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Lupus In 'Overlap' With Other Connective Tissue Diseases

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Connective Tissue Disease And Overlap Syndromes The connective tissue diseases are a family of closely related disorders. They include:

- * rheumatoid arthritis (RA)
- * systemic lupus erythematosus (SLE or lupus)
- * polymyositis-dermatomyositis (PM-DM)
- * systemic sclerosis (SSc or scleroderma)
- * Sjogren's syndrome (SS)
- * various forms of vasculitis.

These diseases have a number of common features:

1. They affect females much more frequently than males.
2. They are "multisystem" diseases, capable of affecting the function of many organs.
3. They "overlap" with one another, sharing certain clinical symptoms, signs, and laboratory abnormalities.
4. Blood vessels are the most common target of injury in all of these diseases.
5. The immune system is abnormal and accounts, at least in part, for the observed tissue damage.

Although lupus most often occurs alone, many people with lupus also have symptoms characteristic of one or more connective tissue diseases. In this circumstance, a physician may use the term "overlap" to describe the illness. The following are several well-recognized overlaps that may affect people with lupus.

Lupus and Rheumatoid Arthritis

Lupus

- * Joint pain (arthralgia) is common.
- * Joint swelling (arthritis) may be present in some cases.
- * The majority of those with lupus experience joint pain without swelling or only intermittent swelling.
- * If a person with lupus develops severe arthritis with joint deformities, he/she should be considered to have rheumatoid-like arthritis.

RA

- * Joint swelling is always present.
- * Joint pain is common but less prominent.
- * Because rheumatoid arthritis is more likely than lupus to cause joint deformities and bone destruction, joint replacement or reconstructive surgery is more often required in RA than in lupus.

Frequency Of Overlap Syndromes In People With Lupus The majority of people with lupus have lupus alone. Between 5 and 30 percent of people with lupus report having overlap symptoms. The likelihood of a person with lupus also having an overlap disease is 15 percent, distributed as follows:

Overlap Disease Rate of Occurrence

Rheumatoid Arthritis 1%
 Polymyositis-Dermatomyositis 2%
 Mixed Connective Tissue Disease 3%
 Scleroderma 4%
 Sjogren's syndrome 5%

Heredity And Overlap

- * It is unusual (less than 10 percent of the time) for a person with lupus to have a close family member (parent, child, brother, or sister) who also suffers from lupus.
- * However, it is common for persons with lupus to have relatives (including grandparents, aunts/uncles, cousins) with other connective tissue diseases such as rheumatoid arthritis, Sjogren's syndrome, scleroderma, etc.
- * These co-occurrences raise the possibility that heredity may be a factor in all of the connective tissue diseases.
- * Most scientists agree that important hereditary associations with these diseases are present in some families. Additional research is needed to shed light on this important question.

Prognosis For People With Lupus And Overlap Syndromes

It is important for those with lupus to be aware of the symptoms that might indicate the development of "overlap" features, since these symptoms and abnormalities may be best managed with treatments not typically used for lupus. Fortunately, when an overlap syndrome is present, the symptoms characteristic of the other connective tissue diseases involved are usually mild and not life-threatening.

Treatment should be individualized and directed at the particular problems present at any given time.

Sjogren's Syndrome and Lupus

Sjogren's syndrome

Henrik Sjögren was a Swedish ophthalmologist and the first to recognize that dry eyes and dry mouth were often found in people with connective tissue diseases.

- * These symptoms are caused by the accumulation of immune system cells (lymphocytes) in and around tear and saliva producing glands.
- * This build-up of cells disturbs the function of these glands and leads to reduced production of tears and saliva.
- * This condition also interferes with the protective mechanisms of the eye and mouth.
- * Eye inflammation and ulcers of the cornea, as well as fungal infections of the mouth (thrush), occur with increased frequency in those with Sjogren's.
- * Rarely, a person with this disorder develops a malignancy, or cancer, of the lymphocytes (lymphoma).
- * Today, Sjogren's syndrome is itself accorded the status of a distinct connective tissue disorder.

Lupus

Sjogren's syndrome also occurs in some people with lupus:

- * These individuals have an increased frequency of sun-sensitive rashes and Sjogren's-related blood antibodies (anti-SSA and anti-SSB antibodies).
- * Women with anti-SSA antibodies are at increased risk of having babies with "neonatal lupus." Symptoms can be as minor as a temporary lupus-like skin rash, or as serious as permanent damage to the electrical system of the heart which results in a very slow heart rate (complete heart block)

Treatments

Treatments for Sjogren's syndrome include:

- artificial tears (usually satisfactory)
- artificial saliva (most often unsatisfactory)
- a saliva stimulant (pilocarpine, Salagen)
- hydroxychloroquine (Plaquenil).

Overlap

In some instances, the physician might have reason to believe that both diseases-SLE and RA-have occurred in the same person.

Treatments

- * When arthritis develops in the course of lupus, treatment with these agents can be helpful:
 - non-steroidal anti-inflammatory drugs (NSAIDs)
 - low doses of cortisone
 - the antimalarial drug hydroxychloroquine (Plaquenil).
- * People with lupus who have typical rheumatoid arthritis are prescribed the standard forms of RA treatment:
 - methotrexate
 - gold
 - D-penicillamine - in some cases, more potent drugs to suppress joint inflammation.

Lupus and Myositis

Lupus

- * Many of those with lupus have muscle pain (myalgia), but few have muscle weakness due to inflammation (myositis).
- * The "muscle weakness" that people with lupus report is most commonly due to fatigue or high doses of cortisone.

Myositis

- * In polymyositis-dermatomyositis, the primary problem is muscle weakness due to muscle inflammation.
- * Weakness especially affects:
 - the hips (inability to rise from a chair or toilet seat, or to climb stairs unassisted)
 - the shoulders (inability to lift a weight onto a high shelf or comb one's hair).
 - typically, there is little or no pain associated with the weakness.
- * People with myositis also have:
 - increased blood levels of creatine kinase (CK), a substance that leaks from injured muscle
 - abnormal electrical activity of muscles detected by electromy-

ogram (EMG)

- muscle biopsy showing muscle cell degeneration and inflammation.

Treatments

* Prednisone or other cortisone-like drugs are most often recommended.

* These may be used with more potent immune-suppressing drugs.

* Cortisone itself, in high doses, may actually cause muscle weakness of the hips and shoulders, very similar to what occurs in myositis. But in this condition, called "steroid myopathy," the CK, EMG, and the muscle biopsy do not suggest inflammation, and recovery of strength promptly follows reduction of the cortisone dose.

Lupus and Scleroderma

Lupus

* The variety of skin rashes seen in lupus are due to inflammation, rather than fibrosis. Features of these rashes are limited to the skin surfaces exposed. They include:

- the facial "butterfly" rash
 - a photosensitivity reaction (rash, hives or blisters) seen immediately after exposure to sunlight or other sources of ultraviolet light.

* An exception is "discoïd" or "cutaneous" lupus, which consists of spots or patches of rash, mostly in sun-exposed areas (face, ears, extremities), which typically cause scarring and skin pigment changes. The appearance of scleroderma and discoïd lupus are entirely different, and should be easily distinguished from one another by your physician.

Scleroderma

* The hallmark of scleroderma (SSc) is thickened skin (sclero=hard; derma=skin).

* These changes are due to the excessive production and uncontrolled "laydown" of collagen, the substance normally present in scar tissue.

* Scleroderma mostly affects the fingers, but also:

- the hands
 - forearms
 - feet
 - face.

* If skin fibrosis (hardening) is widespread, it may extend to:

- the upper arms

- thighs

- chest

- abdomen.

* Other features less common in lupus than in SSc include:

- pulmonary fibrosis: scarring of the lower portions of the lung

- difficulty in swallowing solid foods such as bread or meat

- stomach acid "refluxing": heartburn or indigestion from food coming back up into the esophagus due to sluggish and uncoordinated motion of the muscle layer of the esophagus (esophageal dysmotility).

- finger and hand deformities due to the combination of skin thickening, arthritis, and tendinitis and tenosynovitis (inflammation and scarring of tendons and their lining tissues). These processes ultimately result in limited movement of the fingers.

- Raynaud's phenomenon: fingers turn blue or white with cold. This occurs in 95 percent of persons with scleroderma and in 40 percent of persons with lupus.

Treatment

* The primary treatment approaches to SSc are quite different from those for lupus.

* Therefore, treatment for scleroderma should be individualized and directed at the particular problems present at any given time.

Mixed Connective Tissue Disease

* Some individuals have symptoms and signs of three connective tissue diseases:

1. lupus
2. polymyositis-dermatomyositis
3. scleroderma.

* At any given time, the combination of problems encountered by the patient may vary considerably, from no active disease to features of one, two, or all three of these conditions at the same time.

* These individuals often (but not always) have one specific blood antibody (anti-U1RNP antibody) but not the other antibodies commonly associated with SLE, SSc, or PM-DM.

* Whether this is an entirely separate disease, or a situation in which one person has three diseases, remains uncertain. However, the presence of a single blood antibody is a strong point in favor of a distinctive disease.

Treatment