



A CASE OF ROWELL'S SYNDROME

SANJEEVI KRISHNAN P^{1*}, SHANMUGANANDAN KRISHNAN², AISWARYA
KOSARAJU³, KAVURU NAGA SIRI¹ AND LOHITHA MALLIPEDDI¹

1: Postgraduate student, Department of General Medicine, Sree Balaji Medical College and
Hospital, Chennai

2: Professor, Department of General Medicine, Sree Balaji Medical College and Hospital,
Chennai

3: Senior Resident, Department of General Medicine, Sree Balaji Medical College and Hospital,
Chennai

*Corresponding Author: Dr. P. Sanjeevi Krishnan: E Mail: sanjeevikrishnan96@gmail.com

Received 22nd Feb. 2021; Revised 20th April 2021; Accepted 6th May 2021; Available online 1st Nov. 2021

<https://doi.org/10.31032/IJBPAS/2021/10.11.5743>

INTRODUCTION

Rowell's syndrome is now identified as a subtype of subacute lupus erythematosus (LE) with erythema multiforme-like skin lesions, positive RF, anti-Ro La positivity and speckled pattern of antinuclear antibodies. Ours was a unique case of progression of Rowell's syndrome to ds-DNA negative LN.

Presentation

28-year-old female presented with fever for 1 month and rash over face and forearms for 3 weeks. Initially she developed high-grade

fever. Followed by she developed lesion over the ear. Three days later she developed blisters and red lesions over face and forearms and blackish lesions over forearms. She gave no history suggestive of Raynaud's phenomenon. No history of decreased urine output and hematuria, chest pain, dyspnea, cough or no history of spontaneous bleeding tendencies. She denied any history of drug intake prior to developing similar lesions. There was no significant past or family history.

Cutaneous examination revealed lesions over face, are palpable purpura, scars and targetoid lesions. Purpuric and targetoid lesions were also seen over the arms. Non-tender erythematous macules and targetoid lesions were seen over palms. Crusting erosions and edema were seen over lips. Hair over the frontal area was sparse and brittle. Systemic examination was normal.

Course and Management

CBC and urine examination were normal except for a raised ESR of 30 mm/hour. Anti-nuclear antibody was positive with a titer of 1:160 and showed a speckled pattern. Anti-ds-DNA was negative. Anti-Ro antibody was positive with a titer of 11 U/ml. Rheumatoid Factor (RF) was negative. Histopathological examination of the biopsy from targetoid lesion over the forearm revealed orthokeratosis, focal hypergranulosis, basal cell vacuolar degeneration, lympho-histiocytic infiltrate at the dermo epidermal junction with pigment incontinence diagnosis of Rowell's syndrome was made on the basis of clinical and laboratory findings.

The patient was duly started on T. Hydroxychloroquine 200 mg BD and T. Prednisolone at 30 mg which was gradually

tapered to 5 mg over a period of 6 months. The skin lesions subsided within a month with some post inflammatory hyperpigmentation.

Four months later, she developed recurrence of lesions on face, chest, back, arms and ears. Lichenoid papules were seen over the over face, arms, forearms and hands. Erythematous papules with dusky red hue were seen over the concha of ears and forearms. 24-hour urine protein examination revealed 472 grams of protein per 24 hours. ANA was positive with a titer of 1:80 and a homogeneous pattern. C3 levels were low with a titer of 83.4 mg/dl. She was advised a renal biopsy. Histopathology of the kidney revealed increased mesangial cellularity and increased cellular proliferation in few glomeruli with mild basement membrane thickening. There was no evidence of tubular casts, interstitial inflammation or blood vessel thickening. The patient was started on prednisolone 40 mg, tacrolimus 1.5 mg twice daily and ramipril 10 mg for one month. With the above treatment 24-hour urine protein decreased to 744 mg of protein per 24 hours. The skin lesions subsided completely over a period of 2 months.



Figure 1: Erythematous papules over the face



Figure 2: Erythematous papules on the forearm, hands and face

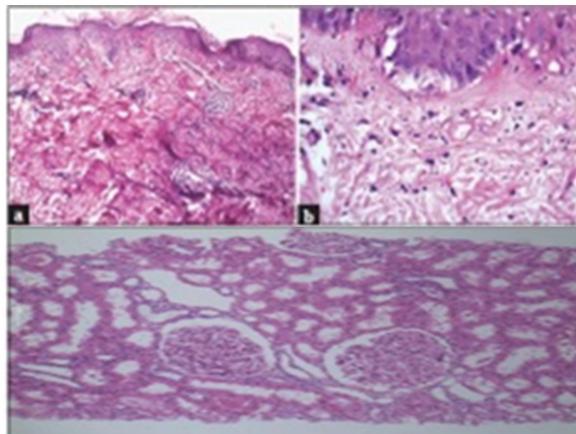


Figure 3: a, b and c

DISCUSSION

Rowell's syndrome has been described with all subtypes of LE (systemic, acute, subacute or discoid) [1]. Dysregulated apoptosis is the cause for manifestation of LE and EM skin lesions. Unidentified HSV, EBV or other viral infections may cross-react with lupus autoantigens and they can initiate the immunologic response, hence triggering the EM like lesions in SLE. Similarly, immune-pathogenetic mechanisms described in both diseases may be responsible for the co-occurrence [2, 3].

Rowell's syndrome -Major criteria

- LE: acute, subacute or discoid
- Erythema multiforme like lesions (with/without involvement of the mucous membranes),
- Speckled pattern of ANA.

Rowell's syndrome -Minor criteria

- Chilblains
- Anti-Ro antibody or anti-La antibody,
- Positive RF.

Our patient satisfied all three major and one minor of the above criteria and also satisfied four of the ACR criteria for SLE.

REFERENCES

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