

Limited Cutaneous Systemic Sclerosis

What is limited cutaneous systemic sclerosis (lcSSc)?

In the early 1900's several physicians noted a group of features often seen when they examined patients with scleroderma (systemic sclerosis). They coined the term CREST syndrome which is now known as limited cutaneous systemic sclerosis. Systemic sclerosis is a form of scleroderma that affects the skin and internal organs. The limited cutaneous subset affects the skin of the extremities but is associated with a set of characteristic features abbreviated as the acronym CREST

What is Crest?

- C - Calcinosis which is an accumulation of calcium below the outer layer of the skin.
- R - Raynaud's phenomenon - a condition in which the blood supply to the extremities, usually the fingers and toes, is temporarily interrupted.
- E - Esophageal (American spelling) involvement, causing difficulty in swallowing or indigestion.
- S - Sclerodactyly is when the skin of the digits becomes thin, shiny and leathery looking. Fingers and toes may become flexed and stiff.
- T - Telangiectasia is the appearance of small blood vessels near the surface of the skin. These can be seen on the fingers, palms, lips, face, tongue and chest wall.

Other features of lcSSc

In addition several other features may be found:

- Non pitting swelling of the fingers
- A "salt and pepper" appearance due to areas of hypopigmentation and hyperpigmentation
- Dilated capillary loops at the base of the fingernails
- Microstomia (small mouth) - caused by tightening of the skin
- Ulceration of skin
- Involvement of internal organs may occur, particularly in the bowels and lungs.

Calcinosis

Calcium deposits, chalky material (termed calcinosis), is often deposited in the soft tissues of the fingers, forearms, buttocks and other body areas can be seen in people with scleroderma, usually of long duration. They can also be seen in people with inflammation of the muscles (myositis, especially when it occurs in childhood) and in those on long term kidney dialysis. Calcium deposits can appear late on in the disease process. After several years, soft-tissue calcium deposits may be found in only about 40% of scleroderma patients and even then an X-ray may be needed to detect them. It is a troublesome complication of scleroderma which develops in some patients and can occur anywhere but is most often seen in the fingers. Calcinosis can be very painful, especially when at a site where it is knocked or pressed upon and may cause secondary problems or ulceration. Calcinosis may be hard, like chalk or semi liquid. Both types sometimes push up through the skin. Calcinotic nodules can form the focus for local infections which may need to be treated with antibiotics. Occasionally surgery is necessary to remove particularly troublesome nodules of calcinosis, although it can sometimes return or develop at new sites.

Raynaud's

Raynaud's is reported in 90-99% of people with scleroderma. In response to cold or emotion, the blood vessels constrict or narrow, and the resulting disturbance in circulation of the blood causes a series of colour changes in the skin - white, blanched or pale, as circulation is reduced, blue as the affected part loses oxygen from decreased blood flow and then red or flushed as blood flow returns and the part rewarms. Finally, as the attack subsides and the circulation returns to normal, usual skin colour is restored. In the 'white' or 'blue' stages, sensations such as tingling, numbness, and coldness may be felt. In the 'red' stage, a feeling of warmth, burning or swelling may be noted. Raynaud's attacks can be very painful.

When Raynaud's occurs by itself, it is most common in young females who are otherwise healthy. In the context of rheumatic disease it may also be present in association with lupus, myositis, or undifferentiated connective tissue disease.

Esophageal problems

(American spelling of oesophagus) - weak or 'lazy' muscle in the gullet or oesophagus, which can result in heartburn or a sense of food sticking in the throat or chest, upon swallowing.

Oesophageal problems such as heartburn, reflux, and stomach hyperacidity are fairly common in the general population but lack of motility in the oesophagus is unusual in healthy individuals. It is not unique to scleroderma, but is present in many other rheumatic conditions such as mixed connective tissue diseases, where weakness of the muscle in the gullet can lead to heartburn, or to a feeling of just-swallowed food or liquids, sticking in the throat or chest.

Oesophageal problems can be found in almost all patients with established lcSSc within 3-4 years of onset, even if there are no symptoms of heartburn or of food sticking in mid chest. This is one of the most treatable complications of scleroderma. Many drugs such as proton pump inhibitors, which are useful in treating peptic ulcers, have been found to be very effective at treating the symptoms and consequences of oesophagitis in scleroderma.

Sclerodactyly

This is hard or tight, shiny skin on the fingers, which may cause them to become flexed and stiff. Sclerodactyly or puffy fingers and tightness of the skin of the fingers occurs in over 90% of people with scleroderma.

Telangiectasia

Small clusters of dilated blood vessels (spider veins) in the skin, which look like red dots or blotches, found most commonly on the hands, face, chest and in the mouth. They can be covered with cosmetic camouflage or treated by laser therapy. These may occur in scleroderma or as part of a disorder called 'hereditary telangiectasias', which is not related to scleroderma.

Telangiectasias may be seen in 50% of people with limited cutaneous systemic sclerosis within the first 2-3 years, and the percentage increases to 80-90% after 10 or more years.

Photographs illustrating symptoms of Limited Cutaneous Systemic Sclerosis



Calcinosis -
finger showing a calcium deposit

Raynaud's -
numb fingers



Esophagus (American Spelling) -
abnormal motility in oesophagus
(radiography)

Sclerodactyly -
tight shiny skin



Telangiectasia -
small red markings seen
here on the mouth

Treating an ulcer

If an ulcer develops as a result of poor circulation or calcinosis, it requires medical attention. To heal, dead tissue should be removed and infection needs to be cleared. Unwanted crusts and dried pus can be removed by soaking the ulcer in luke-warm water for approximately five minutes. The area should then be allowed to dry before applying a dressing. There are several good dressings available but they need to be used properly to get the best results. Treatments available from your doctor may include antiseptic or antibiotic ointments. Sometimes a dry dressing is best, such as gauze or a non-adherent dressing or one of the hydrocolloid adhesive preparations. Alternatively, a calcium alginate dressing derived from seaweed which can be removed from the ulcers by washing with saline solution may be used.

The Lungs in IcSSc

The lungs can be affected in scleroderma in two ways. Firstly fibrosis or scarring can occur and secondly the blood vessels can become thickened, narrowed and scarred, without any other part of the lung becoming affected. This causes pulmonary hypertension (PHT) which usually shows itself from five years onwards in patients with limited cutaneous systemic sclerosis. Although a rare complication it occurs in 1 in 7 people with scleroderma. and should be taken seriously. There are several new drug therapies which are available to help alleviate the symptoms.

Further reading

The RSA has published a wide range of leaflets including Raynaud's, scleroderma, the heart and lungs, the gut, the kidneys, localised scleroderma, sexuality in scleroderma, dental care, foot care, ulcers and skin care



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