Rhupus Syndrome, an Overlap of Systemic Lupus Erythematosus(SLE) and Rheumatoid Arthritis(RA); Case Report

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Introduction

Nearly 25% of systemic symptoms of autoimmune disorders are misdiagnosed and mismanaged and often come with flare-ups or complications. The coexistence of multiple rheumatic diseases makes it more challenging regarding both management and diagnosis. Several types of overlap syndrome are documented in the literature. While two or more rheumatic diseases coexist, one disease often is clinically predominant. Rhupus syndrome, which combines the clinical and laboratory aspects of rheumatic arthritis (RA) and systemic lupus erythematosus (SLE), is one of the uncommon and sporadically documented overlaps. SLE and RA are systemic autoimmune rheumatic illnesses that affect multiple organs and systems and have unique clinical and serological traits. (1)

“ Rhupus” was first coined in 1971 to describe patients who satisfy the criteria for both systemic lupus erythematosus(SLE) and rheumatoid arthritis(RA). (2) it has been defined as a deforming and symmetric polyarthritis accompanied by symptoms of SLE and the presence of antibodies such as ant-ds-DNA, anti-smith, and rheumatoid factor with or without ant-CCP antibody. (a) because of the rarity of the disease the prevalence, pathophysiology, natural history, and radiological and immunological profiles profile of rhupus are poorly described.

There is no known etiology causing the disease but there are limited studies suggesting the combined role of genetic, immunological, hormonal, and environmental factors in the progression of the disease. (3) identification of rhupus is very essential owing that their therapy and outcome differ from those patients having RA or SLE alone. (4)

We report one such rare case below.

Case presentation :

History and physical examination

A 44-year-old female known Rheumatoid arthritis patient on treatment for the past 10 years initially presented with a complaint of multiple Joint pain and lower back pain of 4 months duration. Associated with that she also had a history of morning stiffness of the involved joints which usually stayed for 2 to 3 hours. Up on Physical examination, the initial presentation revealed a tender wrist, left knee, metacarpophalangeal (MCP), and proximal interphalangeal (PIP) joints of her hands but no limitation of movement. photo(figure 1) showed subluxation of the bilateral interphalangeal joint of the thumb.

Investigation and treatment

Laboratory investigations revealed a positive rheumatoid factor and the anti-ccp antibody was negative. The X-rays of the hands (Figure 2A and B) showed subluxation of the bilateral interphalangeal joint of the thumb, (Hitchhiker's thumb deformity).

with these presentations, she was diagnosed with RA. She was initially started on Methotrexate 10 mg po/week and prednisolone 10 mg po/d. Throughout her course of treatment and follow up she has had multiple flare-ups because of poor drug adherence. After 9 years and 3 months of treatment and follow-up, she developed an erythematous non-itchy patch on both cheeks, alopecia, and oral ulcers on the buccal mucosa bilaterally. And physical examination this time revealed chronically swollen PIP and MCP joints in her hands.

On laboratory investigation Urinalysis showed Blood: +2, Protein: +3, RBC 5-8/ hpf, WBC: 3-5/hpf. 24 hours urine protein: 3024 mg/dl. ANA (qualitative): Positive, Anti-dsDNA Antibody: 1424.78 IU/ml (strongly positive), Anti Smith Antibody: 190 Units (Strongly positive).

Based on the above findings, a mixed connective tissue disease of systemic lupus erythematosus on top of rheumatoid arthritis, Rhupus, was considered and she was started on MMF (mycophenolate mofetil) 500 mg PO bid, chloroquine 250 mg PO/day, prednisolone 1mg/kg (55 mg) po/day, and folate 5 mg PO/day.

Outcome and follow-up

Currently, the patient has continued her follow-up and is showing improvement. The improvements include the resolution of the erythematous non-itchy patch on her cheeks and the oral ulcers on her buccal mucosa bilaterally. The swellings on her joints mentioned above have also been decreasing on each subsequent follow-up. The proteinuria has also decreased.

Discussion

Rhupus is one of the overlap syndromes in CTD(connective tissue disease) in which patients present with criteria satisfying both RA and SLE. The prevalence varies from 0.01% to 2%.(3,4). The coexistence of RA and SLE, Rhupus syndrome, is very rare. The absence of specific diagnostic criteria to make a diagnosis makes it very difficult to identify such patients. reports showed the presentation first comes with clinical features of RA followed by SLE, and it is more common in women. 5

RA is an autoimmune inflammatory illness involving joints and other organ system. There is no known etiology but genetic and environmental factors are contributory. Synovial inflammation orchestrated by T-cells, and B-cells, along with the release of pro-inflammatory cytokines by TH1 cells is the key pathophysiologic process.

The exact etiology and pathology of SLE remains unclear. But complicated interaction of environmental, genetic, and immunologic factors is probably involved. The loss of immune tolerance increases the antigenic load, and shifting of Th1 to Th2 immune responses leads to B-cell hyperactivity and the production of pathogenic autoantibodies. The coexistence of these two diseases with different pathophysiologies makes it unique. (6)Previous studies have shown that patients with rhupus were found to have a lower incidence of malar rash, hemolytic anemia, and renal and neurological involvement compared with the SLE. The rhupus group also had a rare incidence of severe renal disorders such as nephrotic syndrome and renal failure. (7)Previous studies have also shown a lower incidence of visceral organ involvement compared with SLE alone. (8)

For nearly 10 years our patient had erosive polyarthritis with episodes of flare-ups. She was on treatment for RA, and after 10 years of treatment, she presented with an erythematous non-itchy patch on both cheeks, alopecia, and oral ulcers on the buccal mucosa bilaterally which are evidence of SLE (systematic lupus erythematosus).

The rare coexistence of those two diseases and the co-occurrence of some of the symptoms such as arthralgia makes it difficult to pick it early.

Management of rhupus is not yet established and is primarily an expert's recommendation. The majority of reports of rhupus had been managed based on the predominating features. Recent preliminary findings showed the promising effect of Bacitracin along with other DMARDs, regarding reducing disease activity and improving quality of life. Further investigation is needed with a larger sample size to confirm the efficacy and safety of bacitracin in rhupus (9)

Conclusion.

Rhupus syndrome is a rare overlap of RA and SLE with variable clinical features satisfying the criteria for both SLE and RA. The treatment and prognosis are different from RA and SLE alone. Therefore, early recognition and initiation of appropriate management are very important for prognosis.

Data Availability Statement

The data that support the findings of this case report are available from the corresponding author upon reasonable request

Ethical Approval

The author's institution does not require ethical approval for the publication of a single case report.

Consent for Publication

The patient provided written informed consent for the publication of details including, history, physical findings, laboratory reports, and imaging results.

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Disclosure

We authors have no conflict of interest.

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