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Scleredema

There are three types of scleredema, a type of cutaneous [mucinosis](#) of unknown cause. Scleredema should not be confused with 'scleroderma', in which the skin is fibrotic ([morphoea](#) and [systemic sclerosis](#)).

Scleredema presents with symmetrical hardening and thickening of the dermal or middle layer of the skin with mucin deposits between collagen bundles. The diagnosis is confirmed by [skin biopsy](#).

The affected areas are firm and woody plaques, sometimes slightly red or brown and often with a 'peau d'orange' (orange-skin) appearance. The face may appear expressionless and there can be difficulty opening the mouth.

All three types of scleredema can restrict movement but otherwise seldom have serious consequences. Occasionally swallowing and speech can be affected and other organs involved – the eyes, tongue, parotid gland, muscles, joints and heart.

Scleredema may be associated with systemic diseases. These include:

- [Diabetes mellitus](#)
- Hyperparathyroidism
- [Sjögren syndrome](#)
- [Rheumatoid arthritis](#)
- Multiple myeloma
- Malignant insulinoma
- [HIV infection](#).

How do the three types of scleredema differ?

Type 1

The acute type of scleredema typically starts with an infection, most often [Streptococcus pyogenes](#), the cause of tonsillitis. It mainly affects middle aged women and children. Hardening of the skin of the face and neck quickly develops and spreads to the upper trunk and arms. It usually improves spontaneously over six months to two years.

Type 2

This type of scleredema is not associated with infection. It starts more slowly and persists. Abnormal levels of an abnormal paraprotein (immunoglobulin) in the blood may occur, sometimes due to multiple myeloma.

Type 3

'Scleredema adultorum of Buschke', also called 'scleredema diabetorum', occurs in diabetics, particularly adult men, and is very persistent. The skin of the neck and upper back slowly thickens over months or years.

Treatment

The best way to treat scleredema is unknown, because of its rarity. Some benefit has been reported with the following:

- [PUVA](#)
- [Cyclophosphamide](#)
- [Oral corticosteroids](#)

- [Ciclosporin](#)
- UVA1 phototherapy
- Electron beam radiation

Related information

On DermNet NZ:

- [Mucinoses](#)

Other websites:

- [Scleredema](#) – 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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