MORPHOEA
LOCALISED SCLERODERMA

A sclerotic skin condition characterised by thickening/hardening (fibrosis) and inflammation of the skin

scleroderma australia

What is morphoea?

Morphoea (also known as localised scleroderma) is a sclerotic skin condition characterised by thickening/hardening (fibrosis) and inflammation of the skin. There are many subtypes of morphoea which vary according to the location of the areas of involved skin. Any subtype of morphoea can also result in deeper involvement of the underlying fat, fascia, muscle, or bone. However, morphoea does not cause fibrosis of the internal organs.

Unlike systemic sclerosis, morphoea does NOT cause:

- Fibrosis and/or vascular damage of the internal organs
- Skin thickening of the fingers and toes (sclerodactyly)
- Specific autoantibodies in the blood (such as an anti-centromere antibody or anti-Scl70)
- Abnormal small blood vessels in the fingers (nail fold capillaries)

Morphoea:

- **is a separate condition to systemic sclerosis**
- **does not cause fibrosis of the internal organs**
- **cannot develop into systemic sclerosis**

What is the cause of morphoea?

The precise cause of morphoea is not known. Morphoea is not an inherited condition, however certain genetic backgrounds are associated with an increased risk. Up to 40% of those with the more severe forms of morphoea will have a personal or family history of other autoimmune conditions. These can include thyroid disease, vitiligo, rheumatoid arthritis and others.

Morphoea can develop after an external trigger such as:

- Repeated friction
- Penetrating wound
- Surgery, radiotherapy
- Injections or vaccinations
- Insect bite or tick bite (the role of Borellia burgdorferi/Lyme disease is controversial)
- Extreme exercise

Trauma-related morphoea may occur at the affected site, or at sites distant and unrelated to the trauma.

What are the subtypes and clinical features of morphoea?

There is much debate about the best way to sub-classify the different types of morphoea. As a consequence, it is common to read sometimes confusing and inconsistent terminology in this field.

When describing morphoea, it is often useful to consider three features:

1 **Where the morphoea is located**

– This is the formal way that we classify morphoea

2 What the skin looks like at the affected sites

- There may be inflammation, with red or bruise-like discolouration
- The skin may feel tight, hard and thickened due to increased collagen deposition
- Discolouration with increased pigmentation may occur, usually after the earlier inflammation starts to settle
- The skin may become indented or concave, due to tissue loss from the fibrotic processes

3 How deep the morphoea is

- We can determine whether the morphoea is affecting the skin only, or extends to the fat, fascia, or rarely the muscles/bones directly underlying the affected skin
- This can be established by certain signs when we look at the skin, by taking a biopsy or performing an MRI
- This information will help inform us what the best treatment will be for your morphoea

A note on eosinophilic fasciitis:

Eosinophilic fasciitis is usually best thought of as a form of generalised (pansclerotic) morphoea which involves the fascia (the connective tissue under the fat). When the fascia is involved the skin can look puckered, with a cobblestone or orange peel appearance. There is usually more superficial skin involvement as well.

WHERE IS THE MORPHOEA?

- **Limited Morphoea** < 2 body sites
- **Generalised Morphoea** > 3 body sites
- **Linear Morphoea** occurring in lines/swirls
- **Mixed Morphoea**

THE AFFECTED SKIN'S APPEARANCE

- **Inflammatory** red, purple skin
- **Sclerotic** shiny, hard
- **Dyspigmented** lighter or darker skin discolouration
- **Atrophic** skin indentation

DEPTH OF THE INVOLVED TISSUE

Determined by the clinical appearance, deep incisional biopsy, and/or MRI

SUPERFICIAL

- **Epidermis/Superficial Dermis**
- **Dermis only**
- **Deep Dermis/Subcutaneous**
- **Fascial**

DEEP

- **Central Nervous System (CNS)**

Limited plaque morphoea

- The most common type of morphoea in adults
- Oval shaped patches occurring on one or two body sites

Linear morphoea

- The most common subtype of morphoea in children
- Occurs on the limbs, trunk or head/face

- Usually on one side of the body only, but can be widespread
- Linear morphoea of the head/face was previously sub-classified as:
  - En coup de sabre; linear line classically on the forehead or scalp
  - Parry Romberg/progressive hemifacial atrophy; loss of the fat, muscle and bone affecting one side of the face

Generalised morphoea

- Is widespread, affecting three or more body sites.
- There are two major patterns:
  - Disseminated plaque morphoea: scattered patches of affected skin, with intervening unaffected skin
  - Pansclerotic morphoea: circumferential, confluent skin tightness, which is usually rapidly progressive and affects most of the body surface area.

Mixed pattern morphoea

- This is when more than one of the above subtypes coexists
- The most common mixed pattern is linear morphoea of a limb, with limited plaque morphoea on the trunk.

Limited morphoea is considered mild, while linear morphoea and generalised morphoea are more severe subtypes.

Symptoms from morphoea

Other than the visible skin changes, morphoea may have no symptoms. When symptoms do occur, they may arise from the skin itself, from the deeper tissues (such as the fascia, muscles or joints), or they may be due to more widespread inflammation.

Symptoms due to skin changes

- Sometimes morphoea may cause itch, pain and/or a dull ache due to inflammation.
- Sclerosis can entrap the skin's superficial nerves resulting in pain, tingling or sometimes mild weakness.
- When morphoea occurs on a hair bearing area, it will usually cause permanent hair loss

Symptoms due to deeper tissue involvement

- Tissue involvement over joints causes joint pain, arthritis or limited joint movement (contractures).
- Teeth and jaw involvement in linear morphoea of the head/face can cause oral and dental problems such as difficulty chewing, jaw locking, or pain.
- Skull and/or brain involvement can cause headaches or in some very rare cases, neurological symptoms such as nerve palsies or seizures.

Systemic symptoms

- In the more severe types of linear or generalised morphoea, non-specific inflammatory symptoms can occur in up to 30% of people. These can include:
  - Fatigue, lethargy
  - Non-specific joint pain and/or inflammation (arthralgia, arthritis)
  - Muscle pain
  - Reflux/heartburn
  - Raynaud phenomenon (cold hands with red/white/blue colour changes)
  - Eye dryness, irritation or blurred vision

In contrast, systemic sclerosis results in direct damage and fibrosis of the lungs, heart, kidneys, and/or gastrointestinal tract. This does not occur in morphoea.

How is morphoea treated?

There is no cure for morphoea. Treatment is aimed at halting ongoing disease activity and progression. This can help minimise permanent changes and scarring from occurring. The type of treatment most appropriate for your morphoea will depend on many things, including:

- The morphoea subtype (limited, linear, generalised, mixed)
- The location of the morphoea (eg head/face, limb, trunk)
- How deep the morphoea extends beyond the superficial skin
- How active the morphoea is
- Whether you have troubling symptoms or not
- The impact the morphoea is having on you
- Your age, goals and personal preferences

All of this information is used to determine whether your morphoea is mild, moderate or severe. This assists with deciding which treatments to commence, and when to commence them.

TREATMENT OF ACTIVE MORPHOEA

MILD MORPHOEA

Topical treatments may be trialled, such as:

- Strong steroid creams
- Other prescription creams such as tacrolimus or calcipitriol

Phototherapy may be suggested, including:

- Narrowband UVB
- Topical or bath psoralen UVA (PUVA, limited availability)
- UVA1 (very limited availability)

MODERATE MORPHOEA

In addition to topical treatments and phototherapy (listed for 'mild' disease), when morphoea is of moderate severity, your doctor may also discuss the pros and cons of oral medications.

These may include:

- Courses of corticosteroids
- Immunosuppressant medications such as Methotrexate or Mycophenolate Mofetil
- Immunomodulatory medications such as Hydroxychloroquine

SEVERE MORPHOEA

When morphoea is severe, oral or injection treatments will be needed. Severe morphoea is usually linear or generalised morphoea, and/or morphoea with involvement of the tissues deep to the skin (such as the fascia or joints). Treatment options include:

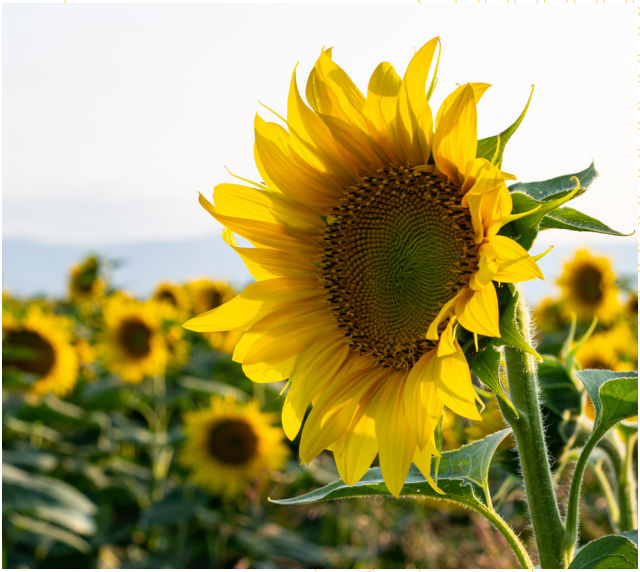
- Oral or intravenous corticosteroids
- Oral or injection immunosuppressant and immunomodulatory medications.

Physiotherapy to improve joint mobility should be undertaken cautiously when the morphoea is active, as the joint/soft tissue trauma induced by physiotherapy exercises may potentially be an ongoing disease trigger.

In some cases, surgery may be of benefit, such as autologous fat transfer, fillers or reconstructive surgery, to improve atrophy/scarring. However any procedures must only be done when there is absolute certainty that the morphoea is no longer active. There is limited availability of specialists who perform these procedures for morphoea.

What is the natural history of morphoea?

Milder forms of the morphoea tend to become inactive within 3–5 years. However more severe subtypes can follow a more protracted course, with many years of waxing and waning degrees of activity and quiescence. Relapse can occur after successful treatment, especially in morphoea that begins in childhood. In these more severe presentations, extended courses of oral or injection treatments of 4 to 5 years or more may be needed to lessen the risk of relapse after treatment.



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