Rhupus Syndrome: A Case Report

ABSTRACT

Rheumatoid arthritis and systemic lupus erythematosus are two extremely common autoimmune illnesses that cause disability and poor quality of life. The innate immune system, a topic in autoimmune disorders that has long been neglected, is becoming more significant and represents a new area of attention for the treatment of these conditions.

People with systemic lupus erythematosus (SLE) who also exhibit symptoms of another rheumatologic disorder, such as rheumatoid arthritis, Sjogren's syndrome, and/or vasculitis, are said to have overlap syndromes.2 Rhupus Syndrome is a rare condition that shares traits with both rheumatoid arthritis (RA) and systemic lupus erythematosus (SLE). Even if there have been case reports before, the condition is still more uncommon in men than in women.
Here, we present the case scenario of a middle-aged male presenting with Rhupus syndrome and muscle weakness as the primary complaints. He was initiated on steroid and his improvement was recorded after a 6 week follow up. Physicians should thus, remain alert to manifestations of autoimmunity and features of overlap syndromes.

Keywords: Rhupus Syndrome, Systemic Lupus Erythematosis, Rheumatoid Arthritis, Overlap Syndromes

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" [1]. "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" [3]. 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. Biochemical parameters are shown in table 1.

Table no.1

INVESTIGATIONS	VALUES	NORMAL RANGE
1.CBC	Hb- 8.2 g/dL Platelet count- 38,000 /L	12-18 g/dL 150-400 × 10^{9} /L
2.ESR	45 mm/hr	0-15 mm/hr
3.C-Reactive Protein	12 mg/L	0-5 mg/L
4.ANA (Anti-nuclear Antibodies)	++ (>1:80)	
5.Creatine phosphokinase	215 U/L	60-350 U/L
6.Serum Thyroid Stimulating Hormone	3.5 IU/mL	0.4-4.0 IU/mL
7. Immunoserology	Anti-dsDNA- +++ Rheumatoid Factor- +++ Anti-CCP- ++ C3,C4- Decreased	
8. HbsAg/ Anti-HCV/ anti-HIV	Negative	

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-Prednisolone in a dose of 60mg/day and Disease-modifying antirheumatic drugs (DMARDs), which in this case was Hydroxychloroquine in a dose of 200mg/day. The patient was followed up after 6 weeks, and showed marked improvement in muscle strength, and was now able to perform his daily activities.

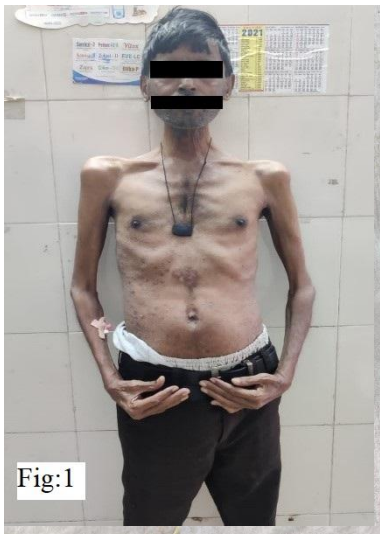
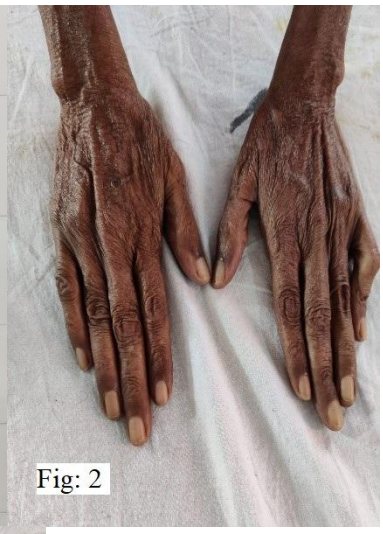


Fig:1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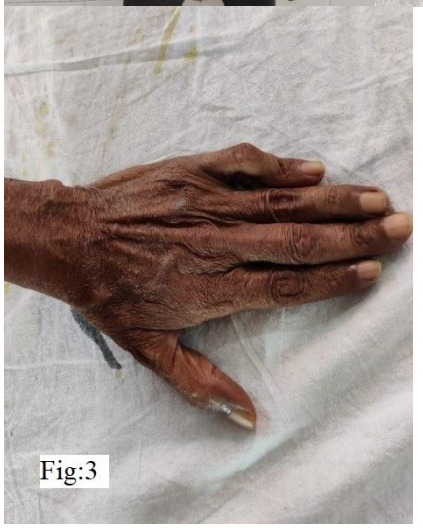
Fig:3

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" [4]. 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”.[3] 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.

A poorly understood and underdiagnosed illness called "rheumatoid arthritis" (RA) and systemic lupus erythematosus (SLE) manifest in the same patient, most frequently consecutively, is called "rhupus" or "rhupus syndrome"5. The haematological problems, cutaneous, serosal, and renal involvement predominate over the typically modest SLE-related involvement. The progression of rhupus arthritis' natural history might lead to the characteristic inflammatory erosions, deformations, and impairment seen in RA.

In the presence of anti-double-stranded DNA (anti-dsDNA) and/or anti-Smith antibodies (anti-Sm), erosive symmetrical polyarthritis, the classic presentation of rheumatoid arthritis (RA), and clinical indicators of SLE are collectively referred to as rhupus. On how to characterize the rhupus syndrome, however, there is still no agreement.
Though cases of Rhupus syndrome have previously been reported, it is a rarity to find this illness in males with muscle weakness as the predominant complaint. [6]

4. CONCLUSION

Overlap Syndromes are an uncommon occurrence. Symmetric polyarthritis of the small and large joints, SLE-like symptoms, and the presence of highly specific autoantibodies (anti-dsDNA or anti-Smith for SLE and rheumatoid factor or anti-CCP Antibodies for RA) are all characteristics of rheumatoid syndrome [7]. Rhupus arthropathy, which is characterized by erosive polyarthritis and an overlap of clinical and immunological signs, is a syndrome that combines rheumatoid arthritis and systemic lupus erythematous [8]. Physicians should remain alert to manifestations of autoimmunity and overlapping disease features.

AUTHORS' CONTRIBUTIONS

Corresponding Author: Literature search, Clinical studies, Data acquisition, Data analysis, Statistical analysis, Manuscript preparation, Manuscript editing, Manuscript review, Guarantor.
Author Ahmad: Data acquisition, Data analysis, Statistical analysis, Manuscript preparation.
Author M. Aslam: Concepts, Design, Definition of intellectual content

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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