TABLE 79-1 CLINICAL MANIFESTATIONS OF SYSTEMIC LUPUS ERYTHEMATOSUS

CONSTITUTIONAL	**NEUROPSYCHIATRIC**
Fatigue	Seizures*
Fever	Cerebritis/aseptic meningitis
Lymphadenopathy	Cerebrovascular disease
Weight loss	Transverse myelitis
Anorexia	Chorea
MUCOCUTANEOUS	Headache
Oral, genital, nasal ulcers*	Cognitive impairment
Angioedema	Autonomic dysfunction
Alopecia	Cranial neuropathy
Photosensitivity*	Peripheral neuropathy
Malar (butterfly) rash*	Psychosis*
Discoid lesions*	Anxiety
Subacute cutaneous lupus	Depression
Tumid lupus	Mood disorder
Panniculitis	**HEMATOLOGIC**
Vasculitis	Leukopenia*
Chilblain	Lymphopenia*
Urticaria	Hemolytic anemia*
Periungual erythema	Nonhemolytic anemia (anemia of chronic disease, iron-deficiency)
MUSCULOSKELETAL	Thrombocytopenia*
Arthritis*	Antiphospholipid antibody syndrome
Arthralgias	**VASCULAR**
Jaccoud's arthropathy	Raynaud's phenomenon
Avascular necrosis	Livedo reticularis
Myositis	Arterial or venous thrombosis
CARDIAC	Vasculitis (almost any location)
Pericarditis*	**OCULAR**
Pericardial effusion	Keratoconjunctivitis sicca
Myocarditis	Episcleritis
Valvular thickening	Scleritis
Libman-Sacks endocarditis	Retinal vasculitis
Atherosclerotic heart disease	Arterial and venous occlusions
PULMONARY	Optic neuritis
Pleuritis*	**GASTROINTESTINAL**
Pleural effusion	Hypomotility
Pneumonitis	Mesenteric vasculitis
Alveolar hemorrhage	Malabsorption
Interstitial lung disease	Protein-losing gastroenteropathy
Bronchiolitis obliterans	pancreatitis
Pulmonary hypertension	Lupus enteropathy
Pulmonary emboli	Thrombosis of mesenteric and hepatic vasculature
Vasculitis	Hepatitis
Acute reversible hypoxemic syndrome	Hepatomegaly
Shrinking lung syndrome	Splenomegaly
RENAL	Pancreatitis
Cellular casts or glomerulonephritis*	Acalculous cholecystitis
Proteinuria or membranous nephropathy or nephrotic syndrome*	Peritonitis
	SEROLOGIC
	Autoantibodies*
	Hypocomplementemia
	Elevated acute phase reactants
	REPRODUCTIVE
	Recurrent spontaneous abortions
	Premature fetal delivery
	Neonatal lupus

*An item in the 1997 American College of Rheumatology classification criteria for SLE.

noninfective Libman-Sacks endocarditis are frequently observed on echocardiography and autopsy studies. Myocarditis should be suspected in patients with cardiopulmonary symptoms and fever.

Renal Manifestations

Nephritis, which manifests with hematuria and proteinuria, is a major cause of morbidity and mortality for SLE patients. The International Society of Nephrology/Renal Pathology Society (ISN/RPS) revisited the 1982 World Health Organization classification of lupus nephritis (classes I through VI). ISN/RPS class IV (i.e., diffuse, proliferative) lupus nephritis is the most common form and has the worst prognosis, but it is also the most amenable to aggressive immunosuppressive therapy.

Neuropsychiatric Manifestations

Neuropsychiatric symptoms are broad, including sensorimotor neuropathy, headache, cognitive dysfunction, mood disorders, psychosis, life-threatening ischemic stroke, cerebritis, and transverse myelitis. In 2010, the European League Against Rheumatism task force recognized that "mild or moderate cognitive dysfunction is common in SLE" and recommended the "management of SLE and non–SLE-associated factors, as well as psychoeducational support to prevent further deterioration of cognitive function."

Hematologic Manifestations

Leukopenia, primarily lymphopenia, anemia, and thrombocytopenia are common in SLE. Anemia is typically results from hemolysis or chronic disease. Antiphospholipid antibodies (APAs) are detected in approximately 33% of SLE patients and are associated with recurrent thromboses, thrombocytopenia, and recurrent spontaneous miscarriages.

Vascular Manifestations

More than 40% of SLE patients have Raynaud's phenomenon. Venous clots (e.g., pulmonary emboli, deep vein thromboses) and arterial clots typically result from APAs. Leg ulcers, gangrene, thrombophlebitis, nail fold infarcts, cutaneous necrosis, and necrotizing purpura may also be seen. Small vessel vasculopathy or vasculitis can occur in any organ system and can be a life-threatening manifestation.

Ocular Manifestations

Keratoconjunctivitis sicca from secondary Sjögren's syndrome (see Chapter 85) is the most common ocular manifestation of SLE. Episcleritis, scleritis, and retinal vasculitis can occur but are less common.

For a deeper discussion of these topics, please see Chapter 266, "Systemic Lupus Erythematosus" in Goldman-Cecil Medicine, *25th Edition.*

DIAGNOSIS AND DIFFERENTIAL DIAGNOSIS

SLE is a clinical diagnosis; no single test or feature is definitively diagnostic of the disease. The clinical disease of most patients evolves over time, and, in most situations, only after several years (and several different physicians' visits) are patients recognized as having SLE.