CONDITION

Primary cutaneous amyloidosis

Primary cutaneous amyloidosis causes abnormal protein deposits in the skin, leading to itchy, discoloured, or raised patches.

WHAT IT IS
Amyloid protein deposits limited mainly to the skin

BODY SYSTEM
Skin

MAIN SYMPTOMS
Itching, dark patches, raised bumps, firm nodules

WHAT CAUSES IT
Usually unknown; sometimes inherited or linked to repeated scratching

HOW IT IS DIAGNOSED
Skin examination and biopsy with amyloid staining

MAIN TREATMENT
Itch control, skin treatments, light therapy, or procedures

CAN IT BE CURED?
No - treatments control symptoms, recurrence common

IS IT AN EMERGENCY?
No - systemic symptoms need prompt assessment

LONG-TERM OUTLOOK
Usually skin-limited and persistent but medically manageable

Overview

Primary cutaneous amyloidosis is a condition in which an abnormal material called amyloid collects in the skin. Amyloid is made from proteins that have folded into an unusual shape and formed deposits.

"Primary" means the deposits begin in the skin rather than resulting from another known condition. "Cutaneous" means related to the skin. Most forms remain limited to the skin and do not affect internal organs.

There are three main forms:

Some people have features of both macular and lichen amyloidosis. This is sometimes called biphasic amyloidosis.

The condition is not contagious and is not skin cancer. It may still matter greatly because itching can disturb sleep, scratching can injure the skin, and changes in appearance can affect confidence. Nodular cutaneous amyloidosis also needs careful assessment because, unlike the more common macular and lichen forms, it can occasionally be associated with amyloid deposits elsewhere in the body or with an abnormal group of antibody-producing cells.

- Macular amyloidosis causes flat, brown or grey-brown patches, often on the upper back
- Lichen amyloidosis causes clusters of small, firm, very itchy bumps, often on the shins
- Nodular cutaneous amyloidosis causes one or more larger, waxy or firm lumps

Symptoms

Symptoms depend on the form of primary cutaneous amyloidosis.

Macular amyloidosis usually appears as flat, rippled or net-like areas of darker skin. The patches commonly develop on the upper or middle back, especially between the shoulder blades. They may also occur on the chest, arms, or thighs. Itching ranges from mild to troublesome.

Lichen amyloidosis produces many closely grouped bumps. They are usually skin-coloured, brown, or grey and may have a rough surface. The shins are a common site, but the thighs, arms, and back can also be affected. Itching may be intense and can lead to repeated rubbing or scratching.

Nodular cutaneous amyloidosis causes firm plaques or lumps. They may look pink, red-brown, yellowish, or waxy. The face, legs, arms, or genital area may be affected. Nodules may contain small visible blood vessels and can sometimes bruise or bleed after minor injury.

Symptoms may include:

The appearance can overlap with eczema, lichen planus, scars, and other skin conditions. A photograph or symptom description alone cannot confirm the diagnosis.

- Long-lasting itching or burning
- Darker or uneven skin colour
- Rough, thickened, or raised skin
- Firm plaques or nodules
- Scratch marks, crusting, or broken skin

Causes and risk factors

In macular and lichen amyloidosis, the amyloid is thought to come from damaged keratinocytes. Keratinocytes are the main cells in the outer layer of the skin. When these cells are injured and break down, part of their protein material may be changed into amyloid and deposited in the upper skin.

The reason this happens is often unknown. Long-term rubbing, friction, or scratching may contribute in some people. Examples include rubbing the skin with rough towels, brushes, or other abrasive materials. However, not everyone with the condition has a history of repeated friction, and scratching may sometimes be a result rather than the original cause.

Some families have an inherited tendency to develop macular or lichen amyloidosis. Changes in genes involved in itch signalling have been identified in certain inherited forms. Having an affected relative does not mean that everyone in the family will develop symptoms.

Nodular cutaneous amyloidosis develops differently. Its amyloid is usually made from parts of antibodies called light chains, produced by plasma cells within the skin. Plasma cells are immune cells that normally make antibodies. This is why nodular disease may lead to blood, urine, or other tests to look for a related plasma-cell condition or amyloid outside the skin.

Primary cutaneous amyloidosis is not caused by poor hygiene, and it cannot be passed from one person to another.

Diagnosis

A healthcare professional will examine the colour, texture, pattern, and location of the affected skin. They may ask when the changes began, how much they itch, whether they have spread, and whether relatives have similar skin changes.

A skin biopsy is often used to confirm the diagnosis. During a biopsy, a small sample of numbed skin is removed and examined under a microscope. Special stains, commonly Congo red stain, can identify amyloid. Under polarised light, Congo red-stained amyloid has a characteristic appearance.

Further testing may be recommended if you have nodular cutaneous amyloidosis, unusual biopsy findings, or symptoms that could suggest internal involvement. Tests may include blood and urine studies for abnormal antibody proteins, blood counts, kidney and liver tests, and other investigations selected for your symptoms.

These tests help distinguish primary cutaneous amyloidosis from systemic amyloidosis. In systemic amyloidosis, amyloid collects in organs such as the kidneys, heart, nerves, or digestive system. Macular and lichen amyloidosis are generally skin-limited. Nodular cutaneous

amyloidosis needs more careful initial assessment and ongoing follow-up.

Treatment and management

Treatment aims to reduce itching, interrupt the itch-and-scratch cycle, improve the skin's texture or appearance, and prevent further injury. No single treatment works for everyone, and deposits may return after apparently successful treatment.

Regular moisturiser can reduce dryness and make rough skin more comfortable. Avoiding abrasive rubbing may help prevent further irritation. Keeping fingernails short and using cool compresses can reduce skin damage during itchy periods.

Prescription treatments may include corticosteroid like prednisone or hydrocortisone creams or ointments to reduce itching and inflammation. Long-term or inappropriate use can thin the skin, so these medicines should be used according to professional advice. Non-steroid anti-inflammatory creams, such as calcineurin inhibitors, may be considered in selected areas.

Keratolytic treatments soften and loosen thick surface skin. They may contain ingredients such as urea or salicylic acid. These products can irritate sensitive or broken skin, and their suitability depends on the site and extent of the condition.

Phototherapy exposes the skin to controlled ultraviolet light in a specialist clinic. Narrowband ultraviolet B or psoralen plus ultraviolet A therapy may be considered when larger areas are affected or topical treatments have not helped. Improvement is not guaranteed, and repeated ultraviolet exposure has longer-term skin risks.

For severe or persistent cases, a dermatologist may discuss oral medicines that affect skin-cell growth or itch. These treatments require monitoring and are not suitable for everyone.

Procedures such as laser treatment, dermabrasion, curettage, or surgical removal may improve selected thick patches or nodules. Scarring, pigment changes, and recurrence are possible. Nodular lesions are often managed with a procedure, but follow-up remains important even after complete removal.

Complications

Repeated scratching can thicken the skin, deepen pigment changes, and cause cracks or sores. Broken skin may become infected, although this is not a usual feature of the amyloid deposits themselves.

Persistent itching can interfere with sleep, concentration, and daily activities. Visible patches or lumps may also affect self-confidence, clothing choices, relationships, or emotional wellbeing.

Treatment can sometimes cause irritation, lighter or darker skin, scarring, or skin thinning. Procedures may remove a visible lesion without preventing amyloid from returning at the same site or developing nearby.

Macular and lichen amyloidosis do not usually progress to internal-organ amyloidosis. Nodular cutaneous amyloidosis carries a small but important possibility of association with a plasma-cell condition or systemic amyloidosis. For this reason, appropriate testing and longer-term review are more important for the nodular form.

Prevention

Primary cutaneous amyloidosis cannot always be prevented. This is especially true when there is an inherited tendency or no identifiable cause.

Reducing repeated friction may help limit irritation or new changes in some people. Avoid vigorously rubbing itchy areas with rough cloths, brushes, or exfoliating tools. Treating itch early may also help interrupt the cycle in which itching leads to scratching, skin injury, and more itching.

These measures may reduce aggravation, but they cannot reliably stop the condition from developing or returning.

Living with it and what to expect

Primary cutaneous amyloidosis often lasts for years. The patches or bumps may remain stable, become more noticeable, or slowly extend to nearby skin. Symptoms can improve and worsen over time, especially if heat, dryness, clothing, or friction affects itching.

Treatment commonly controls symptoms rather than removing all amyloid. You may need to try more than one approach. Even when the skin becomes smoother or less itchy, colour changes can fade slowly and may not disappear completely.

Routine follow-up is useful when symptoms are difficult to control, treatment is causing side effects, or the diagnosis needs to be reviewed. Macular and lichen amyloidosis may not require frequent appointments once symptoms are stable.

Nodular cutaneous amyloidosis generally needs longer-term follow-up. This may include

repeat assessment for symptoms or test results suggesting amyloid outside the skin. The exact schedule is based on the biopsy findings, initial tests, and your overall health.

Most people with skin-limited macular or lichen amyloidosis continue their usual work, exercise, travel, and relationships. Managing itch and finding acceptable skin care can take time, but the condition itself does not usually shorten life.

When to get medical help

Arrange a routine appointment if you develop persistent itchy, darkened, thickened, or lumpy skin that has not been assessed. You should also seek review if a known area is spreading, bleeding repeatedly, becoming painful, or no longer responding to your usual management.

Contact a healthcare professional promptly if you have nodular cutaneous amyloidosis and develop unexplained swelling of the legs, foamy urine, increasing breathlessness, numbness, unusual weakness, unexplained weight loss, or persistent diarrhoea. These symptoms have many possible causes, but they may require assessment for a condition affecting more than the skin.

Seek urgent medical care for rapidly increasing redness, warmth, swelling, pus, fever, or severe pain around broken skin, as these may suggest infection.

Call emergency services for severe trouble breathing, chest pain, fainting, or sudden severe weakness. These are not typical symptoms of skin-limited amyloidosis, but they require emergency assessment regardless of the cause.

Common questions

Is primary cutaneous amyloidosis cancer?

No. Macular and lichen amyloidosis are not cancers. Nodular cutaneous amyloidosis involves local plasma cells, but this does not automatically mean cancer. Testing may be needed to rule out a related plasma-cell condition.

Can skin amyloidosis spread to my organs?

Macular and lichen amyloidosis usually stay in the skin. Nodular cutaneous amyloidosis has a greater, though still uncommon, association with systemic amyloidosis or a plasma-cell condition. Initial testing and follow-up help monitor this risk.

Will the dark patches go away?

They may fade with successful itch control and treatment, but this can be slow. Some pigment

or texture change may remain. Scratching and friction can make the discolouration more noticeable.

Why is it so itchy?

The amyloid deposits and changes in skin nerve signalling may contribute to itch. Scratching then damages more skin and can create an itch-and-scratch cycle. Reducing irritation and treating itch are central parts of management.

Can I pass primary cutaneous amyloidosis to someone else?

No. It is not caused by an infection and cannot spread through touch, sex, shared towels, or household contact. Some forms can run in families because of inherited gene changes, but they are not contagious.

Does removing a nodule cure it?

Removing a nodule can clear that particular lesion, but amyloid may return in the treated area or appear elsewhere. Continued skin checks and any recommended medical follow-up remain important.

Sources

- DermNet New Zealand - Cutaneous amyloidosis
- British Association of Dermatologists - Primary localised cutaneous amyloidosis
- Weidner T, Illing T, Elsner P. "Primary Localized Cutaneous Amyloidosis: A Systematic Treatment Review." American Journal of Clinical Dermatology . 2017;18(5):629-642
- Schreml S, Szeimies RM, Vogt T, Landthaler M, Schroeder J, Babilas P. "Cutaneous amyloidoses and systemic amyloidoses with cutaneous involvement." European Journal of Dermatology . 2010;20(2):152-160
- National Organization for Rare Disorders - Amyloidosis