SCLERODERMA UPDATE

Current Clinical Opinions in Systemic Sclerosis

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Scleroderma is an autoimmune connective tissue disorder with symptoms ranging from mild localized lesions to a systemic disease injuring multiple organs.

What is Scleroderma?

Scleroderma, which is also called systemic sclerosis, is a chronic autoimmune connective tissue disorder in which normal tissue is replaced with thick, hardened, fibrous tissue. Fibrous tissue, which resembles scar tissue, is caused by increased production of the protein collagen. Scleroderma primarily affects the skin, but it can also affect other tissues and organs.

Who is Affected?

In the U.S. scleroderma is considered rare, affecting only 200-300 of each one million persons. Each year 12-13 cases for each million people are diagnosed. About 75 percent of patients are females, and most people are diagnosed between the ages of 30 and 50 years. People of all ages, including children, may be affected although the disease course may be different in children.

Types of Scleroderma

There are two major types or forms of scleroderma:

* A Localized more common form that affects the skin, forming distinct waxy patches known as morphea; localized scleroderma may resolve or stop progressing spontaneously; alternately, morphea can become generalized causing a condition of linear scleroderma in which thickness and scarring extend to the underlying fat, muscle, and rarely bone; another form of localized scleroderma called en coup de sabre can primarily affect the face.

* A Systemic form that affects multiples tissues and organs; systemic scleroderma can affect the skin, lungs, kidneys, heart, blood vessels, muscles, joints, and the gastrointestinal tract; in its severest form, systemic scleroderma can be life threatening.

Subtypes of Systemic Scleroderma

Systemic scleroderma may be diffuse or limited. Patients with both scleroderma and the autoimmune liver disease primary biliary cirrhosis have Reynold's syndrome.
1. Diffuse scleroderma affects many parts of the body, including various tissues and organs and the skin over any part of the body.

2. Limited scleroderma is restricted to specific areas of the body (forearms, hands, legs, feet, and face) and doesn’t cause kidney involvement; in the later stages of the disease, the lungs and the blood vessels leading to the heart may be affected causing conditions of pulmonary fibrosis or pulmonary hypertension; limited scleroderma is also known as CREST syndrome, which stands for:

   * Calcinosis, which causes abnormal calcium deposits in the skin
   * Raynaud’s Phenomenon, which causes an extreme sensitivity to cold in the extremities.
   * Esophageal dysmotility (difficulty swallowing)
   * Sclerodactyly (tightening of the skin on the fingers)
   * Telangectasias (red spots on the skin)

**Causes**

Scleroderma develops in people with certain genes who are exposed to certain environmental triggers. Scleroderma can occur in families, and patients with scleroderma often have a family history of scleroderma or other autoimmune diseases. The Choctaw Native Americans have the highest reported prevalence of scleroderma.

Environmental triggers suspected of inducing scleroderma include repetitive motion, vibrational tasks, silica dust, recreational drugs and industrial solvents. Scleroderma-like conditions have been caused by contaminated rapeseed oil, polyvinylchloride, and contaminated tryptophan. However, most people with scleroderma do not have any connection to these environmental triggers.

**Diagnosis**

Scleroderma is diagnosed on the basic of physical findings, imaging tests and laboratory tests. Patients with scleroderma usually have positive ANA tests with patterns suggestive of scleroderma. Tests for scl70 antibodies help differentiate the type of scleroderma present.

**Treatment**

There are no treatments available to cure scleroderma. However, a variety of immunosuppressive medications such as methotrexate can reduce disease progression and ameliorate specific symptoms such as pain. In addition, vasodilators including calcium channel blockers can reduce symptoms associated with Raynaud’s phenomenon. Low dose naltrexone (LDN) has also been reported to show promise in halting the disease course and in some cases improving symptoms in patients with scleroderma. Used in high doses as a topical agent, naltrexone also improves wound healing.
It’s important to keep the skin well lubricated and protected from the elements. Digital injury should be avoided by using appropriate protection during activities. Exercise is important in maintaining joint mobility and muscle tone. A nutrient-rich diet is important for immune system health, and frequent small meals with food that is easy to chew benefit patients who have gastrointestinal involvement. The recommendations for dietary supplements and for diet and lifestyle in Lupus are also beneficial for patients with scleroderma.

**Resources:**


What You Need to Know About Scleroderma, Rheumatic and Immunologic Diseases, Cleveland Clinic, accessed April 1, 2007.

Scleroderma (Systemic Sclerosis), American College of Rheumatology, accessed April 1, 2007

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