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Cutaneous lupus subtypes

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ABSTRACT

A broad spectrum of dermatologic manifestations, known as cutaneous lupus erythematosus (CLE), may or may not be linked to the development of systemic disease. There are numerous subtypes of cutaneous lupus, such as acute cutaneous lupus (ACLE), subacute cutaneous lupus (SCLE), and chronic cutaneous lupus (CCLE). CCLE encompasses lupus tumidus, chilblain cutaneous lupus, LE profundus (LEP), and discoid lupus erythematosus (DLE). In order to diagnose these diseases, it is necessary to accurately classify the subtype. This is achieved through a combination of physical examination, laboratory studies, histology, antibody serology, and occasionally direct immunofluorescence, while also excluding systemic disease. The treatment of cutaneous lupus involves the provision of appropriate topical and systemic agents, as well as patient education regarding solar protection. In cases where the disease is pervasive, scarring, or treatment-refractory, systemic agents are recommended. In this chapter, we address the classification and diagnosis of the diverse subtypes of CLE and offer a comprehensive update on therapeutic management.

ARTICLE DETAILS

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Cutaneous lupus subtypes

Subtypes.

Subtype	Description	Key Features	Commonly Affected Areas
Acute Cutaneous Lupus Erythematosus (ACLE)	Often associated with systemic lupus (SLE).	Butterfly rash on the face, erythematous rash, photosensitivity.	Face, neck, upper chest, arms.
Subacute Cutaneous Lupus Erythematosus (SCLE)	Characterized by non-scarring rashes.	Annular or papulosquamous lesions, photosensitivity, may develop after sun exposure.	Shoulders, arms, neck, upper torso, face.
Chronic Cutaneous Lupus Erythematosus (CCLE)	Long-lasting and may result in scarring.	Scarring, dyspigmentation, atrophy. Includes several specific forms like DLE and hypertrophic lupus.	Scalp, face, ears, neck.
Discoid Lupus Erythematosus (DLE)	Most common form of CCLE, may lead to scarring.	Disc-shaped, erythematous plaques, scarring, pigmentation changes, alopecia in affected areas.	Scalp, face, ears, neck, and other sun-exposed areas.
Hypertrophic Lupus Erythematosus	A rare variant of DLE with thick, wart-like lesions.	Thick, verrucous (wart-like) lesions, hyperkeratotic plaques.	Face, neck, and extensor surfaces of the arms.
Lupus Panniculitis (Lupus Profundus)	Affects deeper layers of the skin, including subcutaneous fat.	Firm, deep nodules or plaques, can result in atrophy and scarring.	Face, upper arms, buttocks, thighs, breasts.
Tumid Lupus Erythematosus	Characterized by edematous, non-scarring plaques.	Smooth, non-scarring plaques, absence of surface changes, highly photosensitive.	Face, neck, upper trunk.
Chilblain Lupus Erythematosus	Triggered by cold exposure, presents with painful lesions.	Painful, purplish-red papules or nodules, exacerbated by cold and damp conditions.	Fingers, toes, ears, nose.

Acute cutaneous lupus erythematosus (ACLE) is a skin disease that frequently occurs in the third decade of life and is frequently accompanied by active systemic lupus erythematosus (SLE). There are both localized and generalized varieties of ACLE, including the malar rash, which is transient, sun-induced, and nonscarring. Malar rashes may be mistaken for acne rosacea and seborrheic dermatitis; however, the former is characterized by the development of pustules and papules, whereas the latter is located within the nasolabial creases.

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A'maculopapular rash of lupus' or 'photosensitive lupus dermatitis' is a more uncommon generalized form of lupus that manifests as a widespread, often pruritic eruption of symmetric macules and papules that are photosensitive and may resemble a drug rash. Patients may experience diffuse hair depletion, as well as associated mucosal ulcerations/apthae. Rowell's syndrome is the term used to describe lesions that resemble erythema multiforme in patients with ACLE or SCLE.

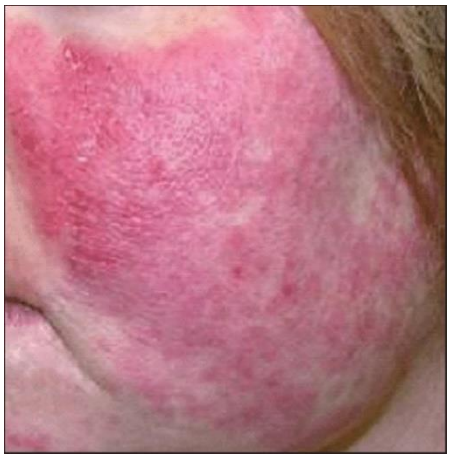

Figure 1. ACLE

Subacute cutaneous lupus erythematosus (SCLE) is predominantly a condition that affects young to middle-aged women. It is characterized by a high degree of photosensitivity, with 70-90% of patients meeting the ACR definition of aberrant photosensitivity. Annular and papulosquamous are the two morphologic variants of SCLE. The annular type is distinguished by scaly annular erythematous lesions, whereas the papulosquamous variant may resemble eczema or psoriasis, as well as pityriasis in certain cases.

SCLE lesions manifest in sun-exposed regions, such as the upper thorax, upper back, and extensor surfaces of the arms and forearms. Lesions generally do not develop below the pelvis, and the central face and cranium are typically spared. The cutaneous lesions do not indurate and resolve without scarring, although they may exhibit hypopigmentation reminiscent of vitiligo.

It is frequently necessary to differentiate between ACLE and SLE. Patients with SCLE typically exhibit only moderate systemic symptoms, with arthritis and myalgias being the most common. An estimated 50% of SCLE patients meet the criteria for SLE. 70% of SCLE patients are anti-Ro (SS-A) positive immunologically, and there is an overlap between SCLE and Sjogren's syndrome. In a 2012 population-based matched case-control study, the most frequently reported perpetrators were terbinafine, tumor necrosis factor-a inhibitors, antiepileptics, and proton pump inhibitors, which are more common in drug-induced SCLE than in other sub-types.

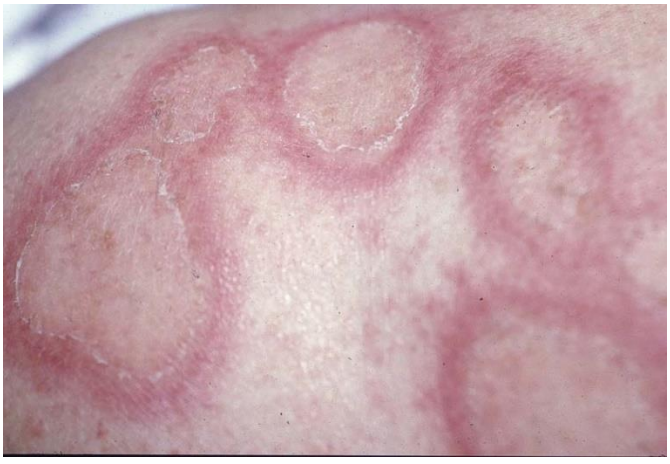

Figure 2. SCLE

DLE, LEP, CHLE, and LET are all components of chronic cutaneous lupus.

Lupus erythematosus (CLE) is a prevalent cutaneous condition that primarily impacts women in their fourth and fifth decades of life. In comparison to other CLE sub-types, it is more prevalent among women in their fourth and fifth decades of life and has a more benign medical course. Patients with generalized DLE are more susceptible to developing systemic disease than those with localized DLE. Localized DLE typically affects the head and neck, with a particular

emphasis on the cranium and ears, whereas generalized DLE is characterized by the extensor forearms and hands and can occur both above and below the neck.

DLE lesions initially manifest as a well-defined, scaly, erythematous macule or papule. These lesions progress to an indurated discoid plaque with an adherent scale that is excruciating to remove. Scarring alopecia is the final consequence of plaques that extend into the hair follicle. Typically, these lesions undergo atrophic development over time, characterized by central depigmentation and peripheral hyperpigmentation. Quamous cell carcinoma may manifest within a DLE lesion, and sun exposure or trauma can exacerbate a condition.

Psoriasis, lymphocytoma cutis, cutaneous T-cell lymphoma, granuloma faciale, polymorphous light eruption, and sarcoidosis. DLE lesions may resemble these conditions. Hypertrophic or verrucous DLE, an uncommon variant of DLE, is characterized by lesions that are exceedingly dense and develop on the face, hands, and extremities. Hyperkeratosis, dilated compact keratin-filled follicles, vacuolar degeneration of the basal keratinocytes, and an intensely inflammatory dermal infiltrate are all observed during a histologic examination of a protracted active DLE lesion. In comparison to other CLE sub-types, DLE patients exhibit a lower incidence of ANA, dsDNA, Sm, U1RNP, and Ro/SSA antibodies serologically.

Chilblain lupus (CHLE) is an uncommon variant of CCLE that resembles frostbite. Lesions manifest as painful, violaceous lesions and nodules in cold-exposed regions, and central erosions or ulcerations may develop on acral surfaces. CHLE is a condition that can be challenging to differentiate from frostbite and is characterized by a decrease in temperature

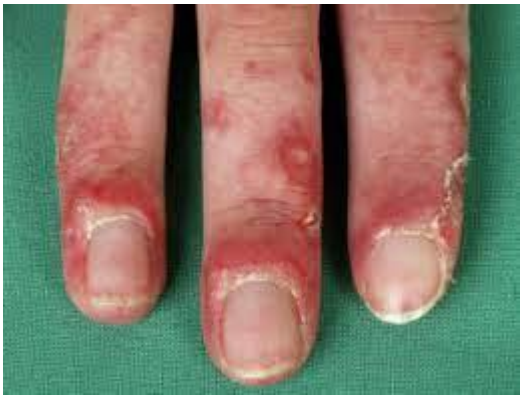

Figure 3. CHLE

A sub-type of CCLE, lupus erythematosus tumidus is characterized by a benign course and extreme photosensitivity, typically affecting males. Systemic agents, topical therapeutics, and sun protection comprise the treatment regimen, with antimalarials serving as the initial approach.

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Subtype	Topical Treatments	Systemic Treatments	Additional Recommendations
Acute Cutaneous Lupus Erythematosus (ACLE)	- Topical corticosteroids	- Antimalarials (e.g., hydroxychloroquine)	- Sun protection
	- Calcineurin inhibitors (e.g., tacrolimus)	- Systemic corticosteroids	- Patient education on sun avoidance
		- Immunosuppressants (e.g., methotrexate)	- Regular monitoring for systemic lupus erythematosus (SLE)
Subacute Cutaneous Lupus Erythematosus (SCLE)	- Topical corticosteroids	- Antimalarials (e.g., hydroxychloroquine)	- Sun protection
	- Calcineurin inhibitors (e.g., tacrolimus)	- Systemic corticosteroids	- Avoidance of photosensitizing drugs
		- Immunosuppressants (e.g., methotrexate, azathioprine)	- Regular monitoring for systemic involvement
Chronic Cutaneous Lupus Erythematosus (CCLE)	- Topical corticosteroids	- Antimalarials (e.g., hydroxychloroquine)	- Sun protection
(Including DLE, Hypertrophic Lupus, Lupus Panniculitis, Tumid Lupus, Chilblain Lupus)	- Calcineurin inhibitors (e.g., tacrolimus)	- Systemic corticosteroids	- Regular skin examinations
	- Intralesional corticosteroids	- Immunosuppressants (e.g., methotrexate, azathioprine, mycophenolate mofetil)	- Patient education on sun avoidance
	- Topical retinoids (for hypertrophic lupus)	- Thalidomide (in refractory cases)	- Treatment of any secondary infections
Discoid Lupus Erythematosus (DLE)	- Topical corticosteroids	- Antimalarials (e.g., hydroxychloroquine)	- Sun protection
	- Calcineurin inhibitors (e.g., tacrolimus)	- Systemic corticosteroids	- Regular monitoring for systemic involvement
	- Intralesional corticosteroids	- Immunosuppressants (e.g., methotrexate, azathioprine)	- Patient education on sun avoidance
		- Thalidomide (in refractory cases)	

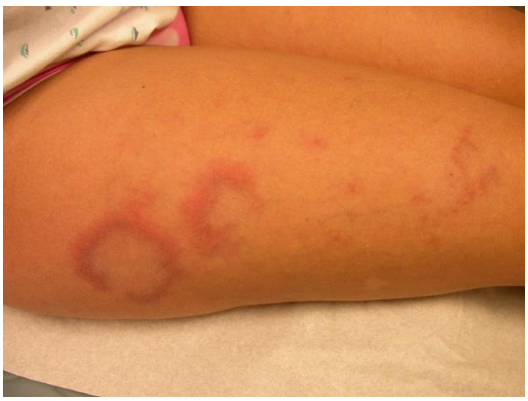

Figure 4. Tumid lupus

EMERGING THERAPIES

Anifrolumab, BIIB059, VIB7734, and iberdomide are emerging therapies that have been shown to be effective in certain forms of CLE through clinical trials. Potential prospective therapeutic targets of interest include other pathways implicated in lupus pathogenesis, including tyrosine kinase 2 (TYK2) and serine/threonine kinase IL-1R–associated kinase (IRAK4). In order to optimize the likelihood of demonstrating clinical benefit, it is crucial to carefully consider the design of clinical trials to evaluate therapies for potential use in CLE. This includes enrolling a sufficient number of patients with moderate to severe baseline disease activity, as evidenced by the skin-specific data from the baricitinib phase 2 and topical SYK inhibitor phase 1B trials. Skin-specific outcome measurements should be included in trials of SLE therapies, as CLE and SLE frequently exhibit disparate responses to therapies.

CONCLUSION

Understanding the diverse subtypes of cutaneous lupus erythematosus (CLE) is crucial for accurate diagnosis and effective treatment. CLE encompasses a range of dermatologic manifestations, from acute cutaneous lupus erythematosus (ACLE) and subacute cutaneous lupus erythematosus (SCLE) to chronic forms like discoid lupus erythematosus (DLE) and lupus tumidus. Each subtype has distinct clinical features, affected areas, and potential systemic associations. Diagnosing CLE requires a combination of physical examination, laboratory tests, histological analysis, antibody serology, and sometimes direct immunofluorescence. Treatment involves topical and systemic agents, with a strong emphasis on patient education regarding sun protection. More severe or treatment-resistant cases may necessitate systemic therapies. Comprehensive knowledge of CLE subtypes allows for tailored therapeutic strategies, ensuring better patient outcomes.

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