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 describes a group of rare conditions in which abnormal proteins, known as amyloid, accumulate in various organs. Amyloid (Latin ‘amylum’ means starch) has a characteristic, ‘starch-like’ appearance under the microscope.

In the forms of cutaneous amyloidosis discussed in this leaflet, only the skin is affected, but usually no other organs. Only very seldom does the rarest of the three main types of cutaneous amyloidosis, nodular localised cutaneous amyloidosis, develop into systemic amyloidosis with internal organ involvement. On the other hand, patients with multiple myeloma, which is a form of cancer of cells in the blood and bone-marrow, can also develop nodular localised cutaneous amyloidosis.

Cutaneous amyloidosis is rare in Western populations and occurs more commonly in South-East Asia, South America and some populations of the Middle East. It starts early in adult life and then tends to persist.

Cutaneous amyloidosis can occur together with or as a result of other chronic skin conditions, most commonly atopic eczema.

What causes cutaneous amyloidosis?
The cause for amyloidosis is not known. In some forms of amyloidosis, chronic scratching and rubbing to certain areas of the skin increase likelihood of amyloid deposition. This can also be seen in chronic atopic eczema.

**Is cutaneous amyloidosis hereditary?**

Cutaneous amyloidosis does not usually run in families, but in about 9% of lichen amyloidosis sufferers, there is a history of the skin condition passing from one parent to a child.

**What are the symptoms of cutaneous amyloidosis?**

There are three main forms of cutaneous amyloidosis with different symptoms.

The most common form is lichen amyloidosis, which produces symptoms similar to eczema and can be very itchy.

Less common is macular amyloid, which may be less itchy. These two forms can sometimes occur together and look very similar, when affected skin is examined under the microscope.

The least common form is nodular localised cutaneous amyloidosis, which may not cause any symptoms, apart from its appearance.

**What does cutaneous amyloidosis look like?**

Lichen amyloidosis presents as groups of little skin coloured, reddish or brown scaly spots, which can merge together to form plaques, most often on the shins and lower limbs. The arms and back can also be affected.

Macular amyloid is most often seen on the upper back in a symmetrical distribution as greyish-brown, slightly thickened skin.

Nodular localised cutaneous amyloidosis can be on any part of the body in the form of isolated, few or many lumps ranging from a few millimetres to centimetres. The lumps can be red, pink or brown. Sometimes, if there are several lumps grouped together, plaques are formed.

Apart from these 3 main types, various other even rarer forms of cutaneous amyloidosis have been described, for example one in which areas of skin become thinner and paler.
How is cutaneous amyloidosis diagnosed?

When cutaneous amyloid is suggested by the appearance of the skin, often a skin sample (biopsy) is taken under a local anaesthetic injection to confirm the diagnosis. Under the microscope, amyloid has a characteristic appearance.

In order to exclude an internal cause for amyloid deposition in the skin, blood and urine tests may be requested. In localised cutaneous amyloidosis, often all these tests are normal.

Can cutaneous amyloidosis be cured?

Unfortunately, amyloidosis cannot be cured, and all currently known treatments aim to control the symptoms.

How can cutaneous amyloidosis be treated?

Many different treatments have been tried, which in itself shows that there is no easy one-fits-all solution.

For lichen amyloidosis and macular amyloidosis, the treatments are similar to what is used in eczema: antihistamine tablets can improve the itching. Different steroid ointments, vitamin D creams (calcipotriol) or courses of ultraviolet light treatment can improve the skin. Steroid ointments under hydrocolloid dressings can help the itching and flatten the skin lesions.

Acitretin (a type of vitamin A tablet) or ciclosporin (an immune-suppressive drug) have helped some patients.

If there are isolated or troublesome lesions of nodular localised cutaneous amyloidosis, they can sometimes be removed with various methods, but unfortunately often grow back. Excision, curettage and cautery (scraping the lump off and stopping the bleeding with heat), as well as taking the skin off with ablative lasers, require the use of a local anaesthetic injection. Cryotherapy (freezing the lump with liquid nitrogen), steroid creams or steroid injections can also be tried to shrink the lumps.

Self care (What can I do?)

As friction can aggravate amyloidosis, any rubbing or scratching of the skin should be avoided as much as possible.
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

For details of source materials used please contact the Clinical Standards Unit (clinicalstandards@bad.org.uk).

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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