SCLERODERMA
SYSTEMIC SCLEROSIS AND THE SKIN

An information guide to managing Systemic Sclerosis

scleroderma australia

Systemic Sclerosis and the Skin

Systemic sclerosis (SSc) has many potential impacts on the skin. The type and severity of the skin involvement will vary and is often related to the underlying subtype of SSc.

Systemic Sclerosis is due to a complex combination of:

- overproduction of collagen ('fibrosis' or 'sclerosis'),
- inflammation caused by overactivity of the immune system ('autoimmunity'), and
- changes in the blood vessels.

These underlying processes are responsible for the varied impacts SSc has on the skin.

UNDERLYING PROCESS	SKIN AFFECTS
Increased Collagen	Skin sclerosis; thick, tight and immobile • Stiff stuck fingers (sclerodactyly) • Tightened mouth opening • Stiff stuck joins (contractures) • Changes in facial shape and features • Speckled brown and white discolouration
Blood vessel changes	• Telangiectasia • Raynaud's • Digital Ulcers
Autoimmune related inflammation	• Itchy Skin • Inflamed and uncomfortable skin

Other: Calcinosis

Many of these skin changes in SSc need separate consideration when it comes to managing your symptoms and improving the skin's function.

Skin tightening

The fundamental abnormality in SSc is over production of collagen. This causes fibrosis. Fibrosis means that the connective tissues in the skin (and other organs such as the lungs), become thick, hard and tight. Almost everyone with SSc will have some skin tightening. However, how thick the skin becomes and the areas involved can vary widely. People with the limited form of SSc usually have skin thickening limited to the face and below the knees and elbows. In contrast, those with the diffuse form of SSc can have more widespread skin thickening also involving the upper arms, thighs and trunk. The skin thickening itself can be mild and subtle, or severe where it is very tight, cannot be pinched and appears smooth and shiny. There is a very rare subtype of SSc which does not involve the skin called "systemic sclerosis sine scleroderma".

It is also important to understand though, that the skin thickening can improve over several years.

Sclerodactyly

The fingers/hands are most commonly involved in all subtypes of SSc. When the skin over the fingers becomes hard and immobile, the fingers may sometimes become 'locked' in a bent position so that they cannot straighten. This is called 'sclerodactyly'. In some people, sclerodactyly can make daily living activities such as writing, doing up buttons or holding a glass of water more difficult. Similarly, the toes can be affected.

Joint contractures

If the skin thickening extends, it can involve the wrists, forearms and elbows. If the skin over joints such as the wrists, ankles and elbows becomes very tight and immobile, the joints contract and it can be hard to move those joints .

Facial skin

Skin tightness can also affect the face. This can alter overall appearance. Our face is a big part of our identity that we physically present to the world and how people recognise us. If that changes, it can therefore understandably be very confronting and difficult to come to terms with. Involvement of the facial skin around the mouth can also decrease how wide the mouth can open. This is called microstomia.

Treatment for skin tightness

Treating fibrosis in SSc is the main goal of treatment. Softening the skin with treatment is key to limiting the functional and cosmetic impacts of skin thickening and complicating joint contractures. Medications used for this aspect of treatment are often called 'anti-fibrotics'. The most commonly used is mycophenolate mofetil.

General measures such as keeping warm, stretching exercises and physiotherapy can also be beneficial at reducing the risk of worsening joint contractures and microstomia.

Due to the skin being generally abnormal, tight and inflamed, it can also be prone to easily breaking down and poor wound healing. Doing everything possible to avoid trauma can really help avoid related complications. Wearing gloves, warm protective shoes, long sleeves and putting on trousers, putting on a thick layer of a greasy moisturiser can all help to protect the skin.

Digital Ulcers

The term 'digital ulcer' usually refers to an ulcer on the fingers or toes which is a consequence of poor blood circulation in SSc. Digital ulcers tend to co-exist with severe Raynaud's (cold hands or feet, with blue, white and/or red skin colour change).

Making lifestyle changes to minimise the risk of digital ulcers is very important. The key is to keep warm! Dress warmly, wear double lined gloves, invest in silver socks/thermal socks and use heat pads inside shoes and gloves.

Important things to avoid include:

- Smoking
- Caffeine
- Medications that constrict the blood vessels (beta blockers, medications for migraine & ADHD)
- Abrupt changes in temperature
- Trauma

Treatment for digital ulcers

There are many medical options to help the blood circulation to the fingers and toes. These include tablets such as calcium channel blocking medications, and in severe cases other options such as sildenafil or intravenous infusions of iloprost can be used.

These treatments can promote ulcer healing and help prevent the formation of new digital ulcers. It is also important to have the ulcer itself attended to with appropriate cleaning, dressings, and care of the surrounding skin. This can also help prevent secondary infection of the digital ulcers.

Telangiectasia

Telangiectasias (often called matt telangiectasias) are small red spots on the skin produced by dilated and 'broken' superficial skin capillaries. In SSc, these classically occur on the face and palmar side of the hands and fingers, but can extend to the neck and chest, or elsewhere. These are a cosmetic concern, but they have no other consequences.

Using green tinted makeup can often help to camouflage the red spots and blend them into the background skin colour.

Vascular light or laser treatments, such as intense pulsed light (IPL) or pulsed dye laser (PDL) can help. Telengiectasia in SSc are often stubborn to treat and repeated treatments may be required. However, repeated laser treatments can result in a good cosmetic outcome in many patients, boosting confidence.

Itchy, dry and inflamed skin

Almost half of people with SSc will experience itchy skin. Itch is such an uncomfortable symptom. It can have a significant impact on daily life and feel impossible to escape.

Itchy skin is generally a sign that the underlying SSc is active. It implies there is ongoing inflammatory changes in the skin, although not everyone with active SSc will have itchy skin.

The skin in SSc is often dry and easily irritated. This alone can contribute significantly to itch and general skin discomfort. Using a regular moisturizer and avoiding excessive showering can be helpful. Switching to a a 'soap free cleanser' which does not contain sodium lauryl sulfate is also important.

Of course, itch can also be caused by a multitude of other causes too, unrelated to SSc. It's important that other possible contributing factors, like low iron, or abnormal thyroid function, are checked and treated. Other skin conditions can also co-exist with SSc, like eczema or rashes from viruses.

Treating itch

Treating itchy skin in SSc can be challenging. General measures such as not overheating, wearing loose cotton clothing over affected areas and keeping the skin well hydrated are all important.

There are also many specific treatments to try. The best options depend on many factors and seeing a dermatologist or talking to your rheumatologist can be very helpful. Steroid creams, phototherapy, antihistamines or other prescription medications (such as mast cell stabilisers or those used for neuropathic itch) are all options that can be tried in consultation with your doctor.

Calcinosis

Cutaneous calcinosis is the deposition of calcium in the skin and/or underlying fat layer. Calcinosis tends to occur over sites of pressure such as joints of the fingers or the elbows. However, it can occur anywhere.

Calcium is very hard, and so when it deposits in the skin it produces firm hard lumps. These are usually small and just under the skin, but larger and deeper 'chunks' of calcium can develop. Sometimes the calcium can 'pop through' the skin as a white chalky material. In some cases this can cause ulceration, with skin break down. However, the biggest problem with calcinosis tends to be pain and discomfort.

Treatment for calcinosis

Importantly, if calcinosis is not causing any pain or symptoms, it is perfectly safe to leave it there without any treatment. Cutaneous calcinosis in SSc is notoriously challenging to treat and high level studies assessing the effectiveness of different treatments are very limited. Simple measures such as avoiding trauma, using protective gloves and ensuring the hands are kept warm are important.

Prescription creams such as topical steroids, medications which block calcium production or release and/or surgical procedures can be tried. Which treatments are applicable will depend on where the calcinosis is, the size of the calcium deposits and how much difficulty it is causing.

Cosmetic procedures

The utilisation of laser devices, cosmetic injectables and other surgical procedures in the treatment of Scleroderma skin disease is expanding and attracting increasing interest. It is important to know that studies and clinical experience in this field, as well as access to most of these treatments, remains very limited, and many procedures are experimental only.

Aside from vascular light and laser options for telangiectasia, other laser interventions in SSc are experimental and not widely utilised. Intense pulsed light can regulate collagen production in the skin and may help microstomia. Ablative CO_2 laser has shown some promise for calcinosis, microstomia and heel contractures.

Injectable fillers can be used to correct contour changes caused by fat loss, particularly around the mouth in SSc. Lipotransfer (+/– fat-derived stem cells)

is gaining increasing utility in SSc. In this procedure, your own (autologous) fat is injected in the areas of tissue loss. This technique can also lessen fibrosis and therefore help improve microstomia. Synthetic nonpermanent dermal fillers such as poly-L-lactic acid are also reported in SSc. The most commonly available synthetic filler in the cosmetic industry is hyaluronic acid. In scleroderma, its use has thus far been limited to morphoea (localised scleroderma).

For difficult to treat Raynaud's or digital ulcers in SSc, Botox injections may also be used, and there are a number of approaches to this.

Importantly, cosmetic injectables pose a theoretical risk of causing disease reactivation in scleroderma due to the physical trauma induced by the treatment, the antigenic stimulation of the injected substance and the collagen inducing property of synthetic dermal fillers. Any such treatments should only be considered with caution and when the condition is stable, or preferably inactive. If you have further questions about cosmetic procedures in SSc, please chat further with your doctor.

In Conclusion

SSc affects the skin in many varied ways. This can impact how we function, our ability to perform daily activities, how we look, how we feel about ourselves and much more. Openly addressing these impacts with your treating doctors is vital, so that each complex aspect of skin involvement can be managed in the best way possible for you.

Author: A/Prof Amanda Saracino
Dermatologist; Melbourne Dermatology Clinic and Emeritus Research Associate Professor; University College London

Scleroderma Australia: PO Box 57, Melton Vic 3337
02 9990 5159 hello@sclerodermaaustralia.com.au

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