

Jaccoud's arthropathy – diagnostic and therapeutic implications

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Summary: A case of isolated chronic severe aortic regurgitation with Jaccoud's arthropathy involving the foot is presented. The interesting feature of the case is the absence of history of acute rheumatic arthritis at any stage of the illness. The diagnostic and possible therapeutic significance of this otherwise uncommon and benign condition are discussed.

Introduction

Polyarthritis of acute rheumatic fever is reversible and leaves no sequelae except for the so-called Jaccoud's arthropathy. This arthropathy is rare and since the original description by Jaccoud¹ only a handful of reports have appeared.

Bittl and Perloff reported an interesting case of this condition and reviewed the Western literature in 1983.² By an analysis of 8 reports covering 31 cases, they concluded that ulnar deviation at metacarpophalangeal joints was a universal phenomenon along with regurgitant lesions in most of the cases. A majority of the cases had developed this arthropathy after recurrent attacks of acute rheumatic fever. We report a case of Jaccoud's arthropathy presenting in an unusual manner, and discuss the diagnostic and therapeutic implications of this entity.

Case report

A 27 year old male presented with complaints of increasing effort dyspnoea and palpitations for 5 years and orthopnoea and paroxysmal nocturnal dyspnoea, and swelling of abdomen and feet for 2 months. There was no history of fever with joint pain or swelling suggestive of acute rheumatic fever in the past. Physical examination revealed a resting heart rate of 110/min. The pulse was regular, collapsing with bisferiens character and blood pressure was 140/50 mm Hg in both upper and 220/60 mm Hg in both the lower extremities. There was

evidence of gross congestive cardiac failure. Examination revealed cardiac enlargement with a hyperdynamic apex beat and a long murmur of aortic regurgitation. A left ventricular third heart sound and Austin Flint murmur were also audible. There was no evidence to suggest rheumatic activity. Examination of major joints did not reveal signs of acute inflammation. However, gross deformity was seen at the metatarsophalangeal joints of the left foot with medial deviation of the phalanges. This deformity was almost completely correctable. Anterior subluxation was present at both first metacarpophalangeal joints which was also correctable without any discomfort to the patient (Figures 1 and 2). The patient was not troubled by these deformities and on questioning said that gradual increase had occurred in these deformities in the past 2 years. There was no feature to suggest Marfan syndrome or any other heritable disorder of connective tissue. A clinical diagnosis of rheumatic heart disease, chronic severe aortic regurgitation with congestive cardiac failure and Jaccoud's arthropathy was made.

Investigations for rheumatic activity yielded negative results. Tests for antinuclear and rheumatoid factors were also negative. Electrocardiogram, chest X-ray, and two-dimensional echocardiogram showed findings consistent with the diagnosis of severe aortic regurgitation. X-ray of the left foot showed medial deviation of 65° at first, 40° at the second, and 20° at the third metatarsophalangeal joints. There was no swelling, obliteration of joint spaces, or destruction of articular cartilages. It was possible to reduce the deformities manually and the involved joints attained almost normal configuration (Figure 3).

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Figure 1 Left great toe and thumb showing medial deviation and anterior subluxation, respectively.

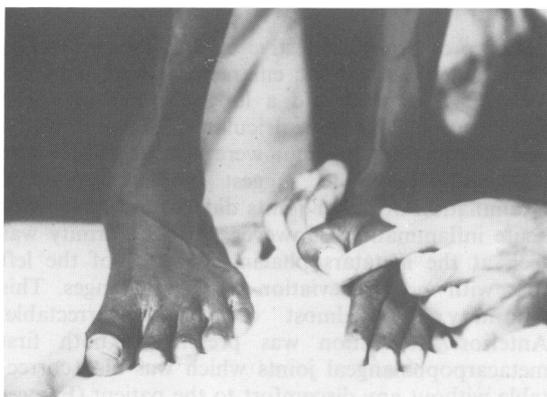


Figure 2 Left great toe and thumb after reduction of deformities.



Figure 3 X-ray of left foot showing metatarsophalangeal joints before (left) and after (right) reduction.

Discussion

Jaccoud's arthropathy, despite its rarity, is well known to cardiologists. Apparently it has no diagnostic, prognostic or therapeutic significance. In the review of Bittl and Perloff,² of 31 cases the average age at presentation was 46 years (range 14-67 years) and the female:male ratio was 2:1. Valvular incompetence was present in 27 of the 29 cases where the cardiac status was mentioned with mitral regurgitation in 21 and aortic regurgitation in 17 cases. Mitral stenosis was present in 5 and aortic stenosis in 6 cases. Reversible ulnar deviation was present at the metacarpophalangeal joints in all and more than 60% (19/31) cases had had at least 2 episodes of acute rheumatic fever. Studies for anti-nuclear and rheumatoid factors were negative in all.

Our case presented in a manner which is different from this description. Firstly, there was no history of acute arthritis at any stage of illness. We have not come across any report where Jaccoud's arthropathy has been reported to occur without a history of acute rheumatic fever in the past. Possibly this is an indicator that acute arthritis has nothing to do with this chronic type of arthropathy which develops later on for reasons not known.

Secondly, the involvement of foot joints was more than that of the hand joints. There is only a passing mention of foot joint involvement in this process in the western literature. In the two cases reported by Zvaifler, the predominant feature was the ulnar deviation of the extensor tendons. Similar but much milder changes were present in the toes in one case; while in the other, bilateral hallux valgus deformity was observed.³ Reports from our country show that this is a constant feature in Indian patients.⁴⁻⁶ This has been attributed to the bare-foot habit of rural Indian dwellers, but looking at the consistency of this finding we postulate that this may be one of the differences in the manifestation of rheumatic disease in developing countries like ours, compared to what is seen in the developed western countries. Some differences are well documented and include a difference in the pattern of acute rheumatic fever. In the developing countries the incidence of carditis is much more and that of arthritis is lower as compared to developed countries where the pattern is reversed.⁷ Another difference which we all are aware of is the early occurrence of severe mitral stenosis with severe pulmonary hypertension requiring early surgery (juvenile or accelerated type) in our patients.^{8,9}

One may question the rheumatic origin of the cardiac involvement in our case in the absence of a history of acute rheumatic fever at any stage of illness. But, it is well recognized that a history of acute rheumatic fever may be absent in about one

half of the cases of established rheumatic valvular lesions. Development of Jaccoud's arthropathy has helped to prove the rheumatic origin of cardiac involvement in our case in retrospect.

At times it is difficult to determine the aetiology of the lesion in cases of isolated aortic regurgitation. Heritable disorders of connective tissue cause aortic regurgitation with a frequent absence of skeletal manifestations. When the question of valve replacement arises, especially in Marfan's syndrome, it is preferable to put in a composite graft (aortic valve prosthesis with part of ascending aorta) than an aortic valve alone because of the

problem of 'valve dehiscence' with the latter approach. Development of Jaccoud's arthropathy clarified that the aetiology of the valvular lesion was indeed rheumatic in our case and, therefore, simple valve replacement would be sufficient when he comes to surgery.

Thus, this case demonstrates that Jaccoud's arthropathy can result without prior history of acute rheumatic fever with arthritis, and that this benign and uncommon condition may, at times, provide important diagnostic as well as therapeutic guidelines.

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