Vesicular Eruption in a Woman with Systemic Lupus Erythematosus

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Introduction

- Bullous systemic lupus erythematosus (BSLE) is a rare autoimmune blistering disorder that presents as an acute onset of tense vesicles and bullae over normal or erythematous skin.[2] The vesiculobullous eruption may be associated with photodistribution or photoexacerbation and tends to occur on trunk and extensor surfaces of the upper extremity. [2,3] Pruritus is either mild or absent. Dyspigmentation may result after lesion resolution, however scarring or the formation of milia do not usually occur. [1,2]
- The vesiculobullous eruption appears in around 5% of patients with a known diagnosis of systemic lupus erythematosus (SLE).[1,2] The incidence of BSLE is greater in middle aged, African-American females.[1] In some cases, BSLE is the initial clinical presentation leading to a SLE diagnosis.[1]
- The development of BSLE may be associated with active extra-cutaneous manifestations of SLE, so a prompt diagnosis of BSLE is crucial to prevent any further complications of SLE.[1,2]
- We present a case of a vesicular eruption in a woman with a previous diagnosis of SLE.

Case Presentation

- A 41-year-old female presented to the dermatology clinic for evaluation of an itchy blistering rash located on her face and neck. She did not experience any fevers, chills or arthralgias.
- Physical exam revealed scattered tense vesicles along her left lateral eyebrow, upper sternum and left inferior central malar cheek (Figure 1). Hypopigmented macules were appreciated along the left cheek and chin (Figure 1).
- Medical history was positive for SLE managed with hydroxychloroquine.
- Punch biopsies from the left lateral eyebrow and upper sternum were obtained for histopathology and direct immunofluorescence (Figures 2A and 2B).
- Several months ago, she was seen by another specialist for a similar rash of scattered non-grouped vesicles and bullae along her right cheek and submandibular area, which responded well to oral dapsone therapy.

Discussion

- This patient had a previous diagnosis of SLE and repeated vesiculobullous eruptions along her face and upper trunk which resolved with subsequent hypopigmentation. The rash responded well to dapsone therapy.
- The vesiculobullous lesions of BSLE result from a disruption in the adhesion of the dermal-epidermal junction. The disruption is secondary to antibody formation against the noncollagenous type 1 (NC1) and type 2 (NC2) domains of type VII collagen in the anchoring fibrils of the epidermal basement membrane zone. [2,4] A characteristic neutrophilic infiltrate in the superficial dermis enhances the pathogenesis at the dermal epidermal junction via neutrophil-mediated proteolysis.[4]
- Hematoxylin-eosin staining of the lesion revealed the classic subepidermal blistering with a neutrophil predominant infiltrate and dermal edema.
- The direct immunofluorescence of the lesion was positive for deposition of IgG10 and IgA (Figures 2A and 2B) in addition to IgG25, IgM, C3 and C5 along the basement membrane.
- The differential diagnoses in this case were ruled out based on clinical and biopsy findings, allowing for SBLE to be diagnosed and the initiation of oral dapsone therapy.

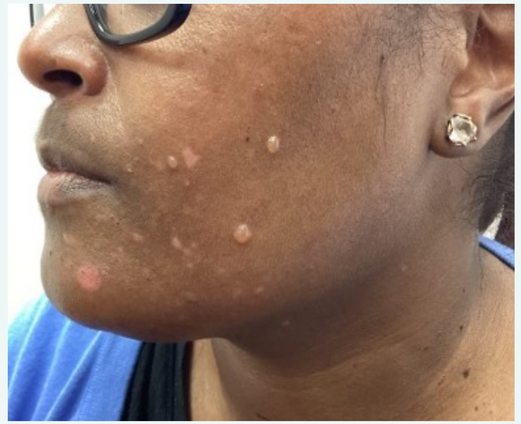

Figure 1: Clinical image shows tense vesicles and hypopigmented macules along the left inferior central malar cheek. She has Fitzpatrick type IV skin.

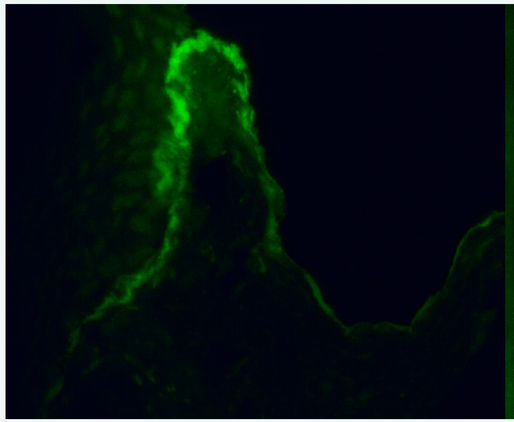

Figure 2A: Direct immunofluorescence of biopsy from left lateral eyebrow demonstrating deposition of IgG10 along the basement membrane

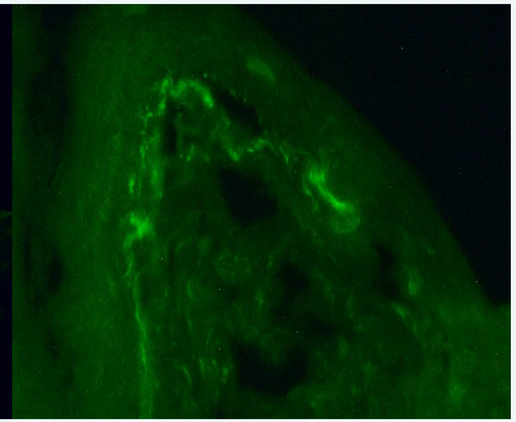

Figure 2B: Direct immunofluorescence of biopsy from left lateral eyebrow demonstrating deposition of IgA along the basement membrane

Conclusion

- In a patient with a history of systemic lupus erythematosus presenting with a vesicular rash, consider the diagnosis of Bullous Systemic Lupus Erythematosus

References

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