

Cutaneous Manifestations in Systemic Autoimmune Diseases

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As the largest and most accessible organ of the body, the skin offers many clues in the diagnosis of autoimmune diseases. Awareness that various alterations in the integument of the skin may be an indication of systemic disease is obviously important in the care of the total patient.

Perhaps the classic autoimmune disease, which has an important dermatologic component, is systemic lupus erythematosus (SLE). Four of the eleven criteria for SLE established by the ACR involve skin changes (butterfly rash, discoid lupus, photosensitivity, and nasopharyngeal ulcers) and skin manifestations may be the presenting sign of systemic lupus in over 20% of patients so affected. Nevertheless, acute involvement of the skin from lupus requires attention to other organ systems (i.e., CNS, or renal). Involvement of these organs may be life threatening. Typical skin involvement is variable. Classically but rarely, patients may have abrupt onset of a malar rash, which initially looks urticarial, but later becomes hyperpigmented. Other dermatologic manifestations include lupus panniculitis with deep firm nodules, which may ulcerated, dermal vasculitis, livedo reticularis, leg ulcers, and rheumatoid nodules. An intermediate form of lupus, which falls between discoid and SLE, is subacute cutaneous LE. The skin lesions are photosensitive and occasionally psoriasiform associated with minimal scarring.

Skin manifestations also represent some of the common signs and pathologies that are noted in vasculitis. Hypersensitivity angitis or leukocytoclastic angitis is the major vasculitis syndrome involving skin. Systemic features are often important aspects of these diseases. Different kinds of vasculitis may originate from different sizes of vessel involvement and presented with different skin features.

Dermatomyositis, an immune-mediated disorder of unknown etiology that could associate with a malignancy, consists of an inflammatory myopathy combined with characteristic cutaneous findings. Some patients with dermatomyositis also have clinical features that overlap with those of SLE or scleroderma. Manifestations of scleroderma range from cutaneous involvement alone to multisystem internal disease. Morphea, progressive systemic sclerosis, eosinophilic fasciitis, eosinophilia myalgia syndrome, and mixed connective tissue disease may present with characteristic features of skin involvement.

Patients with systemic onset of juvenile rheumatoid arthritis (Still's disease) and adult Still's disease frequently develop erythematous macules that may be migratory and transient, typically occurring briefly in late afternoon or early evening and often associated with a temperature elevation. These lesions usually occur on the trunk and proximal extremities. They may be surrounded by a zone of pallor, and larger lesions can have areas in the center that are uninvolved, in addition, the skin manifestations of Sjogren's syndrome and seronegative spondyloarthritis will be discussed as well.