Jaccoud’s Arthropathy with Radial Shaft Deformity in a Patient with a History of Mild Carditis and Sydenham’s Chorea

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Abstract

Background: Jaccoud’s arthropathy was first described in 1869 in a patient with recurrent attacks of acute rheumatic fever and recurrent migratory polyarthritis. Typical features include characteristic joint deviation with metacarpophalangeal subluxation, ulnar deviation, swan neck deformity of the fingers, Z deformity of both thumbs and hook formation, and alignment disorders without noticeable erosions on radiographs. This report describes a patient with Jaccoud’s arthropathy who presented with long bone deformity.

Case presentation: A 12-year-old-girl was diagnosed with chorea at 8 years of age and was treated with haloperidol. The chorea resolved after 3 months of treatment. An echocardiogram showed thickening of the mitral valve leaflets and mild mitral regurgitation. Three years later, the patient presented with progressive symmetric deformity of the wrist joints. She reported no pain or limitation of movement. Physical examination showed no cardiac murmur. There was varus deformity of both wrist joints. There was no tenderness or limitation of movement. The patient’s antistreptolysin O test was negative at first presentation and on this visit. Her rheumatoid factor was negative but the antinuclear antibody test was positive. An echocardiogram showed complete resolution of the mitral regurgitation. A wrist x-ray showed marked symmetric curving of the distal radius and varus deformity of the wrists, joint deviation with metacarpophalangeal subluxation, and ulnar deviation of the hand. There was no Z deformity of the thumbs.

Conclusion: Common presentation of Jaccoud’s arthropathy is involvement of the wrist, metacarpophalangeal joints, and joints of the feet. Our patient had an unusual presentation of deformity of the distal radial bone, which to the best of our knowledge, has not been previously described.

Key words: Jaccoud’s arthropathy, acute rheumatic fever, systemic lupus erythematosus, carditis, chorea

Introduction

Jaccoud’s arthropathy was first described in 1869 in a patient with recurrent attacks of acute rheumatic fever and recurrent migratory polyarthritis [1]. Since then, several case reports describing this condition in patients with acute rheumatic fever have been published [2-5]. This rare deforming arthropathy has also been described in several connective tissue diseases [6-8] and other clinical conditions [9,10]. It has been reported that about 5% of patients with Systemic Lupus Erythematosus (SLE) have Jaccoud’s arthropathy [11]. Typical features include characteristic joint deviation with metacarpophalangeal subluxation, ulnar devia-
tion, swan neck deformity of the fingers, Z deformity of both thumbs and hook formation, and alignment disorders without noticeable erosions on radiographs [6]. Deformity of the long bones is generally rare. Here, we describe a 12-year-old girl who had chorea followed by Jaccoud's arthropathy with deformity of the distal radius.

Case Presentation

A 12-year-old girl was diagnosed with chorea at 8 years of age after presenting with irregular, jerky, rapid, and involuntary movements that involved the face, trunk, and distal extremities. The parents reported that the movements used to disappear with sleep. At first presentation 4 years prior, an echocardiogram showed slight thickening of the mitral valve leaflets with mild mitral regurgitation. There was no migratory polyarthritism or arthralgia. The patient's antistreptolysin O (ASO) test was negative at first presentation and on this visit. She was started on haloperidol. She was also started on monthly benzathine penicillin prophylaxis to prevent recurrence of rheumatic fever. The chorea resolved after 3 months of treatment with haloperidol. The patient began having progressive painless deformity of both wrist joints 3 years after the chorea resolved. She did not have pain or swelling of the wrists, or any limitation of wrist movement. The patient reported no cardiac symptoms. There was also no clinical evidence for recurrence of acute rheumatic fever in the past 3 years.

The patient appeared comfortable upon physical examination. Her vital signs were all within normal limits. Her weight was 22 kg and her height was 122 cm. Examination revealed no pallor. The heart sounds were well heard and there was no murmur. There was gross symmetric deformity of the distal radius and varus deformity of the wrist.

Figure 1. Pictures showing gross deformity of the forearm and wrist joints.

Figure 2. X-ray of the radius, ulna, and wrist joints showing gross deformity of the distal radius and varus deformity of the wrist.

Figure 3. X-ray of the hands showing varus deformity of the wrist joint, but normal metacarpophalangeal and interphalangeal joints.
of the distal forearms and wrist joints (Figure 1). There was no swelling or tenderness, no sign of fracture, and no limitation of motion at the wrist, metacarpophalangeal, or interphalangeal joints. An echocardiogram done 4 years after diagnosis of carditis showed complete resolution of the mitral regurgitation. The serological tests for anti-nuclear antibody (ANA) and rheumatoid factor were positive and negative, respectively. The anti-phospholipid antibody test was not done (not available). A wrist x-ray showed gross deformity of the distal radius (Figure 2), and an x-ray of the hands showed ulnar deviation of the fingers. There was no Z-deformity of the thumbs (Figure 3).

Discussion

In its original description, Jaccoud's arthropathy was noted to follow many recurrent attacks of acute rheumatic fever with recurrent migratory polyarthritis [1,5]. However, it has also been reported in patients without active arthritis [4,12]. In contrast to rheumatoid arthritis, the gradual development of deformity of the hands or feet occurs without symptoms, with little evidence of active synovitis, and with the maintenance of functional capacity [4,5]. This condition has been described as severe joint deformity with no or minimal osseous destruction [13]. Although it has been reported that bone erosions do not occur in Jaccoud's arthropathy, recent findings from ultrasound and magnetic resonance imaging have shown otherwise [14,15].

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 hook like erosion [16]. “Hook” erosions may occur on the radial palmar aspect of the metacarpal heads and can be differentiated from the marginal erosions of rheumatoid arthritis [17,18]. The condition is not associated with swelling, obliteration of joint spaces, or destruction of articular surfaces [12]. Erythrocyte sedimentation rate and rheumatoid factor tests are generally negative in patients with Jaccoud's arthropathy. Although the hands are the most common site of involvement, this condition has also been reported to involve the feet [12,13,19].

One important differential diagnosis that should be considered is Madelung's deformity. This is a condition characterized by ulnar and dorsal curvature of the distal radius due to deficient growth of the volar and ulnar aspect of the distal radial physis, increased inclination of the distal radial joint surface, triangulation of the corpus with proximal and volar migration, and prominent dorsal subluxation of the ulnar head [20,21]. The deformity usually presents in late childhood or adolescence, is more common in females, and is usually bilateral [22]. A number of anatomical changes occur in Madelung deformity, which result in biomechanical alterations leading to a decreased range of movement, decreased grip strength, and often pain [22]. In most patients, there is a hereditary component. In our patient, the absence of pain or functional impairment and lack of significant articular abnormalities on x-rays made Madelung's deformity an unlikely diagnosis. In addition, the fact that Sydenham's chorea and carditis preceded the condition favored Jaccoud's arthropathy as the most likely diagnosis. The fact that the ASO titer was negative and ANA was positive in our patient may suggest SLE as the likely diagnosis rather than acute rheumatic fever. Chorea could be the first and sole manifestation of SLE [23]. However, the fact that the mitral regurgitation was resolved speaks for acute rheumatic fever as a cause of the chorea and mild carditis in this patient. ASO titer could be negative in about 20% of patients with acute rheumatic fever [24]. In either case, presentation of Jaccoud's arthropathy is expected to be similar.

Conclusion

The common presentation of Jaccoud's arthropathy is involvement of the wrist, metacarpophalangeal joints, and joints of the feet. The metacarpal and metatarsal bones are the ones most commonly affected. Our patient had an unusual presentation of deformity of the distal radial bone, which to the best of our knowledge, has not been previously described.

Consent: Written informed consent was obtained from the parents.

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Competing interests

The authors declare that they have no competing interests.

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References