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Cutaneous Manifestations as Indicators of Systemic Autoimmune Diseases

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Abstract | **Review Article**

Cutaneous manifestations are frequently among the earliest clinical signs of systemic autoimmune diseases, often preceding or coinciding with internal organ involvement. As a readily accessible and observable organ, the skin serves as a valuable diagnostic window into systemic immune dysregulation. This review explores the most common and diagnostically relevant skin findings associated with systemic lupus erythematosus, dermatomyositis, systemic sclerosis, Sjögren's syndrome, mixed connective tissue disease, vasculitides, and related disorders. Key dermatologic clues—including malar rash, Gottron's papules, heliotrope rash, digital ulcers, Raynaud's phenomenon, livedo reticularis, and purpura—are discussed in relation to their pathophysiology and systemic implications. By highlighting these cutaneous features, this article underscores the importance of early dermatologic recognition in the diagnosis, classification, and management of systemic autoimmune conditions. Greater awareness among clinicians of these skin signs can lead to earlier diagnosis, more targeted laboratory evaluation, and improved patient outcomes through timely interdisciplinary care.

Keywords: Systemic autoimmune diseases (SAIDs), Cutaneous manifestations, Early diagnosis, Skin signs, Dermatologic recognition, Multi-organ damage.

INTRODUCTION

Systemic autoimmune diseases (SAIDs) represent a complex group of disorders characterized by aberrant immune responses directed against the body's own tissues, resulting in chronic inflammation and multi-organ damage. These conditions, which include systemic lupus erythematosus (SLE), systemic sclerosis, dermatomyositis, Sjögren's syndrome, and others, often present with diverse clinical features, making early diagnosis and management particularly challenging (Ferreira *et al.,* 2022). Among the various organ systems involved, the skin is frequently one of the earliest and most visibly affected, offering a critical window for early recognition of systemic disease.

Cutaneous manifestations are not only common in SAIDs, but they often precede systemic involvement, correlate with disease activity, and serve as useful indicators for disease classification, prognosis, and therapeutic decisions (Chang & Werth, 2020). In some cases, these skin signs may be the first—and sometimes the only—clues pointing to a potentially life-threatening systemic condition. As a result, dermatologic assessment plays a pivotal role in the diagnostic process and should not be overlooked in clinical evaluation.

The skin's accessibility allows for direct clinical examination, photographic documentation, dermatoscopic evaluation, and histopathological confirmation via biopsy. Unlike internal organ manifestations, which often require invasive procedures or advanced imaging techniques for detection, cutaneous signs are readily observable and can be evaluated repeatedly over time. This unique feature underscores the diagnostic and prognostic importance of skin manifestations in the context of autoimmune disease (Okawa *et al.,* 2015).

Historically, classification criteria for several SAIDs have incorporated dermatologic findings. For example, the American College of Rheumatology (ACR) and the European League Against Rheumatism (EULAR) criteria for SLE include malar rash, discoid rash, and photosensitivity as key diagnostic features (Merola *et al.,* 2019). Similarly, the presence of heliotrope rash and Gottron's papules in dermatomyositis, or sclerodactyly and telangiectasias in

systemic sclerosis, often directs clinicians toward specific diagnoses and serologic testing pathways (Callen, 2020; Varga & Denton, 2015).

Furthermore, the cutaneous involvement in SAIDs is often reflective of the underlying immunopathology. For instance, immune complex deposition, complement activation, vasculopathy, and lymphocytic infiltration all contribute to distinct dermatologic patterns (Veeramani *et al.,* 2023). Recognition of these patterns not only facilitates diagnosis but may also help in monitoring disease activity and response to treatment. In diseases such as lupus and dermatomyositis, cutaneous flare-ups may mirror systemic relapses, emphasizing the skin's role as a barometer of systemic disease burden (Maddukuri *et al.,* 2021).

Despite this, the dermatologic manifestations of autoimmune diseases are frequently under-recognized, particularly by non-dermatologists. A multidisciplinary approach—combining the expertise of dermatologists, rheumatologists, pathologists, and primary care providers—is essential to ensure timely identification and appropriate management of patients with SAIDs (Sunderkötter *et al.,* 2018).

This review aims to comprehensively examine the key cutaneous manifestations associated with major systemic autoimmune diseases, emphasizing their clinical significance, diagnostic utility, and implications for patient care. By enhancing awareness and understanding of these skin findings, clinicians across specialties can improve early detection, initiate prompt treatment, and ultimately enhance patient outcomes**.**

Cutaneous Clues in Systemic Autoimmune Diseases

The skin often serves as a mirror of internal disease, particularly in the setting of systemic autoimmune disorders. Early recognition of cutaneous signs may lead to timely diagnosis and treatment, potentially preventing severe complications. This section reviews the most relevant dermatologic manifestations associated with major systemic autoimmune diseases, emphasizing their clinical implications.

1. Systemic Lupus Erythematosus (SLE)

SLE is a prototypic multisystem autoimmune disease with a wide spectrum of cutaneous involvement. Skin manifestations occur in over 70% of patients and are categorized into lupus-specific and lupus-nonspecific lesions. The most classic is the malar rash, a fixed erythema over the cheeks and nasal bridge, often exacerbated by sunlight. Discoid lesions, which are chronic and scarring, typically appear on the face, scalp, and ears (Ferreira *et al.,* 2022). Photosensitivity, oral ulcers, and non-scarring alopecia are also included in diagnostic criteria (Chang & Werth, 2020). These features may correlate with disease activity and aid in classification and prognosis.

2. Dermatomyositis

Dermatomyositis is an idiopathic inflammatory myopathy characterized by specific skin findings that often precede or occur independently of muscle weakness. Hallmark signs include the heliotrope rash—a violaceous discoloration of the eyelids—and Gottron's papules, which are flat-topped violaceous papules over the knuckles (Callen, 2020). Other features include shawl sign, V sign, and mechanic's hands. The presence of cutaneous ulceration or calcinosis is often associated with more severe disease and increased malignancy risk (Maddukuri *et al.,* 2021). Skin biopsy and capillaroscopy may support diagnosis.

3. Systemic Sclerosis (SSc)

SSc involves progressive fibrosis of the skin and internal organs. Cutaneous features often dominate the early presentation, including Raynaud's phenomenon, sclerodactyly, and digital ulcers (Varga & Denton, 2015). Telangiectasias, calcinosis cutis, and mask-like facial tightening are also common. These findings not only reflect disease progression but also guide subclassification (diffuse vs. limited SSc). Nailfold capillaroscopy reveals characteristic vascular changes and is a useful non-invasive tool in evaluation (Cutolo *et al.,* 2018).

4. Sjögren's Syndrome

Although classically defined by sicca symptoms, Sjögren's syndrome may present with diverse skin findings. These include xerosis, annular erythema, palpable purpura, and cutaneous vasculitis (Ramos-Casals *et al.,* 2019). Vasculitic lesions are associated with cryoglobulinemia and systemic involvement. In some patients, annular erythema resembles subacute cutaneous lupus and may be misdiagnosed. Biopsy of purpuric lesions often reveals leukocytoclastic vasculitis (Seror *et al.,* 2021).

5. Mixed Connective Tissue Disease (MCTD)

MCTD is characterized by overlapping features of SLE, systemic sclerosis, and polymyositis, with high titers of anti-U1 RNP antibodies. Common cutaneous findings include Raynaud's phenomenon, malar-like rash, sclerodactyly, and telangiectasias (Kasukawa *et al.,* 2018). Digital swelling is often an early sign. Dermatologic overlap with lupus and scleroderma frequently complicates diagnosis and requires serological confirmation.

6. ANCA-associated Vasculitis

This group includes granulomatosis with polyangiitis and microscopic polyangiitis. Palpable purpura, livedo reticularis, ulcers, and nodules are common cutaneous features, particularly in small-vessel vasculitis (Sunderkötter *et al.,* 2018). Histologically, they show leukocytoclastic vasculitis, sometimes with granulomatous inflammation. Skin findings may precede systemic signs such as pulmonary or renal involvement, making early biopsy crucial for diagnosis.

7. Antiphospholipid Syndrome (APS)

APS is a prothrombotic disorder marked by vascular events and pregnancy morbidity. Cutaneous signs like livedo reticularis, digital gangrene, ulcers, and purpura may precede systemic thrombosis (Asherson & Cervera, 2003). Livedo racemosa is more specific than livedo reticularis and often suggests deeper vascular compromise. Skin findings in APS warrant prompt testing for antiphospholipid antibodies to prevent systemic complications.

8. Rheumatoid Arthritis (RA)

Although RA primarily affects joints, skin manifestations can occur, especially in patients with high disease activity. Rheumatoid nodules—firm, subcutaneous lumps—are typically found on pressure areas. Vasculitic ulcers, pyoderma gangrenosum, and neutrophilic dermatoses may also be seen in advanced cases (Molina-Ruiz & Requena, 2015). These skin findings are more frequent in seropositive RA and may signal extra-articular disease.

9. Psoriatic Arthritis

Psoriatic arthritis is associated with psoriatic plaques, typically with silvery scale on the scalp, elbows, and knees. Nail changes like pitting and onycholysis are common and correlate with joint involvement (Mease *et al.,* 2020). Cutaneous findings often precede arthritis and are essential for diagnosis. Dactylitis may be accompanied by erythema and swelling, and axial disease may manifest without skin lesions.

10. Behçet's Disease

Behçet's is a multisystem vasculitis that commonly presents with oral and genital ulcers, erythema nodosum-like lesions, and pseudofolliculitis (Davatchi *et al.,* 2017). The pathergy test—skin hyperreactivity to trauma—is a useful clinical sign. These cutaneous findings are not only diagnostic but also help assess disease activity and recurrence risk.

CONCLUSION

Cutaneous manifestations provide valuable insights into the presence, activity, and severity of systemic autoimmune diseases. Recognizing dermatologic signs such as malar rash, heliotrope eruption, digital ulcers, and vasculitic purpura can facilitate early diagnosis, appropriate serologic testing, and timely referral to specialists. Given their diagnostic significance, clinicians must maintain a high index of suspicion when evaluating patients with unexplained or persistent skin findings. Dermatologists, rheumatologists, and internists should work collaboratively to optimize outcomes and improve quality of life in patients with systemic autoimmune conditions.

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