

## Pruritic eruption on the anterior shins

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**A** 32-year-old woman presented with a long history of pruritic eruption on her anterior shins (*Figures 1 and 2*). *What is your diagnosis?*



**Figure 1.** Macular hyperpigmentation on the anterior shins.



**Figure 2.** Closer view of macular hyperpigmentation demonstrating a rippled distribution.

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**DIAGNOSIS:** Primary localized cutaneous amyloidosis.

## DISCUSSION

Primary localized cutaneous amyloidosis is a rare idiopathic dermatosis that is considered a spectrum in which the less itchy rippled hyperpigmented *macular* variant (Figure 1) occurs at one end and the extremely pruritic hyperpigmented papular *lichen* variant occurs at the other. A rare bullous variant of lichen amyloidosis exists as well (1). The diagnosis can usually be made based on clinical examination and history.

Clinically, pruritus is the presenting symptom in most patients. The most common areas of involvement include the shins, arms, and back. Familial or racial predisposition and friction may have a pathogenic role (2, 3).

The association of cutaneous amyloidosis and multiple endocrine neoplasia type 2A (MEN 2A or Sipple syndrome—a rare autosomal dominant disease characterized by medullary thyroid carcinoma, pheochromocytoma, and primary hyperparathyroidism) has been reported (4). Additionally, some authors advocate screening patients with hereditary cutaneous amyloidosis for medullary thyroid carcinoma to determine the true frequency of this syndrome (5).

## Pathophysiology

Special studies have found that amyloid in localized cutaneous amyloidosis contains keratin epitopes, which suggests derivation of the fibrillar component from keratin intermediate filaments (6) (Table). Recurrent trauma (rubbing, scratching) may lead to keratinocyte trauma and result in amyloid deposition (2, 9). Furthermore, some authors consider lichen amyloid a variant of lichen simplex chronicus in which scratching leads to necrosis of keratinocytes and eventually to the formation of amyloid in the papillary dermis (9).

## Laboratory findings

Histopathologically, both macular and lichen amyloidosis are similar and vary only with respect to the appearance of the epidermis and the quantity of amyloid present (10). Both feature deposits of amyloid in the papillary dermis that are derived from keratin peptides of necrotic keratinocytes; however, macular amyloid deposits may be scant and require special stains such as Congo red or direct immunofluorescence, while lichen amyloid deposits may fill the dermal papillae. The epidermis in lichen amyloidosis is hyperkeratotic and hyperplastic, visually similar to the epidermal changes of lichen simplex chronicus.

## Treatment

Therapies for primary localized cutaneous amyloidosis must first control symptoms and then address the cosmetic concerns of patients. Historically, dimethyl sulphoxide was used to control pruritus; however, it helps only a minority of patients and, contrary to early beliefs, it does not have amyloid-dissolving properties (11, 12). Various therapeutic modalities such as systemic antihistamines, topical or intralesional corti-

**Table. Composition of amyloid in various types of amyloidosis\***

Type	Subtype	Composition of amyloid
Localized cutaneous	Nodular	Immunoglobulin light chains produced by local plasma cell infiltration
	Lichen	Keratinocyte tonofilaments
	Macular	Keratinocyte tonofilaments
Systemic	Primary	Immunoglobulin light chains resulting from plasma cell dyscrasia
	Secondary	An apoprotein complex resulting from chronic inflammation; no cutaneous lesions are seen

\*From references 7, 8.

costeroids, etretinate, narrow-band ultraviolet B phototherapy, and dermabrasion/epidermal scraping have been useful (3, 9, 13–16).

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