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The red face revisited: Connective tissue disorders

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Abstract Red face is not a rare finding in patients with connective tissue disorders. The malar eruption is the most frequent cutaneous manifestation of systemic lupus erythematosus (LE). This condition is more apparent among fair-skinned individuals, and it usually appears after sun exposure. A very important clinical sign is that nasolabial folds remain free of any erythematous or other changes. With subacute cutaneous LE, sun exposure can provoke a red face that resembles the malar eruption of systemic LE. The typical clinical findings of chronic cutaneous LE are the discoid lesions. There is a clinical form of chronic cutaneous LE called *erythema perstans faciei.* This form is purely erythematous, and it usually appears on the face. Other rare "red face" forms of chronic cutaneous LE are LE tumidus and LE telangiectaticus.

Red face is not typical of systemic sclerosis, but facial telangiectasias are frequent, especially with CREST (calcinosis, Raynaud phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia) syndrome. The differential diagnoses of other red face manifestations are easy due to the additional findings. Telangiectasias are accompanied by calcinosis, sclerodactyly, digital ischemia, and Raynaud disease. Many studies mention telangiectasias as markers of the severity of the systemic sclerosis, the disease duration, any pulmonary arterial hypertension, and any esophageal involvement.

Purple- or violet-colored upper eyelids are the hallmark and one of the first clinical signs that is helpful for the diagnosis of dermatomyositis. This violaceous to dusky erythema can extend over the whole face and the upper aspects of the trunk. Erythematous changes on the face that are different from those of the heliotrope sign which occurs with dermatomyositis may be observed in both sun-exposed skin and non–sun-exposed skin. Malar and facial erythema, linear extensor erythema, V-sign or shawl sign, and other photodistributed eruptions can also appear.

Red face with lupus erythematosus

The skin lesions of patients with lupus erythematosus (LE) can present as lupus-specific or lupus-nonspecific findings. Lupus-specific skin changes occur with chronic cutaneous, subacute cutaneous, and acute cutaneous LE.[1] Localized acute LE has commonly been referred to as the classical "butterfly eruption."[2]

Systemic lupus erythematosus

Systemic LE (SLE) involves cutaneous changes that are listed in the criteria established by the American Rheumatology Association.[3] These include malar eruption, discoid lesions, photosensitivity, and oral mucosal lesions.

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The malar eruption is the most frequent cutaneous manifestation of SLE. It is an erythematous and very slightly edematous lesion that is located on both cheeks and across the bridge of the nose. It is more obvious among affected fair-skinned individuals, and it usually appears after sun exposure. These lesions will then disappear, and they leave no scars or pigmentation. Malar eruption may be mistaken for sunburn, the flush of rosacea, or photosensitive dermatitis. If the erythema persists for a few weeks or if new erythema appears elsewhere, the true diagnosis is the malar eruption of SLE. A very important clinical sign is that the nasolabial folds remain free of any skin changes; this is the main clinical differentiation between malar eruption and seborrheic dermatitis. The differentiation of the malar eruption sign of SLE from other connective tissue diseases may be very difficult and sometimes impossible. Clinical differential diagnoses include—in addition other conditions—telangiectatic rosacea, superficial fungal infections, photosensitive dermatitis, erythema emotivum, rubeosis diabeticorum, and the eruption of some viral infections. The histological findings depend on the type of clinical representation. The main pathologic findings are thinning of the epidermis; hydropic degeneration of the basal layer with disruption of the dermoepidermal junction; edema of the dermis in upper portion; sparse infiltration of lymphocytes in most of the dermis; and fibrinoid degeneration of the connective tissue. With direct immunofluorescence, deposits of immunoglobulins G and M, as well as of C3, are detected at the basal membrane zone; these deposits are found both in lesions and in uninvolved skin. The histopathologic findings are compatible with both SLE and dermatomyositis, but direct immunofluorescence is negative in patients with dermatomyositis.

Subacute cutaneous lupus erythematosus

Patients with subacute cutaneous LE have disseminated, nonscarring, photodistributed eruptions. There are two major subtypes of the disease: erythematous annular, which resembles erythema annulare centrifugum, and erythematous squamous, which resembles psoriasis or lichen planus. Subacute cutaneous LE belongs to the group of well-defined photodermatoses.

Sun exposure can provoke a red face that resembles the malar eruption of SLE,[4] and it may also provoke systemic involvement in some patients. This condition also involves a differential diagnosis that includes rosacea, photodermatitis, rubeosis, erythema emotivum, dermatomyositis and so on.

Chronic cutaneous lupus erythematosus

Chronic cutaneous LE is limited to the skin only. The typical lesions of this condition are the discoid lesions. Because these lesions are the most common skin changes, sometimes the term *chronic discoid LE* is used as a synonym for *chronic cutaneous LE*. The discoid lesions begin with red papules or maculae that are covered with silvery scales. A closer view of the scales shows follicular keratosis, which is a very important clinical finding for this disease. Later, atrophy and scarring appear after the active stage of the disease.

The histology of chronic cutaneous LE includes the following findings: atrophic epidermis, compact hyperkeratosis with follicular plugging, and hydropic degeneration of the cells of the basal layer. In the upper portion of the dermis, there are edema and mucin; during the later stages of the disease, sclerosis, dilatation of the blood vessels, and dense lymphocytic infiltrate around the vessels of the upper and deep vessels and the adnexa are also present. With the use of direct immunofluorescence, deposits of immunoglobulins G and M as well as C3 are detected at the basal membrane zone, but these are found in the skin only.[5]

There is a clinical form of the chronic cutaneous LE called *erythema perstans faciei*. This form is purely erythematous, and it usually appears on the face. The lesions are erythematous macules or plaques with little scaling and very little edema. There is a lack of severe inflammation, and the pathologic process is superficial. The lesions disappear with minimal atrophy. The acute stage of these lesions resembles the malar eruption of SLE, and it is very difficult to differentiate between these conditions.[6]

Some rare variants of chronic cutaneous LE present the clinical picture of a red face. LE edematosus has been described as erythematous and edematous, without well-demarcated plaques, follicular plugging, or scarring.[7] The lesions are sometimes multiple, and they regress without residual changes. The diagnosis may be difficult because the histological and immunologic findings are not always compatible with chronic cutaneous LE.

Another rare form of chronic cutaneous LE is LE tumidus (Figure 1). This rare form is characterized by erythematous, succulent, edematous, nonscarring plaques in sun-exposed areas.[8] Follicular plugging and scarring are usually absent. The lesions can be widespread; they affect all photoexposed areas (ie, back, neck, arms, and shoulders), but they occupy the face predominantly. They sometimes disappear spontaneously, but they can also coalesce to form annular plaques that resemble the annular type of subacute cutaneous LE. Patients with LE tumidus never show systemic involvement. This is one of the most photosensitive subtypes within the spectrum of LE, and Ro/SSA antibodies are detected in all patients. Due to the peculiarities of the clinical and histopathological findings, this variant needs to be differentiated from polymorphous light eruption and Jessner lymphocytic infiltration. Additional differentiation from reticular erythematous mucinosis is also required.

LE telangiectaticus is another rare form of chronic cutaneous LE (Figure 2). The most important clinical feature of this variant is erythema. A closer view reveals that this erythema is caused by telangiectasias. The lesions involve the face, and they can also occur on the extremities and the back. The telangiectasias are usually reticulate,[9] and follicular plugging and scarring are usually absent. The most important clinical differential diagnosis of this subtype