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Case Report

Bullous lupus erythematosus: A rare variant of acute cutaneous lupus erythematosus

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Abstract

Acute cutaneous lupus erythematosus (ACLE) is a variant of acute autoimmune disease with distinctive cutaneous manifestations. In addition to its typical manifestations, it can also manifest as bullous lesion, which can be difficult to distinguish clinically from other dermatological disorders. We reported a 40-year-old lady who presented with painful vesicobullous eruptions on malar region since 1 week ago. Patient refused to be examined further and not compliance to the treatment given. She then consumed traditional medicine and admitted to the hospital 2 weeks later with extensive vesicobullous lesions in the face, scalp, trunk and extremities. Patient showed complete remission and excellent clinical response after corticosteroid treatment. This report showed the importance of early diagnosis as well as prompt treatment can result in a better prognosis.

Key words
Bullous lupus erythematosus; Rare variant; Cutaneous lupus.

Introduction

Acute cutaneous lupus erythematosus (ACLE) is an acute LE variant with distinctive skin manifestations. In the literature, ACLE is reported to be 8 times more common in women. Typical cutaneous manifestation of localized ACLE is malar rash/butterfly erythema, which can be found in 20% to 60% of patients with LE. Generalized ACLE lesions can be found in the form of morbiliform, maculopapular, erythema, erosion and ulceration lesions.[1]

In addition to typical ACLE rash, there are other less common variants, one of which is the bullous type LE. Bullous lesions in cutaneous lupus erythematosus patients are extremely rare.[2] In a study by Chanprapaph *et al*., reported that out of 5149 patients diagnosed with cutaneous LE and systemic lupus erythematosus (SLE), only 10 patients (0.19%) were diagnosed with bullous type LE. In the literature, vesicobullous lesions in LE patients are generalized in nearly 80% of patients with predilection sites on the extremities (80%), face (70%) and trunk (70%).[3]

Early lesion of bullous LE can be difficult to distinguish from other dermatological disorders such as herpes simplex infection or bullous impetigo. This allows doctors who are less familiar with the lesion to misdiagnose this type of LE.[2]

Case Report

A 40-year-old woman came to the Dermatology and Venereology polyclinic of USU Hospital Medan with chief complaint of sudden onset of painful fluid filled blisters on the cheek and nose area since one week ago. Initially, it presented with painful reddish patches which then

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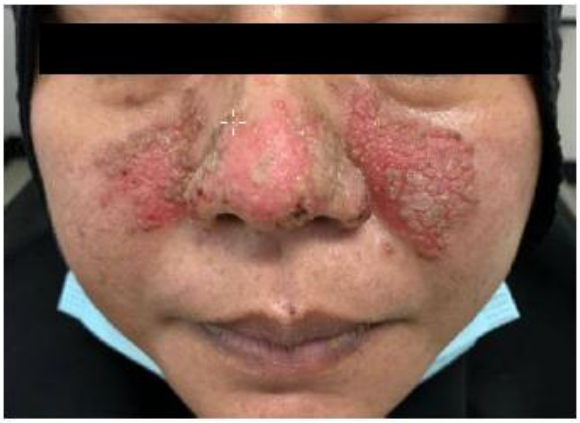

Figure 1 Patient's clinical manifestations during her first visit to the hospital.

developed into fluid filled blisters. Some of these blisters coalescent and increasing in numbers overtime. Some of the blisters then burst and leaving a crust **(Figure 1).** Otherwise, there are no other complaints in this patient. There is no history of previous disease and no previous history of drug or food allergies. There is no history of similar diseases in the patient's family.

On physical examination, patient's general condition is good, compos mentis, good nutritional status, body weight 55kg and height 153cm, blood pressure 130/85 mmHg, pulse 88 bpm, respiratory frequency 16×/minute and body temperature 36.8°C.

Upon dermatological examination, multiple vesicobullous lesions on an erythematous background, erosion and brownish crusts were observed in the malar region and nasal region.

Patient refused to be examined further and was not compliance to the treatment given. She then consumed 3 types of traditional medicine and admitted to the hospital 2 weeks later with extensive vesicobullous lesions in the face, scalp, trunk and extremities. Upon dermatological examination, generalized skin lesions in the form of multiple bullae, erosion and blackish-brown crusts were observed in the facial, oral, scalp region, trunk and extremities. Non-scarring alopecia areata lesion was found on the scalp, which associated with thick yellowish plaque on an erythematous base in the parietal region of the scalp **(Figure 2)**. Patient was then admitted and further serological investigations were conducted.

The patient was differential diagnosed with bullous type ACLE and Steven-Johnson Syndrome/SJS. Laboratory results showed hemoglobin 11.4 g/dL*, MCV 76.40 fL*, hematocrit 35%*, leukocytes 11.67x103/µL*, platelets 556x103/µL*, liver function and kidney function within normal limits, blood glucose levels within normal limits.

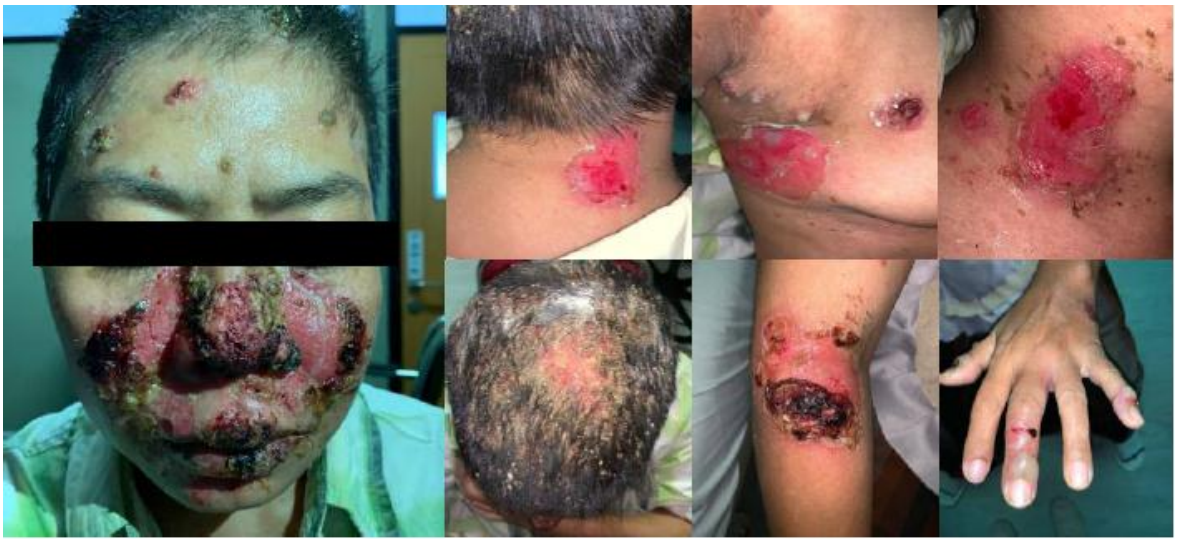

Figure 2 Patient's clinical manifestations during her second visit to the hospital.

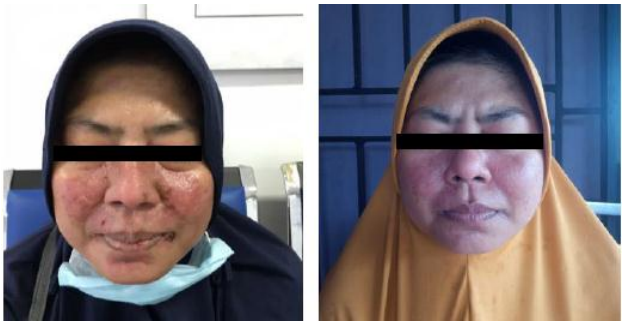

Figure 3

The patient's urinalysis results showed no proteinuria. Antibody titre ANA test result were positive but ds-DNA antibody was negative. The patient refused further histopathological examination. The provisional diagnosis of this patient was bullous type ACLE. Patients were then given intravenous Methylprednisolone 125mg once daily, oral Erythromycin 500mg qid, Paracetamol drip 1 gram bid and Ranitidine 50mg bid. Normal saline compresses were applied to the wet erosion lesion for 15 minutes every 4 hours and topical fusidic acid cream twice daily.

Patients showed significant clinical improvement during hospitalization, no new bullae lesions appeared during hospital treatment and IV doses of methylprednisolone were tapered off gradually. After 8 days of hospital care, patient was discharge with oral dose of Methylprednisolone 20mg/day and 2% Ketokonazol scalp solution twice weekly for her scalp. Patients were advised to get adequate rest, avoid excessive sun exposure, apply SPF 50 sunscreen and was advised not to consume her traditional medicines.

On her first follow up, one week after discharge from hospital, there were no new complaints from patient. Significant clinical improvement was seen and no new bullae appeared **(Figure 3)**. The dose of methylprednisolone tablets was tappered again to 12mg/day (for 4 days), followed by 4mg/day (for the subsequent 3 days), after which the methylprednisolone tablet was stopped. Complete remission was observed in this patient on her third follow up, two weeks after her first follow up **(Figure 3)**.

Discussion

Bullous type LE is a rare variant of LE and its etiology is still not understood clearly. However, the pathogenesis of bullous LE is thought to be related to the presence of autoantibodies to collagen VII or other components of the dermal-epidermal junction. Type VII collagen plays an important role as an anchoring fibril that connects the epidermis to the dermis. Circulating autoantibodies are addressed to non-collagen domains type 1 and 2 (NC1 and NC2) of type VII collagen found in the basement membrane. This results in impaired adhesion of the basement membrane and dermis, resulting in the appearance of subepidermal blisters. This rare entity is usually observed in ACLE with increased systemic disease activity.[2-5]

Bullous LE is usually associated with subepidermal blisters with disseminated vesicobullous lesions that arise on sun exposed skin. It can also present with erosion, crust and mucous membrane lesions. Sometimes lesions healed with residual hyperpigmentation, scarring, and milia. The diagnosis of bullous LE can be established through clinical manifestations, supporting examinations, histopathological examination and immunoflorescence tests.[5] Histopathological findings in bullous LE showed subepidermal bullae with superficial perivascular infiltrates containing neutrophils accompanied by neutrophilic microabscesses in the papillary dermis. Direct immunofluorescence/ DIF will show deposition of IgG and/or IgM, IgA, and C3 in the dermal-epidermal junction.[2]

In this case, the patient was differential diagnosed with bullous type LE and Steven

Johnson Syndrome/SJS. Differentiating these two entities is a challenge because clinically they are very difficult to distinguish.[2] Some aspects that support our diagnosis include no consumption history of SJS high-risk drugs and her skin lesions have appeared before the patient took the traditional supplements, initial vesicobullous lesions in the malar area and subacute course of her illness.

According to the literature, administration of dapsone combined with glucocorticoids and anti-malarials is the first-line therapeutic option for bullous LE and is associated with improvement in almost 100% cases. However, its use is limited when the patient has a history of anemia, hepatitis, a history of allergy to dapsone and in pregnant women.[6]

In some cases, bullous LE appeared to be an early manifestation of SLE. Therefore, patients need to be educated about the early signs and symptoms of organ involvement, protection from sun exposure and the importance of regular follow-ups.[5]

Conclusion

We reported a 40-year-old woman with bullous lupus erythematosus without systemic involvements. Patient showed complete remission and excellent clinical response after corticosteroid treatment. This report showed the importance of early diagnosis as well as prompt treatment can result in a better prognosis.

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