Scleroderma

What is Scleroderma?

Scleroderma means "hard skin". It is a disease of vascular and connective tissue where the skin and many other parts of the body can become thick and hard. There are two kinds of Scleroderma: localized and systemic. The localized type tends to be milder and affect mainly the skin and subcutaneous tissue, while the systemic type may affect the internal organs as well as the skin and may be life threatening.

What Causes Scleroderma?

The exact cause of Scleroderma is unknown.

1. Genes are important and patients are born with the increased susceptibility to developing the disease.
2. The presence of autoantibodies suggests that it is an autoimmune disease – a disease where the body is attacked by its own immune system.
3. The blood vessels are abnormal and there is an increased production of the connective tissue resulting in the thickening of the skin and internal organs.
4. Research suggests that exposure to some environment factors may trigger the disease in people who are genetically predisposed to it. Suspected triggers include viral infections, certain adhesive and coating materials, and organic solvents such as vinyl chloride or tricholorethylene.

Who gets Scleroderma?

Although scleroderma strikes every age, sex and ethnic background, most commonly it appears between the ages of 25 and 55. Women are affected about 4 times as frequently as men. The frequency increases with age in both sexes. The overall occurrence is 30 people per 100,000. Although scleroderma is not directly inherited, it is believed that there is a slight predisposition to it in families and with a history of rheumatic diseases.

What are the symptoms of Scleroderma?

The signs and symptoms depend on the organ involved. The skin is most frequently involved and hardening of the skin is one of the most visible manifestations of the disease. The internal organs like the lung, kidney, heart, stomach and intestines may also be involved.

The skin is thickened and may look shiny. There are three phases of skin changes:

1) Edematous phase – the skin including the subcutaneous tissue become swollen, there is loss of wrinkle, cold skin which usually starts from the hands then spread upwards.

2) Sclerotic phase – in which the skin become hard causing difficulty in finger banding, mouth opening, and expressionless.

3) Fibrotic phase – the skin become fibrotic which fixed to the underling subcutaneous tissue and bone causing wood-like limbs, risk of digital ulcer . Patches of the skin may become darker or lighter in colour.

There may be prominent tiny blood vessels (telangiectasia) on the skin and small chalky lumps (calcinosis) under the skin. Sometimes ulcers form at the bony prominences. Many of the patients are sensitive to the cold and their fingers will turn white or blue when exposed to the cold (this is known as Raynaud's phenomenon) when she/he goes into an air-conditioned room. Swelling of the hands and feet may be associated with aching especially in the morning. The joints may hurt and if proper exercises are not done, the hands may permanently stay crooked.

Involvement of the lungs causes the patient to feel breathless especially on exercise. Eight out of ten patients who have systemic scleroderma also have a problem with their lungs that leads to scarring (fibrosis). This scarring is caused by inflammation in the lungs. The inflammation and scar tissue cause the lungs to work less well because the lungs are “stiffer” and less able to transfer oxygen into the blood stream. As a result, it may be more difficult for patients to walk or perform normal activities without getting shortness of breath. Quality of life may decrease because normal activities once enjoyed may not be possible due to increased lung stiffness or lack of oxygen.

About 15 to 20 percent of people with systemic sclerosis develop heart problems, including scarring and weakening of heart (cardiomyopathy), inflamed heart muscle (myocarditis), and abnormal heart beat (arrhythmia). Patients may experience shortness of breath, decrease exercise tolerance or palpitation.

Involvement of the intestines may give the patient heartburn or a feeling of abdominal pains. There may also be abdominal swelling with poor appetite, vomiting and loss of weight. Involvement of the kidneys may result in sudden high blood pressure with headache and vomiting.

Seventy percent had kidney involvement, which may cause loss of renal function, frothy urine, high blood pressure or acute renal failure.

The Diagnosis of Scleroderma

Scleroderma affects different patients differently. The diagnosis of scleroderma may require several visits to the doctor over a period of time. You will be asked questions about your symptoms, such as

1) fingers sensitive to cold,
2) thickening of skin,
3) shortness of breath,
4) trouble swallowing,
5) stiffness of the hands,
6) joints or bone pain.

If you have four or more of the above symptoms, you may have scleroderma. Your doctor will then perform a thorough physical examination and send you for various tests to determine the extent of your disease. These tests include X-rays, electrocardiogram, urine and blood tests and sometimes other specialized tests. The specific blood tests may show the presence of autoantibodies. Autoantibodies are produced by the body's immune system which has mounted a "rebellion" against itself. The two common antibodies found are Antitopoisomerase-1 (anti-Scl-70) and anticentromere antibodies. Anti-Scl-70 antibodies appear in the blood of up to 40 percent of people with diffuse systemic sclerosis. Anticentromere antibodies are found in the blood of as many as 90 percent of people with limited systemic sclerosis.

For the purposes of classifying patients in clinical trials, American College of Rheumatology in 1980 developed a diagnostic criteria for systemic scleroderma:

<u>Major criteria:</u>

1. Proximal scleroderma

Symmetric thickening, tightening and induration of skin proximal to the hands, can spread to extremity, face and whole body.

<u>Minor criteria:</u>

1. Sclerodactyly: Thickened skin limited to fingers only
2. Loss of substance from finger pad
3. Bibasilar pulmonary fibrosis

If one major or two or more minor criteria are present, then a person shall be said to have systemic sclerosis

Treatment

Scleroderma is a systemic disease with multi-organ involvement. Treatments depend on the symptoms and the extent of the disease. In addition to doctors, professionals like nurse practitioners, physical or occupational therapists, psychologists, and social workers may play a role in your care. Dentists, orthodontist, and even speech therapists can treat oral complications.

1. Raynaud's phenomenon

Raynaud's phenomenon is common in scleroderma. It can be uncomfortable and can lead to painful skin ulcers on the fingertips, which may be precipitated by cold or emotional stress. The following measures may make you more comfortable and help prevent problems:

- Don't smoke
- Dress warmly, with special attention to hands and feet
- For severe cases, consider using calcium channel blockers as mention above
- Protect skin ulcers from further injury or infection by applying nitroglycerine paste or antibiotic cream. Severe ulcerations on the fingertips can be treated with bioengineered skin

2. Stiff and painful joints

In diffuse systemic sclerosis, hand joints can stiffen because of hardened skin around the joints or inflammation of the joints themselves. The following measures may help:

- Exercise regularly
  Ask physical therapist about an exercise plan to increase and maintain range of motion in affected joints. Swimming can help maintain muscle strength, flexibility, and joint mobility.
- Consult occupational therapist to learn a new technique in performing daily tasks, such as lifting and carrying objects or opening doors, in ways that will put less stress on tender joints
- Use acetaminophen or nonsteroidal anti-inflammatory drugs to help relieve joint or muscle pain

3. Skin problems

When too much collagen builds up in the skin, it crowds out sweat and oil glands, causing the skin to become dry and stiff. To ease dry skin, the following methods are useful:

- Apply oil-based creams and lotions frequently, and always right after bathing
- Apply sunscreen before to protect against further damage by the sun's rays
- Use humidifiers to moisten the air in colder winter climates
- Avoid very hot baths and showers, as hot water dries the skin

4. Dry mouth and dental problems:

Dental problems are common in people with scleroderma for a number of reasons: 1) tightening facial skin can make the mouth opening smaller and narrower, which makes it hard to care for teeth; 2) dry mouth due to salivary gland damage speeds up tooth decay; 3) damage to connective tissues in the mouth can lead to loose teeth. You can avoid tooth and gum problems in several ways:

- Brush and floss your teeth regularly
- Have regular dental checkups
- If decay is a problem, as your dentist about fluoride rinses or prescription toothpaste that remineralize and harden tooth enamel
- Regular facial exercises to help keep your mouth and face more flexible
- Keep your mouth moist by drinking plenty of water, using sugarless gum or hard candy. If dry month still bothers you, ask your doctor about a saliva substitute or need of saliva stimulator called pilocarpine hydrochloride (Salagen)

5. Gastrointestinal problems

Systemic sclerosis can affect any part of the digestive system. You may experience heartburn, difficulty swallowing, early satiety, or intestinal complaints such as diarrhea, constipation, gas or difficulty in absorbing nutrients. The followings may help in improving the symptoms:

- Eat small, frequent meals
- Raise the head of your bed with blocks, and stand or sit for at least an hour (preferably two or three) after eating to keep stomach contents form backing up into the esophagus
- Avoid late-night meals, spicy or fatty foods, and alcohol and caffeine, which can aggravate GI distress
- Chew foods well and eat moist, soft foods

6. Lung problems

Lung involvement is a serious complication in scleroderma. It comes in two forms: pulmonary fibrosis (hardening or scarring of lung tissue because of excess collagen) and pulmonary hypertension (high blood pressure in the artery that carries blood from the heart to the lungs). Early detection of lung involvement may give a higher chance of treatment success. Work closely with your medical term and do the following:

- Watch out for signs of lung disease – fatigue, shortness of breath or difficulty breathing, and swollen feet. Report these symptoms to your doctor
- Have your lungs closely checked during the early stages of skin thickening or any new symptoms arise, using imaging (X-ray or CT scan of lung) and standard lung function tests. These test are needed to find the problems at the earliest and most treatable stages
- Get regular flu and pneumonia vaccines as recommended by your doctor.

7. Heart problems:

Fifteen to twenty percent of people with systemic sclerosis develop heart problems; these include cardiomyopathy, myocarditis and arrhythmia as mentioned above. The mainstay of treatment is to report the related symptoms early and regular monitor the cardiac function. Specific treatment ranges from drugs to surgery, and varies depending on the nature of the condition.

8. Kidney problems

About 15 to 20 percent of people with systemic sclerosis develop severe kidneys problems, including loss of kidney function. Uncontrolled high blood pressure can quickly lead to kidney failure. Things you can do to minimize the problem:

- Check your blood pressure regularly, and if you find it high, call your doctor
- If you have kidney problems, take your prescribed medications faithfully. Angiotensin-converting enzyme inhibitors mentioned above markedly decrease the risk of scleroderma-related kidney failure.

What is the outlook?

Scleroderma is a chronic disease of unknown etiology. The symptoms of scleroderma vary greatly from individual to individual, and the effects of scleroderma can range from very mild to life-threatening. The seriousness will depend on what parts of the body are affected and the extent to which they are affected. In fact, only minority had serious and lift-threatening complication with involvement of major internal organ. Prompt and proper diagnosis and treatment by qualified

physicians may minimize the symptoms of scleroderma and lessen the chance for irreversible damage.

Working in partnership with your doctor, you can do much to make yourself feel better and keep your disease under control. With some adjustments to your life-style many can return to a full-time job or to whatever you had been doing before your illness.

Suggestions

Living with scleroderma can place you on a roller coaster of emotion. Here are some suggestions:

- Maintain normal daily activities as best as you can
- Pace yourself and be sure to get the rest that you need
- Stay connected with friends and family
- Continue to pursue hobbies that you enjoy

Keep in mind that your physical health can have a direct impact on your mental health. Denial, anger and frustration are common with chronic illness. Talk with your family, friend and doctor if you're feeling depressed.