Case Report

Pustular and crusted lesions in systemic lupus erythematosus: A case report

Allam P. Reddy, Ramachandran Ramakrishnan, Murali Narasimhan

Department of Dermatology, Venereology and Leprosy, SRM Medical College and Research Centre, Chengalpattu, Tamil Nadu, India

Abstract

Systemic lupus erythematosus (SLE) is the prototype of an autoimmune disease with various manifestations in the skin and several other organs. Subacute cutaneous lupus erythematosus may present with annular and psoriasiform lesions. There have been case reports of pustular lesions in SLE. Herein, we describe a known case of SLE, who developed pustular and crusted lesions which complicated the course of the disease. We considered Acute Generalised Exanthematous Pustulosis, Subcorneal Pustular Dermatosis, Pustular Psoriasis, Pustular Vasculitis, and Pustular Folliculitis as our initial differential diagnosis. A potassium hydroxide mount from crusted lesions showed scabies mites. With these findings, a diagnosis of SLE complicated with crusted scabies was made. She was managed with multiple systemic immunosuppressant and antiscabetic measures following which clinical improvement was seen with remission of her lesions.

Keywords: Crusted scabies, pustular lesions, systemic lupus erythematosus

Introduction

SLE is a systemic autoimmune disease with a broad range of cutaneous manifestations which can be divided into LE-specific and LE nonspecific lesions.[1] Among the LE-specific manifestations, papulosquamous lesions may occur in SCLE and pustular lesions may be nonspecific manifestation of LE. AGEP, pustular psoriasis, and SPD are common differentials for such pustular lesions. Norwegian scabies can occur in patients on chronic immunosuppressive therapy. Herein, we describe a known case of SLE, whose course was complicated by the development of pustular and crusted lesions. We chose to report this case for the diagnostic challenge posed and the therapeutic complexity.

Address for correspondence: Dr. Ramachandran Ramakrishnan, Department of Dermatology, Venereology and Leprosy and SRM Medical College and Research Centre, Kattankulathur, Chengalpattu, Tamil Nadu, India.
E-mail: drram.srkn@gmail.com

Received: 05-04-2024 **Revised:** 30-05-2024
Accepted: 03-06-2024 **Published:** 18-11-2024

Access this article online

Quick Response Code:

Website: http://journals.lww.com/JFMPC

DOI: 10.4103/jfmpc.jfmpc_569_24

Case History

A 15-year-old female patient, who was a known case of SLE for the past 1 year, presented with crusted skin lesions all over the body for 3 months. The lesions started over her abdomen and gradually progressed to involve both her extremities. Pustular lesions developed a month later. She had a documented history of arthralgia, photosensitivity, and palatal erosions over the past year and was on prednisolone, MMF, and other supportive measures. Examination showed multiple thick crusted plaques over the erythematous base over the chest, abdomen, upper back, arms, and thighs [Figures 1a and 2a]. Multiple crusted papules and pustules were also present on both arms, forearms, dorsum of hands, web spaces, thighs, pubic area, and lower limbs [Figure 3]. Her face was completely spared. She had tenderness over the elbow joints.

Investigations

Initial screening investigations were done. Her haemoglobin was 9 g/dl, ESR was raised (93 mm/hr), and the CRP

How to cite this article: Reddy AP, Ramakrishnan R, Narasimhan M. Pustular and crusted lesions in systemic lupus erythematosus: A case report. J Family Med Prim Care 2024;13:5374-7.

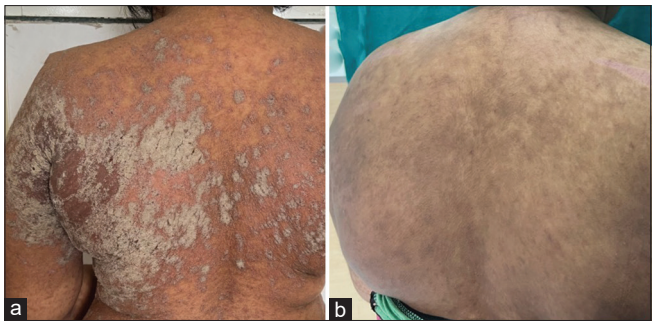

Figure 1: (a) Thick crusted plaques over erythematous base over upper back extending to extensor aspect of arms and (b) lesions over the back resolved with postinflammatory hyperpigmentation after 6 months of therapy

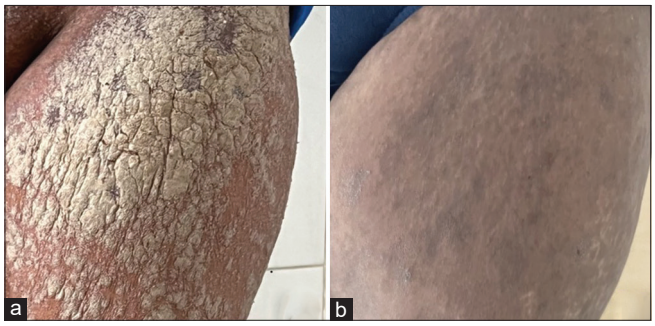

Figure 2: (a) Thick crusted plaques noted over thighs and (b) lesions resolved with postinflammatory hyperpigmentation over thighs after 6 months of therapy

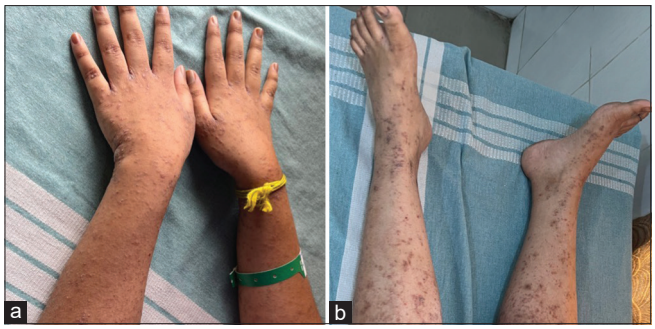

Figure 3: (a) Multiple papules and pustules noted over the extensor aspect of bilateral forearms, dorsum of hands, and web spaces of bilateral hands, and (b) multiple hyperpigmented papules and plaques noted over the bilateral anterior aspect of legs

was positive (11.2 mg/dl). She was also found to have hyperkalemia (5.5 mg/dl) and hypomagnesemia (1.7 mg/dl). KOH mount was done from the crusted lesions in the web spaces and back, which showed scabies mites [Figure 4]. Blood and urine culture showed no growth, whereas pus culture showed MRSA. ANA profile showed positivity (4+, by IF method) and positive for antibodies against ds-DNA, ribosomal proteins, and nucleosomes. The direct Coombs test showed positivity, and C3 (42 mg/dl) and C4 (7.37 mg/dl) levels were low. Skin biopsy was done from one of the pustular lesions which showed mild hyperkeratosis, parakeratosis and irregular acanthosis, neutrophilic collection, and a burrow-like structure within stratum corneum, along with structures suggestive of empty egg shells. A spongiform neutrophilic collection was noted in the upper stratum malpighi [Figure 5] and dilated capillaries seen in the papillary dermis. With the above findings, we arrived at a diagnosis of crusted scabies with pyoderma, along with flare-up of SLE.

Discussion

Based on clinical features, differential diagnosis of pustular psoriasis, AGEP, and SPD were considered.

AGEP is an eruption commonly associated with drug exposure resulting in an acute pustular rash with fever and leucocytosis. It is commonly associated with aminopenicillins, hydroxychloroquine, and terbinafine.[2] It is described as T-cell-related sterile neutrophilic inflammatory response to a drug that usually occurs within 48 hours.[3] Histopathologically, characterized by intraepidermal pustules with edema in the papillary dermis containing neutrophilic and eosinophilic infiltrates.

SPD is a rare yet relapsing benign condition typically affecting elderly individuals. The lower half of the pustule contains purulent material, whereas the upper half contains a clear fluid (hypopyon pustule).[4] Histopathologically, subcorneal pustules containing a small number of neutrophils and minimal acantholysis are seen.

Pustular psoriasis is a rare systemic skin disease that is characterized by pustules over an erythematous base. It is classified into generalized and localized types.[5] Risk factors include sudden withdrawal of systemic steroids, infections, medications, and vaccinations.[6]

Crusted scabies, also known as Scabies Norvegi Boeki or Norwegian scabies,[7] is caused by *Sarcoptes scabiei var hominis*. The failure of the immune system to suppress the proliferation of the mite is an important pathogenetic basis behind crusted scabies. Hyperkeratosis of the skin in crusted scabies is possibly related to raised levels of IL-4.[8] Itching, the hallmark of scabies, is minimal or absent in crusted scabies.[9] The microscopic examination of skin scrapings in a normal saline mount reveals eggs, mites, and scybala.[10] Histopathologically, there are numerous mites and irregular structures consistent with scybala and eggshells inside burrows within the stratum corneum. Massive hyperkeratosis, orthokeratosis and acanthosis, psoriasiform hyperplasia, exocytosis of eosinophils and neutrophils, and intraepidermal microabscess may be seen.[11]

According to European guidelines, oral ivermectin 200 micrograms/kg is given on days 1, 2, and 8. In severe cases, auxiliary doses on days 9, 15, 22, and 29 are given.[12]

The coexistence of SLE flare and crusted scabies, though explainable due to the iatrogenic immunosuppression, still poses a therapeutic challenge. Since she had an SLE flare with crusted scabies, along with pyoderma, we had to augment

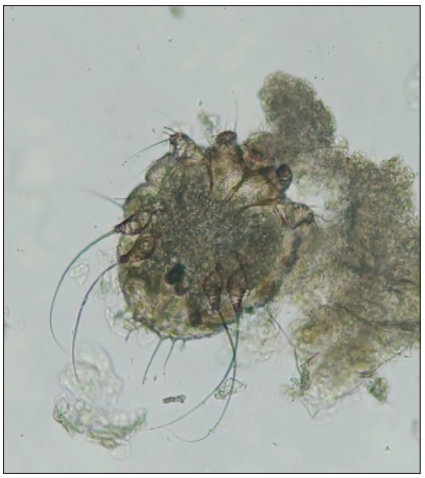

Figure 4: Potassium hydroxide mount showing scabies mite

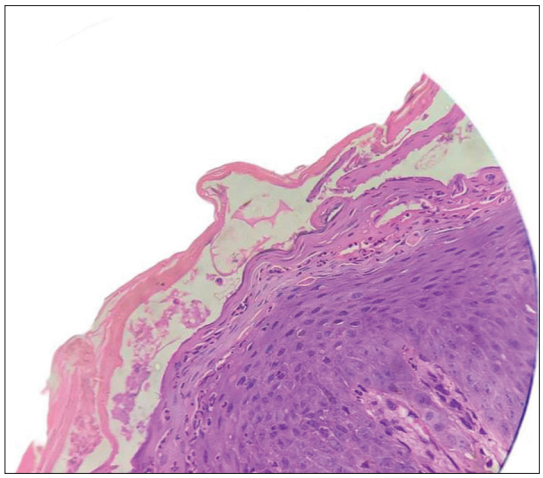

Figure 5: Histopathological picture showed mild hyperkeratosis, parakeratosis, irregular acanthosis, neutrophilic collection, and a burrow-like structure within the stratum corneum, with structures suggestive of empty egg shells. It also showed dilated capillaries in the papillary dermis (400X magnification, H and E staining)

immunosuppressive therapy under the cover of intravenous antibiotics and multiple doses of oral Ivermectin.

Treatment

The patient was administered six doses of ivermectin 12 mg (on days 1, 2, 8, 9, 15, and 22) and a single short contact external application of 5% permethrin cream for 2 hours. The patient was started on intravenous linezolid injection based on pus culture and sensitivity report. Intravenous dexamethasone 8 mg per day was initiated and tapered to 30 mg daily oral prednisolone at discharge. MMF 1 gram per day was continued.

Outcome and follow-up

The thick crusted plaques resolved after ivermectin therapy, and the patient's family was also treated with ivermectin. Her arthralgia was resolved, and complement levels were normalised gradually. The patient is currently under our follow-up, with no new skin lesions and old lesions resolving with PIH, while on daily oral prednisolone 5 mg and MMF 1 gram [Figures 1b and 2b].

Key message

- This case was diagnostically challenging as pustular and crusted lesions in SLE are rare. Despite scabies being a common condition, diagnosing crusted scabies requires a high index of clinical suspicion.
- Since crusted scabies is highly contagious and can pose an epidemiological challenge, the need for early diagnosis and appropriate treatment by a primary care physician holds paramount significance. It can mitigate the otherwise negative impact of such an infestation on the overall well-being of the community.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

List of abbreviations

Abbreviation	Definition
SLE	Systemic Lupus Erythematosus
SCLE	Subacute cutaneous lupus erythematosus
LE	Lupus Erythematosus
ESR	Erythrocyte Sedimentation Rate
CRP	C-reactive protein
KOH	Potassium hydroxide
IF	Immunofluorescence
MMF	Mycophenolate mofetil
.AGEP	Acute Exanthematous Generalised Pustulosis
SPD	Subcorneal Pustular Dermatosis
ANA	Anti-nuclear antibody
ds-DNA	double-stranded Deoxyribonucleic acid
C3 and C4	Complement 3 and 4
MRSA	Methicillin-Resistant Staphylococcus aureus
IL-36RN	Interleukin 36 Receptor Antagonist
PIH	Post inflammatory hyperpigmentation
H and E staining	Hematoxylin-eosin (H and E) staining
mg	milligrams

References

1. Gheisari M, Baghani M, Ganji R, Zahedi K. Annular pustular crusted plaques as unusual cutaneous presentation of systemic lupus erythematosus. Clin Case Rep 2023;11:e7475. doi: 10.1002/ccr3.7475.

2. Heelan K, Sibbald C, Shear NH. Cutaneus reactions to drugs. In: Kang S, Amagai M, Bruckner AL, Enk AH, Margolis DJ, McMichael AJ, editors. Fitzpatrick's Dermatology. 9th ed. New York: McGraw Hill; 2019. p. 751-2.
3. Navarini AA, Valeyrie-Allanore L, Setta-Kaffetzi N, Barker JN, Capon F, Creamer D, *et al.* Rare variations in IL36RN in severe adverse drug reactions manifesting as acute generalized exanthematous pustulosis. J Investig Dermatol 2013;133:1904-7.
4. Bhargava S, Kumar U, Kroumpouzos G. SPD: Comprehensive review and report of a case presenting during pregnancy. Int J Women's Dermatol 2020;6:131-6.
5. Tsuchida Y, Hayashi R, Ansai O, Nakajima M, Oginezawa M, Kawai T, *et al.* Generalized pustular psoriasis complicated with bullous pemphigoid. Int J Dermatol 2019;58:e66-e67.
6. Boehner A, Navarini AA, Eyerich K. Generalized pustular psoriasis - A model disease for specific targeted immunotherapy, systematic review. Exp Dermatol 2018;27:1067-77.
7. Parish LC, Lomholt G. Crusted scabies alias Norwegian scabies. Int J Dermatol 1976;15:747-8.
8. Morsy TA, Romia SA, al-Ganayni GA, Abu-Zakham AA, al-Shazly AM, Rezk RA. Histocompatibility (HLA) antigens in Egyptians with two parasitic skin diseases (scabies and leishmaniasis). J Egypt Soc Parasitol 1990;20:565-72.
9. Roberts LJ, Huffam SE, Walton SF, Currie BJ. Crusted scabies: Clinical and immunological findings in seventy-eight patients and a review of the literature. J Infect 2005;50:375-81.
10. Dia D, Dieng MT, Ndiaye AM, Ndiaye B, Develoux M. Crusted scabies in Dakar apropos of 11 cases seen in a year. Dakar Med 1999;44:243-5.
11. Weedon D. Arthropod-Induced Diseases. In Weedon's skin pathology. 3rd ed. Australia: Elsevier Health 2010. p.655-657.
12. Salavastru CM, Chosidow O, Boffa MJ, Janier M, Tiplica GS. European guideline for the management of scabies. J Eur Acad Dermatol Venereol 2017;31:1248-53.