



PRIMARY LOCALISED CUTANEOUS AMYLOIDOSIS

What are the aims of this leaflet?

This leaflet has been written to help you understand more about cutaneous amyloidosis. It tells you what this condition is, what it is caused by, what can be done about it, and where you can find out more about it.

What is cutaneous amyloidosis?

Amyloidosis or 'amyloid' is a group of rare diseases in which abnormal proteins, accumulate in various organs including the skin. The name 'amyloid' was given to the condition over 150 years ago because the deposits looked like starch under the microscope. (In Latin *amylum* = starch).

In primary localised cutaneous amyloidosis (PLCA) amyloid deposits only occur in the skin. The exception to this is a rare disease called nodular amyloid, which can be associated with amyloid deposits in other body organs and myeloma (a form of bone marrow cancer).

PLCA is uncommon in Europeans and occurs more frequently in people who originate from South-East Asia, South America and the Middle East. It usually starts in adult life and tends to persist for many years.

There are three main forms of PLCA: **Macular amyloidosis**, **lichen / papular amyloidosis** and **Nodular amyloidosis** (the rarest form).

What causes cutaneous amyloidosis?

The cause of PLCA is not known. The commonest trigger is repeated scratching and rubbing of the skin such as in chronic atopic eczema.

Is cutaneous amyloidosis hereditary?

PLCA does not usually run in families, but in about 1 in 10 people there is a history of the skin condition passing from one parent to a child.

What are the symptoms of cutaneous amyloidosis?

Macular and papular (lichen) amyloidosis are usually very itchy, but nodular amyloidosis does not usually cause any symptoms.

What does cutaneous amyloidosis look like?

Macular amyloidosis is most often seen on the upper back in a symmetrical distribution as greyish-brown, slightly thickened skin. The pigment may have a rippled appearance.

Lichen amyloidosis appears as clusters of small skin coloured, reddish or brown scaly spots, which can merge together to form raised thickened areas especially on the shins and lower limbs. The arms and back can also be affected.

Nodular localised cutaneous amyloidosis can affect any part of the body in the form of one, a few or many lumps ranging from a few millimetres to centimetres. They can be red, pink or brown. Sometimes, several lumps fuse together to form raised areas of skin (plaques).

How is cutaneous amyloidosis diagnosed?

When a diagnosis of cutaneous amyloid is suspected a skin biopsy (tissue sample) may be taken under local anaesthetic to confirm the diagnosis. Pathologists can do special tests (stains) on the skin sample to check for the characteristic appearance of amyloid deposits which are found in the dermis (deeper skin layer). Other tests including blood and urine tests may be done if it is necessary to investigate whether someone has an associated disease, especially myeloma.

Can cutaneous amyloidosis be cured?

Unfortunately, there is no cure for PLCA. The aim of treatments is to reduce the symptoms.

How can cutaneous amyloidosis be treated?

Many different treatments have been tried, but large studies are lacking and there is no preferred therapy.

For lichen amyloidosis and macular amyloidosis, the treatments are similar to those used for eczema. They include antihistamine tablets, corticosteroid creams and ointments, vitamin D ointment (e.g. calcipotriol) and ultra-violet light ([phototherapy](#)). Occluding the skin with a hydrocolloid dressing after application of a steroid ointment or cream can improve the effectiveness, reduce irritation and flatten the skin lesions.

Oral medication options include [acitretin](#) (a derivative of vitamin A), [colchicine](#), [ciclosporin](#) (an immune-suppressive drug) and low doses of amitriptyline (an antidepressant drug).

Lesions of nodular amyloidosis can be removed surgically if causing symptoms, but they often re-grow. Excision, curettage and cautery (scraping the lump off and stopping the bleeding with heat or destroying skin with 'ablative' lasers require a local anaesthetic injection. [Cryotherapy](#) can be done without local anaesthetic. Steroid injections may help to shrink the lumps.

Self-care (What can I do?)

Friction can aggravate lichen and macular amyloidosis, so try to avoid rubbing or scratching the skin. A non-prescription menthol-containing cream can be used for instant itch relief. Habit reversal techniques may also help to reduce scratching behaviour. Any associated dryness of the skin should be managed with Regular reduce of moisturisers and soap substitutes

Where can I get more information about cutaneous amyloidosis?

Web links to detailed leaflets:

<http://www.dermnetnz.org/systemic/amyloidosis.html>

<http://emedicine.medscape.com/article/1102770-overview>

This leaflet aims to provide accurate information about the subject and is a consensus of the views held by representatives of the British Association of Dermatologists: individual patient circumstances may differ, which might alter both the advice and course of therapy given to you by your doctor.

This leaflet has been assessed for readability by the British Association of Dermatologists' Patient Information Lay Review Panel

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