Scleroderma: The hard facts about hard skin

By Arunima Sivanand

Scleroderma refers to hardening of the skin and connective tissue (the words comes from the Greek scleros meaning "hard" and derma meaning "skin"). The term is used to refer to a group of disorders that range from mild to life-threatening.

Types of scleroderma

There are two types of scleroderma—localized and systemic. Localized scleroderma is limited anatomically and affects only the skin. Systemic scleroderma, which is also known as systemic sclerosis (SSc), affects the internal organs as well as the skin.

Localized scleroderma

Localized scleroderma can be classified as morphea or linear. In the morphea form, the skin is affected in patches; while in the linear form, the affected areas appear as vertical lines.

Systemic sclerosis

There are two main types of SSc—limited cutaneous and diffuse cutaneous. Limited cutaneous SSc presents on the face or lower half of limbs. Diffuse cutaneous SSc can present on the trunk and limbs, in addition to the face. The larger the area affected by SSc, the greater the likelihood of internal organ involvement.

Patients with limited cutaneous SSc may have symptoms such as Raynaud's phenomenon, whereby their fingers turn bluish when exposed to the cold or reddish when exposed to warmth. They may also experience heartburn if the disease affects the esophagus (the tract connecting the mouth to the stomach). In addition, small red spots, known as telangiectasias, can appear on the skin. The condition can also result in calcinosis (the presence of small calcium deposits under the skin) and sclerodactyly (where the skin on the fingers is thickened). Patients with limited cutaneous SSc are also at risk of pulmonary hypertension, which presents as shortness of breath upon exertion.

Patients with diffuse cutaneous SSc are at risk of developing pulmonary fibrosis, a respiratory disease in which scars form in the lung tissues, leading to shortness of breath and cough. Diffuse cutaneous SSc can also lead to kidney disease, resulting in hypertension, or increased blood pressure. Bowel disease can also occur, leading to bloating, constipation or diarrhea.

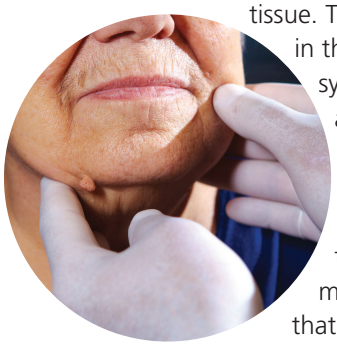
Risk factors

Sex and age: Scleroderma is about four times more likely to occur in women than men. It usually develops between the ages of 25 and 55 years.
Genetic factors: Scleroderma is known to be more common in families with other autoimmune diseases (e.g., rheumatoid arthritis), suggesting a possible genetic component.
Exposure to toxins: Environmental

exposure to drugs, toxins and other products (e.g., silica) has been reported to increase the risk of scleroderma.

Causes

Scleroderma is an autoimmune disease. Normally, the immune system protects the body from foreign agents that could be harmful, such as bacteria. In autoimmune diseases, however, the immune system loses its ability to differentiate foreign agents from the person's own tissue. This results in the immune system attacking and destroying the body's own tissues. However, the exact mechanisms that result in the autoimmunity behind scleroderma remain unknown.



In localized scleroderma, the immune system causes the overproduction of collagen in affected areas of the skin. Collagen is a structural protein that makes up the body's connective tissues, so its overproduction results in abnormalities in the texture and function of the skin.

In SSc, the immune system again causes collagen overproduction. In addition to affecting the skin, however, this leads to fibrosis in internal organs such as the bowels, kidneys, heart and lungs. The tissue damage caused by the fibrosis reduces the flexibility of the organs and results in them malfunctioning. In SSc, the immune system also produces autoantibodies that attack and damage the body's own tissues, including blood vessels.

Making a diagnosis

If you suspect you have scleroderma then you should visit your family doctor, who may refer you to a rheumatologist or dermatologist. You will be asked about your history and the physician will perform a physical examination. You might also be referred for a blood test and skin biopsy to help make the diagnosis. Because scleroderma can mimic other rheumatic conditions with skin and systemic manifestations, it is often misdiagnosed as another autoimmune disease such as lupus.

Managing scleroderma

As yet there is no cure for scleroderma, limiting disease management to the alleviation of symptoms. Physicians may address specific symptoms and prescribe medications such as antacids for heartburn, immunosuppressants for immune function and antihistamines for itching.

Support groups

As scleroderma affects a person's appearance, the psychological impact of the condition can be significant. There are several support groups that can provide information on the disease and opportunities to connect with other people with this condition. Scleroderma Canada has a list of support groups for each region in Canada, available at scleroderma.ca/support/find-a-support-group.php. CS

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5 lifestyle changes to make

1. **Stay active**—Exercise improves the circulation, keeps joints flexible and relieves stiffness.
2. **Quit smoking**—Nicotine narrows the blood vessels and affects lung function, which can worsen the symptoms of scleroderma.
3. **Avoid the cold**—Wearing thick socks and gloves ensures the skin is not exposed to extremely low temperatures, which causes blood vessels to constrict.
4. **Moisturize**—Using oil-based lotions or creams containing lanolin, especially after bathing, can restore moisture to the skin.
5. **Manage stress**—Mindfulness, meditation or other methods can keep stress in check and contribute to overall well-being.