Diffuse skin lesions in a patient with systemic lupus erythematosus: A case report

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Summary

Systemic lupus erythematous (SLE) is a persistent autoimmune disease with systemic and life- threatening manifestations, in which cutaneous disease is the second most common manifestation, after joint inflammation. Skin lesions in SLE is originally divided into two groups: LE-specific lesions and LE-nonspecific lesions. The term "LE-specific lesions" includes 3 distinct categorizes: Acute cutaneous lupus erythematosus (ACLE), subacute cutaneous lupus erythematosus (SCLE) and chronic cutaneous lupus erythematosus (CCLE) [4]. By another method of classification, specific skin lesions can also be classified as localized or generalized lesions. The most common localized lesion is malar, or butterfly rash, which is reported in 20-60% of patients with SLE. In comparision to localized lesions, generalized lesions, which are indiscriminately referred to as lupus maculopapular rash, are relatively rare. This form of lesion is characterized by well-demarcated erythematous macules and/or papules located on the extensor side of limbs, sparing the knuckles. In this article, we report a case of a female patient having generalized cutaneous lesions. She was initially diagnosed with SLE and received treatment at Department of Allergy, Center of Dermato-Venerology and Allergy, 108 Military Central Hospital.

Keywords: Systemic lupus erythematosus, lupus cutaneous disease.

1. Background

Lupus erythematous (LE) is an autoimmune disease with a diverse array of clinical manifestations, from cutaneous to life-threatening systemic illnesses. Skin lesions, in some cases, can be isolated; whereas others, can be combined with other disorders and be considered as a symptom of SLE. Skin illness is observed in 70-80% of patients with SLE at any time throughout their disease process; or it can be the initial manifestation in approximately 25% of patients with SLE [3].

Clinical manifestations of LE-specific lesions are diverse, including localized acute skin lesions (butterfly rash), generalized acute skin lesions, psoriasis-like subacute skin lesions [6], annular erythema in lupus tumidus [2], chronic discoid LE skin lesions [7], panniculitis in lupus profundus, depigmentation and dry necrosis in the distal side of fingers,… LE-nonspecific skin lesions consist of nonscarring alopecia, Raynaud phenomenon, livedo reticularis, vasodilation, LE-nonspecific bullous lesions,…

However, cutaneous manifestations in SLE can be combined by many isolated and distinguished form of lesion. We report a clinical case of a patient with generalized skin disease which is combined by several distinct types of lesion.

2. Case presentation

A 24-year-old female patient with a history of skin disease for 2 years, was initially diagnosed with atopic dermatitis. A month before admitted to the

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hospital, the onset was mild- itchy, scattered erythema located on the face, palms and trunk. Concurrently, she suffered from fatigue, anorexia, significant weight loss (7kg/month), chest discomfort, shortness of breath and alopecia. When being hospitalized, she had high fever (39 to 40 degrees Celsius), low body mass index (16.8), paleness and did not show any sign of joint inflammation, while cutaneous lesions were plentiful and catogorized into many different types:

Scaly, small (less than 3mm in diameter), erythematous macules located on head and face.

Annular, erythematous macules and papules with hypopigmented center on the trunk.

Vasculitis-like lesions, subcutaneous hemorrhage, Gottron papules, interleaving with dry necrotic ulcers on the dorsal of hands.

Alopecia areata.

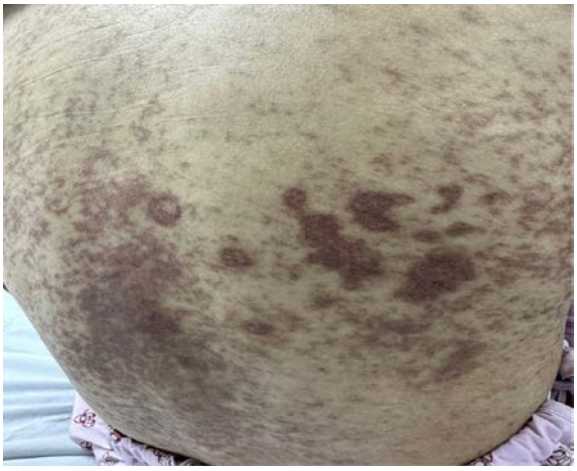
Ulcers on mucous membrane of lips and palate.

The laboratory tests demonstrated many significant changes: anemia (RBC: 2.85T/L, HGB: 88g/L, HCT: 0.269L/L, direct Coombs test: Positive 3+, Indirect Coombs test: Positive 3+), thrombocytopenia (PLT: 117G/L), hypoalbuminemia (albumin: 18.4g/l), elevated liver enzymes (AST: 419.1U/l, ALT: 65.7U/l), elevated muscle enzymes (CK: 1340U/l, LDH: 1060U/l), proteinuria (24-hour urine protein test: 0.73g/24h), low C3 and C4 (Complement C3: 44mg/dL, C4: 2.6mg/dL), high levels of antinuclear antibodies (ANA: positive (> 12)), high level of anti-double stranded DNA antibodies and other changes in immunology tests (dsDNA: Positive (51.6U/ml), Anti SSA: Positive, Anti SSB: Positive, Anti Sm: Positive, Anti Jo1: Negative, Anti Scl 70: negative, Anti CENP: Negative).

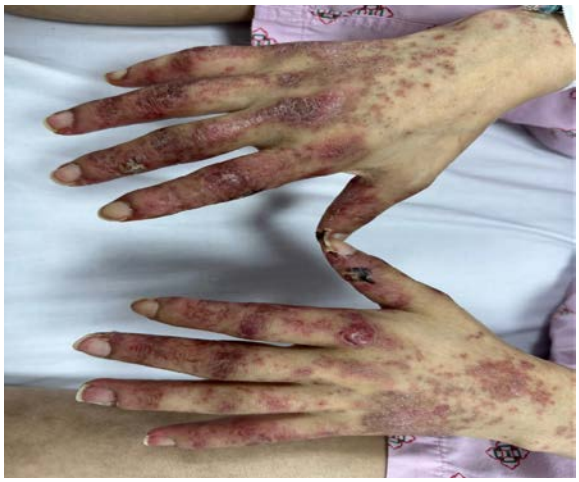
Electromyography: No abnormality in the transmission rate, low amplitude, flat electrical activity, with spontaneous potentials.

Histopathological examination of the skin was not specific. It revealed the infiltration of monocytes and neutrophils in stratum corneum; keratinoid corneal degeneration in the epidermis; lymphocytic infiltration in the dermis and skin appendages; no degeneration of the basement membrane observed.

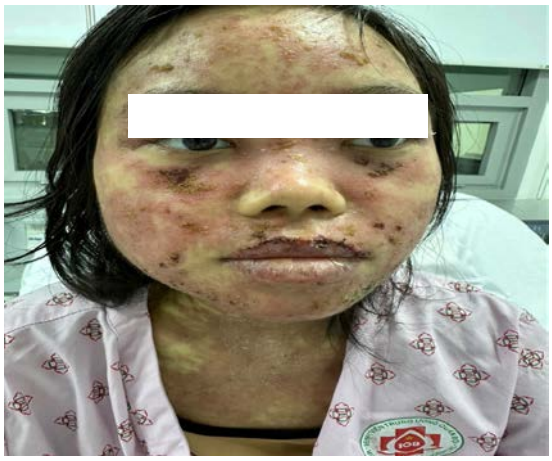
The patient was diagnosed with SLE and treated immediately with methylprednisolone 2mg/kg/d, MMF 2g/d and HCQ 200mg/d. After 1 week of treatment, patient's body temperature became normal; cutaneous manifestations improved remarkably with most of skin lesions crusting and regressing. Our patient was discharged after 17 days, with the dose of methylprednisolone tapered to 16mg/d while the dose of MMF and HCQ remained 2g/d and 200mg/d respectively.



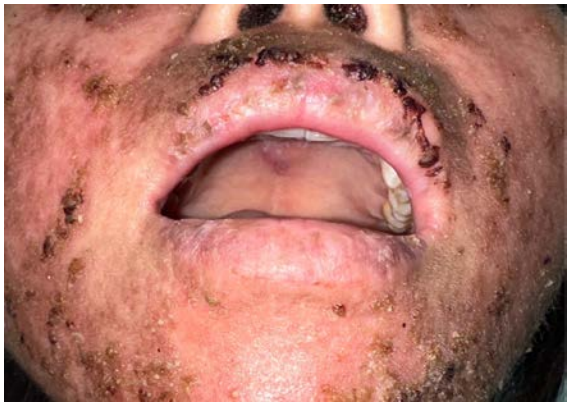
Picture 1. Annular, erythematous macules and papules with hypopigmented center on the trunk



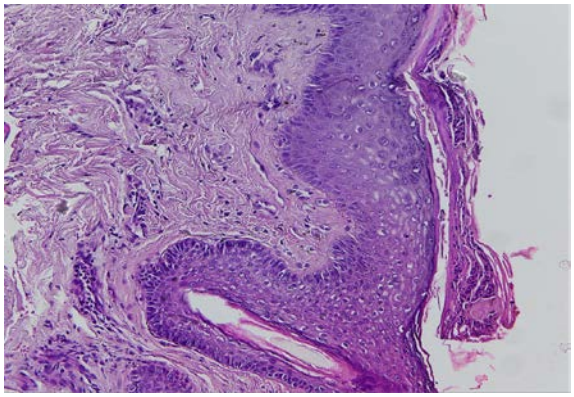
Picture 2. Vasculitis-like lesions, subcutaneous hemorrhage, Gottron papules, interleaving with dry necrotic ulcers on the dorsal of hands.



Picture 3. Scaly, small, erythematous macules located on head and face



Picture 4. Ulcers on mucous membrane of lips and palate



Picture 5. Nonspecific histopathological image.

3. Discussion

SLE is a systemic disease that affects many different organs in the body. The skin is the second most commonly affected organ after joint damage [1], [3]. While LE-specific lesions have their own roles in diagnosis process, LE-nonspecific lesions contribute notably on assessing the level of disease activity and monitoring during treatment [9]. In this patient, there was an acute, aggressive onset, with widespread skin lesions and multi-organ injury. The patient's manifestation included many types of specific and non-specific skin lesions such as: Non-scarring alopecia, vasculitis, diffuse erythematous macules, papules all over the body, and some lesions resemble Gottron papules [3]. At first, the diagnose was quite confusing, due to the fact that there were various types of lesion which can be observed in distinct diseases, accompanied by atypical pathological images. Firstly, in this patient, the disease scored 24 points accroding to 2019 EULAR/ACR criteria for SLE [5]. She had strong ANA positivity, fever, autoimmune anemia, non-scarring alopecia, mouth ulcers, proteinuria > 0.5g/24h, positive aβ2GPI antibodies, low C3 and C4, positive anti-dsDNA antibody, positive anti Sm antibody. These above symptoms played an irreplaceble role on diagnose progress, while the criteria of EULAR/ACR had a sensitivity of 96.1% and specificity of 93.4%. Therefore, the diagnose of SLE was reasonable and trustworthy. On the other hand, due to the apperance of Gottron papules and elevated muscles enzymes, diagnosis of dermatomyosis should be taken into consideration. However, electromyography and antibody testing, moreover, helped rule out dermatomyositis [3]. Immediately after the diagnosis of autoimmune disease was made, the patient was initially treated with methylprednisolone 2mg/kg/day for 3 days. At this stage, the disease improved insignificantly. We added MMF 2g/day and HCQ 200mg/d, the response was more remarkable, the skin lesions tended to regress without leaving scars, concurrently, the whole body's condition improved considerably, the patient had normal temperature and less fatigue.

Compared with localized skin lesions, generalized skin lesions in patients with systemic lupus erythematosus (SLE) are rare [7], occurring in

only 5-10% of patients [3]. However, these two types of lesions have relatively similar pathological images, characterized by the image of interface dermatitis. Treatment of skin lesions includes therapies such as: systemic corticosteroids, topical corticosteroids, antimalarials and other immunosuppressive agents [8].

Accurate diagnosis is the key for optimal disease management. However, in rare cases of generalized LE cutaneous disease, precise diagnosis is quite challenging and can be mistaken for many other skin disorders with nonspecific and polymorphous lesions. Differential diagnoses should be taken into consideration:

Drug-induced photosensitivity [1]: The patient has a history of previous drug use. There are certain drugs that often cause this condition, and at the same time the onset of the disease is more acute.

Dermatomyositis [1]: In addition to skin damage, muscle damage also appears, expressed through muscle weakness, increased muscle enzymes, and abnormal electromyography.

Pemphigus erythroderma [1]: Usually excluded if the patient has multi-organ involvement.

Atopic dermatitis [1]: Diagnosed by exclusion after examining all organs and taking a thorough medical history. Atopic dermatitis is characterized by persistent, long-lasting, chronic skin disorder, without accompanying organ damage and with accompanying atopic factors.

4. Conclusion

We reported this case because there are very few reports of SLE patients with generalized acute skin lesions, consisting of both typical and atypical manifestations. Diffuse skin lesions in SLE is a rare condition, including many different forms of lesion. Therefore, misdiagnosis is completely able to occur. Accurate diagnosis and optimal treatment not only help improve skin damage, but also slow down the disease process, minimalize disease activity, thereby prolonging patient's life as well as improving the quality of life.

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