Localised Scleroderma (Morphoea)

Localised scleroderma causes one or more patches of hard skin. There are different types. The common plaque type usually causes no problems (apart from sometimes an unsightly appearance) and tends to fade in time. The linear type can cause problems to underlying tissues such as muscles and bones. Often no treatment is needed for the plaque type but treatment may be advised in more severe cases and for the linear type.

What is localised scleroderma?

Scleroderma is a condition that causes areas of the skin to become harder than usual. Hence the name scleroderma, which means hard skin. There are two kinds of scleroderma.

- **Localised scleroderma (also known as morphoea or morphea)** only affects the skin. In some cases it can spread to the tissues underneath the skin, such as muscles and bones.
- **Systemic sclerosis** affects the skin but may also involve the body’s internal organs. See separate leaflet called Scleroderma - Systemic Sclerosis for more details.

The rest of this leaflet is only about localised scleroderma.

What are the symptoms of localised scleroderma?

Localised scleroderma usually comes on gradually. Areas of the skin may be thickened and discoloured, and hair may be lost over the area of scleroderma. There are different types of localised scleroderma. The most common types are:

- **Plaque morphoea** - these are oval patches that can range from 2-20 cm across. They start off mauve in colour, then gradually become white. Older patches may become brown. The surface is smooth, shiny and hairless. One to three plaques may develop in different areas of the skin. This type mainly affects adults and usually has no other symptoms or problems, although the affected skin can look unsightly.

- **Superficial morphoea** - is similar to the plaque type. It is typically seen as symmetrical mauve-coloured patches, usually in skin folds of the groin, armpits or under breasts. This type is most commonly found in middle-aged women.
- **Linear scleroderma** - usually occurs on an arm or leg of a child. It is a long, narrow area of thickened skin. In more severe cases the tissues just beneath the skin are affected which may cause scarring (contractures) underneath the skin.
- **En coup de sabre** - this is a deep form of linear scleroderma affecting the scalp and temple. The name comes from its shape, meaning 'like a sword cut'. Hair is lost over the affected skin, and the skull bone may be shrunk underneath it.
- **Generalised morphoea** - this is a rare type where there are more plaques spread around different areas of the body.
Are there any complications of localised scleroderma?

Usually, localised scleroderma does not cause any complications. It is not linked to the systemic sclerosis form of scleroderma and it does not involve internal organs.

Some types of localised scleroderma may affect not only the skin but the tissues directly underneath it such as bone or muscle. This only happens in the deeper types of scleroderma (linear scleroderma and en coup de sabre). In this situation, the scleroderma can affect growth in the underlying tissues and so may cause some degree of deformity. About one third of people with deep localised scleroderma have this type of problem. Rarely, if the affected skin is located on the head, the brain or eye can be affected.

What causes localised scleroderma?

The cause is not clear. What is known is that cells called fibroblasts make too much of a protein called collagen. The collagen gets deposited in the skin, causing scarring and thickening (fibrosis).

It is not known why the fibroblasts produce too much collagen in the areas of affected skin. It is probably some fault with the immune system. It is sometimes seen after the development of diseases in which the immune system attacks the body's own cells (autoimmune conditions), such as lichen sclerosus and lichen planus. It can also occur after tick bites (Lyme disease), measles, radiotherapy, local injury to the skin and pregnancy. Most of the time, however, there is no obvious cause.

How common is localised scleroderma?

Localised scleroderma is uncommon. The exact number of people affected is not known. However, it is thought that quite a number of cases are not diagnosed. This is because many people who have a small plaque or two of localised scleroderma causing no symptoms may not report it to a doctor. It is two or three times more common in women than in men. It can occur at any age and is as common in children as it is in adults. About 3 in one million children in the UK and Ireland develop localised scleroderma every year. Children are nine or ten times more likely to develop localised scleroderma than systemic sclerosis.

How is localised scleroderma diagnosed?

Localised scleroderma can usually be recognised by its appearance. The diagnosis can be confirmed by a biopsy. A biopsy is a procedure in which a small sample of skin is removed under local anaesthetic and examined under the microscope. Sometimes blood tests can give a clue but there is not a specific blood test for this condition. In some cases an ultrasound scan or a magnetic resonance imaging (MRI) scan may be used to assess whether the scleroderma is affecting tissues underneath the skin. X-rays are occasionally used to check that children's bones are growing properly.

What is the treatment for localised scleroderma?

The most common form, plaque morphoea, does not always need treatment. This form often does not cause symptoms and often gradually improves or clears away after a number of years. However, strong creams or ointments are sometimes used as they may help to stop it spreading. Creams which may be used include:

- Cream or ointment containing calcipotriol.
- Tacrolimus ointment.
- Imiquimod cream.
- Steroid creams or ointments.

For other types of localised scleroderma, treatment will vary depending on the individual situation, the severity of the condition and whether underlying tissues are affected. One of the creams or ointments above may be used in some cases. If it is too widespread or too deep or too severe, one or more of the following treatments may be used:

- Ultraviolet light therapy.
- High-dose steroids.
- Medicines affecting the immune system, such as methotrexate.
- Physiotherapy or surgery may help if the skin is very tight or if there is a deformity or scar underneath the skin.

What is the outlook (prognosis)?

With the plaque morphoea type of localised scleroderma, in many cases the plaques last 3-5 years before softening and fading. The affected areas of skin may gradually go back to normal; however, a brown stain may remain and sometimes a small depressed area of skin. In some cases the plaques remain for many years.

Linear-type localised scleroderma tends to persist for longer but may improve after several years. It may come and go and may flare up after a long time of being settled. However, linear-type localised scleroderma, especially the en coup de sabre subtype, may become more extensive with time. This may cause severe contractures that result in limited movement and permanent disability of an arm or leg. Damage to underlying brain and eye tissue is a potential, serious complication of the en coup de sabre subtype.

Further reading & references