

Case Report

A Case Report of Rhupus Syndrome: An Overlap Syndrome of Rheumatoid Arthritis and Systemic Lupus Erythematosus

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ABSTRACT

Rhupus an extremely uncommon condition is used to describe patients who have features of both rheumatoid arthritis and systemic lupus erythematosus. We report a 23 year old female who initially presented with features of rheumatoid arthritis and later developed features of systemic lupus erythematosus. Antibodies were positive for both rheumatoid arthritis and systemic lupus erythematosus. The patient was diagnosed as Rhupus Syndrome as the patient had manifestations of both rheumatoid arthritis and systemic lupus erythematosus.

Key words: Rhupus Syndrome, Rheumatoid Arthritis, Systemic Lupus Erythematosus.

INTRODUCTION

Rhupus is a term used to describe patients with co existence of rheumatoid arthritis (RA) and systemic lupus erythematosus (SLE). It is a rare clinical condition and was first described by Schur et al. [1] Rheumatoid arthritis prevalent in 0.5-1.0% of adult population [2] and SLE prevalent in 10-400/100,000 [3] are not uncommon while as the coexistence of two in the same patient is a rare phenomenon, which has been estimated between 0.01% and 2%. [4-6] The first clinical observations that helped to identify the Rhupus syndrome was described by Toone, [7] who found the presence of LE cell phenomenon in patients with RA, which was considered an exclusive feature seen in SLE patients. Later it was verified that some patients with an

initial diagnosis of rheumatoid arthritis (RA) showed erosive arthritis and developed symptoms or signs of SLE, and the patients with initial feature of SLE later developed RA; but the former was more common. [8] We present a 23 year old female as a case of Rhupus Syndrome who initially developed features of Rheumatoid Arthritis and later developed clinical and serological features of SLE.

CASE REPORT

A 23 year old female presented with complaints of pain and swelling of small joints of hand from past 6 months. There was a history of morning stiffness for more than one hour duration. On examination the 2nd, 3rd and 4th metacarpophalangeal joints (MCP's) of right hand and 2nd and 3rd

MCP's of the left hand were swollen and tender. In addition to this left elbow was also swollen and tender. RF (Rheumatoid Factor) and anti-CCP (anti-cyclic citrullinated antibodies) antibodies were more than 5 times upper limit of normal. ESR (Erythrocyte Sedimentation Rate) and CRP(C Reactive Protein) was raised. X ray of the hands was showing osteopenia as shown in figure 1.

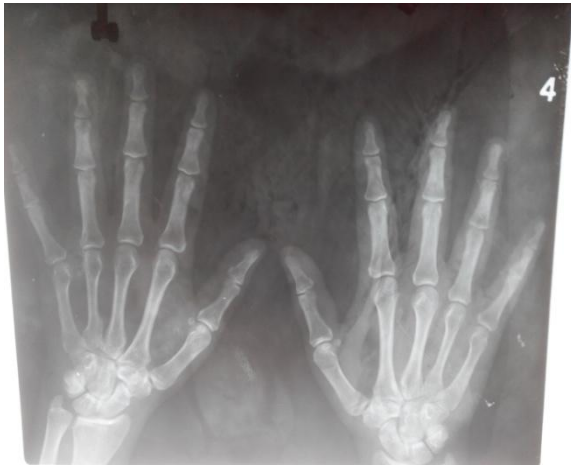


Figure 1: X-ray of the hands showing osteopenia.

A patient was diagnosed as rheumatoid arthritis using 2010 ACR/EULAR (American College of Rheumatology/European League against Rheumatology) Criteria and was put on methotrexate and hydroxychloroquine. One year after being diagnosed as RA patient developed butterfly rash on face as shown in figure 2. In addition to this patient also complained of photosensitivity and excessive hair fall with receding of hair line.



Figure 2: Butterfly rash on face which is a feature of SLE.

Patient also complained of pain during chewing food. On oral examination patient has irregularly shaped, raised white plaques with area of surrounding erythema as shown in figure 3.



Figure 3: Oral Ulcers

Labs were showing anemia with thrombocytopenia. ESR was raised. ANA (Anti Nuclear Antibodies), anti-ds DNA and anti-smith were more than 10 times normal.

Using 2010 ACR/EULAR criteria and 2012 SLICC(Systemic Lupus International Collaborating Clinics) criteria, patient was fulfilling both the criteria with RF and anti-CCP positive ; and ANA, anti-ds DNA and anti-smith also positive. Patient was diagnosed as Rhupus Syndrome was put on prednisolone 1 mg/kg body weight and Hydroxychloroquine.

DISCUSSION

The co existence of RA and SLE in a same patient called a Rhupus Syndrome is a rare clinical entity reported in 0.01 and 2%. [4-6] Rhupus syndrome is characterized by symmetric polyarthritis of the small and large joints which is erosive on radiography and accompanied by clinical signs and symptoms of SLE and by the presence of specific auto antibodies with high specificity (anti-dsDNA antibody or anti-Smith for SLE and Rheumatoid factor or anti-citrullinated peptide antibodies for RA) [9] as in our patient. But in our patient there was osteopenia but no clear erosions as disease duration was only one year, and for

erosions it usually takes more than 2 years. [10]

Some studies mention that rhus arthritis was another variant of systemic lupus erythematosus arthritis. [11,12] However others like Amezcua Gyerra et al. [13] support the possibility that Rhus is an overlap between RA and SLE, because highly specific autoantibodies for RA and for SLE (anti-dsDNA and anti-Smith antibodies) are detected in coexistence. It also mentions that the features of RA are preceded by the features of SLE.

From the analysis of the HLA-DR molecules, the genetic characterization of patients with a diagnosis of rhus showed clear differences with those present in patients with RA and SLE, supporting the possibility of an independent entity. [4,5,14] It was observed that these patients were distinguished by the presence of alleles HLA-DR4, DR2, DR6 and DR1, presenting a greater frequency of DR1 than patients with RA and less frequency of DR3 than patients with SLE. [5] A study in China observed that the mean age of rhus was 36.8 years. Patients developed SLE 7.7 years after initial presentation of RA and SLE associated severe organ damages other than hematopoietic abnormalities was less frequent. [15] Features of RA are dominated by polyarthritis in all cases and rheumatoid nodules in around 40% of the cases. [5] SLE is usually manifested by cutaneous (butterfly skin rash, photosensitivity and alopecia), hematological (leukopenia and thrombocytopenia), serosal (pleural and pericardial effusion) and mucosal involvement [16] The clinical behavior of individual diseases RA and SLE when they coexist in Rhus is not clearly known. [17,18]

CONCLUSION

Rhus is an overlap of RA and SLE as it is associated with clinical, radiological and immunological features of both RA and SLE. Although rare but it should be looked in both patients of RA and SLE as its

disease course and therapy varies from both RA and SLE

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