

## The Coexistence of Systemic Lupus Erythematosus and Rheumatoid Arthritis in a Young Male Patient; A Rare Clinical Condition: Rhupus Syndrome

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**ABSTRACT** The coexistence of systemic lupus erythematosus (SLE) and rheumatoid arthritis (RA) is called Rhupus syndrome and an extremely rare overlap syndrome. It is more common in women, as in SLE and RA, and the majority of cases in the literature are female patients. This case report was planned to be presented for the reason that rhupus syndrome is both rare and was detected in a male patient.

**Keywords:** Rheumatoid arthritis, systemic lupus erythematosus

Rhupus syndrome; is a term used to describe the coexistence of systemic lupus erythematosus (SLE) and rheumatoid arthritis (RA). Clinically, symmetrical deforming arthritis of small and large joints is together with signs and symptoms of SLE. High specificity autoantibodies (anti-dsDNA, anti-Sm, anti-CCP) are associated with this clinical condition.<sup>1,2</sup> The coexistence of SLE and RA has a very low incidence (0.01-0.2%) in patients with arthritis and the incidence is <2% in patients with connective tissue diseases.<sup>2</sup> It is more common in women as it is in SLE and RA. When we look at the literature, most of the publications in the form of case reports are composed of female patients.<sup>2</sup> This case report was planned to be presented for the reason that rhupus syndrome is both rare and was detected in a male patient. A written informed consent was obtained from the patient.

### CASE REPORT

A 24-year-old male patient referred to our rheumatology outpatient clinic due to an increase in existing joint complaints. The patient is being treated in another outpatient clinic with a diagnosis of rheumatoid arthritis and he is treated with prednisolon 5 mg/day. On rheumatologic examination; he had arthralgia, photosensitivity, arthritis and morning stiffness in hand joints. Joints examination revealed tenderness with swelling of right hand 3<sup>rd</sup> proximal interphalangeal joint, left hand 2<sup>nd</sup> proximal interphalangeal joint and there was sensitivity and mild effusion in the left knee joint. He had not any joint deformity and restriction of movement. The results of the investigations are summarized (Table 1).

**TABLE 1:** Tests and results.

White Blood Cell Count	7600/ $\mu$ L
Hemoglobin Level	14 g/dL
Platelet Count	317000/ $\mu$ L
Creatinine	0,82 mg/dL
Erythrocyte Sedimentation Rate	67 mm/h
CRP	39 mg/L (0-8 mg/L)
Rheumatoid Factor	22 IU/mL (0-20 IU/mL)
Anti-Cyclic Citrulline Peptide	15 U/mL (0-5 U/mL)
Antinuclear Antibody	4 + homogeneous
Anti-dsDNA	3 +
C3c	0,67 g/L (0,79-1,52 g/L)
C4	0,11 g/L (0,16-0,38 g/L)
Urine analysis	Normal

The clinical and laboratory findings of the patient were consistent with both SLICC and 2010 ACR/EULAR RA classification criteria, so the patient was diagnosed with Rhupus syndrome (SLE+RA).<sup>3,4</sup> Extraarticular involvement was not detected in the patient. The patient was started treatment with methotrexate 15 mg/week, folic acid 5 mg/week, hydroxychloroquine 400 mg/day, methylprednisolone 8 mg/day. Methylprednisolone therapy was discontinued with dose reduction. The patient continues outpatient clinic controls without any signs or symptoms.

## DISCUSSION

Rhupus syndrome is a term used to describe the coexistence of systemic lupus erythematosus (SLE) and rheumatoid arthritis (RA). Clinically, symmetrical deforming arthritis of small and large joints is together with signs and symptoms of SLE. High specificity autoantibodies (anti-dsDNA, anti-Sm, anti-CCP) are associated with this clinical condition.<sup>1,2</sup> The term of rhupus was first used by Peter Schur in 1971 for describe patients who satisfy the criteria for both SLE and RA.<sup>2,5</sup> It is very rare that two or more connective tissue disorders occur together. In particular, the incidence of SLE and RA association is very low (0.01-2%) in patients with arthritis and the incidence is <2% in patients with connective tissue diseases.<sup>1,2,6</sup> Rhupus syndrome is more common in women, as in SLE and RA. When

we look at the literature, most case reports are composed of female patients.<sup>2,6</sup>

Although the pathogenesis of Rhupus syndrome is still not fully understood, it is clear that autoantibodies play a key role in the formation of the disease, as in SLE.<sup>1</sup> In addition, the assumptions about pathogenesis, antibody positivity (anti-dsDNA, anti-Sm, anti-CCP), radiology findings and treatment support the idea that Rhupus is an overlap syndrome of SLE and RA.<sup>1</sup>

In a publication of Liu et al., it is stated that the age at onset of rhupus is younger than that of RA, but similar to that of SLE.<sup>7</sup> Clinically, the first findings of rhupus syndrome are generally RA related symptoms and characterized by arthritis.<sup>7,8</sup> Symptoms of SLE manifested after an average of 4-9.2 years.<sup>7,8</sup> The incidence of baseline symptoms consistent with SLE or SLE and RA comorbidity is 7.8%, which is quite low.<sup>7</sup>

In Rhupus syndrome, the most frequent clinical manifestations of RA are erosive symmetric polyarthritis and rheumatoid nodule in 40% of cases. The most common SLE characteristics are usually skin related (malar rash, alopecia and photosensitivity), hematological (leukopenia, thrombocytopenia), serositis (pleural and pericardial effusion) and mucosal compromise.<sup>6</sup> Rhupus patients compared with SLE patients; Tani et al. observed that, renal involvement was significantly less in the Rhupus group, and there was no difference in neuropsychiatric, cutaneous, and hematological involvements and serositis. In the same report, it was found that the cumulative erosive burden was significantly higher in patients with rhupus than in SLE patients and similar to RA patients.<sup>9</sup>

Treatment of Rhupus syndrome usually consists of DMARDs such as methotrexate and leflunomide, which are used in combination with low-medium dose corticosteroids.<sup>2</sup> Case reports have been published that mycophenolate mofetil and cyclosporin A are effective in the treatment of rhupus.<sup>6,10</sup> Tumor necrosis factor inhibitors showed little effect on Rhupus or SLE, and may even lead to disease aggravation, despite reports of their success in RA treatments.<sup>2</sup> There are case reports on

the use of rituximab and abatacept in the treatment of Rhupus. It is mentioned that these treatments are promising.<sup>2,6,11</sup>

The prognosis of Rhupus syndrome is often related to the severity of organ involvement. However, the prognosis is typically better than SLE, worse than RA.<sup>2</sup>

This case report was planned to be presented for the reason that rhupus syndrome is both rare and was detected in a male patient. So, it is important to bring up this syndrome in patients whose clinical and laboratory findings are appropriate with Rhupus, even if it is in a male patient. Thus, the disease can be diagnosed early and the prognosis can be improved with appropriate treatment.

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### Conflict of Interest

*No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.*

### Authorship Contributions

*This study is entirely author's own work and no other author contribution.*

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