

Localised Scleroderma (Morphoea, Linear and En Coup de Sabre)

LOCALISED SCLERODERMA (MORPHOEA)

Localised scleroderma is also known as morphoea. The word 'scleroderma' means 'hard skin'. Morphoea plaques are limited to the skin alone and there is no spread internally - in this way the condition is different from systemic sclerosis. The skin changes of morphoea may last for many years before they resolve. Unfortunately, morphoea cannot be cured but in most people it is not a serious condition. It may progress for several years on and off but then tends to get better by itself, and in some cases the skin may return to normal. The cause of morphoea is at present not known, but its hardness is due to an excessive build up of collagen, a protein found within the skin, which contributes to its strength.

Morphoea usually occurs for no reason although on occasion patients with the condition may describe an event such as skin trauma, a tick bite or a viral infection which is considered a trigger in their particular case. Furthermore, morphoea has been associated with other immune related conditions of the skin such as lichen planus, vitiligo and lupus. Due to this association and because some drugs that suppress the immune system seem to work in the treatment of morphoea, it is thought that an altered activity of the immune system within the skin may be of importance in the cause of the condition.

PLAQUE MORPHOEA, LINEAR MORPHOEA AND EN COUP DE SABRE

Morphoea usually appears between the ages of 20 and 50 years, more often in females, and is most commonly seen in the linear form in children. Please see our leaflet "Childhood Onset Scleroderma". Morphoea can appear for the first time during pregnancy. It is rarely seen in people with dark skin and does not usually run in families.

In plaque morphoea, the most common form, slightly raised, oval or round areas of thickened, waxy looking skin develop slowly and spread outwards. There are usually no symptoms associated with the development of these plaques although sometimes there is an itchy sensation to them. The middle of the plaques are white or ivory in colour but the edges in new plaques often have a reddish or purplish appearance to them. Older plaques of morphoea tend to look like brown bruises and no longer have a clearly defined edge.

The skin may or may not be softer than when the plaque was "new". The plaques may lose their hairs and sweat glands and the skin can look and feel dry.

Sometimes morphoea can occur in a line and usually affects a limb in a unilateral fashion. This is known as linear morphoea. When a whole limb is affected, structures deep to them (fat, muscle and even bone) may be at risk of growing poorly. If the linear morphoea crosses a joint, contractures can form that limit the movement of joints. In one type of linear morphoea, a narrow groove runs up one side of the forehead into the scalp. This type of linear morphoea is known as "en coup de sabre".

If the morphoea blisters, as is possible but uncommon, the affected skin can break down and ulcers may form. These ulcers often heal poorly and are susceptible to becoming infected. Rarely, morphoea plaques can cover much of the skin surface area (generalised morphoea). When this happens over the trunk, it can cause restriction of skin movement and difficulty with expansion of the chest when breathing.

Treatment

Plaque morphoea does not always need treatment, as it may get better by itself. However, strong steroid creams or ointments are sometimes used as they may help to reduce inflammation and prevent progression. They are applied thinly once a day and can be used safely for many weeks. They may also be used under dressings or be injected into the edges of plaques. Other creams including a group of drugs called the Vitamin D analogues have been described to have some effect on plaque morphoea. Immunosuppressive ointments including tacrolimus are also sometimes recommended.

To suppress immune activity within the skin affected by morphoea, and in rare or severe cases, oral or intravenous steroids may be given as treatment. Other treatments such as hydroxychloroquine, cyclosporin, methotrexate or mycophenolate mofetil have also been described as useful in the treatment of morphoea. Light therapy such as PUVA (using a psoralen medicine followed by ultraviolet A light) may also be used as a treatment in generalised morphoea.

UVA1 is another form of light therapy but it is not widely available and its value in the treatment of morphoea is still not established.

If any contractures of joints occur, surgery and long-term physiotherapy can be of use. Plastic surgery may be a possible form of treatment for deeper forms of morphoea, in particular en coup de sabre.

At all times, the side effects of treatment versus the condition itself must be weighed up by the patient and his or her doctor. If the disease is mild or not progressing then it may be appropriate not to apply or take specific therapy. In extensive morphoea or when there is interference with growth, as can occur in childhood, then treatment should be much more intensive.

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