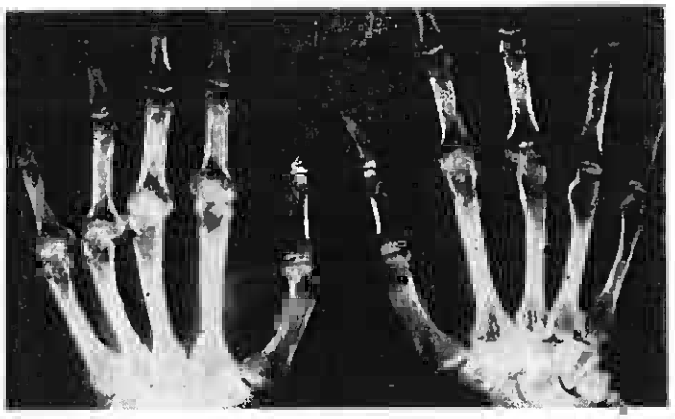

Fig. 8. X-rays of the hands of Case 2 show subluxation of the metacarpophalangeal joints and peri-articular soft tissue swelling.

Relevant laboratory investigations results were: Haemoglobin of 12 gm.%, Erythrocyte Sedimentation Rate of 22 mm./hour (Westergren), Rheumatic Arthritis factors and Lupus Erythematosus cells were negative, and the serum uric acid was 3·2 mgm.%.

The electrocardiogram showed atrial fibrillation and right ventricular hypertrophy. Chest X-ray showed gross cardiomegaly, large left atrium pulmonary haemosiderosis, and septal lines (Fig. 7). X-rays of the hands (Fig. 8) showed subluxation of the metacarpophalangeal joints affecting the right second to fifth, and left second to third metacarpophalangeal joints. There was soft tissue swelling involving the metacarpophalangeal joints. X-rays of the spine were normal.

Cardiac catheterisation done on 27th February 1973 confirmed severe mitral incompetence and mitral stenosis, and moderate aortic incompetence. The left ventriculogram showed severe mitral incompetence, aneurysmal dilatation of the left atrium and the aorta stretched over the enormously dilated left atrium.

DISCUSSION

In the case described by Jaccoud (1869), the joint deformities were characterised by marked ulnar deviation of the fingers, hyperextension of the proximal interphalangeal joints and was unassociated with bone destruction. Bywaters (1950) reviewed the literature at that time, described 3 cases of his own, and suggested the features which were characteristic of this entity:—

1. There is a history of repeated severe attacks of rheumatic fever, distinguished from rheumatoid arthritis by the presence of heart involvement, chorea, migratory polyarthritis and rheumatic nodules in some cases.
2. Recovery from initial attacks may be complete but repeated attacks may produce

residual stiffness, especially in the metacarpophalangeal joints from which the deformity arises.

3. The characteristic deformity is due to periarticular, fascial and tendon fibrosis rather than a synovitis.
4. The deformity is characteristically that of ulnar deviation, most marked in the fourth and fifth fingers. In the early stages of the disease, the ulnar deviation can be corrected voluntarily. There may be hyperextension of the proximal interphalangeal joints, mild flexion at the metacarpophalangeal joints which may have periarticular soft tissue swelling.
5. Tendon crepitus in the metacarpophalangeal and proximal interphalangeal joints may be elicited.
6. The joints disease is usually inactive with few or no symptoms, good function and little evidence clinically or biochemically of active synovitis. The erythrocyte sedimentation rate is normal in most cases.
7. Radiologically the earliest bone change is erosion of the metacarpal heads on the most palmar and radial part of their circumference, in the antero-posterior projection, producing a hook-like lesion.

Zvaflier (1962) suggested an additional criteria viz. the rheumatoid factor should be negative.

These two patients with severe aortic and mitral valve disease, mild joint disease and a history of severe, repeated attack of rheumatic arthritis as in Case 1 meet the criteria for Jaccoud's arthritis, a rare sequel of rheumatic fever. Both patients were misdiagnosed as having co-existing rheumatoid arthritis, although the lack of symtoms and serological abnormalities were recognised. With the decreasing incidence of rheumatic fever and the effective prophylaxis against recurrent attacks, the development of chronic post rheumatic arthritis would in future be non-existent.

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