

## CASE REPORT

### RHUPUS SYNDROME: A CASE REPORT

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**ABSTRACT:** Rhopus syndrome, overlap of Rheumatoid Arthritis (RA) & Systemic Lupus Erythematosus (SLE), is an extremely uncommon condition. Further involvement of lung (pneumonitis) is rare. We are reporting a case of 28yr old female with multiple joint pain for 10-12yrs, diagnosed & treated as rheumatoid arthritis for 4 months. She presented with increased severity of joint pain along with high grade fever, chest pain, and decreased appetite. On further investigation, she was found +ve for ANA (Anti-nuclear antibody) & Anti-ds DNA. Her counts were normal, Chest X ray PA showed right lower zone opacity. Repeated sputum examination, & also c/s of sputum revealed no organism (gram +/-ve or AFB). The diagnosis of rhopus was confirmed by combined manifestations of RA and SLE & diagnosis of pneumonitis was confirmed on basis of clinical, pathological & radiological evidence.

**KEYWORDS:** Rheumatoid arthritis, Systemic lupus erythematosus, Rhopus syndrome, Lupus pneumonitis, ENA (Extractable Nuclear Antigens), Anti dsDNA.

**INTRODUCTION:** Autoimmune diseases Rheumatoid arthritis (0.5-1.0% of adult population)<sup>1</sup> and SLE (10-400/100,000)<sup>2</sup> are not uncommon and are considered to be separate entity. The coexistence of two or more connective tissue diseases in the same patient is a rare phenomenon, particularly for the coexistence of SLE and RA, which has been estimated between 0.01% and 2%.<sup>3,4,5</sup> Joint compromise in systemic lupus erythematosus (SLE) is one of the common manifestations of this disease, with only a small fraction of patients (~5%) developing deformity in the form of Jaccoud's arthropathy. Less than one percent of patients with SLE develop erosive disease which is indistinguishable from rheumatoid arthritis (RA), an entity known as rhopus.<sup>6</sup> The term "rhopus" is traditionally used to describe patients with coexistence of rheumatoid arthritis (RA) and systemic lupus erythematosus (SLE).

Toone et al. Performed the first clinical observations that helped to identify this disease,<sup>7</sup> and the term rhopus was proposed by Schur in 1971.<sup>8</sup>

Rhopus syndrome is a rare clinical condition and there are only a small number of well-documented cases in the literature.

If rhopus represents an overlap of SLE and RA, a subgroup of SLE with an intense joint expression or a completely different entity is still a matter of debate. There is evidence to sustain the existence of rhopus as a true overlap syndrome.<sup>9</sup>

Further most common pulmonary manifestation of RA are peural diseases, may produce pleuritic chest pain and dyspnea. Most common pulmonary manifestation of SLE is pleuritis. This manifestation may respond to NSAID; when severe patients may require a brief course of steroid therapy.

We are reporting a case of rhopus syndrome with right sided pneumonitis without pleural effusion, a rare condition with very few reported cases.

## CASE REPORT

**CASE REPORT:** A Hindu married female patient, of 28yrs age on treatment with analgesics and dexona tab/inj for multiple joint pain, for 10-12 yrs, diagnosed as a case of Rheumatoid Arthritis 4 months back with anti-ccp antibody (strongly positive) done outside & on treatment with NSAIDS, hydroxychloroquine, methotrexate presented with complain of fever on/off for 45 days, Increased Multiple joint pain with swelling of knee, ankle, elbow & small joint associated with difficulty in movement for 45 days, Chest pain and shortness of breath for 1 months, Decreased appetite for 45 days.

On general examination, following were the findings: pt febrile temp 102°F, moderate pallor, blood pressure 110/70 mmHg, pulse 100/min, regular, respiratory rate 24/min, alopecia, oral ulcer, multiple bilateral symmetrical small & large joint swelling & tenderness. There was no skin nodule, skin rash, lymphadenopathy, hepato-splenomegaly, puffy fingers, sclerodactyly, or no history of Raynaud's phenomenon, dysphagia, hemoptysis or any bleeding episodes, muscle weakness, seizures or abnormal behavior. Chest examination revealed impaired percussion notes in right infrascapular areas with decreased vesicular breath sound and right basal crepitation's.

**INVESTIGATION RESULTS:** Total leucocyte count (TLC) - 8000/microl, DLC - N 65%, L 30%, E 5%, M 0%, Blood sugar random-113 mg/dl, Creatinine-0.9mg/dl, Hb 9.6g/dl, normocytic normochromic, Antigen for MP -ve, Widal / typhidot -ve, Montoux test -ve, RK 39 test for kala-azar -ve, HIV / HBS / HCV -non reactive, R/E of urine straw coloured acidic, sugar nil, cell -3-4 / HPF; C/S of urine - sterile, C/S of blood - sterile. Chest x ray PA showed right lower zone opacity. X-ray of both hands showed juxta articular osteopenia, loss of carpometacarpal and metacarpophalangeal joint spaces. Usg of abdomen was normal. Usg of chest showed- small consolidation in right lower zone, no fluid. CRP +ve(increased), **ANA +VE**. After +ve ANA report & on the basis of symptoms we advised for ENA (Extractable Nuclear Antigens) qualitative profile of serum which showed Anti dsDNA +ve.

<b>EXTRACTABLE NUCLEAR ANTIGENS (ENA), QUALITATIVE PROFILE,SERUM</b>	
<b>Test Name</b>	<b>Results</b>
U1-nRNP/Sm	Negative
Sm	Negative
SS-A	Negative
Ro- 52	Negative
SS-B/La	Negative
Scl - 70	Negative
PM-Scl	Negative
Jo - 1	Negative
CENP-B	Negative
PCNA	Negative
ds-DNA	<b>Positive</b>
Nucleosomes	<b>Borderline</b>
Histones	<b>Borderline</b>
Rib- P Protein	Negative
AMA- M2	Negative
<b>Table 1: Extractable Nuclear Antigens (ENA), Qualitative Profile</b>	

## CASE REPORT

We diagnosed as a case of rhupus syndrome with right sided pneumonitis without pleural effusion.

Tab Prednisolone (40 mg) once daily was started along with methotrexate, HCQS & iron supplement.

Fever subsided within 48 hrs and patient was discharged with medication and advised for regular follow up in OPD

After 3 week pt visited the OPD and was feeling better & X-ray chest postero-anterior view showed complete clearance of opacity, no relapse noted during tapering period, patient kept at prednisolone 10 mg daily.

**DISCUSSION:** The diagnostic criteria for rhupus was suggested by inflammatory symmetrical erosive polyarthritis, positive RA factor / anti ccp clinical features suggestive of SLE and positive anti-dsDNA or anti-smith auto-antibodies.<sup>10</sup>

AND Pneumonitis (?lupus pneumonitis, non-infective) after excluding all possible infective causes.

A retrospective study in China observed that the mean age of rhupus was 36.8 years; SLE developed about 7.7 years after initial presentation and SLE associated severe organ damages other than hematopoietic abnormalities were less frequent.<sup>11</sup>

Features of RA are dominated by erosive polyarthritis in all cases and rheumatoid nodules in around 40% of the cases.<sup>5</sup> SLE is usually manifested by cutaneous (butterfly skin rash, photosensitivity and alopecia), hematological (leukopenia and thrombocytopenia), serosal (pleural and pericardial effusion) and mucosal involvement.<sup>5</sup>

We diagnosed the case as rhupus by clinical manifestations of RA and SLE with positive ANA and ds-DNA.

### CONCLUSION:

- Polyarthritis is common problem in rhupus syndrome.
- Many patients with multiple joint pains are either undiagnosed or undertreated.
- Every patient should be properly investigated in case of either SLE or RA who are symptomatic even on standard treatment for the RA or SLE and having features of other diseases.

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## CASE REPORT

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