Scleroderma

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Scleroderma (Systemic Sclerosis)

Scleroderma (also called systemic sclerosis or SSC is a rare disease characterized most often by skin thickening. Scleroderma also may involve internal organs. This often leads to intestinal, kidney, lung and heart problems. Scleroderma that affects other organs may be referred to as CREST syndrome. It is a chronic disease, which means it lasts a long time.

Limited Cutaneous Disease (scleroderma affecting the skin)

Limited cutaneous scleroderma is characterized by skin sclerosis or thickening. The sclerosis is restricted to the hands, fingers and, to a lesser extent, the face and neck. People with limited cutaneous scleroderma are less likely to develop lung disease. The lung disease often seen with scleroderma is called interstitial lung disease (ILD). When scleroderma is restricted to the hands, ILD often does not occur early in the course of disease. ILD may develop later in the disease process.

Diffuse Cutaneous Disease (scleroderma affecting the skin)

Diffuse cutaneous scleroderma is characterized by extensive skin sclerosis or thickening. The sclerosis involves the arms, legs and torso. It often spares the upper back. People with diffuse cutaneous scleroderma are at an increased risk of developing ILD early in the course of their disease.

Who gets scleroderma?

Scleroderma is a rare disease. About 10-20 new cases per 1 million people are diagnosed each year. People of all races and ethnic backgrounds get scleroderma, but about 75 percent of people with the disease are women. It can occur at any age. However, scleroderma most commonly begins when people are between the ages of 30 and 60 years old.

What causes scleroderma?

While the cause of scleroderma is unknown, we do know that it is an autoimmune disease. This means the body's natural immune system does not behave normally. Instead of serving to fight off infections from bacteria, viruses and the like,

the immune system of a person with scleroderma attacks its own body. This damages blood vessels.

The body's response to damage to its own blood vessels is to make specific proteins, called collagen. The collagen repairs the damage by forming diffuse scars (fibrosis) throughout the blood vessels. The excess collagen can deposit in the skin, in turn causing the many skin changes seen with scleroderma. In addition, the fibrosis (growth of scar tissue) of the blood vessels leads to involvement of many of the internal organs.

Twenty-five to sixty percent of people with scleroderma are often diagnosed with ILD. This number is higher when more of the skin is affected. Ten to fifteen percent of people with scleroderma may be diagnosed with pulmonary hypertension. This is an increase in the blood pressure in the lungs.

What are common symptoms of scleroderma?

Because scleroderma primarily targets the skin, its most obvious symptoms show themselves as skin problems. Due to the nature of the disease, however, which can also affect internal organs, other symptoms may occur.

Common skin symptoms include:

- Thickening, hardening and tightness
- Swelling of hands and toes
- Hair loss in involved skin
- Sores over fingertips
- Generalized itching
- Color changes of hands and feet with cold exposure (known as Raynaud's)

Other common symptoms include:

- Fatigue
- Arthritis
- Muscle pain and weakness
- Dryness of the eyes and mouth
- Heartburn, bloating and other digestive symptoms
- Shortness of breath
- Cough
- High blood pressure (known as hypertension)
- Pulmonary hypertension

These other common symptoms may vary, depending on the organs affected. The lung disease often seen with scleroderma is interstitial lung disease (ILD). Pulmonary hypertension may also occur with scleroderma. Pulmonary hypertension refers to a condition in which high blood pressure exists within the vessels of the lungs. It is important to identify and treat the organs affected with scleroderma.

How is scleroderma diagnosed?

It is often difficult to diagnose scleroderma. A specialist in autoimmune diseases is often required to establish the diagnosis. This specialist is a rheumatologist. The diagnosis of scleroderma is made based on the careful analysis of many factors.

A thorough history and physical examination are essential. There are also certain laboratory studies that can be helpful when considering the diagnosis. These may include specific blood and urine tests. Other tests, including specialized lung and heart evaluations, are needed to determine if there is any internal organ involvement from scleroderma.

It is important to note that a diagnosis cannot be made based on any specific blood test alone.

How is scleroderma managed?

Early recognition of scleroderma is essential. This will allow for prompt treatment. It is important to recognize that there is no cure for scleroderma. In addition, because it is a chronic disease, people often require medical therapy for many years to keep scleroderma under control.

Goals of therapy vary for each person, because the various organs involved in a given person with scleroderma guide treatment. There are a number of effective organ-specific treatments available for people with scleroderma. Combinations of anti-inflammatory and immunosuppressive medicines are often needed. These manage the underlying problems with the immune system. Various other medications are often needed to control the skin and internal organ problems associated with this disease. In particular, chemotherapy may be required to control underlying lung problems. If a person is diagnosed with ILD or pulmonary hypertension, specific treatment is recommended to treat these.

In addition to medication therapy, management of scleroderma may include:

- Partnering with your health care provider to formulate a comprehensive treatment plan
- Learning more about scleroderma
- Living a full life that includes adopting a healthy lifestyle involving
    - Regular exercise
    - Healthy eating
    - Rest
    - Support from others
- Giving up smoking
- Avoiding infections
- Taking medications as prescribed

Many people benefit from physical therapy and rehabilitation. Under the guidance of rehabilitation therapists, people with scleroderma often learn how to appropriately rest, exercise, strengthen and maintain joint and muscle function.

Visit our website for more information about support groups, clinical trials and lifestyle information.

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