THE AUSTRALASIAN COLLEGE
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Amyloidosis

Also known as ... Cutaneous Amyloidosis

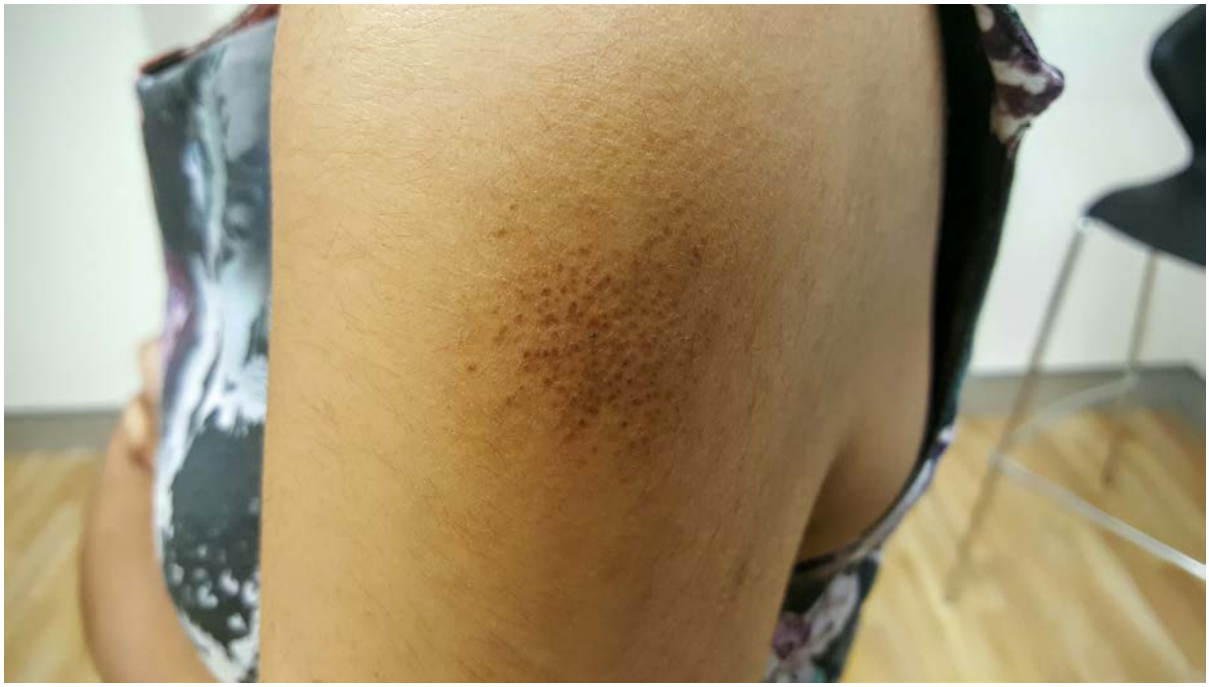
What is Amyloidosis?
Amyloidosis is a group of conditions characterised by the deposition of amyloid protein in organ systems. About 18 different types of amyloid protein have been identified.

There are three main types of amyloidosis:

- **Primary cutaneous amyloidosis** is a condition where amyloid protein is deposited in apparently normal skin without associated deposits in internal organs.

- **Primary systemic amyloid** may develop as a consequence of an underlying abnormality of the plasma cells in the bone marrow or plasma cell cancer (multiple myeloma). In systemic amyloidosis, the skin is involved along with many other organ systems such as the heart, the nerves, the kidneys and the liver.

- **Secondary systemic amyloidosis** is seen as a complication of a severe chronic infection such as tuberculosis, or a chronic inflammatory disorder such as rheumatoid arthritis or inflammatory bowel disease. Long-term haemodialysis may be associated with systemic amyloid. In rare cases, it can also be associated with inherited disorders.



Lichen amyloidosis - image reproduced with permission of Dr Davin Lim

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What causes Amyloidosis?
In ***localised cutaneous amyloidosis***, the amyloid protein is produced by the skin cells and the protein accumulates only in the skin.

Amyloid protein can also accumulate in internal organs. When the amyloid affects more than one body organ or system, it is referred to as ***systemic amyloidosis***.

What does Amyloidosis look like?
In ***macular amyloid,*** the skin does not appear or feel thickened. However, it may show increased pigmentation and the affected area is often itchy. Sometimes the skin does become thickened and this is referred to as ***popular amyloid.***

Lichen amyloidosis is the most common form of localised skin amyloid. In this type, there are raised, persistent, individual very itchy lumps and groups of lumps (plaques) most commonly on the fronts of the legs or forearms. These areas are often pigmented.

Nodular amyloid is much less common. The biopsy findings are different. The amyloid proteins deposited in the skin do not come from the skin cells. This form may progress to systemic amyloid.

How is Amyloidosis diagnosed?
A medical history and skin examination may suggest a diagnosis of cutaneous amyloidosis.

A skin biopsy is then taken to test for the amyloid protein and exclude other skin problems that may resemble cutaneous amyloid.

How is Amyloidosis treated?
The aim of treatment is to break the itch-scratch-itch cycle.

General measures are recommended to reduce itchiness of the skin. These include:
- Minimise irritants, such as abrasive clothing, or drying soaps
- Moisturise regularly
- Use cooling topical applications such as menthol cream to help distract from the sensation of itch.

Specific treatments include:
- Strong topical corticosteroid creams with or without wrapping or wet wrap treatment
- Corticosteroids may be injected into the affected area in severe localised cases
- For macular amyloid, capsaicin cream can be effective if applied regularly for some weeks
- Phototherapy, NBUVB or PUVA treatment may help
- Ablative laser treatment
- Dermabrasion
- Rarely acitretin

What is the likely outcome of Amyloidosis?
Persistent treatment is needed as cutaneous amyloidosis often fluctuates in severity and is quite persistent. Skin irritation and itchiness is often recurrent so repeat treatment may be required.

This information has been written by Dr Amanda Saracino, Dr Lauren Young, Dr Adrian Mar and Dr Adriene Lee

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