

Localised Scleroderma

What is localised scleroderma?

Unlike systemic sclerosis, localised scleroderma seldom affects the internal organs of the body. There are two types of localised scleroderma, morphoea and linear scleroderma. One of the important differences between morphoea and linear scleroderma is that linear scleroderma tends to involve not only the skin but also the deeper tissues with fixation to muscle and bone.

Plaque morphoea and linear scleroderma are the main types but deep morphoea (morphoea panniculitis) is when there is a scarring process in the fat tissues and the overlying skin can become tethered to the deeper tissues. Atrophic morphoea or atrophoderma, is where there is a loss of deep tissue under the skin causing a dented appearance often without any skin thickening.

Morphoea is three times more common in women than men. It most frequently occurs between the ages of 20-40 years but 15% of cases occur in children. Localised scleroderma usually comes on gradually.

Morphoea



Early morphoea showing inflamed margin

Morphoea is the name given to localised patches of hardening (sclerosis) which can affect the skin. So why is it not called scleroderma, which as you may know means hardening of the skin? Probably because it helps to have a different name to distinguish

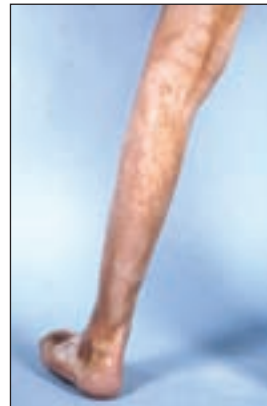
morphoea from the more generalised sclerosis, which affects both the skin and other organs in systemic sclerosis. Although there is some overlap, morphoea and systemic sclerosis are better regarded as separate entities.

Morphoea usually appears spontaneously, as asymptomatic hard patches in the skin ranging from 2 to 20 cms across. These initially have a faint purplish colour, fading centrally to develop a waxy, ivory appearance with purple coloured edges.

The patches are round or oval and may be multiple, affecting almost any part of the body. They appear most commonly on the trunk, but can occur on the face, arms or legs. Older patches may become brown. Sometimes the lesions in morphoea are deeper, involving tissue beneath the skin. The surface appears to be smooth, shiny and hairless and one or more plaques may develop in different areas of the skin. This type mainly affects adults and usually has no other symptoms or problems. Only rarely does morphoea become generalised.

Linear scleroderma

As the name suggests, in linear scleroderma, the skin affected is in a line, usually along an arm or leg. The skin may feel tight, with a loss of the normal plumpness of the skin, which becomes shiny, often ivory coloured, or sometimes darkened. Linear scleroderma can also affect the underlying muscle, fat and bone leaving a scarred appearance.



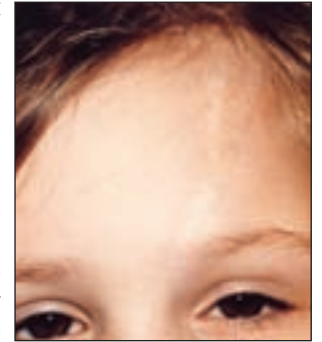
Linear morphoea

This is important to recognise in growing children, as normal growth may be reduced in the affected area. This can eventually lead to the affected arm or leg being shorter than the unaffected side. If the lesion crosses over a joint such as an elbow or knee, the tightening of the skin may prevent the joint from straightening or bending fully, and can become fixed in one position. This is called a contracture.

As linear scleroderma affects not only the skin but also the subcutaneous tissue and fat under the skin lesion, there is a risk of it causing serious deformities, particularly when it causes a joint to bend and failure of the whole limb to grow. Sometimes it will be necessary to have a splint, particularly for night use to prevent the knee bending or to hold the wrist in a good position.

En coup de sabre

One well known but uncommon deep form affects the face and scalp. This is called 'en coup de sabre' (the cut of a sword). This form of scleroderma is more common in children, beginning usually before the age of ten and affects both boys and girls. En coup de sabre is a type of linear morphoea which affects the face and often the scalp. It can spread into deeper tissues of the skin, into muscle and even bone. In this form, a characteristic linear depressed groove of scarring extends from the forehead back into the scalp and sometimes down onto the face.



En coup de sabre

The rest of the fat, muscle and bone on the same side of the face can waste away (atrophy) leaving a pinched appearance to one side of the face (hemifacial), called 'hemifacial atrophy'. This is also sometimes called the 'Parry Romberg Syndrome'.

Do we know the cause?

The cause is unclear. What is known is that cells called fibroblasts make too much of a protein called collagen. The collagen gets deposited in the skin causing fibrosis (scarring and thickening). It is not known why the fibroblasts produce too much collagen in the areas of affected skin but it is probably due to a weakness in the immune system.

Investigations have not uncovered any consistent causes. Different morphoea subtypes often coexist in the same patient, suggesting that the causative processes are similar. Morphoea may occur at the site of previous radiation therapy for cancer. Infections such as Epstein-Barr virus, varicella, measles and borreliosis have been reported to precede the onset of morphoea and have been proposed as possible triggers.

Management

In the management of local disease it is important for all lesions to be carefully noted and also at times to measure the size of a skin lesion as well as the length of the affected limb and the opposite limb. Monitoring in this way is very important to assess the state of the illness and its response to treatment.

Treatment

Patients with localised scleroderma are usually monitored by a dermatologist and sometimes a rheumatologist. Most lesions of morphea improve with time - typically over 3-5 years - and eventually clear almost completely, but residual pigmentation may persist. Treatment is not generally indicated, although local steroid applications may be used to hasten clearance. Treatment varies according to the extent of the localised scleroderma, the areas involved and the age of the patient. Not every patient with morphea or linear scleroderma will require treatment. Therapy is however, indicated for patients whose lesions are increasing in number or where there is growth impairment or functional incapacity.

The aim of drug treatment is to stop the ongoing inflammation which causes these skin lesions. Drug treatment aims to stop the skin lesions progressing and to soften the skin a little, but at present it cannot completely remove the lesions. Since there is clear evidence of inflammation in the start of the lesions, and that suppression of the inflammation early in the disease leads to less atrophy and growth problems, anti-inflammatory treatment is the treatment of choice. Methotrexate has been shown to be effective and is in use in large centres.

Plaque morphea, does not always need treatment. This form often does not cause any symptoms and can improve gradually or clear away after a number of years. However, strong steroid creams or other immunosuppressive ointments are sometimes used as they may help to prevent progression.

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.

Counselling and open discussions can help family and friends understand any physical and psychological problems associated with the condition.

Orthopaedic surgery is occasionally used for the release of joint contractures, lengthening of limbs and correction of deformities in linear and deep morphea patients.

One or more of the following treatments may be used:

- Topical therapy with immunosuppressive ointments or creams containing vitamin D.
- Ultraviolet light therapy (PUVA, UVA-1 or broadband UVA).
- High dose steroids by intravenous infusion or orally.
- Medicines affecting the immune system such as methotrexate, ciclosporin or hydroxychloroquine.
- Physiotherapy or surgery may help if the skin is very tight or if there is a deformity or scar underneath the skin. It is particularly important in children with linear scleroderma. A physiotherapist will teach the child and parents a rigorous exercise programme to be followed at home.

Newer treatments

Some newer treatments are laser therapy, photodynamic therapy and autologous tissue injection. These are still under research.

Further reading

The RSA has published a wide range of leaflets on Raynaud's and scleroderma which are available on request or visit our website (see details below).



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