SKIN MANIFESTATIONS IN SYSTEMIC LUPUS ERYTHEMATOSUS: A CASE REPORT AND REVIEW OF THE LITERATURE

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Introduction: The skin is the second most frequently affected organ in Systemic Lupus Erythematosus (SLE). Approximately 75% of the patients present cutaneous manifestations at some point during the course of the disease. We present a case of a patient with SLE and multiple skin lesions and a review of the available literature about this topic.

Case report: A 51-year-old obese male patient with a history of asthma was admitted to the hospital because muscle weakness associated with myalgia and skin lesions of recent onset. On physical examination, he presented weakness of proximal upper and lower limbs, scaly erythematous macules in elbows (**Figure 1**), retroauricular area and scalp and painful crusted erythematous-violaceous mucosal lesions on upper and lower lips (**Figure 2**). Laboratory results showed tricytopenia, accelerated ESR, increased CPK, proteinuria > 1g/24hs, ANA > 1/1280 with a homogeneous nuclear pattern, anti DNA > 1/320, hypocomplementemia and presence of lupus inhibitor. Pleural and pericardial effusion were observed on imaging studies. Skin biopsy reported interface dermatitis (**Figure 3**) and renal biopsy: Lupus Nephritis class IV-V. Treatment with high-dose glucocorticoids and cyclophosphamide was started, with a good response.

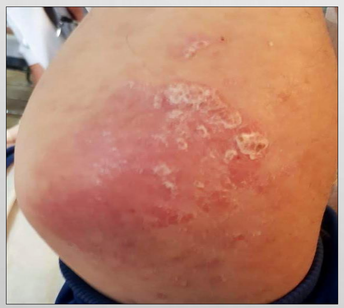


Figure 1: Scaly erythematous macules on the elbows.

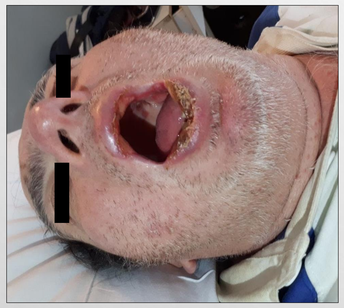


Figure 2: Erythematous-violaceous mucous lesions with crusts on the upper and lower lips.

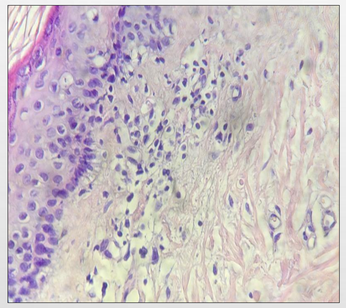
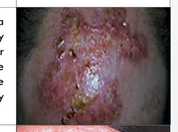
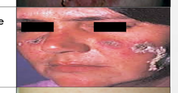
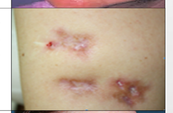
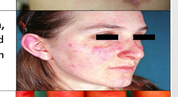
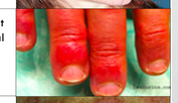
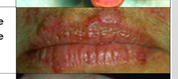
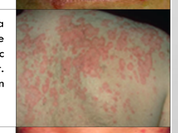
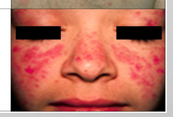


Figure 3: Skin biopsy: Mild spongiosis, vacuolar degeneration of the basal keratinocytes, and leukocyte exocytosis.

Table: Specific skin lesions of Cutaneous Lupus Erythematosus.

Cutaneous Lupus Erythematosus (CLE)		Clinical features	Histology	
CHRONIC CLE	*Discoid Lupus Erythematosus (DLE)*	It is the most common variant. It includes 2 forms: localized (head and neck) and generalized. It consists of erythematous plaques with follicular hyperkeratosis that progresses to atrophy with pigmentary changes and scars. It can cause scarring alopecia.	Lichenoid reaction pattern (interface dermatitis) and a superficial and deep dermal infiltrate of inflammatory cells, with a significant perianexal infiltrate. Vacuolar or hydropic degeneration of the basal layer with civatte bodies (apoptotic keratinocytes). DIF: thick Ig deposit in the basement membrane area in 50-90% of cases (particularly IgG and IgM).	
	Lupus Erythematosus Hypertrophic/ Verrucosus	It consists of thick, attached scales that are replaced by massive hyperkeratosis. They frequently are localized on the extensor surface of the forearms, face and upper trunk and present a torpid evolution.	Hyperkeratosis, follicular plugs, elongation of the rete ridges, irregular acanthosis and papillomatosis.	
	Lupus Profundus or Lupus Paniculitis	It consists of Inflammation of the adipose panniculus that when resolved develops deep depressions in the skin. It is characterized clinically by painful, inflammatory-looking nodules or plaques. The overlying skin can be normal or erythematous. It involves the proximal region of the extremities, shoulders, face, buttocks, scalp and less frequently, the trunk.	Lobular panniculitis with prominent lymphocytic infiltrate and mucin deposition between collagen bundles. Lymphocytic nuclear dust is frequently detected. Development of fibrosis is typically observed.	
	Lupus Erythematosus Tumidus or Intermittent CLE	It consists of erythematous plaques with an edematous appearance in photo-exposed areas that heal without scarring. They are asymptomatic or slightly itchy and recurrent.	Epidermal changes (atrophy, vacuolar degeneration, hyperkeratosis, follicular tamponade). Perivascular and perianexial lymphocytic infiltrate and abundant mucin deposition in dermis.	
	Lupus Pernio or Chilblain Lupus	It consists of erythema or purple plaques on the toes, heels, feet soles and ears. It ocurrs almost exclusively in women during winter.	Vacuolar degeneration and thickening of the basement membrane of the epidermis. Perivascular and perianexal lymphocytic infiltrate and abundant mucin deposit.	
	Mucosal Lupus	It affects oral, genital mucosa and lip semi-mucosa (Grinspan's sign). It is difficult to differentiate it from lichen planus and leukoplakia.	Hyperkeratosis with keratotic plaques, atrophy of the spinous layer, deep inflammatory infiltrate, edema of the lamina propria.	
SUBACUTE CLE (SCLE)		It consists of erythematous macules or papules which evolve into either scaly papulosquamous psoriasiform lesions or annular patches and plaques (papulosquamous or psoriasiform variant and annular variant respectively). Mixed forms are also found. It can symmetrically affect photoexposed areas that heal without scarring. Other forms include vesiculobullous erythema, exfoliative erythroderma, lupus gyratum repens and toxic epidermal necrolysis-like SCLE.	Basal vacuolar changes, epidermal atrophy, dermal edema and superficial mucin deposition are usually more intense than in DLE. Pauci-inflammatory, vacuolar, lymphocytic interface dermatitis. Civatte bodies are quite prominent. The positive lupus band test (60%), is usually less thick when compared to DLE.	
ACUTE CLE (ACLE)		It is the classic butterfly erythema generally associated with SLE. It consists of erythematosus macules, papules and plaques in central areas of the face that respect nasolabial folds and periorbital regions. It can affect other photoexposed areas and heals without leaving sequelae.	Vacuolar changes involving the basal keratinocyte cell layer. Edema, small hemorrhages and a mild infiltrate of inflammatory cells, principally lymphocytes, are seen in the upper dermis. Thickening of the basement membrane.	

DIF: Direct immunofluorescence.

Discussion: According to Gilliam and Sontheimer classification, the cutaneous manifestations of Lupus Erythematosus (LE) are divided into specific and nonspecific lesions based on histopathological findings. Specific lesions are characterized by interface dermatitis and nonspecific lesions don't present a distinctive histological pattern and aren't exclusive of cutaneous LE. They include photosensitivity, diffuse alopecia, nail abnormalities, splinter hemorrhages, livedo reticularis, livedo racemosa, Raynaud's phenomenon, periungual telangiectasias, gangrene, purpura and vasculitis. . Clinical and histological characteristics of the specific lesions of cutaneous LE are described in the table.

Conclusion: Knowing the broad spectrum of skin manifestations of SLE allows establishing an accurate diagnosis and adequate treatment.

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