Case study

Rhupus Syndrome- A Case Report

ABSTRACT

Rhupus Syndrome is a rare entity with features of both Systemic Lupus Erythemtosis (SLE) and Rheumatoid Arthritis (RA). Though case reports have previously been described, it is still rarer to find the illness in the Male population. Here, we present the case scenario of a middle-aged male presenting with Rhupus syndrome and muscle weakness as the primary complaints. Physicians should thus, remain alert to manifestations of autoimmunity and features of overlap syndromes.

Keywords: Rhupus Syndrome, Systemic Lupus Erythematosis, Rheumatoid Arthritis

1. INTRODUCTION

Overlap between two or more autoimmune diseases is a common phenomenon, and some studies suggest that these patients are different from patients with a single disease in terms of presentation, prognosis and treatment strategies. Rhupus Syndrome is a rare combination of SLE and RA, and is characterized by the presence of erosive arthritis together with signs and symptoms of systemic lupus erythematosis. Rhupus Syndrome is a rare clinical entity which has an estimated prevalence rate of 0.09%, out of which the predominance of the male gender is still rarer. In this scenario, we describe the case of Rhupus Syndrome in a middle aged male patient presenting with muscle weakness as the primary complaint.

2. PRESENTATION OF CASE

This is a Case Report of a Middle-aged Male presenting with Rhupus Syndrome in the General Medicine OPD of Jawaharlal Nehru Medical College, Aligarh. A 55-year-old male presented to us with the complaints of bilaterally symmetrical proximal upper and lower extremity weakness progressively increasing over the past six years, along with a history of swelling and pain of small joints of hands, oral ulcers and fever.

The weakness progressed such that, one month before hospital admission, he reported increasing fatigue and difficulty in standing from a seated position even with the support of his hands, in climbing stairs and difficulty in combing his hair.

On Examination, power was decreased in the upper limbs with only 1/5 power at the shoulder and 3/5 at elbow in all ranges of motion; in the lower limbs power was decreased to 3/5 at the hip and 4/5 at knee bilaterally in all ranges of motion with absent triceps and biceps reflexes and generalized wasting of muscles of all four limbs. Bilateral Wrist joints showed presence of ulnar deviation, with boutonnière deformity of the Left little finger and Hitch-Hike deformity of the Right thumb. Routine investigations showed anemia with thrombocytopenia with an elevated ESR, CRP and ANA. CPK and TSH were in the normal range.

Immunoserology demonstrated highly positive anti-ds DNA, Positive RA factor, Positive Anti-CCP and Hypo complementaemia. Biopsy of the Gastrocnemius showed endomysial and perimysial lymphocytic inflammation suggestive of Inflammatory Myopathy. Electromyography of bilateral upper and lower extremities was consistent with myopathic processes.

The patient was begun on oral treatment of Steroid and DMARD and was followed up after 6 weeks, and showed marked improvement in muscle strength.

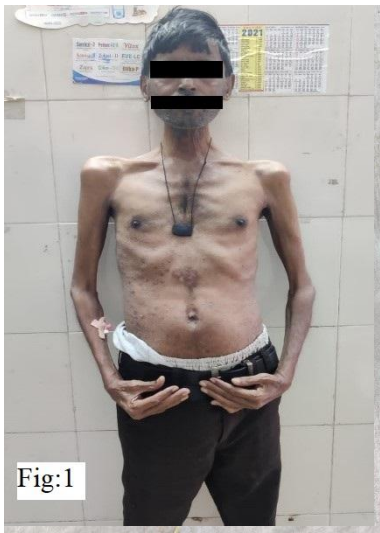
Fig:1


Fig: 2

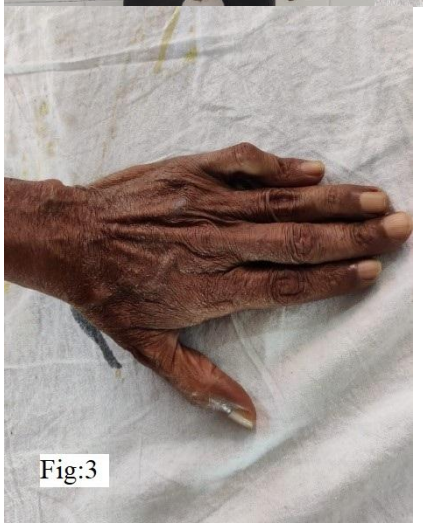
Fig:3

Figure 1- shows wasting of the muscles of the upper limb, including the biceps, triceps and deltoid muscles. Figure 2 & 3 – shows boutonnière deformity of left little finger and hitch-hike deformity of left thumb.

3. DISCUSSION

Arthritis is a common manifestation in many systemic autoimmune diseases, in systemic lupus erythematosus (SLE) a mild symmetric synovitis affecting little and medium sized joints is frequently observed at disease onset and is usually treated with low dose steroids and antimalarial drugs. Late deformities are also described in up to 35% of SLE patients, these are typically reducible and non-erosive defining the so called "Jaccoud's arthropathy"[1]. In rare cases (up to 3–5%) a severe, erosive and deforming arthropathy, clinically indistinguishable from rheumatoid arthritis (RA) can be observed; this clinical entity is traditionally known as "rhupus" to describe patients with coexistence of SLE and RA[2].

"Rhupus" or "rhupus syndrome" is a poorly described and underdiagnosed disease in which features of both rheumatoid arthritis (RA) and systemic lupus erythematosus (SLE) appear in the same patient, most often sequentially. The SLE-related involvement is usually mild, dominated by hematological abnormalities and skin, serosal and renal involvement. The natural history of rhupus arthritis follows an RA-like pattern and can progress towards typical inflammatory erosions, deformations and disability. [3]

The term rhupus refers to the coexistence of erosive symmetrical polyarthritis, typical manifestation of rheumatoid arthritis (RA), and clinical signs of SLE in the presence of anti double-stranded DNA (anti-dsDNA) and/or anti-Smith antibodies (anti-Sm). However, to date, no consensus exists on how to define the rhupus syndrome.[4]
Though cases of Rhupus syndrome have previously been reported [5], it is a rarity to find this illness in males with muscle weakness as the predominant complaint.

4. CONCLUSION

Overlap Syndromes are a rare phenomenon. Rhupus Syndrome is characterized by symmetric polyarthritis of the small and large joints and symptoms of SLE along with the presence of specific autoantibodies with high specificity (anti-dsDNA or anti-Smith for SLE and rheumatoid factor or Anti-CCP Antibodies for RA) [6]. Rhupus arthropathy is an overlapping syndrome of rheumatoid arthritis and systemic lupus erythematosus that is defined by erosive polyarthritis accompanied by an overlap of clinical and immunological symptoms.

Physicians should remain alert to manifestations of autoimmunity and overlapping disease features.

CONSENT

As per international standard or university standard, parental(s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

The protocol of the study was approved by the Institutional Ethical Committee and the study was conducted as per the standards of Good Clinical Practice and the Helsinki Declaration.

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