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A case of toxic epidermal necrosis–like cutaneous eruption as the first manifestation and clue to the diagnosis of systemic lupus erythematosus: A case report

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Abstract

We present a rare case of a 61-year-old woman presenting with a widespread erosive eruption on her torso and extremities. Although the lesions were histologically compatible with toxic epidermal necrolysis, clinically the patient was hemodynamically stable, had no mucosal involvement and had no relevant medical history or potentially incriminating medications. Further investigations uncovered a new diagnosis of systemic lupus erythematosus, with this toxic epidermal necrolysis–like eruption being the first presentation of the disease. This case highlights the importance of broadening the differential diagnosis in patients presenting with acute widespread cleavage of the epidermis, using the spectrum of acute syndrome of apoptotic pan-epidermolysis as a reference.

Keywords

Dermatology, systemic lupus erythematosus, toxic epidermal necrolysis, acute syndrome of apoptotic pan-epidermolysis

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Introduction

Toxic epidermal necrolysis (TEN) is a potentially fatal adverse drug reaction characterized by rapidly progressive painful mucocutaneous erosions, widespread flaccid bullae and hemodynamic instability.[1] The majority of cases appear to be drug-related; however, a similar clinicopathological presentation can be seen in the absence of drug hypersensitivity.[2] Although rare, vesiculobullous eruptions in the context of systemic lupus erythematosus (SLE) can present as areas of sheet-like epidermal detachment resembling TEN.[1] In this report, we describe a TEN-like cutaneous eruption as the first manifestation and clue to the diagnosis of SLE.

Case report

A 61-year-old woman presented to the emergency room with a 3-week history of a progressive painful skin eruption that began on her torso and spread to her extremities. She reported anorexia, fatigue and one episode of fever the day prior. She denied any night sweats, chills or recent weight loss. Her previous medical history included hypertension, unspecified arthritic pain and a history of superficial phlebitis in the preceding year. She denied any history of recent infections, Raynaud's phenomenon, oral ulcers, photosensitivity or symptoms of serositis. She had been taking amiloride for hypertension for the past 10 years and denied any new medications.

On examination, the patient was afebrile and appeared non-toxic, with normal vital signs. Skin examination revealed widespread erythematous papules coalescing into dusky annular plaques with central erosions, distributed on the trunk and extremities (Figure 1). Her face, palms and soles were spared. There was no evidence of mucosal or joint involvement, nor any palpable lymphadenopathy.

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Figure 1. Initial presentation on arrival to the emergency room.

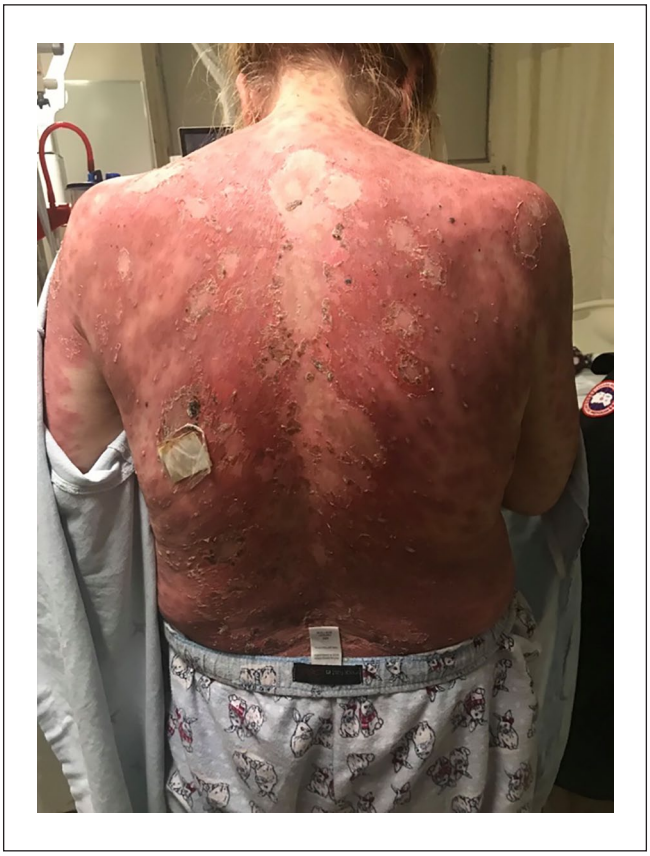

Figure 2. Peripheral sheet-like detachment and desquamation on follow-up the next day.

The next day, the patient remained afebrile and hemodynamically stable, without any mucosal involvement. However, peripheral sheet-like detachment and desquamation involving 30% of her body surface area were observed, with positive Nikolsky sign (Figure 2).

The differential diagnosis at this time included classic TEN, TEN-like LE, TEN-like presentation of linear IgA bullous dermatosis (LABD), pseudoporphyria, generalized fixed drug eruption (GFDE) and drug reaction with eosinophilia and systemic symptoms (DRESS). A skin biopsy revealed prominent epidermal necrosis with minimal superficial perivascular lymphocytic infiltrates without eosinophils (Figure 3). Immunofluorescence was unremarkable. Pseudoporphyria was deemed unlikely given the absence of an offending medication and characteristic histopathological findings.[3] GFDE likewise became less likely due to the lack of dermal infiltration of eosinophils and melanophages and no pertinent medication history.[4] LABD was excluded due to the absence of characteristic findings on immunofluorescence.[5] Given the absence of eosinophils and dermal edema on histopathology and a low score on the RegiSCAR criteria, DRESS was excluded.[6] Although the histopathology was most consistent with a diagnosis of TEN, the clinical picture did not correlate with classic TEN, given lack of mucous membrane involvement and the general well-being and hemodynamic stability of the patient.

Laboratory investigation was significant for normocytic anaemia, neutropenia, lymphopenia and proteinuria. Autoimmune workup revealed a positive anti-nuclear antibody (ANA) at 1:160 (nucleolar fluorescence pattern), positive dsDNA and positive anti-RNP. All other autoantibodies were negative, including anti-Ro, anti-La, anticardiolipin and lupus anticoagulant. Complements were normal and parvovirus B19, hepatitis B and C serologies were negative.

Our patient met the Systemic Lupus Erythematosus International Collaborating Clinics (SLICC) classification criteria for SLE.[7] Based on the clinical, histological and autoimmune laboratory findings, the diagnosis of SLE with TEN-like cutaneous presentation was made. Although a drug reaction was not suspected, amiloride was discontinued as blood pressure was normalized and controlled. The patient was admitted for close observation and received prednisone 40 mg orally daily. Wound dressings were applied over denuded areas and a medium-potency corticosteroid cream was applied to erythematous areas on the body. In addition, hydroxychloroquine was started at 400 mg orally daily by rheumatology. Cutaneous symptoms improved remarkably within a week and on 1-month follow-up, the eroded skin had completely healed, with remaining post- inflammatory hyperpigmentation (Figure 4). Cytopenia and arthritic pain had also resolved at this time. On 6-month follow-up, prednisone was stopped, and the patient remained only on hydroxychloroquine, with no recurrence of cutaneous symptoms and no active systemic involvement of SLE.

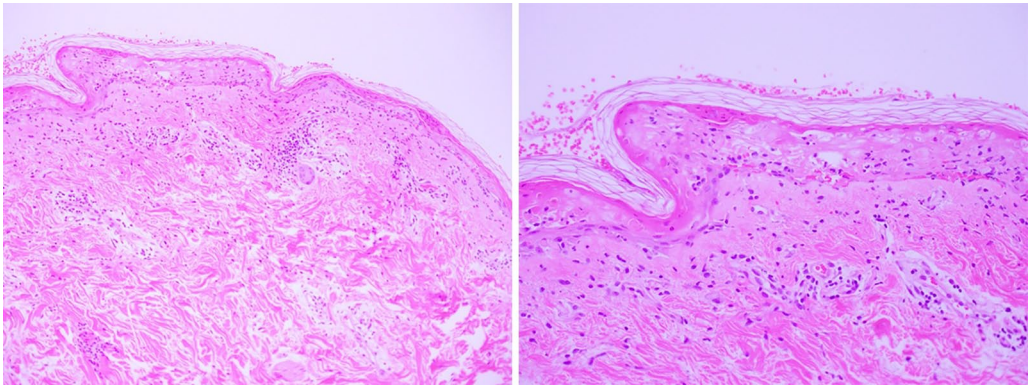

Figure 3. Skin biopsy with prominent epidermal vacuolar degeneration and extensive epidermal necrosis with minimal superficial perivascular dermal lymphocytic infiltrates without eosinophils.

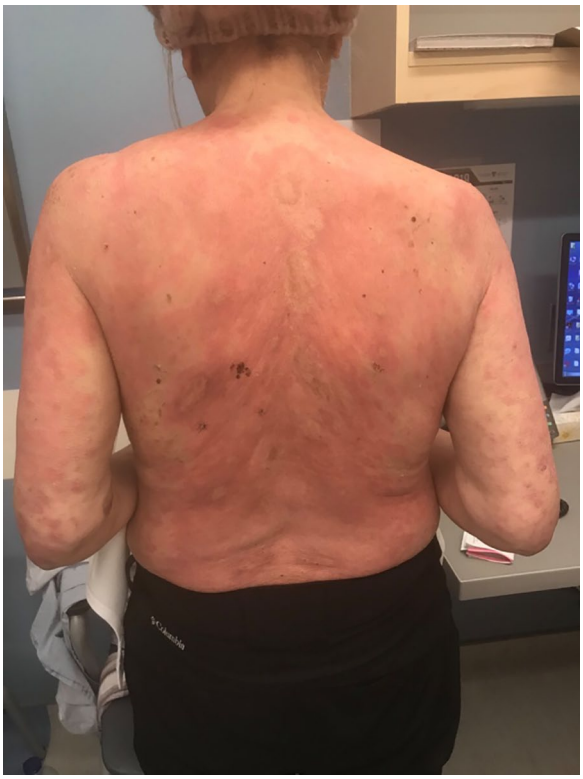

Figure 4. Remaining post-inflammatory hyperpigmentation on 1-month follow-up.

Discussion

The term ASAP (acute syndrome of apoptotic pan-epidermolysis) has been proposed by Ting et al.[2] to describe a spectrum of conditions with TEN-like presentation, in which massive life-threatening cleavage of the epidermis results from a hyperacute apoptotic injury. This presentation may be drug-induced, as in classic TEN, or due to other causes like lupus erythematosus (LE), acute graft-versus-host disease and pseudoporphyria. Differentiation between these causes is important as treatment of the cutaneous manifestations can vary depending on the underlying cause of the eruption.[2]

There are less than 50 reported cases of TEN-like presentation in the context of LE, with 60% of them occurring in patients with a previously established diagnosis of SLE. Only 15 cases report this cutaneous eruption as the first manifestation of SLE.[8]

Rutnin and Chanprapaph[9] classify vesiculobullous diseases in LE patients into four categories: LE-specific and aspecific vesiculobullous diseases, LE-related autoimmune bullous diseases, and LE in association with non-autoimmune conditions. Our patient's clinical presentation, as well as specific histologic and serologic findings, meets Rutnin et al.'s diagnostic features for LE-specific vesiculobullous diseases, specifically TEN-like LE.

Clinically, distinguishing between TEN-like LE and classic TEN can be difficult. This difficulty is compounded in the case presented, as the TEN-like presentation preceded the diagnosis of SLE. Although the widespread sheet-like desquamation and histological changes of apoptosis of the epidermis were compatible with a diagnosis of TEN, the diagnosis of TEN-like LE was favoured due to the absence of high-risk drug ingestion, lack of mucosal involvement, hemodynamic stability of the patient and discovery of positive autoimmune serology.[10]

While classic TEN is associated with a high morbidity and mortality, TEN-like LE has been associated with a low mortality in the treated cases reported.[1,8,11,12] Unfortunately, there is no consensus regarding treatment as there are so few cases reported.[11,13,14] Systemic corticosteroids have been shown to be effective but also associated with resistance in several patients.[13] In a review of 43 cases of TEN-like acute cutaneous lupus erythematosus (ACLE), Romero et al.[8] describe combination of systemic corticosteroids with

hydroxychloroquine (8 cases), intravenous immunoglobulin (10 cases) and mycophenolate mofetil (4 cases), resulting in clinical resolution. Plasmapheresis was described as a successful treatment in three cases.[1] We report successful treatment with systemic corticosteroids and hydroxychloroquine, resulting in complete resolution of skin lesions in our patient after 1 month.

In conclusion, this is one of few reported cases of TEN-like skin eruption occurring as the first manifestation of SLE.[1,8,9,11–15] TEN-like dermatoses should be included in the differential of acute sheet-like epidermal detachment, especially in the absence of mucosal involvement, high-risk drug ingestion and hemodynamic instability. Positive autoimmune serology may further support the diagnosis of TEN-like LE.

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Informed consent

Written informed patient consent was obtained for publication of images and case report content.

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