Received: 5 August 2021 | Revised: 3 September 2021 | Accepted: 9 September 2021

DOI: 10.1002/cia2.12207

Journal of Cutaneous Immunology and Allergy

WILEY

CORRESPONDENCE

Stevens-Johnson syndrome/toxic epidermal necrolysis-like acute cutaneous lupus erythematosus in a patient with systemic lupus erythematosus

Dear Editor,

A 33-year-old man with a 3-year history of treatment with hydroxychloroquine for mild cutaneous lupus erythematosus (CLE) progressed painful erythema 1 week prior to his visit to us. Two weeks before the onset, he was diagnosed with acute sinusitis and took carbocisteine and clarithromycin. He developed erythema with vesicles dominantly across a photodistributed area, crusts on the lips and nostrils, and erosions on the oral mucosa (Figure 1A-C). Ophthalmologic examination revealed only mild blepharitis and conjunctivitis.

Initially, we considered a diagnosis of Stevens-Johnson syndrome (SJS); however, chilblain-like lesions on every finger and toe appeared subsequently. A skin biopsy from the upper arm revealed severe interface dermatitis, perifollicular lymphocyte infiltration, and necrotic keratinocytes (Figure 1D,E). Laboratory investigations revealed leukopenia (white blood cells, 2560/μl); decreased levels of C3 (36.4 mg/dl) and C4 (6.0 mg/dl) complement; positive antinuclear antibody at a titer of 1:160; and positive anti-ds DNA, anti-RNP, anti-Sm, and anti-La/SS-B antibody. Kidney biopsy performed due to abnormal urinalysis led to a diagnosis of lupus nephritis class 2. According to these data, he was diagnosed with an acute exacerbation of systemic lupus erythematosus (SLE) probably exacerbated by the infection. Drug-induced lymphocyte stimulation tests for hydroxychloroquine, carbocisteine, and clarithromycin were negative. Steroid pulse therapy was administered, and subsequently, prednisolone was gradually tapered with the reintroduction of hydroxychloroquine. The skin and mucous rash have improved and have not flared up.

Cutaneous lupus erythematosus is morphologically classified as lupus-specific or nonspecific based on histological findings of

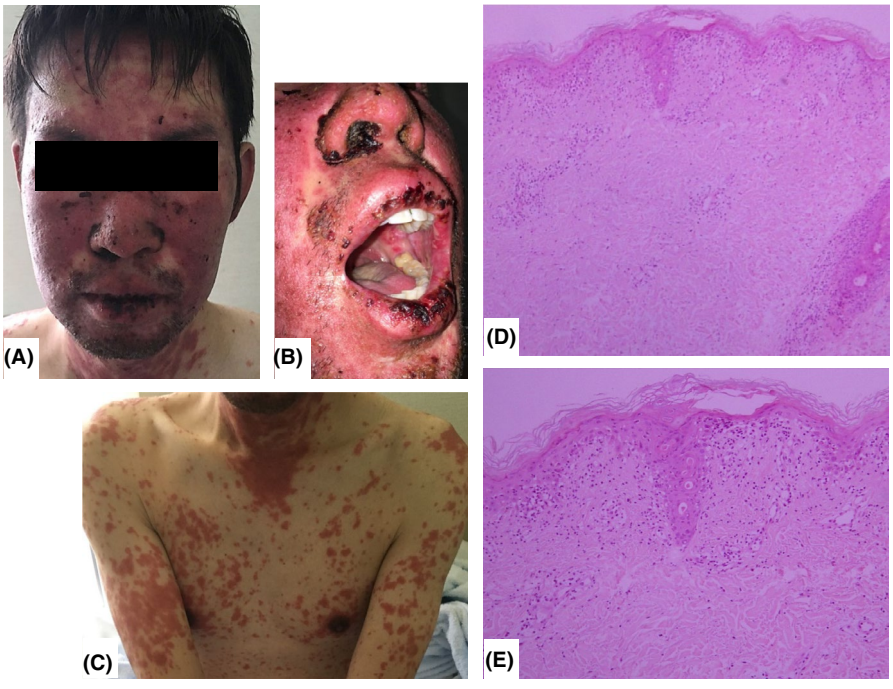

FIGURE 1 Clinical findings on initial examination. (A) Erythematous plaque with vesicles on the whole face and V-neck area. (B) Crusting on the lip and nostrils and erosions on the oral mucosa. (C) Exudative erythematous areas of varying size on the upper arm and trunk. Histopathological findings of skin biopsy of the upper arm. (D) Severe vacuolar degeneration, perifollicular lymphocytes infiltration (hematoxylin and eosin stain [HE]; magnification, ×40). (E) Numerous necrotic keratinocytes (HE; magnification, ×200)

This is an open access article under the terms of the Creative Commons Attribution-NonCommercial-NoDerivs License, which permits use and distribution in any medium, provided the original work is properly cited, the use is non-commercial and no modifications or adaptations are made.
© 2021 The Authors. *Journal of Cutaneous Immunology and Allergy* published by John Wiley & Sons Australia, Ltd on behalf of The Japanese Society for Cutaneous Immunology and Allergy.

interface dermatitis.[1] SJS/toxic epidermal necrolysis (TEN)-like acute CLE (ACLE), a hyperacute and severe form of CLE, is included in lupus erythematosus (LE)-specific vesiculobullous diseases. LE nonspecific vesiculobullous diseases include bullous SLE and other vesiculobullous skin disorders, such as autoimmune bullous disease,[2] SJS, and TEN.[3]

Rutnin and Chanprapaph[4] described that SJS/TEN-like LE occurs in patients with a history of recent onset or exacerbation of LE and usually starts in photodistributed areas, and mucosal involvement is absent or minimal. These characteristics were also observed in our patient. Histopathological findings of skin biopsy and subsequent clinical features, such as chilblain-like lesions and lupus nephritis, also supported our diagnosis.

Romero et al.[5] reviewed TEN-like ACLE and mentioned that the diagnosis of TEN-like ACLE is often made retrospectively, after correlating the clinical course, serologic profile, and histopathology. When the patient has a history of prior culprit drug exposure or infectious disease, it is difficult to distinguish between SJS/TEN-like ACLE and SJS/TEN. Although an increased incidence of SJS/TEN in patients with SLE has been reported, Hsu et al.[6] found that individuals with SLE had an odds ratio of 5.34 for SJS and an odds ratio close to one for TEN. These reports suggest that patients with TEN-like ACLE are correctly identified, but SJS-like ACLE is still prone to be misclassified as SJS. We should take into consideration that ACLE may present with SJS-like findings when vesicular lesions are seen in SLE patients.

DECLARATION SECTION

Approval of the research protocol: This study has been approved by the research ethics committee of Sakai City Medical Center.
Informed Consent: Verbal informed consent was obtained from the patient for publication of this case report and accompanying images.
Registry and the Registration No. of the study/trial: N/A.
Animal Studies: N/A.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

Aya Tanaka MD, PhD[1]
Shota Bun MD[1,2]

[1]*Department of Dermatology, Sakai City Medical Center, Sakai-shi, Japan*
[2]*Department of Dermatology, National Hospital Organization Osaka National Hospital, Osaka-shi, Japan*

Correspondence
Aya Tanaka, MD, PhD, Department of Dermatology, Sakai City Medical Center, 1-1-1, Ebaraji-cho, Nishi-ku, Sakai City 593-8304, Japan.
Email: tanaka-a@sakai-hospital.jp

ORCID

Aya Tanaka https://orcid.org/0000-0003-0737-3363

REFERENCES

1. Gilliam JN, Sontheimer RD. Distinctive cutaneous subsets in the spectrum of lupus erythematosus. J Am Acad Dermatol. 1981;4(4):471–5.
2. Sontheimer RD. The lexicon of cutaneous lupus erythematosus – a review and personal perspective on the nomenclature and classification of the cutaneous manifestations of lupus erythematosus. Lupus. 1997;6(2):84–95.
3. Torchia D, Romanelli P, Kerdel FA. Erythema multiforme and Stevens-Johnson syndrome/toxic epidermal necrolysis associated with lupus erythematosus. J Am Acad Dermatol. 2012;67(3):417–21.
4. Rutnin S, Chanprapaph K. Vesiculobullous diseases in relation to lupus erythematosus. Clin Cosmet Investig Dermatol. 2019;12:653–67.
5. Romero LS, Bari O, Forbess Smith CJ, Schneider JA, Cohen PR. Toxic epidermal necrolysis-like acute cutaneous lupus erythematosus: report of a case and review of the literature. Dermatol Online J. 2018;24(5):13030/qt5r79d67k.
6. Hsu DY, Brieva J, Silverberg NB, Silverberg JI. Morbidity and mortality of Stevens-Johnson syndrome and toxic epidermal necrolysis in United States adults. J Invest Dermatol. 2016;136(7):1387–97.

How to cite this article: Tanaka A, Bun S. Stevens-Johnson syndrome/toxic epidermal necrolysis-like acute cutaneous lupus erythematosus in a patient with systemic lupus erythematosus. J Cutan Immunol Allergy. 2022;5:32–33. https://doi.org/10.1002/cia2.12207