Systemic lupus erythematosus

Systemic lupus erythematosus (SLE) is a chronic inflammatory autoimmune disease associated with autoantibody production in response to nuclear and cytoplasmic antigens. SLE has protean manifestations and follows a relapsing and remitting course. More than 90% of cases of SLE occur in women, frequently starting at childbearing age.

SLE can involve almost any organ and may present in many different ways. This can make it difficult to establish a diagnosis and an extensive work-up may be needed to determine the full extent of involvement and to exclude other possible etiologies for the manifestations.

Other subtypes of SLE:

- Pediatric systemic lupus erythematosus
- Systemic lupus erythematosus nephritis or Lupus nephritis
    - Lupus nephritis is clinically evident in 50-60% of patients with systemic lupus erythematosus (SLE), and it is histologically evident in most SLE patients, even those without clinical manifestations of renal disease.
- Bullous systemic lupus erythematosus (BSLE)
    - Bullous systemic lupus erythematosus is an autoantibody-mediated subepidermal blistering disease that occurs in patients with systemic lupus erythematosus.
- Drug-Induced Lupus
    - About 10% of cases of SLE can be attributed to drugs.
    - TNF-α inhibitors, such as adalimumab, etanercept, and infliximab.
    - calcium channel antagonists, thiazide diuretics, ACE inhibitors, and terbinafine.
    - Minocycline, procainamide and hydralazine

Signs and symptoms

SLE can affect almost any organ system, although it mainly involves the skin, joints, kidneys, blood cells, and nervous system. Its presentation and course are highly variable, ranging from indolent to fulminant.

More common features include involvement of the skin and mucus membranes, joints, kidneys, CNS, serous membranes, cardiovascular system, and hematologic cell lines.

Fatigue and depression are frequent symptoms and can adversely affect quality of life. Arthritis or arthralgias are experienced by 83% to 95% of patients with SLE. SLE may present differently in men and women. For example, men tend to get SLE at an older age and are more likely to have renal and hematologic involvement, but have fewer dermatologic features. Race and ethnicity may also affect the specific manifestations.

In **childhood**-onset SLE, several clinical symptoms are more commonly found than in adults, including malar rash, ulcers/mucocutaneous involvement, renal involvement, proteinuria, urinary cellular casts, seizures, thrombocytopenia, hemolytic anemia, fever, and lymphadenopathy.

In **adults**, Raynaud pleuritis and sicca** are twice as common as in children and adolescents. In a **woman** of childbearing age, the classic presentation of a triad of [1]fever, [2]joint pain, and [3]rash should prompt investigation into the diagnosis of SLE.

Acute emergencies in patients with systemic lupus erythematosus (SLE) include the following:

- Severe neurologic involvement
- Systemic vasculitis
- Profound thrombocytopenia with a thrombotic thrombocytopenia (TTP)–like syndrome
- Rapidly progressive glomerulonephritis
- Diffuse alveolar hemorrhage

Diagnosis

The diagnosis of SLE is based on a combination of clinical findings and laboratory evidence. Familiarity with the diagnostic criteria helps clinicians to recognize SLE and to subclassify this complex disease based on the pattern of target-organ manifestations.

The American College of Rheumatology (ACR) criteria, proposed summarize features that may aid in the diagnosis. ***The presence of 4 of the 11 ACR criteria may be enough to diagnose SLE.*** Patient is they classified as having SLE in the presence of biopsy-proven lupus nephritis with antinuclear antibodies (ANA) or anti–double-stranded DNA (anti-dsDNA) antibodies or if 4 of the diagnostic criteria, including at least 1 clinical and 1 immunologic criterion, have been satisfied.

ACR mnemonic of SLE diagnostic criteria

The following are the ACR diagnostic criteria in SLE, presented in the "***SOAP BRAIN MD***" mnemonic:

- **S**erositis
- **O**ral ulcers
- **A**rthritis
- **P**hotosensitivity
- **B**lood disorders
- **R**enal involvement
- **A**ntinuclear antibodies
- **I**mmunologic phenomena (eg, dsDNA; anti-Smith [Sm] antibodies)
- **N**eurologic disorder
- **M**alar rash
- **D**iscoid rash

Treatment

Management of SLE often depends on the individual patient's disease severity and disease manifestations, although hydroxychloroquine has a central role for long-term treatment in all SLE patients.

Medications used to treat SLE manifestations include the following:

- Nonbiologic DMARDS:
    - Azathioprine, mycophenolate, cyclosporine
    - Chemotherapeutic agents: cyclophosphamide, methotrexate
    - Antimalarials (eg, hydroxychloroquine)
- Biologic DMARDs
    - Belimumab
    - Rituximab (off-label)
    - IV immune globulin
- Corticosteroids: short-term use recommended
    - eg, methylprednisolone, prednisone
- NSAIDS
- Vitamin D may be useful
- Using sunscreens

Antiphospholipid Syndrome

In patients with systemic lupus erythematosus (SLE), the presence of antiphospholipid antibodies is common. Therefore, it is important to evaluate these patients for risk factors for thrombosis, especially in pregnancy. ow-dose aspirin in individuals with SLE and antiphospholipid antibodies is potentially useful for primary prevention of thrombosis and pregnancy loss.

Secondary prevention of thrombosis in nonpregnant patients with SLE and thrombosis associated with antiphospholipid syndrome can be managed with long-term use of oral anticoagulants. In pregnant patients with SLE and antiphospholipid syndrome, unfractionated or low-molecular-weight heparin and aspirin may reduce the risk of pregnancy loss.

Laboratory testing (FYI)

The following are useful standard laboratory studies when SLE is suspected:

- CBC with differential
- Serum creatinine
- Urinalysis with microscopy

Other laboratory tests that may be used in the diagnosis of SLE are as follows:

- ESR or CRP level
- Complement levels
- Liver function tests
- Creatine kinase assay
- Spot protein/spot creatinine ratio
- Autoantibody tests

Imaging studies (FYI)

The following imaging studies may be used to evaluate patients with suspected SLE:

- Joint radiography
- Chest radiography and chest CT scanning
- Echocardiography
- Brain MRI/MRA
- Cardiac MRI

Procedures (FYI)

Procedures that may be performed in patients with suspected SLE include the following:

- Arthrocentesis
- Lumbar puncture
- Renal biopsy

*** Sicca means dryness, and as a term, it may refer to:*

- *Sjögren syndrome*
- *Dry eye syndrome (DES) a.k.a. keratoconjunctivitis sicca (KCS) or keratitis sicca.*
- *Dry mouth (Xerostomia)*