WHAT IS SCLERODERMA?

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Literally, the term scleroderma means “hard skin”. It is a progressive disease of the vascular and immune systems and a severe connective tissue disorder. In scleroderma, some unknown factor triggers the over production of collagen (body protein), causing thickening, hardening and scarring of the skin and other organs. This often affects the small blood vessels that carry blood to many parts of the body.

Scleroderma is also known as systemic sclerosis.

TYPES OF SCLERODERMA

Localized Scleroderma
- Morphea
- Linear Scleroderma

Systemic Scleroderma
- Limited*
- Diffuse
- Sine

LOCALIZED SCLERODERMA

This form of scleroderma affects some areas of the skin, but does not affect the internal organs.

Morphea is the most common form of localized scleroderma and is characterized by oval patches of inflamed, often discoloured skin. The trunk, face, and extremities may be involved.

In linear scleroderma, a band or bands of skin hardens or thickens on the trunk and/or extremities.

SYSTEMIC SCLERODERMA

This form of scleroderma involves not only the skin but also internal organs, most commonly the digestive, circulatory, pulmonary, and muscular systems. Systemic scleroderma is divided into three forms: limited, diffuse and sine.

Sine may resemble either limited or diffuse systemic sclerosis causing damage in lungs, kidneys and/or blood vessels. However, unlike other forms of systemic sclerosis, the skin in sine is not usually involved.

*Limited scleroderma is often referred to as CREST. This is an acronym that stands for a combination of symptoms:

C – Calcinosis – small white calcium lumps forming under the skin

R – Raynaud’s Phenomenon – poor circulation in the fingers and/or toes. Small blood vessels in the fingers tend to narrow and decrease blood flow causing patients to be unduly sensitive to cool temperatures. This narrowing can also be a response to stress or emotion. Fingers often turn white, then blue

E – Esophageal dysfunction - difficulty swallowing, heartburn, or regurgitation

S – Sclerodactyly – skin of fingers and sometimes toes become thick and shiny. Affected digits may be difficult to move and may become fixed in a bent position

T – Telangiectasia – small clusters of dilated blood vessels in the skin especially on the face and fingers and palms of hands

The CREST syndrome commonly manifests itself slowly over a period of ten to twenty years. Usually it involves the skin first, then the esophagus, lungs, and bowels.

Diffuse scleroderma is the most serious internally involved form of the disease. It involves many of the internal organs of the body: esophagus, the digestive tract, kidney, heart, and/or lungs. The skin involvement includes the face, neck, torso, and both hands, arms, feet and legs.

Sine may resemble either limited or diffuse systemic sclerosis causing damage in lungs, kidneys and/or blood vessels. However, unlike other forms of systemic sclerosis, the skin in sine is not usually involved.

SYMPTOMS

Scleroderma is highly individualized, so it affects patients in different ways. Symptoms and severity of the symptoms differ greatly.

SKIN:

The most commonly identified symptom of scleroderma is the gradual thickening and tightening of the skin. Ulcers, especially on the fingertips, are common. They can be slow to heal because of poor circulation.

MUSCLE WEAKNESS:

Muscles weaken and can become painful.

DIGESTIVE SYSTEM AND GASTROINTESTINAL TRACT:

Swallowing difficulties can result from the narrowing of the esophagus. Digestive difficulties range from poor absorption of nutrients to slow movement of food. The weakened muscles of the gastrointestinal tract can lead to a "backwash of stomach acid."

DENTAL:

Because of tightening of facial skin, dental health may be compromised. Dry mouth can cause difficulties in swallowing, which in turn may lead to an increase in tooth decay.

KIDNEY:

Early signs of kidney damage may include high blood pressure and an excess of protein in the urine (detected by a urine test). Renal crisis is a severe complication of scleroderma and unless treated promptly may lead to kidney failure.

RAYNAUD'S PHENOMENON:

This is a very common symptom and is generally believed that 98% of scleroderma patients have Raynaud's Phenomenon. (See the information on CREST).

JOINTS:

Joints can become stiff and sore, similar in many respects to arthritis.

SJOGREN'S SYNDROME:

Sjogren's Syndrome is dry eyes and mouth due to a decrease in secretions of the tear ducts and salivary glands.

LUNGS:

A build up of fibrosis (scarring) in the lungs and/or weakened respiratory muscles leads to shortness of breath and persistent coughing. The fibrosis affects oxygen absorption and may lead to Pulmonary Arterial Hypertension.

HEART:

The muscles that surround the heart may become thickened and scarred, decreasing heart contractions. This can cause chest pains and irregular heartbeats.

NON–SPECIFIC SYMPTOMS:

These include extreme fatigue, general weakness, weight loss, hair loss and vague aches in muscles, bones and joints.

* Report any change in symptoms or new systems to your doctor.

CAUSES

While the cause of scleroderma is not known, there are several theories being studied which involve different systems of the body: the immune system, the vascular system, and the connective tissues. Some research indicates an infectious component with a virus like organism acting as a trigger. It is generally believed that scleroderma is neither contagious nor inherited. There has been some indication of a genetic predisposition to the disease with an environmental trigger.

WHO DEVELOPS SCLERODERMA?

Although scleroderma strikes every age, sex, and ethnic background, more than 80% of patients are women between the ages of 30 to 50. There is no authoritative incidence rate of scleroderma available, but the most conservative estimate is 9,000 cases and responsible estimates are as high as 40,000 cases.

TREATMENT

Although there is no known cure for scleroderma, symptoms can be moderated with medication and lifestyle changes. Some medications are aimed at specific symptoms, while others are aimed at decreasing the activity of the immune system. Because of the advances in treatment, patient survival has improved a great deal over the past years.

We would like to thank **Dr. Margaret Larché, MBChB, MRCP(UK), PhD** for her assistance with this information pamphlet.

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