

Drug-induced Rowell syndrome due to omeprazole

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Background

- Rowell syndrome is a rare entity consisting of overlapping features of erythema multiforme (EM) and cutaneous lupus erythematosus (CLE) with characteristic serological markers.
- Typically considered idiopathic, there have been rare case reports of drug-induced presentations, which may be more difficult to manage.¹

Case Report

- A 68-year-old female who had recently been diagnosed with an esophageal ulcer presented with a two-week history of a pruritic, primarily photodistributed eruption.
- One month prior to presentation, she had started a course of the proton-pump inhibitor (PPI), omeprazole.
- Physical exam demonstrated annular and targetoid, dusky pink to red papules and plaques with necrotic centers on the face, neck, lower arms and legs and superior trunk.
- Labs found +ANA 1:640 and both SS-A/Ro and SS-B/La were >8. RF was 47, and infectious studies including HSV/Mycoplasma were negative.
- Biopsy demonstrated an acute interface dermatitis with epidermal necrosis. There was adnexal involvement and increased dermal mucin. Direct immunofluorescence (DIF) was negative.
- Based on these findings, the patient met criteria to be diagnosed with Rowell syndrome,² with EM and subacute CLE overlap.



Figure 1. Initial presentation of Rowell syndrome (A) The cheeks, chin, anterior neck and upper chest of our patient demonstrated dusky, annular and targetoid necrotic pink and red papules and plaques with central pseudobullae. **(B)** The lateral neck demonstrated pink edematous targetoid papules, many with a central necrotic zone.

References

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Treatment

- Omeprazole was discontinued and substituted with famotidine.
- Oral prednisone at a dose of 1 mg/kg/day as well as topical corticosteroids led to improvement.
- Prednisone was tapered and initially transitioned to cyclosporine. This was tolerated poorly and her rash persisted, so hydroxychloroquine was started instead. Her eruption resolved.
- Gastroenterology felt strongly that the patient needed a PPI for her esophageal ulcer, so she was cautiously trialed on pantoprazole, which she has tolerated without recurrence of her eruption.

Conclusions

- Rowell syndrome can be diagnostically challenging, both because of its rarity and its inherently overlapping features with other conditions. However, presence of both EM- and CLE-like lesions with a positive ANA, SS-A/SS-B, reactive RF and negative DIF point to the diagnosis.
- Some proposed diagnostic criteria for Rowell syndrome have included lack of a pharmacologic trigger as a criterion.³ Yet our case and others demonstrate that, like CLE, Rowell syndrome likely can be induced by a culprit medication.
- Our patient has been able to tolerate the alternative PPI pantoprazole, suggesting that drugs in the same class as the culprit medication may be cautiously retried when absolutely necessary.