LUPUS OVERLAP SYNDROMES

The Family of Autoimmune Systemic Rheumatic Diseases

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Systemic Lupus Erythematosus (SLE) belongs to a family of autoimmune rheumatic disorders with features that often overlap.

The SLE Family

Systemic lupus erythematosus (SLE) belongs to a family of related autoimmune rheumatic disorders, including dermatomyositis, Sjogren's Syndrome, scleroderma, polymyositis, mixed connective tissue disease, and rheumatoid arthritis. All of these disorders are capable of affecting multiple organs, and they are all associated with a variety of autoantibodies that rise and fall depending on the clinical disease pattern. Systemic lupus erythematosus is 9 times more common in women than men and 9 times more likely to occur in Afro-Caribbeans and Asians than in white patients. Because symptoms in systemic rheumatic diseases often overlap, the diagnosis of SLE is not always clear-cut. In addition, patients may have autoimmune overlapping disorders.

Diagnosing SLE

The diagnosis of SLE requires the presence of at least four of the following criteria during the course of the disease:

- Malar rash
- Photosensitivity
- Arthritis
- Discoid rash
- Oral ulcers
- Positive autoantibody tests (serositis)
- Renal (kidney) disorder
- Hematological disorder (for instance, anemia)
- Presence of antinuclear antibodies
- Neurological disorder
• Immunological disorder

SLE may be difficult to diagnosis when it is unexpected, for instance in young women and in men. Lupus should be considered when certain clinical features, such as arthralgia (with pronounced morning stiffness), mucocutaneous (mucous membranes or skin) manifestations, and fatigue, either occur simultaneously or appear to evolve over time. About 20 percent of patients with arthralgia will develop a non-erosive deformity of their fingers called Jaccoud-type arthropathy, which is caused by affected tendons. In its early stages, this deformity is reversible although it can become permanent and require surgical intervention.

Signs and Symptoms in SLE

The classic malar rash of SLE occurs in the shape of a butterfly with wings grazing each cheek. This reddened rash may be confused with more common conditions of rosacea and parvovirus, which causes a "slapped cheek" rash. Rapid hair loss is seen in active SLE and can lead to a condition of alopecia. Unless the scalp is scarred, hair growth returns as the disease remits.

Raynaud's syndrome occurs in about half of newly diagnosed lupus patients, but it is less common and less severe than in patients with scleroderma or related conditions. Most patients who present with Raynaud's phenomena will not develop SLE. However, if they have positive tests for antinuclear antibodies (ANA) they are likely to develop a connective tissue disease. The type of ANA pattern helps determine the specific condition. Rising ANA titers with falling levels of complement, C3 and C4, often herald the onset of kidney disease in patients with SLE.

In systemic lupus, anemia is common in active disease. Low white blood cell counts (leucopenia) and low lymphocyte counts (lymphocytopenia) are also common. These hematological changes are helpful in distinguishing lupus from rheumatoid arthritis, a condition that is not usually associated with anemia. C-reactive protein is helpful in differentiating flare from infection in SLE. In flares, the C-reactive protein is usually normal and it rises in infection.

In addition, lupus anticoagulant, despite its name, may occur alone in antiphospholipid syndrome, in patients with SLE, and in patients with other autoimmune rheumatic disorders.

Resources:


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