

## BRIEF ARTICLES

## A Rare Presentation of Toxic Epidermal Necrolysis-like Acute Systemic Lupus Erythematosus

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### ABSTRACT

A 25-year-old woman with a history of systemic lupus erythematosus (SLE) was transferred from an outside hospital with a worsening painful generalized rash and oral ulcerations for the prior three weeks due to concern for Stevens-Johnson Syndrome / toxic epidermal necrolysis. Exam revealed denuded erythematous plaques covering over 80% of the patient's body surface area and a 4.2 x 2.6 cm ulcerated plaque of the superior hard palate. Histology demonstrated parakeratosis and compact hyperkeratosis overlying an atrophic epidermis with vacuolar interface change, prominent keratinocyte dyskeratosis, and a thickened basement membrane zone (BMZ). The superficial dermis had a mild predominantly lymphocytic perivascular infiltrate and superficial dermal mucin deposition. Direct immunofluorescence was positive for IgG (granular, BMZ), C3 (granular, BMZ), and IgA was negative. Labs were remarkable for positive ANA (1:160), ds-DNA, anti-Smith, anti-RNP, low C3/C4, with negative anti-SSA/SSB. The clinicopathological correlation was most consistent with the diagnosis of TEN-like acute systemic lupus erythematosus. Our patient improved with treatment for her acute systemic lupus erythematosus. This case highlights a challenging clinicopathologic differential diagnosis.

### INTRODUCTION

Acute systemic lupus erythematosus presenting as toxic epidermal necrolysis (TEN-like ASLE) is rare with less than 25 reported cases.<sup>1</sup> Differentiating ASLE and TEN can be difficult, as both share clinical and histological findings such as diffuse desquamation, mucosal erosions, and keratinocyte necrosis. Prodromal systemic

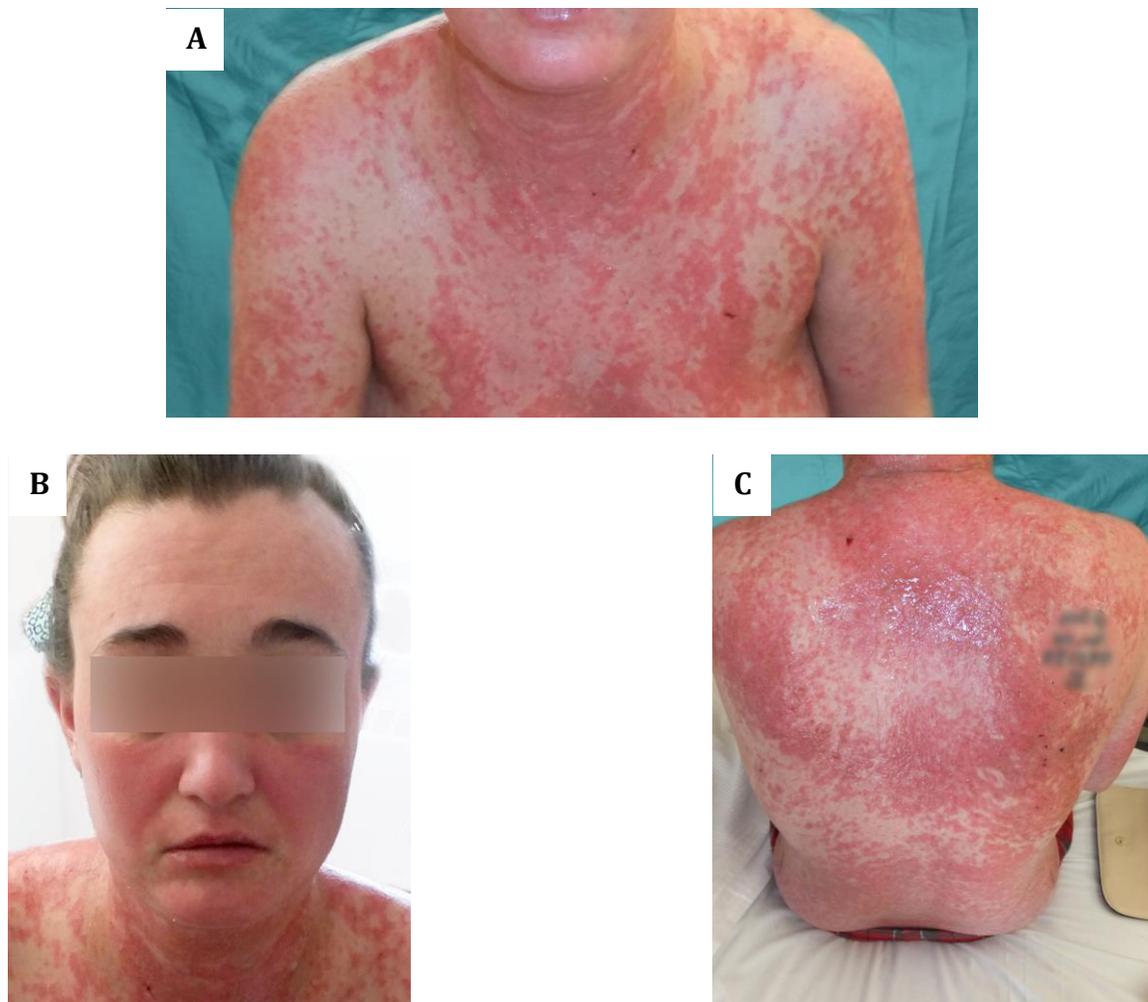
symptoms (fatigue, myalgias, and polyarthritis), positive ANA and ENA, photodistributed eruption, and a prolonged disease course characterize ASLE, while an identifiable trigger and more acute onset (less than 9 days) favor TEN. Histopathologically, vacuolar interface change, limited keratinocyte apoptosis,

dense lymphocytic infiltrate, and dermal mucin suggest ALSE, whereas a basket-weave stratum corneum, sparse lymphocytic infiltrate, and full thickness epidermal necrosis are consistent with TEN.<sup>1-5</sup>

## CASE REPORT

A 25-year-old woman with a history significant for systemic lupus erythematosus (SLE) was transferred from an outside hospital with a worsening painful

generalized rash and oral ulcerations for the prior 3 weeks due to concern for Stevens-Johnson Syndrome / toxic epidermal necrolysis. Prior to admission, she noted a diffuse and painful rash, large oral ulcerations, fatigue, and muscle/joint pain. Initial evaluation revealed an ill-appearing female of Fitzpatrick skin type I. Involving approximately 80 % body surface area (BSA), including the face, torso, arms, and legs, were diffuse confluent eroded erythematous reticulated plaques with palmar and plantar sparing (Figure 1).



**Figure 1.** Generalized eroded, erythematous plaques of the chest (A), trunk (B), and back (C) coalescing into larger plaques involving approximately 80% body surface area.

Examination of the mouth revealed a 4.2 x 2.6 cm ulcerated plaque on the superior hard palate (Figure 2). A lesional punch biopsy was performed showing areas of parakeratosis and compact hyperkeratosis overlying an atrophic epidermis with vacuolar interface change and prominent keratinocyte dyskeratosis and apoptosis. The basement membrane was thickened. Within the superficial dermis, there was a mild, predominantly lymphocytic perivascular infiltrate. There was also superficial dermal mucin deposition. These findings were most consistent with an interface dermatitis (Figure 3). Direct immunofluorescence was positive for IgG (granular, BMZ) and C3 (granular, BMZ),

and IgA negative. Labs were remarkable for positive ANA (1:160), ds-DNA, Anti-Smith, Anti-RNP, low C3/C4, with negative Anti-SSA/SSB. The clinicopathological correlation was most consistent with the diagnosis of TEN-like ASLE. Our patient improved with a loading dose of hydroxychloroquine, 200mg three times daily for three months following by 5mg/kg/day thereafter, methylprednisone 1g IV every 24 hours for the first three days and then she was transitioned to oral prednisone 60mg daily which was tapered over approximately 6 months, mycophenolate mofetil 1g twice daily, and dapsone 100mg daily.



Figure 2. Clinical image demonstrating a 4.2x2.6 cm ulcerated plaque on the superior hard palate.

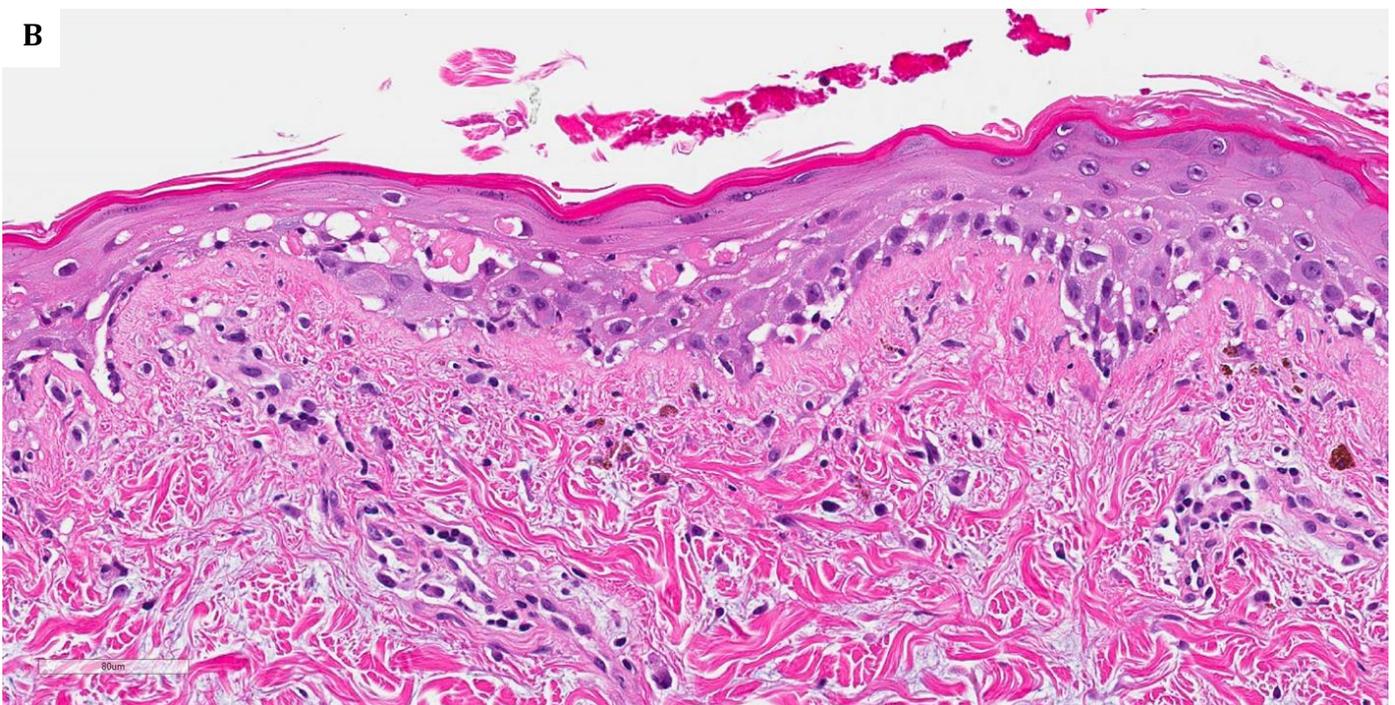
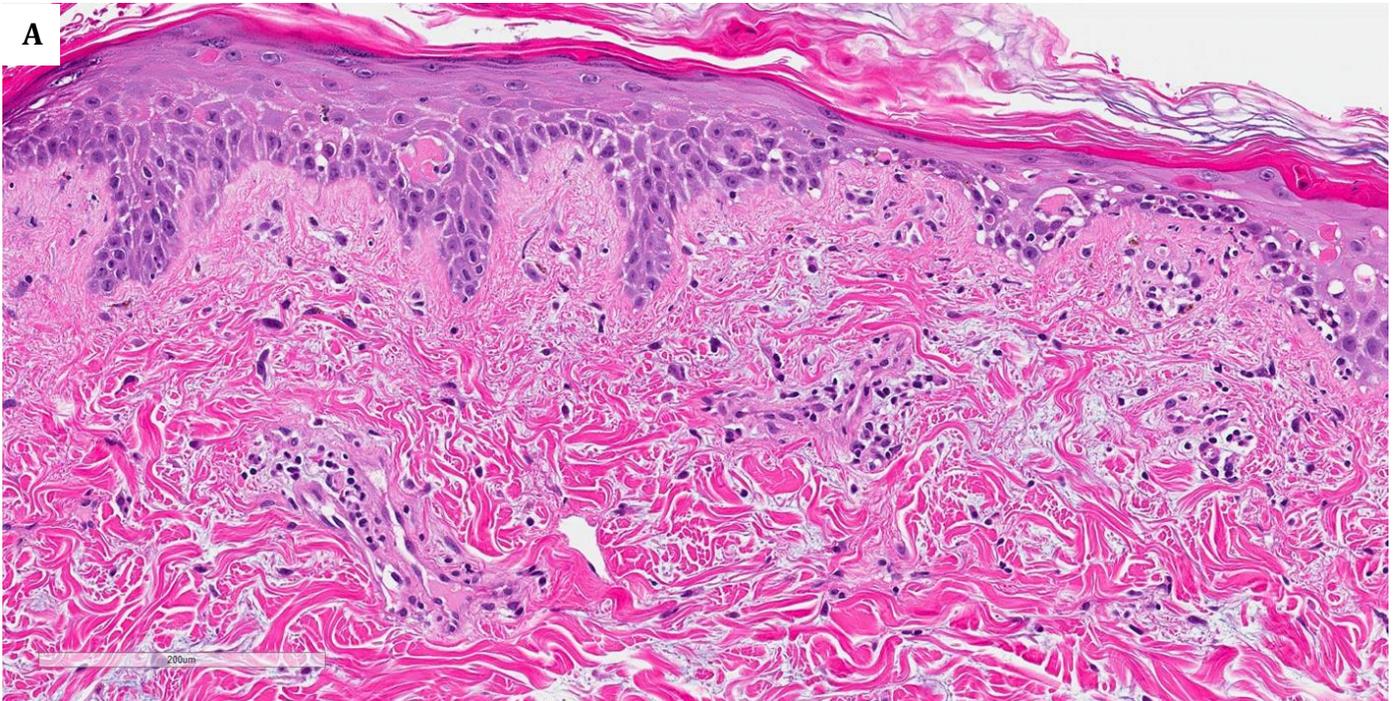


Figure 3. Histologic sections demonstrating parakeratosis and compact hyperkeratosis overlying an atrophic epidermis with vacuolar interface change (A), prominent keratinocyte dyskeratosis and apoptosis (B). Within the superficial dermis, there is a mild, predominantly lymphocytic perivascular infiltrate and mucin deposition.

## DISCUSSION

TEN-like ASLE is rare. TEN is characterized by erythema with severe necrosis and desquamation of the epidermis and epithelia of mucous membranes with greater than 30% BSA involvement. The onset is typically acute, with maximal extension of lesions within hours to days.<sup>2</sup> Both ASLE and TEN are inflammatory and interface dermatoses, and share clinical and pathologic findings such as generalized desquamation, mucosal erosions, and keratinocyte necrosis.<sup>1</sup> Prodromal systemic symptoms, positive ANA and ENA, photodistributed eruption, and a prolonged disease course characterize ASLE<sup>3-5</sup> whereas an identifiable trigger and more acute onset (less than 9 days) favor TEN.<sup>1</sup> Histologically, vacuolar interface change, limited keratinocyte apoptosis, dense lymphocytic infiltrate, and dermal mucin suggest ALSE, while a basket-weave stratum corneum, sparse lymphocytic infiltrate, and full thickness epidermal necrosis are more consistent with TEN.<sup>1</sup>

## CONCLUSION

There is both clinical and histological overlap of ASLE and TEN. Differentiating these two entities has practical and important implications for patients, treatment, and counseling. We present a rare case of TEN-like ASLE that required treatment with systemic immunomodulatory and immunosuppressant medications. This case highlights a challenging clinicopathologic differential diagnosis important for dermatologists and dermatopathologists to be aware of.

**Conflict of Interest Disclosures:** None.

**Funding:** None.

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