

# Anifrolumab Use in Rowell Syndrome: A Case Presentation with Marked Improvement

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## INTRODUCTION

- **Rowell syndrome (RS)** is a rare and severe variant of **cutaneous lupus erythematosus (CLE)**
- The cutaneous eruption of RS resembles **erythema multiforme (EM)** with features of **systemic lupus erythematosus (SLE)**
- **Therapeutic options** include those used to treat SLE, such as **corticosteroids**, **hydroxychloroquine (HCQ)**, **immunosuppression**, and **biologics**
- **Anifrolumab** is a monoclonal antibody that selectively binds and inhibits the **type 1 interferon- $\alpha$  receptor 1 (IFNAR1)**
- In 2021, anifrolumab received FDA-approval for **SLE treatment**

### Objective:

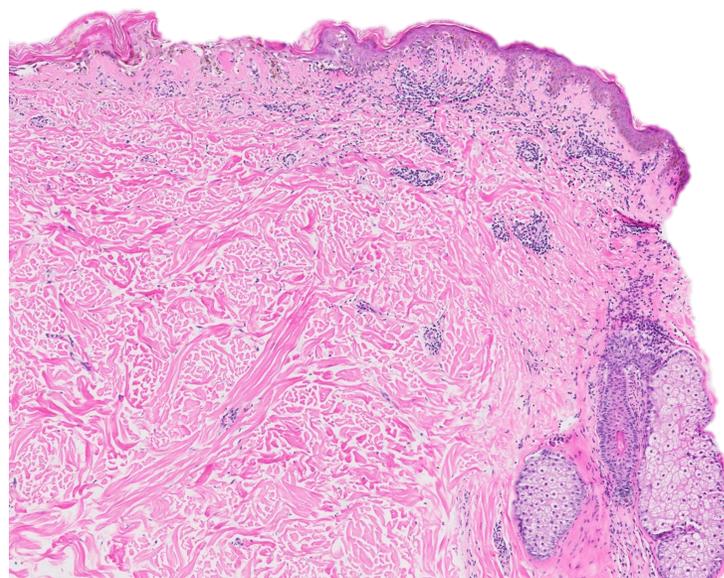
To highlight the use of anifrolumab for the treatment of RS, a rare variant of CLE

## CASE PRESENTATION

- **43-year-old** African-American female
- Presented with a **diffuse, blistering rash** on the dorsal arms, chest, and upper back
- Present for **3 months, following sun exposure**
- Complex medical history, including:
  - **SLE with secondary Sjogren's syndrome** (+ANA, +SSA, +SSB, +RF),
  - **Anti-phospholipid antibody syndrome**
  - **Uveitis**
  - **HIV on HAART**
  - **Breast cancer** on tamoxifen
- **SLE was well controlled** using **HCQ and azathioprine**
- As her SLE was in **remission**, these medications were **discontinued**
- Upon discontinuation, she developed the blistering rash above, following sun exposure
- **Clobetasol, triamcinolone, and oral prednisone** were trialed **without improvement**
- **On exam:** (figures 1 and 2)
  - Dusky, targetoid plaques with blisters and crusting (**consistent with EM**)
  - Erythematous discrete plaques with central clearing (**consistent with LE**)
  - Hyperpigmented patches with serpiginous borders (**consistent with PIPA**)



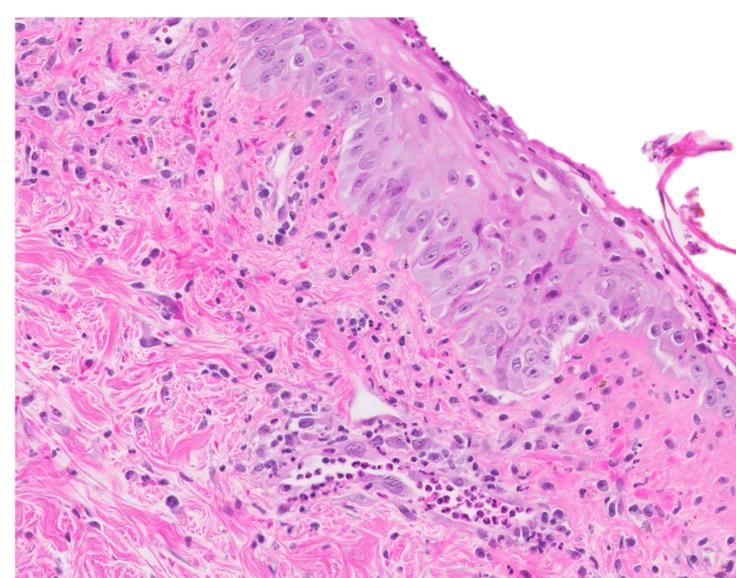
**Figure 1:** Hyperpigmented patches with serpiginous borders and erythematous plaques with central clearing are visible



**Figure 3:** Punch biopsy of left chest lesions demonstrates interface dermatitis with superficial and deep perivascular and periadnexal inflammation on H&E



**Figure 2:** Dusky, targetoid plaques with blisters and crusting on the right arm and chest during admission for treatment of RS following prednisone withdrawal



**Figure 4:** Repeat punch biopsy 5-months later reveals interface dermatitis with scattered necrotic keratinocytes on H&E

## EVALUATION & MGMT

- The microscopic differential included connective tissue disease, and drug eruption secondary to Lyrica, which was recently started
- Given her exam findings and serologic history, a **diagnosis of RS** was favored
- She was started on a **new course of prednisone** and **resumed HCQ**
- Despite changes to her regimen, including rituximab, mycophenolate, and IVIG, her CLE **continued to flare**
- **Steroids could not be tapered**
- She was **transitioned to anifrolumab**
- She **tolerates it well** and has had **no skin flares** while receiving monthly infusions for **six months**, despite **tapering her steroid** to discontinuation

## DISCUSSION

- RS is characterized by the presence of lesions of **EM** and **LE**
- RS is also associated with a **unique immunologic pattern:**
  - **+ANA** with speckled pattern
  - **+Anti-Ro/SSA** or **Anti-La/SSB**
  - **+Rheumatoid Factor (RF)**
  - **Negative DIF** on EM-like lesions
  - **Necrotic keratinocytes** on biopsy of EM-like lesions
- **Treatment of RS** is like that of **SLE or DLE** occurring independently:
  - Antimalarials
  - Systemic corticosteroids
  - Immunosuppressives (MMF, CsA, azathioprine)
  - Biologics (BAFF, anti-IL12/23, JAK inhibitors)
- **Anifrolumab decreases** levels of **type 1 interferon**, which is found in high levels in patients with SLE
- This patient had **severe RS** resistant to several therapies, with a **complex PMHx**
- Given the patient's **excellent response** to an emerging therapy used as **monotherapy**, we recommend that **anifrolumab** be considered in cases of **refractory RS**

## REFERENCES

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