IMMUNOLOGY

# Unusual Presentation of Jaccoub's Arthropathy in Rhupus Syndrome

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## ABSTRACT

***Introduction:*** **Rhupus syndrome is a rare condition in which patient presented with coexisting rheumatoid arthritis (RA) and systemic lupus erythematosus (SLE) with erosive arthropathy. In this case, we aim to highlight a case of Rhupus syndrome presenting with Jaccob's arthropathy which is a deforming, non-erosive arthropathy.**

***Case Presentation:*** **A 49 years old woman presented to the Rheumatology clinic with multiple joint pain involving the shoulder, neck, elbow, ankle, proximal metacarpophalangeal joints (MCPJ) and interphalangeal joints for the past 6 months. The pain was associated with stiffness and restricted movement especially during early morning which increases in severity. There was loss of weight and loss of appetite. She had a family history of rheumatoid arthritis and systemic lupus erythematosus (SLE) among her family members. She was initially diagnosed with rheumatoid arthritis and started on the disease-modifying anti-rheumatic drugs (DMARDS). During consecutive follow-up, her full blood count (FBC) showed persistent leucopenia with positive ANA and ENA, hence a diagnosis of Rhupus was made.**

***Conclusion:*** **Patients with rhupus syndrome may present with overlapping features of rheumatoid arthritis and SLE, hence a high clinical suspicion is required to diagnose a patient with rhupus syndrome.**

## KEY WORDS

**jaccoub's arthropathy, rhupus syndrome**

## INTRODUCTION

Jaccoub's arthropathy (JA) is a deforming, non-erosive arthropathy; first described by a French physician FS Jaccoud in 1869[1)]. It has been identified in various autoimmune diseases particularly in systemic lupus erythematosus (SLE) patients, affecting around 10-35% SLE patients[2)].

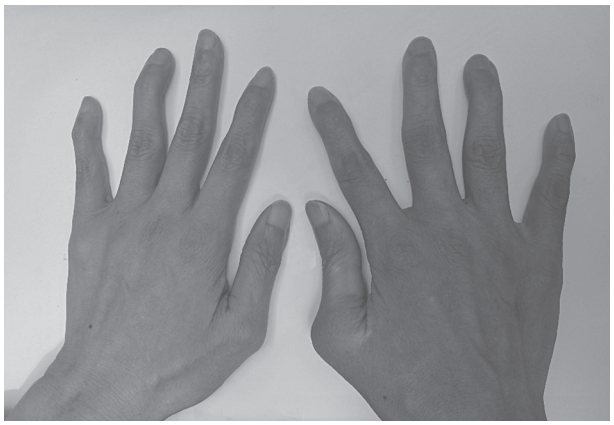

**Figure 1:** Both hands showing swan neck deformity; hyperextension of the PIP and flexion of the DIP joints

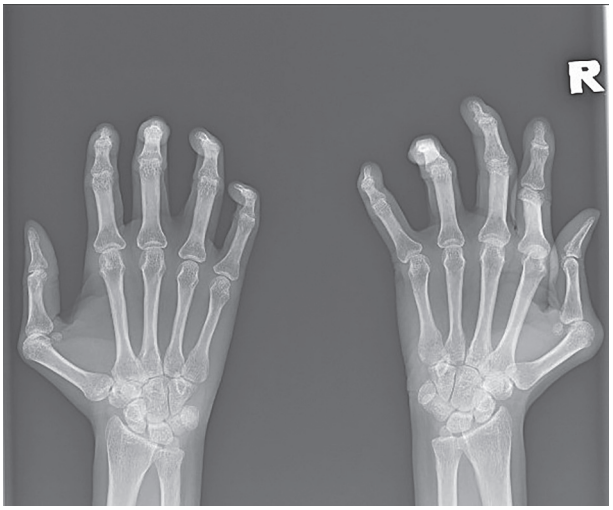


**Figure 2:** This radiographic film of both right and left hands shows subluxation of the proximal interphalangeal joint of the first finger without evidence of bony erosions.

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Besides that, it has been recognized in other autoimmune diseases including systemic sclerosis, dermatomyositis, vasculitis, infectious and neoplastic pathologies[1]. Clinically, JA can involve all joints but it mainly affects the hands, others including feet, knees, and shoulder joints. Patients usually presented with severe deformities of the hand including swan neck and boutonniere deformities, Z-deformity of the thumb, ulnar deviation, and multiple non-erosive subluxations[3].

Rhupus syndrome is a term used to describe patients with coexisting rheumatoid arthritis (RA) and systemic lupus erythematosus (SLE). In this report, we describe a case of rhupus syndrome presenting with jaccoub's arthropathy.

## CASE PRESENTATION

A 49 years old woman with underlying hypertension presented to the Rheumatology clinic with multiple joint pain involving the shoulder, neck, elbow, ankle, proximal metacarpophalangeal joints (MCPJ), and interphalangeal joints for the past 6 months. The pain was associated with stiffness and restricted movement especially during early morning which increases in severity. There was loss of weight and loss of appetite. She lost about 5 kg in 3 months. She had a family history of rheumatoid arthritis and systemic lupus erythematosus (SLE) among her family members.

She was initially diagnosed with rheumatoid arthritis and started on the disease-modifying anti-rheumatic drugs (DMARDS). During consecutive follow-up, her full blood count (FBC) showed persistent leucopenia with positive ANA and ENA, hence a diagnosis of Rhupus was made.

Laboratory investigation showed persistent low white cell count, positive anti-nuclear antibody (ANA) with titre > 1: 320 of homogenous pattern, anti-cyclic citrullinated peptide (ACCP) antibodies positivity 45.6 IU/ml and rheumatoid factor of 96 IU/ml. Her dsDNA was persistently negative with a normal erythrocyte sedimentation rate (ESR) of 29 mm/h. Her extractable nuclear antigen (ENA) showed positivity against SSA/Ro 60 kD and SSA/Ro 52 kD.

She was started on Hydroxychloroquine 200 mg BD, Methotrexate 20mg/week, Celebrex 200 mg PRN, prednisolone 5mg OD, and sulfasalazine 1 g BD. Her total white count improved with the treatment given.

## DISCUSSION

Jaccoub's arthropathy is a deforming, non-erosive arthropathy commonly seen in SLE. The prevalence of Jaccoub's arthropathy in SLE is about 10-35%[2]. It was previously described to occur after rheumatic fever, however, it can also occur in other rheumatic conditions, for example, systemic sclerosis, dermatomyositis, and non-rheumatic condition such as infection and neoplastic pathologies[1]. In this report, we described an unusual presentation of Jaccoub's arthropathy in Rhupus syndrome.

Rhupus syndrome is a rare clinical condition, first described by Peter Schur in 1971 where the patient presented with coexisting features of rheumatoid arthritis and SLE[4]. Elena et all. defined rhupus as referring to the coexistence of erosive symmetrical polyarthritis with a typical manifestation of RA and clinical signs of SLE with positive anti-dsDNA, and anti-Sm[5]. However, up to date, there is no clear consensus on how to define or diagnose rhupus syndrome.

The incidence of Rhupus in patients with arthritis is very low about 0.01% to 0.2%[6]. A study by Silvia et all, reported low prevalence of 0.46% in the study population[7]. The exact pathophysiology of the condition is not well understood, however, it is believed that the clinical presentation, laboratory and serological findings, and radiological manifestation will be overlapped of those of rheumatoid arthritis and SLE.

In this case, the patient presented with Jaccoub's arthropathy, which is a deforming, but non-erosive arthropathy compared to the erosive arthropathy seen in rhupus syndrome. The pathology behind it is thought to be due to ligamentous laxity of the joints. This differentiating features can be appreciated by observing the plain radiograph to see the presence or absence of bony erosions. As for rhupus syndrome, the patient will usually present with an erosive bone lesion as expected for patients with rheumatoid arthritis.

The extractable nuclear antigen (ENA) showed positivity against SSA/Ro 60 kD and SSA/Ro 52 kD. Ro60 and Ro52 are cellular proteins with a molecular weight of 60 and 52 kD respectively. They are situated in different cellular compartments, with Ro60 confined to the nucleus and nucleolus while Ro52 confined to the cytoplasm. Unlike anti-La antibodies which are specific for Sjogren's syndrome and SLE, anti-Ro antibodies are detected in a wide range of autoimmune diseases, including SLE with 32 percent and rheumatoid arthritis with 15 percent. The prevalence of the anti-Ro60 and anti-Ro52 antibodies in SLE was reported as 49 and 43 percent respectively[8]. The presence of anti-Ro52 antibodies is suggestive of more severe disease.

The diagnostic challenge faced in rhupus syndrome is due to the uncertainty in disease characterization to establish the presence of the rhupus, thus reflecting the lack of consensus and criteria in defining the disease. Rottenberg *et al*. reported that rheumatoid arthritis most often preceded rhupus[9]. This is in agreement with the study by Jing Li et all. which found out that most of the rhupus patients (83.9%) were diagnosed with RA at the onset of disease[6].

In summary, rhupus syndrome is a rare clinical condition with overlapping features of RA and SLE. Thus, the harmonization in the definition and classification criteria is needed for early diagnosis, for choosing the proper management and improving the outcome of the patient.

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