

Medicine for Managers

Dr Paul Lambden BSc MB BS BDS FDSRCS MRCS LRCP DRCOG MHSM



Scleroderma

Scleroderma, also known as Systemic Sclerosis, is a group of rare, chronic diseases in which normal tissue is replaced by thick, dense fibrous tissue, which leads to hardening and tightening of the skin. In normal circumstances, the body's immune system defends against disease and infection, but in scleroderma, it triggers other cells to produce too much of the fibrous protein collagen, similar to scarring.

Scleroderma may be categorised as either localised or systemic.

Localised scleroderma is the more common form of the disease and normally only produces patchy changes localised to the skin.

They may appear as streaks on the skin and they are often mild and may resolve over time, obviating the need for treatment.

Systemic Scleroderma may be broken down into **diffuse** and **limited**.

The diffuse form affects many parts of the body including the heart and blood vessels, lungs, kidneys, gastro-intestinal tract, joints and muscles. It impairs normal bodily function and, in its most serious form, is life-threatening.

The limited form is also known as **CREST** syndrome, thanks to a mis-spelling by the Americans.

Calcinosis (abnormal calcium deposits in skin)
Raynaud's phenomenon
Esophageal dysfunction (difficulty swallowing)
Sclerodactyly (skin tightening on the fingers)
Telangectasia (red spots on the skin)

Causes and Risk

The cause of scleroderma is unknown but it may run in families and in most cases there is an identifiable family history. In some people symptoms may be triggered by exposure to certain medications, drugs or viruses.

Some chemicals encountered at work have also been implicated. People who develop scleroderma may also have symptoms of another auto-immune disease such as rheumatoid arthritis.

It results from over-production of collagen. It is four times more common in women than men and the overall incidence is approximately 2-0-2.5/100,000 of the adult population. The



peak age of development is between 35 and 55. There is also a paediatric form which is rare.

The Symptoms

These vary according to the individual and depend on the parts of the body affected.

Most people with the disease experience skin thickening and tightening, which normally affects the fingers, hands, feet and face.

It may also involve other areas of skin. It may be accompanied by symptoms of itching and some swelling.

Affected skin looks shiny because of tightness and may become darker in colour. Some people develop spots (**telangiectasia**) on the skin, which are caused by dilatation of capillaries appearing as red or purplish clusters, which may have a spidery appearance.

Calcium deposits may also appear in the skin (**calcinosis**) which may appear as small lumps, particularly on the tips of the fingers, and which can be seen on X-ray.

Raynaud's phenomenon is common in scleroderma. It occurs because of an exaggerated sensitivity in the small blood vessels of the fingers and toes in response to cold stimuli and in some cases to emotional disturbances.

Patients displaying Raynaud's phenomenon may notice their fingers or toes become white (with constriction), blue (with reduction in blood movement) or red (with subsequent blood vessel dilation)

Digestive Problems can affect any part of the

gastro-intestinal tract. Difficulty swallowing and heartburn are more common symptoms, but bloating, diarrhoea or constipation with incontinence may be a problem.

In patients with more severe manifestations of the disease, complications may occur.

- **Fingers and toes.** Extreme disturbance of blood flow may result in the tip of the digit developing sores or areas of skin dying.
- **Heart** may display heart failure or abnormal heart rhythm may occur.
- **Lungs** Scarring in the lungs may result in difficulty breathing
- **Blood pressure** increase because of narrowing and loss of elasticity of blood vessels
- **Kidneys** may become scarred leading to kidney failure
- **Joints** may be stiff and become less flexible because of the tightening of the skin overlying them

Diagnosis

Scleroderma may be difficult to identify and diagnosis is therefore often delayed. It may be mistaken for rheumatoid arthritis.

In general the diagnosis is made by a combination of physical examination augmented by blood tests and immune studies.

Elevated antinuclear antibodies are found in 95% of patients with scleroderma.

Other tests will include electrocardiogram, radiography and CT scanning of the lung to identify any damage and other organ-specific testing if symptoms are present or there is suspicion that the organ(s) may be involved.

Treatment

The disease cannot be cured or arrested. Any treatment is based on symptom control:

- **Control of blood pressure**
- **Drugs to suppress the immune system** to reduce scarring and thickening of soft tissues
- **Digestive system** symptom control
- **Infections** may develop in damaged skin and secondary to lung involvement. Antibiotic should be available early as needed and vaccinations may protect against lung infections.
- **Pain relief** as required

Apart from medication, the patient can take steps to better manage the disease. These will include:

- Protecting the skin to keep warm and well clothed during the colder months
- Dietary changes to avoid heartburn, drink plenty, have a high fibre diet to control constipation and lift the bed head to minimise acid reflux into the oesophagus.
- Dental care regularly is important because some patients also have a dry mouth which promotes dental decay.

It is disappointing that no cure has yet been found for scleroderma but, with good management and attention to a healthy lifestyle, augmented by medical intervention when needed, many people can have healthy and active lives.

Support and assistance can be obtained at:

Scleroderma and Raynaud's UK (SRUK)

e-mail – info@sruk.co.uk

website – www.sruk.co.uk

Contact – 0203 893 5998

Helpline 0800 311 2756

paullambden@compuserve.com