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Scleromyxoedema, lichen myxoedematosus & papular mucinosis

These terms are used to describe uncommon forms of cutaneous [mucinosis](#), in which a gooey material is deposited in the skin for unknown reasons.

Scleromyxoedema

Scleromyxoedema (American spelling 'scleromyxedema') is a generalized form of lichen myxoedematosus (also known as papular mucinosis). Scleromyxoedema mainly develops in middle age and is equally common in females and males. Affected areas develop multiple papules (small bumps) and gradually become shiny and thickened. Itch tends to be mild.

The limbs (hands, forearms, thighs), the head and neck, and upper trunk are most often involved. Deep furrows across the brow can occur. As the condition progresses skin movement can be reduced, particularly at joints and around the mouth.

Scleromyxoedema is nearly always associated with monoclonal gammopathy, in which abnormally high levels of an immunoglobulin called a paraprotein are found in the blood. Usually these are IgG lambda light chain molecules. A small increase in the number of plasma cells in the bone marrow may be found.

Other affected organs may include the heart, muscles, kidneys, lungs, nervous system and joints with potentially serious consequences. Spontaneous improvement can occur but is very uncommon. Scleromyxoedema may rarely be associated with multiple myeloma, a malignancy of the plasma cells, worsening the prognosis.

Nephrogenic fibrosing dermopathy

Nephrogenic fibrosing dermopathy is a recently recognised fibrosing disorder originally identified as a scleromyxoedema-like cutaneous disease in patients with renal disease. It may also cause fibrosis of internal organs, called nephrogenic systemic fibrosis, which may ultimately prove fatal. The condition has been linked to exposure to gadolinium-containing contrast material during magnetic resonance imaging.

Localised lichen myxoedematosus

Localised lichen myxoedematosus is less serious than the generalised form:

- Skin lesions are not as widespread
- Internal organs are not involved
- There are no abnormal protein levels in the blood

Subtypes of lichen myxoedematosus include:

- Discrete lichen myxoedematosus
- Acral persistent papular mucinosis
- Self-healing cutaneous mucinosis
- Cutaneous mucinosis of infancy
- Nodular lichen myxoedematosus.

The cause is unknown but a few cases have been linked with [HIV infection](#), Hepatitis C virus, exposure to toxic oil and contaminated L-tryptophan.

Investigations

[Skin biopsy](#) is the main diagnostic test to assess for suspected mucinosis.

Blood tests may include:

- Paraproteins (serum protein electrophoresis)
- Thyroid function
- Auto-antibodies including antinuclear factor

If paraprotein is present referral to a haematologist and a bone marrow biopsy may be considered.

Patients who have widespread skin involvement may be referred to a general physician for assessment of internal organ involvement.

Treatment

Treatment aims to reduce the underlying plasma cell proliferation. Scleromyxoedema is usually treated with chemotherapy medicines, which have potentially serious adverse effects.

- Melphalan
- [Cyclophosphamide](#) (with or without a [corticosteroid](#) such as prednisone)
- Chlorambucil
- 2-chlorodesoxyadenosine
- [Methotrexate](#)
- Extracorporeal photochemotherapy
- [Intravenous immunoglobulin](#)

Other options include [PUVA](#), UVA1 phototherapy, [isotretinoin](#), plasmapheresis, [dermabrasion](#) and electron beam radiation.

[Topical corticosteroid creams](#) are of limited value for localized scleromyxoedema.

Related information

On DermNet NZ:

- [Mucinosis](#)
- [Scleredema](#)

Other websites:

- [Scleromyxedema Survivors](#)
- [Lichen myxedematosus](#) - emedicine dermatology, the online textbook

Books about skin diseases:

See the [DermNet NZ bookstore](#)

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DermNet does not provide an on-line consultation service.

If you have any concerns with your skin or its treatment, see a [dermatologist](#) for advice.

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