What is Scleroderma?

Scleroderma (also known as systemic sclerosis) literally means “hard skin”. The disorder is characterised by abnormalities of the blood vessels, connective tissue and the immune system. It can affect the skin, digestive system, heart, lungs, muscles and joints.

What causes scleroderma is currently unknown. Some unknown factor triggers an overproduction of collagen (a type of protein that is the main component of the connective tissue) causing thickening, hardening and scarring of the skin and organs throughout the body.

What happens in Scleroderma?

In scleroderma, the body’s faulty immune system stimulates the fibroblast cells to produce excess collagen. How the body manages and ‘stores’ the excess collagen gives rise to the two major classifications of scleroderma: Limited Scleroderma and Diffuse Scleroderma with different patterns of involvement and symptoms.

Limited Scleroderma

In limited scleroderma, the skin involvement is less widespread and is typically confined to the fingers, hands, lower arms, lower legs and the face. Limited scleroderma is sometimes called CREST syndrome, which represents the initial letters of 5 common symptoms:

Calcinosis: Calcium deposits form under the skin around the elbows, knees or fingers, causing the affected regions of the skin to appear as hard, whitish areas. These deposits can feel tender and sometimes have a tendency to break through the skin, forming painful ulcers.

Raynaud’s phenomenon: Excess collagen deposits cause narrowing of small blood vessels and decreased blood flow to the extremities, such as the fingers and toes, which often turn white/blue in response to cold temperatures or emotional stress. In extreme cases, this may lead to finger ulcers. Around 90% of patients with limited and diffuse scleroderma will experience this.

Esophageal dysfunction: When scleroderma involves the muscles of the digestive system, some patients may develop an abnormally wide esophagus, experiencing heartburn, acid reflux, or swallowing difficulties.

Sclerodactyly: Excess collagen is deposited as areas of thick and shiny skin on the fingers and sometimes the toes. The layer of hard skin often makes it difficult to bend or straighten the fingers.

Telangiectasia: Small clusters of dilated blood vessels that appear as red spots may develop on the hands and face. These spots are not usually painful, but can cause cosmetic problems.

Limited scleroderma commonly manifests itself slowly over a period of 10 to 20 years. Raynaud’s phenomenon often precedes the skin involvement by many years, followed by other areas of involvement. One of the late manifestations may include heart/ lung involvement called pulmonary arterial hypertension (increased blood pressure in the artery that carries blood from the heart to the lungs).

Diffuse Scleroderma

Skin involvement in diffuse scleroderma is more widespread and has a much faster onset and progression than limited scleroderma. More importantly, people with diffuse scleroderma have a higher risk of developing hardening of the internal organs such as the lungs, kidneys and heart.

Who gets affected?

Scleroderma is not a common condition. An estimated 20 to 200 people per million are affected worldwide, with the age of onset usually between 30 to 50 years. Women are 4 times more likely to develop the condition than men.

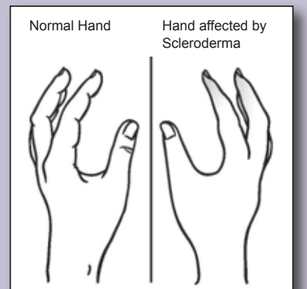
What are the risk factors?

Scleroderma is not contagious, so you cannot “catch it” or “give it” to someone else. It is not hereditary, so most patients do not have relatives with scleroderma, and they do not “pass on” scleroderma to their children. Whilst there are some susceptibility genes, which increase the risk of getting scleroderma, the genes themselves do not cause the disease. An environmental trigger, in a person who is at risk, may result in the disease manifesting.

What are the symptoms?

The first sign of scleroderma is usually the development of areas of thickened or hard skin around the joints of the fingers and toes. This hardened skin tends to lead to skin tightening that may decrease joint mobility. There may be intense skin itching and inflammation.

Patients may also experience symptoms such as joint pain and swelling (similar to arthritis), shortness of breath or persistent coughing (if the lungs are affected), fatigue with very high blood pressure (if the kidneys are affected), chest pains or irregular heart beat (if the heart is affected), heartburn, swallowing difficulties, bowel constipation, diarrhoea or weight loss (if the digestive system is affected) and dry mouth and eyes (if salivary glands and tear ducts are affected). Dental health may be compromised due to difficulty in opening the mouth caused by tight facial skin and dry mouth may lead to tooth decay.



With Scleroderma the skin on the hands become stiff and tight, and later stages the pads on the fingers may become thinned.

How is Scleroderma diagnosed?

Depending on the symptoms, scleroderma can be diagnosed by many different specialists, including rheumatologists, dermatologists or internists. The most common test for scleroderma is an immunological blood test that detects antibodies produced by the body that are specific for scleroderma. Occasionally a skin biopsy (in which a sample of the skin is taken for examination under a microscope) or examination of the small blood vessels of the nailbed under the microscope is required.

Other tests may include X-rays, CT scan, echocardiogram (heart scan), lung function test, endoscopy (to look at the oesophagus and stomach) and urine tests, depending on whether other internal organs are affected.

What is the treatment?

There is currently no cure for the underlying cause of scleroderma, which is the overproduction of collagen. Specialised and dedicated scleroderma clinics are shown to result in better outcome. Sustained and increasing research is therefore necessary. Treatment is based on alleviating the different symptoms of the condition and limiting the damage to the internal organs through medication.

Regular moderate exercise is important to improve joint flexibility and cardiovascular health. Consult your doctor or physiotherapist about the type of exercise you can do.

Pain relief for stiff and swollen joints is usually possible with non-steroidal anti-inflammatory drugs. Areas of dry and irritated skin can be helped by a regular moisturiser. Antihistamines may help with intense itching of the skin.

For Raynaud's phenomenon, it is important to keep warm and to stop smoking. Medication to improve blood may be prescribed, while antibiotics can protect skin ulcers from getting infected. Proper wound care to promote ulcer healing can help to slow down the progression in scleroderma.

For oesophageal involvement, H2 blockers, proton pump inhibitors or prokinetics may be prescribed. Simple measures such as avoiding acidic foods, eating smaller and more frequent meals, elevating the head of the bed and avoiding lying down for at least 3 hours after a meal can help to decrease symptoms of acid reflux.

When a patient is diagnosed with diffuse scleroderma, medication may be prescribed to decrease the activity of the immune system. When internal organs are affected by the condition, other specific treatments may be needed.

Information modified from National Arthritis Foundation, Singapore

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