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CASE REPORT

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Annular pustular crusted plaques as unusual cutaneous presentation of systemic lupus erythematosus

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Key Clinical Message

Systemic lupus erythematosus (SLE) is a systemic autoimmune disease that affects several organs including the skin. Clinical cutaneous symptoms of SLE come in a broad range, consisting of both non-specific and specific cutaneous lesions. Except for cases of amicrobial pustulosis of the folds, generalized pustular psoriasis, acneiform eruptions, pustular vasculitis, Wells' syndrome, subcorneal pustular dermatosis, and neutrophilic dermatosis, there are no reports of pustular lesions linked to SLE. The unusual cutaneous features of our patient were annular plaques with pustules and crusts on the margins.

KEYWORDS

cutaneous lupus, pustule, systemic lupus erythematosus

1 | INTRODUCTION

Systemic lupus erythematosus (SLE) is an autoimmune disorder with a chronic course. The broad range of SLE cutaneous manifestations is present in 70%–85% of the affected patients. There are two categories of SLE cutaneous lesions, namely lupus erythematosus (LE)-specific, including acute cutaneous manifestations of LE (ACLE), subacute CLE (SCLE), and chronic CLE (CCLE), and LE-non-specific. The classification criteria of systemic lupus erythematous based upon EULAR including cutaneous lesion (malar rash, discoid rash, photosensitivity), oral ulcers, arthritis, serositis, renal, neural and hematologic disorders, immunologic disturbances, and abnormal titer of ANA.[1] Interface dermatitis, lichenoid infiltration, basal cell vacuolation, and mucinous deposition are characteristics of the histopathology of LE-specific lesions.[2] Pustular appearance is one of the least often documented presentations reported in the setting of amicrobial pustulosis of the folds (APF),[3] generalized pustular psoriasis,[4] acneiform eruptions,[5] pustular vasculitis,[6] Wells' syndrome,[7] subcorneal pustular dermatosis,[8] and neutrophilic dermatosis.[9] Here, we describe annular lesions with marginal pustules in a patient whose condition was subsequently identified as SLE.

2 | CASE REPORT

An 85-year-old woman presented to our clinic with new-onset cutaneous eruptions for 7 months. Over the previous 2 months, a variety of symptoms and issues, including confusion, malaise, fever, poor food intake, and debilitation had arisen. Her only noteworthy prior medical history was a long-lasting hypertension treated with metoprolol.

At the presentation, the patient was ill but afebrile. Her heart rate was 90 bpm with an irregularly irregular rhythm. Other vital signs were a respiratory rate of 17 per minute, a blood pressure of 140/80 mm Hg, and a 96% oxygen saturation at rest in room air. The skin examination revealed

generalized erythematosquamous papules and plaques as well as an erythematous rash over the cheeks and the nasal bridge and chilblain. Most of the plaques were polycyclic and annular in shape, with crusts and pustules on the edges of the sheens, chest, and back (Figure 1). Similar plaques covered the hands, although the knuckles were noticeably spared. Along with regular arterial pulses and capillary refill time, a dark blue discoloration of the plantar surface and toes was observed. Mucosal surfaces were normal.

The initial laboratory testing showed sodium levels of 129 mEq/L and hypokalemia (2.7 mEq/L). Intravenous fluid administration was initiated. Specimens of pustule secretions were collected for bacterial smear and culture. Other laboratory test results revealed lymphopenia but normal white blood cell count, platelet count, hemoglobin, mean corpuscular volume, C-reactive protein, and erythrocyte sedimentation rate. Pustule culture and smear did not reveal any growth. Hypocomplementemia, positive levels of an anti-nuclear antibody (7.5), anti-SSA, and anti-SSB were all seen on the immunological panel. The coronavirus disease 2019 (COVID-19) polymerase chain reaction came back negative, and all other laboratory test results were normal.

Atrial fibrillation and multifocal atrial tachycardia were present on the electrocardiogram. A unilateral left-sided pleural effusion was evident on chest computed tomography. Although thoracentesis was not possible owing to volume limitations, pleural effusion was verified by ultrasonography.

Electroencephalogram and brain magnetic resonance imaging were carried out due to the reduced level of consciousness. While the former had non-specific moderate diffuse encephalopathy, the latter displayed numerous signal alterations of white matter associated with atherosclerotic manifestations.

For histologic and direct immunofluorescence (DIF) investigation, two lesional (the rim of annular chest lesions including pustules) and one perilesional punch biopsies of 4.0 mm were obtained. Lesional DIF showed focal scant granular deposition of IgM (2+) along the dermoepidermal junction (DEJ) and perilesional DIF revealed focal scant granular deposition of IgM (3+) and IgG (2+) along the DEJ. The microscopic manifestations of lesional histology included neutrophilic exocytosis into the epidermis, subcorneal intraepidermal neutrophilic pustule formation, diffuse interface lichenoid reaction, and civatte body formation. The dermis showed modest extravasation of red blood cells, swelling of the endothelial cells, and superficial and deep infiltration of polymorphonuclears and lymphocytes in the perivascular and perifollicular regions. Periodic acid–Schiff staining only

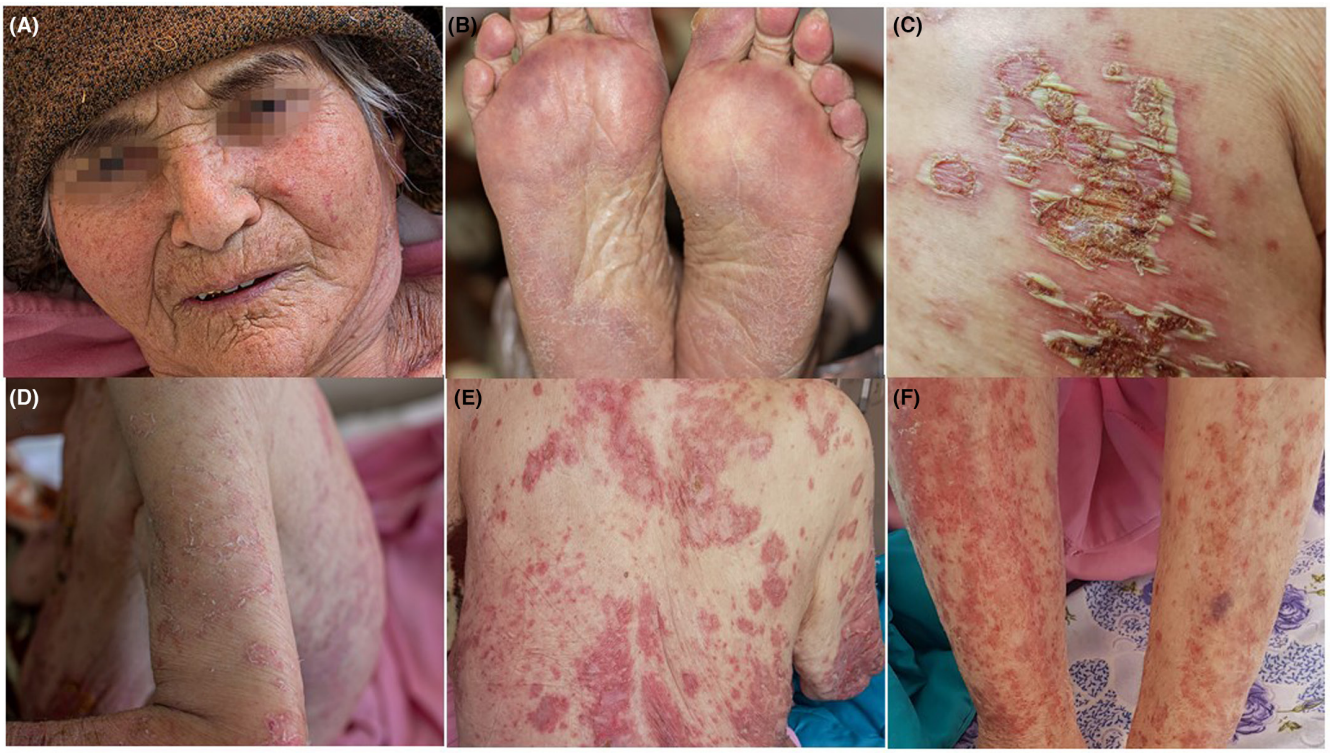

FIGURE 1 Erythematous rash over the cheeks and the nasal bridge (A), chilblain (B), and generalized erythematosquamous papules and plaques with polycyclic and annular in shape, with crusts and pustules on the edges on the chest (C), arms (D), back (E), and sheens (F).

showed mild focal basement membrane thickening, and Alcian blue staining revealed marked interstitial dermal mucin in the superficial and mid-dermis. In brief, the superficial and deep perivascular-perifollicular inflammation with acute small vessel vasculitis, interface lichenoid reaction, and subcorneal-intraepidermal pustule formation were suggestive of collagen vascular diseases such as SLE (Figure 2).

Based on the Systemic Lupus International Collaborating Clinics (SLICC) criteria, the index patient was identified as having SLE. The fulfilled criteria included malar rash, subacute cutaneous lupus, pleural effusion, lymphopenia that had been detected on multiple occasions, the presence of anti-nuclear antibody (ANA), hypocomplementemia, and acute confusion. Accordingly, the patient received prednisolone 20 mg PO and hydroxychloroquine 200 mg BD, which led to the patient's regaining complete consciousness and resolution of the cutaneous lesions over 2 weeks except chilblain lesions. Remedy of patient was maintenance with hydroxychloroquine for 2 years and prednisolone tapered for 1 year.

3 | DISCUSSION

Skin manifestations are present in 80% of SLE patients; however, in only a quarter of them, these cutaneous presentations are a sign of disease onset.[10] The index patient first presented with a variety of cutaneous manifestations and later fulfilled the diagnostic criteria of SLE in our further evaluations. These diverse skin manifestations included a combination of three subtypes of CLE: malar rash and photosensitivity (ACLE), annular papulosquamous lesions (SCLE), and chilblain (CCLE). In addition, the presence of multiple marginal pustules on the edge of annular papulosquamous lesions was a unique finding. Our patient also had pleural effusion, lymphopenia, and hypocomplementemia, all of which pointed to the diagnosis of SLE. Furthermore, the index case was elderly. It is worth noting that although SLE is more common among women of childbearing age, 10%–20% of SLE patients are seniors.[11]

Rarely have pustular lesions been reported in SLE patients. Using the available literature, we were able to locate 25 instances of SLE with pustular lesions (Table 1).

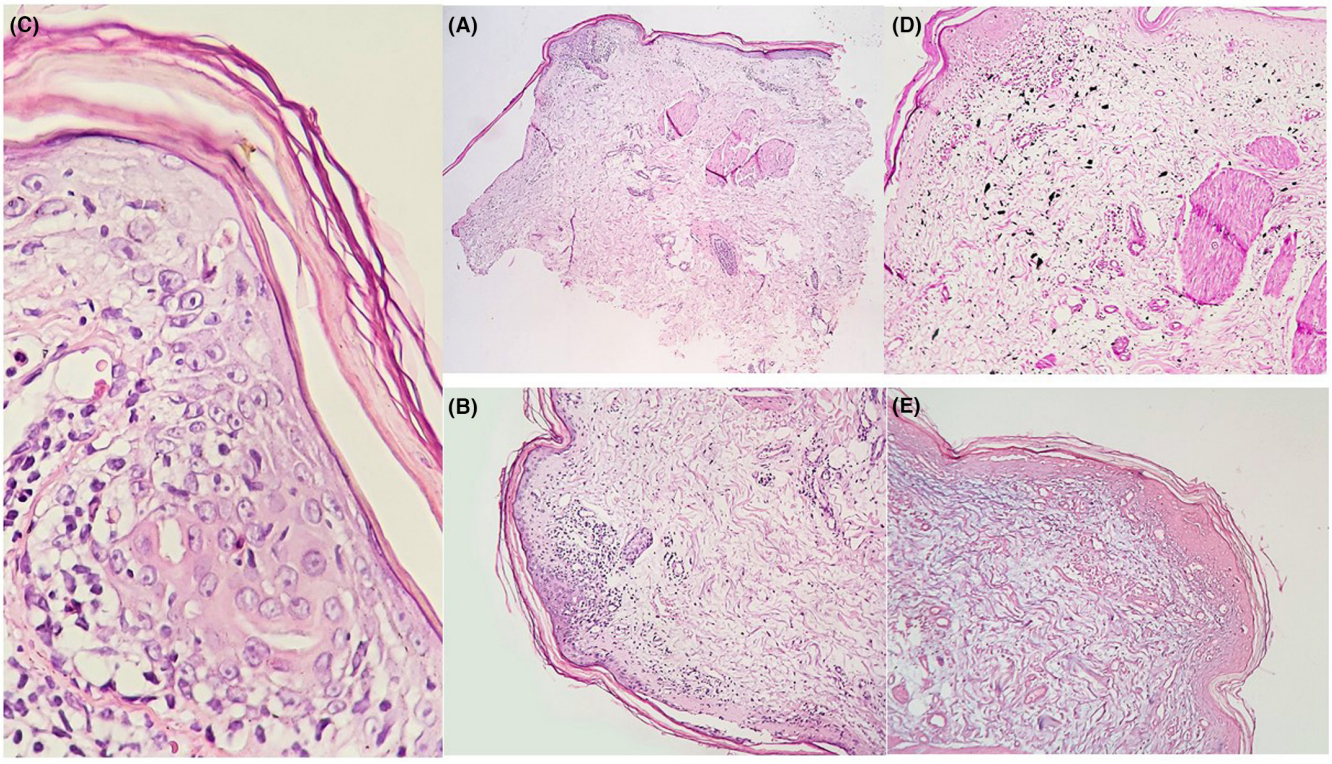

FIGURE 2 Hematoxylin and eosin stain showed neutrophilic exocytosis into the epidermis, subcorneal intraepidermal neutrophilic pustule formation, diffuse interface lichenoid reaction, and civatte body formation. The dermis showed modest extravasation of red blood cells, swelling of the endothelial cells, and superficial and deep infiltration of polymorphonuclears and lymphocytes in the perivascular and perifollicular regions. (A–C) Periodic acid–Schiff staining only showed mild focal basement membrane thickening, and Alcian blue staining revealed marked interstitial dermal mucin in superficial and mid-dermis. (D and E) In brief, the superficial and deep perivascular-perifollicular inflammation with acute small vessel vasculitis, interface lichenoid reaction, and subcorneal-intraepidermal pustule formation were suggestive of collagen vascular diseases such as SLE.

TABLE 1 Cases of systemic lupus erythematosus with pustular lesions.

Author, year	Number of patients	Clinical manifestations
Schissler et al., 2017[12]	18	Amicrobial pustulosis of the folds
Shindo et al., 2019[4]	1	Generalized pustular psoriasis
Hannah et al., 2017[5]	1	Acneiform eruption
Martin et al., 2006[6]	1	Pustular vasculitis
Yin et al., 2012[7]	1	Wells' syndrome
Freire et al., 2017[8]	1	Subcorneal pustular dermatosis
Saulsbury et al., 1984[13]	1	Subcorneal pustular dermatosis
Larson et al, 2014[14]	1	Neutrophilic dermatosis

The pattern of pustular lesions in none of these cases was similar to that of our patients. The 24-year-old woman described by Hannah et al.[5] who had papules, pustules, and vesicles with hemorrhagic crusts in an annular pattern on the face, the frontal scalp, hard palate, and shins was the only example in which the lesions were almost similar to those of our patient. Nevertheless, in this case, there were dermis-centered scattered eosinophils in the pathology as well as a dense neutrophilic inflammatory infiltrate.[5] Thus, our patient's lesions were located elsewhere and had a distinct pathology.

On the contrary, two cases with subcorneal neutrophilic pustule development in the pathology and subcorneal pustular dermatosis as the clinical presentation were comparable to our case.[8,13] Pustular skin lesions were the presenting feature of SLE in one of these individuals.[8] It was intriguing to find that clinical alterations supporting SLE, such as dermal mucin deposition and lichenoid response, were present at the same time as subcorneal neutrophilic pustule development. There was also a polymorphonuclear infiltration in the superficial and deep dermis, as well as some lymphocytes.[8,13] In the case described by Martin et al., the purpuric bases on the limbs had pustular and necrotic lesions that showed pathological alterations in favor of pustular vasculitis as well as an increase in mucin levels in the dermis. SLE was detected by a serology study; however, the pathology report did not include any more SLE features.[6]

Prednisolone in addition to hydroxychloroquine was used to treat our patients. We discovered a dramatic improvement in annular pustular lesions and malar rash immediately after initiating this treatment regimen; nonetheless, the chilblain persisted for another 6 months.

We also did a thorough search in the literature to find similar cases of SLE in which cutaneous manifestations preceded the onset of the disease or cutaneous manifestations were presenting signs. APF was reported in 17 cases of SLE by Schissler et al.[12] In another case, SLE occurred 9 years after the diagnosis of APF.[15] Moreover, neutrophilic dermatosis might be the first indicator of underlying SLE in up to a third of patients.[14] Likewise, Hannah et al.[5] reported a neutrophilic dermatosis linked to SLE which presented as an acneiform eruption in a 24-year-old woman. Subcorneal pustular dermatosis has also been reported in a 6-year-old boy as a dermatological manifestation antecedent to the diagnosis of SLE.[8] This has been previously described by Saulsbury and Kesler as well.[13] In another study on 39 children, cutaneous manifestations were the presenting signs 28 (72%).[16]

4 | CONCLUSION

Cutaneous manifestations may appear before the onset of SLE or they may serve as the first symptoms. Pustules may form on the edge of SLE annular lesions; thus, they should not be overlooked. In these cases, further evaluations are warranted for the diagnosis of SLE.

AUTHOR CONTRIBUTIONS

Mehdi Gheisari: Supervision; writing – review and editing. **Moein Baghani:** Data curation; writing – original draft. **Raziyeh Ganji:** Data curation; writing – original draft. **Khatere Zahedi:** Data curation; writing – original draft; writing – review and editing.

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CONFLICT OF INTEREST STATEMENT

The authors confirm that there is no conflict of interest.

DATA AVAILABILITY STATEMENT

The corresponding author will provide the data that support the findings of this study upon reasonable request.

ETHICAL APPROVAL

This report has received ethics approval from the Ethics Committee of Shahid Beheshti University of Medical Sciences.

CONSENT

In compliance with the journal's patient consent policy, the patient's written informed consent was acquired for the publication of this report.

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