Rheumatologic Skin Disease

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KEYWORDS

• Rheumatology • Dermatology • Skin • Lupus • Dermatomyositis • Morphea

KEY POINTS

- The cutaneous presentation of lupus is variable and attention to the clinical features assists in defining the chances of systemic disease.
- In dermatomyositis serologic studies help identify the associated conditions of lung disease and malignancy.
- Photoprotection and topical therapy are important therapeutic interventions in lupus and dermatomyositis.
- Clinical findings can differentiate morphea from systemic sclerosis. An important association with morphea is the common co-occurrence of genital lichen sclerosus.

LUPUS ERYTHEMATOSUS

Lupus is a chronic autoimmune disease with the potential for multiorgan disorder and prominent involvement of the skin. There is great variety in the clinical features and severity from one patient to the next. There are several distinct and recognizable patterns of skin involvement that are specific for lupus.

Patient History

Lupus preferentially affects young women but can occur at any age. The most common clinical presentation involves skin rashes and constitutional symptoms, with fatigue and musculoskeletal complaints predominating.[1,2] The skin manifestations of lupus are classified as acute cutaneous lupus, subacute cutaneous lupus, and chronic cutaneous lupus. Chronic cutaneous lupus includes several subtypes, the most common being discoid lupus.[3] Less common forms of chronic cutaneous lupus are lupus panniculitis, chilblain lupus, and tumid lupus. Some controversy exists about the relationship of tumid lupus to the rest of the lupus spectrum.[3]

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Lupus is characterized by relapses and remissions. It is expected that the clinical pattern that develops early in the disease will predominate over the course of the illness. Recognition of the type of skin disease can help predict systemic disease because the subsets of cutaneous lupus relate to systemic disease differently (Fig. 1). Patients with lupus also present with skin findings that are not specific to lupus, and these include pernio, vasculitis, photosensitivity, alopecia, livedo, and bullous lesions.

It is important to carefully consider medications, because they can be a trigger for lupus. Subacute cutaneous lupus erythematosus (SCLE) is a form of lupus commonly attributed to medicines. More than 40 medications are reported to cause SCLE; of these, the most common are hydrochlorothiazide, diltiazem, angiotensin-converting enzyme inhibitors, and terbinafine.[4] In recent years it has been recognized that the tumor necrosis factor (TNF) inhibitors cause multiple presentations of autoimmunity, including lupus.[5,6] Patients often have systemic and cutaneous findings, although the systemic findings may be more prominent. In contrast with drug-induced lupus caused by procainamide, hydralazine, and minocycline, antihistone antibodies may not be present. Drug-induced forms often resolve when the offending medications have been stopped, although it may take months.

Clinical Findings

Acute cutaneous lupus

This condition presents as classic malar erythema, known as a butterfly rash (Fig. 2). It is present in about 40% to 50% of patients at the diagnosis of systemic lupus.[2] Erythema spreading over the cheeks and nose, sparing the sun-protected areas like the nasolabial fold, is characteristic. There can be extension onto the forehead and chest. In a few patients a more generalized eruption accompanies this rash, involving the extensor arms and hands and often localized to the interphalangeal skin and sparing the skin over the knuckles. Erythema and small papules tending toward confluence are present. Small amounts of scale may be found. The clinical course of the rash can worsen with sun exposure or reappear with systemic disease flares.

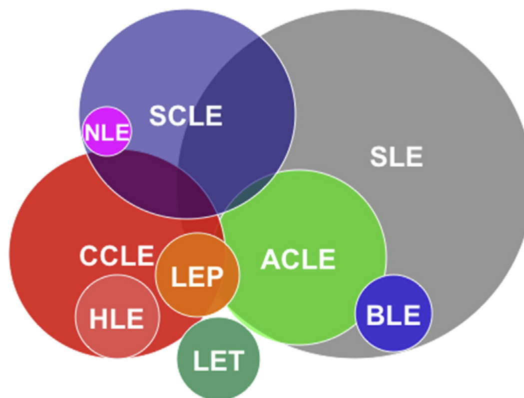

Fig. 1. Relationship of lupus subsets to systemic disease. ACLE, acute cutaneous lupus erythematosus; BLE, bullous lupus erythematosus; CCLE, chronic cutaneous lupus erythematosus; HLE, hypertrophic lupus erythematosus; LEP, lupus erythematosus profundus; LET, lupus erythematosus tumidus; NLE, neonatal lupus erythematosus; SCLE, subacute cutaneous lupus erythematosus; SLE, systemic lupus erythematosus. (*Courtesy of* J. Callen, MD, Louisville, KY.)

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