RESEARCH LETTER

Jaccoud's Arthritis

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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, the most typical of which is migratory polyarthritis. Permanent joint deformity is not one of the criteria used in the diagnosis of rheumatic fever (1).

In 1950 Bywaters reviewed the available literature on the condition and credited Jaccoud with its first clinical description. Bywaters also enumerated the diagnostic criteria for the diagnosis (2). Weintraub and Zvaifler described the characteristic features that distinguish chronic post-rheumatic-fever, arthritis from rheumatoid arthritis associated with either rheumatoid or rheumatic heart disease (2). The characteristic deformities of chronic post-rheumatic- fever arthritis are found in the hand and consist of flexion of the metacarpophalangeal (MCP) joints associated with ulnar deviation of the fingers, most marked in the fourth and fifth digits (3,4,5). We are describing the case of a patient with rheumatic valvular disease who had comparable deformities of the hands and fingers and who fulfilled all of the criteria suggested by Bywaters (2).

A nineteen year old female presented with pain and swelling of multiple joints including hip, knee, ankle, shoulder, elbow and the hand joints, predominantly the MCP, PIP and the distal DIP joints. The joint pains were non migratory. She also complained of dyspnea on exertion and palpitations but had no orthopnea or paroxysmal nocturnal dyspnea. She gave no history of sore throat, cough & expectoration, rash or abnormal body movements. She had a history of migratory polyarthritis involving the knee, ankle, wrist & elbow joint when she was five years of age with an antecedent history of sore throat and fever. She complained of progressively increasing dyspnea on exertion and palpitations since then.

On examination she was febrile, had congested throat and inflamed tonsils. She had tachycardia, loud S1 & a harsh pansystolic murmur radiating to the left axilla and back with a systolic thrill at the apex. All the involved joints were tender with restricted mobility. She had flexion deformity of the MCP joints; spindling of the PIP joints and ulnar deviation of the fingers. In addition, subcutaneous nodules (0.5-1cm) on olecranon process of right arm were noted. Routine hematological and biochemical parameters were normal except for elevated ESR. ASO titre & CRP levels were significantly raised. Blood & throat swab culture showed no growth. Rheumatoid factor, anti CCP antibodies & antinuclear antibodies were negative. Chest X ray showed cardiomegaly and radiographs of hand showed marked ulnar deviation of the fingers at the MCP joints and hyperextension of the PIP joints of the 2nd, 3rd, 4th, and 5th digits unaccompanied by clinical evidence of bone destruction. Echocardiography showed thickened and calcified mitral leaflets, dilated left atrium and ventricle and severe mitral regurgitation without any vegetations.

Jaccoud (1869) first described the chronic arthritis appearing after frequent and severe attacks of rheumatic fever which he named chronic fibrous rheumatism (6). 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. 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

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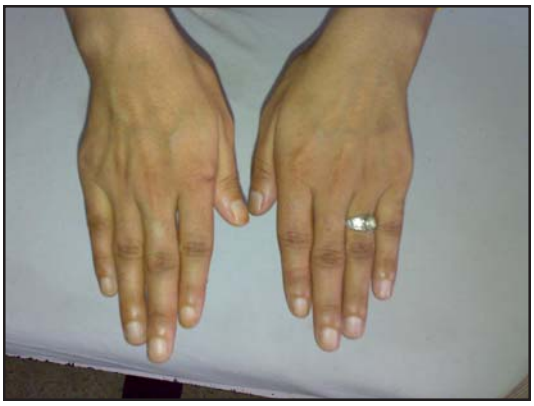

Fig 1- Flexion Deformity of MCP Joints, Spindling of the PIP & Ulnar Deviation of the Fingers

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 (2):

(1). A history of 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."

Jaccoud's arthropathy is suggested to arise due to connective tissue dysplasia resulting in soft tissue impairment in different rheumatic and nonrheumatic diseases (7). Concomitant hypocomplementemic urticarial vasculitis, Jaccoud's arthropathy and valvular heart disease this rare combination of manifestations.

Recently also valvular heart disease in patients with hypocomplementemic urticarial vasculitis syndrome associated with Jaccoud's arthropathy ha sbeen reported, where in authors recommended that Patients with HUVS and associated JA should be evaluated for the presence of valvular heart disease. The latter is probably a nonrheumatic, inflammatory, and degenerative process, mediated by immune complex, as well as cellular immune mechanisms (8,9).

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. The alternative diagnosis of Rheumatoid arthritis combined with rheumatic disease was considered, but 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.

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