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Lupus erythematosus (LE)

By Fabiola Pabón-González, MD, Mariana Sadurní-García, MD, and Gerardo Caussade, MD

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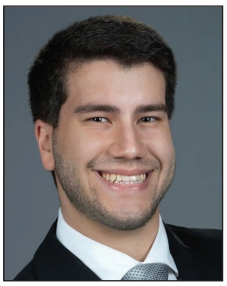

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1. Systemic and cutaneous forms

Disease	Key clinical description	Distribution	Pathology	Associations/ risk	Drugs/ triggers	Labs/ serologies	Treatment pearls
Systemic lupus erythematosus (SLE)	Multisystem autoimmune disease; flares correlate with systemic activity; 80% + skin manifestation; strong genetic component	Variable; skin + internal organs	Interface dermatitis; lupus band on non-lesional skin, mucin deposition in dermis	Female > male; ↑ in Black patients; flares in pregnancy	UV exposure, infections, smoking, estrogen, vitamin D deficiency	ANA (99%); anti-dsDNA (60%) can be used to monitor disease activity, anti-Smith (10-30%) is highly specific, ↓C3/C4	Varies with disease severity. Mild: Hydroxychloroquine (HCQ) + NSAIDs; Moderate-severe: prednisone + steroid-sparing agent; Severe: high dose prednisone vs. IV steroids + cyclophosphamide or MMF. Belimumab, rituximab or volcosporin if refractory
Acute cutaneous LE (ACLE)	Butterfly malar erythema; transient; spares nasolabial folds; photosensitive	Face > chest/ back; hands (knuckles spared)	Vacuolar interface dermatitis; sparse perivascular lymphocytic infiltrate in upper dermis; dermal mucin; DIF: + LBT	Cutaneous subtype with strongest association with SLE	UV exposure	Anti-dsDNA often ↑; SLE panel/ standard labs to search for EOD	Skin lesions respond by treating systemic disease; sun protection
Subacute cutaneous LE (SCLE)	Photosensitivity; no scarring after healing, but with dyspigmentation Two clinical variants: Annular (polycyclic annular plaques) or Papulosquamous (psoriasiform plaques); arthralgias in 70%	Lateral face (central face spared), neck, chest, upper back/ extensors	Epidermal atrophy, vacuolar interface dermatitis; BMZ thickening; <mucin deposition than ACLE; DIF: + LBT in 60–85%	Female > male; ↑ in white patients; 30-50% meet SLE criteria; Sjögren overlap; HLA-B8 (strongest genetic association), HLA-DR3	Thiazides, terbinafine, TNF-a inhibitors, PPIs, CCBs	Anti-Ro/ SSA (75–90%), anti-La/ SSB (30–40%), ANA speckled; associated with complement deficiencies	Antimalarials + sun protection; stop offending drug if one present
Chronic cutaneous LE (CCLE)	Persistent, typically scarring lesions; umbrella term (DLE, LET, panniculitis, etc.)	Head/neck > trunk	Dense perivascular and periappendageal infiltrates (findings vary by subtype)	Female > male; DLE are majority of CCLE, 5–20% of patients with DLE → SLE; ↑ in Black patients; smoking is a risk factor; UVR triggers (UVB>UVA)	UV exposure, smoking	Varies	Topical treatment, HCQ; escalate per CLE algorithm

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2. Chronic cutaneous lupus subtypes

Disease	Key clinical description	Distribution	Pathology	Associations/ risk	Drugs/ triggers	Labs/ serologies	Treatment pearls
Discoid lupus erythematous (DLE)	Erythematous macules or plaques → then develop induration, scale, atrophy, scarring with central hypopigmentation and peripheral hyperpigmentation; carpet "tack-like" spines on undersurface of scale; cicatricial alopecia + follicular plugging; dyspigmentation → common sequela	Face, scalp, ears; ± mucosa (25%)	Interface dermatitis; superficial and deep dermal inflammatory infiltrate; BMZ thickening; lacks eosinophils; orthrohyperkeratosis; follicular plugging; mucin; DIF: LBT + in 75%	↑ risk of progression to SLE	UV exposure, smoking	ANA + in 5–25%	HCQ ± topical therapy and intralesional triamcinolone; sun protection
Localized DLE	As DLE	Above neck only	As DLE	Low SLE risk (~5%)	—	—	Standard DLE care
Widespread/ Disseminated DLE	As DLE	Above and below neck	As DLE	↑SLE risk (~20%)	—	More serologic abnormalities	Systemic therapy; close follow-up
Hypertrophic (verrucous) lupus erythematous	Thick, verrucous hyperkeratotic plaques; indurated borders	Extensor forearms, face, upper half of body (sun-exposed areas)	Marked orthohyperkeratosis; endophytic buds of hyperplastic follicular epithelium; pseudoepitheliomatous hyperplasia mimics SCC	↑ SCC risk	—	—	Treat as DLE; retinoids useful in resistant disease
Lupus erythematosus tumidus (LET)	Edematous, firm, erythematous plaques (may have central clearing); non-scarring; no scale	Face, trunk	No epidermal change (lack interface dermatitis); notable perivascular and periadnexal inflammatory infiltrate within the dermis; massive dermal mucin	Similar to REM and Jessner's, but Jessner's displays CD8+ infiltrate predominance and no mucin. Tumid LE shares histology with REM but lacks reticular eruption on the mid-back and chest.	Photosensitivity	Often ANA -; increased IFN absent compared to SLE, DLE, and SCLE. Low prevalence of cutaneous lesions Ig deposition	Strong HCQ response; intralesional triamcinolone; photoprotection
Lupus panniculitis/profundus	Firm, tender, depressed subcutaneous nodules/plaques that heal with atrophy; overlying DLE findings in lupus profundus	Face, upper arms/trunk, thighs, breasts	Subcutaneous lobular lymphocytic panniculitis and hyaline (waxy pink) fat necrosis; dermal mucin; DIF: LBT + 35–70%	~15% w/ SLE; panniculitis may precede it	—	—	HCQ; follow CLE treatment algorithm

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Disease	Key clinical description	Distribution	Pathology	Associations/ risk	Drugs/ triggers	Labs/ serologies	Treatment pearls
Chilblain lupus erythematosus	Red to dusky papules and plaques on fingertips, ears, calves, and heels; chronic and relapsing course; arthralgias	Acral (fingers, toes, rims of ears, calves, heels)	Shows features of chilblains (papillary edema, perivascular lymphohistiocytic infiltrate) and DLE; DIF: + LBT	TREX1/ SAMHD1 mutations (familial)	Cold exposure	ANA ±	Warmth if cold exposure is related; HCQ; CLE protocol
DLE–lichen planus overlap	Overlap of DLE and lichen planus, often affecting palms and soles	Palmoplantar involvement	Features of lichen planus and lupus erythematous	Consider drug-induced LP-like eruption in SLE patients on antimalarials	Antimalarials (drug-induced type)	—	Treat both DLE and lichen planus
Mucosal lupus erythematosus	Usually seen with cutaneous DLE; plaque with central erythema & white keratotic rim (usually on hard palate); lip discoid lesions	Buccal mucosa, hard palate, vermillion lip (lower>upper)	Hyperkeratosis with vacuolar-lichenoid interface dermatitis and perivascular lymphocytes; DIF: + LBT	↑ SCC risk; ulcers increase risk of systemic involvement	—	—	Potent topical corticosteroid treatment; systemic involvement → HCQ

3. Special forms

Disease	Key clinical description	Distribution	Pathology	Associations/ risk	Drugs/ triggers	Labs/ serologies	Treatment pearls
Neonatal LE (NLE)	Develops during 1st week of life; photosensitive SCLE-like annular, polycyclic erythematous plaques; "raccoon eyes"; resolves ~6mo with dyspigmentation and telangiectasias but no scarring	Face, periorbital region and scalp mainly	As SCLE; DIF: (+) ~50%	Female predominance; cardiac abnormality 70% (3rd degree heart block in 30–40%), hepatobiliary disease	—	Mother anti-SSA/ Ro (52 and 60) ± SSB/ La; Infant almost always anti-SSA/RO and less frequently anti-U1RNP; cytopenias, ↑ LFTs	Sun protection + topical steroids; maternal steroids or HCQ may reduce fetal heart block risk, but established block is irreversible and most require pacemaker
Bullous SLE	Eruption of tense subepidermal bullae typically overlying erythematous, edematous base; rapid dapsone response	Sun & non-sun areas (face, neck, upper trunk, extremities); usually affects mucosa	Subepidermal bulla; neutrophils at DEJ; DIF: linear/ granular IgG,M,A/C3 along basal membrane; resembles neutrophil-rich bullous pemphigoid or epidermolysis bullosa acquisita (EBA)	Predominant in female and Black patients; SLE flare (must have diagnosis)	—	HLA-DR2; positive ANA (100%), dsDNA, Sm, Ro/La; antibodies against NC1/2 domains of VII collagen	Dapsone = response in 1-2 days (differentiates from EBA); immunosuppressants if refractory

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Disease	Key clinical description	Distribution	Pathology	Associations/ risk	Drugs/ triggers	Labs/ serologies	Treatment pearls
Rowell syndrome	Erythema multiforme-like targetoid lesions in the setting of DLE, ACLE, or SCLE	Face, trunk, limbs; depending on the setting (DLE, SCLE, ACLE)	Erythema multiforme and lupus erythematous changes	Depends on the setting of the syndrome (DLE, ACLE, or SCLE)	—	Anti-Ro/ SSA+, ANA+	Systemic corticosteroids
Toxic epidermal necrolysis-like lupus erythematosus	Severe epidermal necrosis secondary to lupus flare or trigger (e.g. UV radiation, drug); patient with underlying ACLE or SCLE	Sun-exposed areas	Epidermal necrosis	Photosensitivity; referred to as a form of acute syndrome of apoptotic pan-epidermolysis (ASAP)	UV exposure excess	ACLE and SCLE serologies	Supportive; control underlying lupus erythematous
Drug-induced SLE (DI-SLE)	Lupus-like syndrome; develops >1 year after initiating offending drug; minimal skin involvement	Systemic, minimal skin involvement except in the setting of TNF-a inhibitors	—	Systemic disease (fever, myalgias, weight loss, malaise, serositis), 90% have arthritis/arthralgias; lacks renal/CNS	Procainamide, hydralazine, quinidine, isoniazid, methyldopa, chlorpromazine, TNF-a inhibitors, minocycline	Anti-histone+ (95%); anti dsDNA– (except TNF-a inhibitors); minocycline (- anti-histone and + ANCA against MPO/elastase)	Resolve days to weeks after discontinuation of drug; symptomatic management

4. Complement deficiencies associated with lupus

Disease	Key clinical description	Distribution	Pathology	Associations/risk	Drugs/ triggers	Labs/ serologies	Treatment pearls
Primary C2 deficiency	Mild SLE (absent renal disease); adult onset; photosensitivity, SCLE-like lesions; ↑ infections (S. pneumonia)	—	Defective phagocytic clearance; UV-damaged apoptotic keratinocytes express antigens on cell surface	Female predominance; most common hereditary complement defect; most common cause of complement-associated SLE; however, only 10-20% with homozygous mutations develop SLE	—	CH50↓ (screening test); ANA low or absent; anti-Ro/ SSA+	Available vaccines for prevention of infections; standard SLE treatment if developed
Primary C1q/r/s & C4 deficiencies	Childhood onset; severe renal disease; palmoplantar keratoses (C4 deficiency only); risk of infection of encapsulated bacteria (*S. pneumonia*)	—	Impaired phagocytic clearance	High risk of developing SLE (C1q~90%)	—	CH50↓; low C1q/C4	SLE management; infection prophylaxis including vaccines
Anti-C1q autoantibodies	Arise in 30-50% with SLE; strong lupus nephritis link	—	Acquired antibodies to C1q	Acquired; seen in ~100% patients with hypocomplementemic urticarial vasculitis	—	Anti-C1q+	Treat SLE, nephritis, and/ or HUV

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Abbreviations: SLE = systemic lupus erythematosus; CLE = cutaneous lupus erythematosus; NLE = neonatal lupus erythematosus; ACLE = acute cutaneous lupus erythematosus; SCLE = subacute cutaneous lupus erythematosus; CCLE = chronic cutaneous lupus erythematosus; LBT = lupus band test; HCQ = hydroxychloroquine; UV = ultraviolet; EBA = epidermolysis bullosa acquisita; HUV = hypocomplementemic urticarial vasculitis; LET = lupus erythematosus tumidus; EOD = end organ damage; PPIs = proton pump inhibitors; CCBs =calcium channel blockers

References:

1. Alikhan, A., & Hocker, T. L. H. (2017). *Review of Dermatology* (1st ed.). Elsevier.

2. Bolognia, J. L., Schaffer, J. V., & Cerroni, L. (2024). *Dermatology: 2-Volume Set* (5th ed.). Elsevier.