Answer to Dermacase

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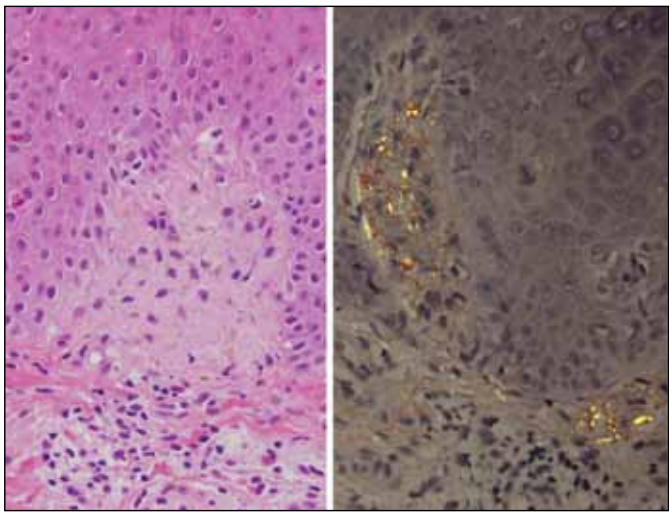
5. Lichen amyloidosis



Lichen amyloidosis (LA) is a heterogeneous disorder resulting from extracellular deposition of one of several biochemically unrelated proteins that share certain characteristic staining properties. The most characteristic property is apple-green birefringence of Congo red–stained preparations that can be observed under polarizing light. Primary localized cutaneous amyloidosis is classically categorized into 2 types—lichen and macular amyloidosis. The latter usually affects the upper back and limbs, and consists of small brownish macules distributed in a rippled pattern. It is more common among Central and South American, Middle Eastern, and non-Chinese Asian populations. Lichen amyloidosis is more common in Southeast Asian populations than in Western populations.[1] Men are affected more often than women, and the age of onset is predominantly in the fifth or sixth decades of life. Clinically, LA usually presents as a group of intensely itchy, brown, hyperkeratotic papules coalescing into large thickened plaques, predominantly located on the pretibial surfaces. Other areas of the body such as the chest, abdomen, arm, back, and thigh can also be affected. The precise pathogenesis of LA has not been fully elucidated. However, several lines of evidence, such as positive immunohistochemical staining with antikeratin antibodies and sulfhydryl groups found in the deposits, suggest that these amyloid depositions are probably derived from degenerated epidermal keratinocytes.[2] In addition, prolonged friction from a rough nylon towel and the use of horsehair gloves or artificial, rough sponges have been reported to precipitate the occurrence of cutaneous amyloidosis.[3] These observations further substantiate the speculation that focal epidermal damage elicited by repeated trauma might result in damage of keratinocytes, which subsequently converge into amyloid depositions in the papillary dermis. Recently, a growing number of reports indicate that primary cutaneous amyloidosis might be associated with various autoimmune disorders, such as systemic lupus erythematosus, rheumatoid arthritis, Hashimoto thyroiditis, sarcoidosis, and immunoglobulin A nephropathy.[4] Most patients in these reports had extensive lesions or had lesions distributed in atypical locations. This unusual connection raised the possibility that a common immune-mediated mechanism might be present in a subset of these patients with primary localized cutaneous amyloidosis.

Diagnosis

Diagnosis of LA depends on identification of characteristic skin findings—namely, intensely itchy, hyperpigmented lichenoid papules, typically appearing bilaterally on the pretibial surfaces. Definitive diagnosis requires histologic confirmation. Histopathologically, in LA, there are depositions of hyalinized amorphous materials within the papillary dermis, and Congo red stain under polarizing light gives these depositions apple-green birefringence. In addition, the histology of LA usually combines features of both macular amyloidosis and lichen simplex chronicus (LSC)—irregular acanthosis, and hyperkeratosis of the overlying epidermis that is usually secondary to chronic rubbing.

Differential diagnosis. Lichen planus (LP) is a distinct inflammatory skin disorder that predominantly affects flexural areas of the arms and legs.[5] The typical clinical features can be summarized into an easily recalled mnemonic: 6 Ps—*purple*, *pruritic*, *planar* (flat-topped), *polished* (shiny surface), *polygonal*, and *papules*. Well-developed lesions might have fine whitish reticulated lines on top of the lesion; this network is referred to as *Wickham striae*. It is a highly characteristic finding of LP and can be better observed with the aid of a magnifying lens after applying oil or water. Based on the morphology, configuration, and site of involvement, LP can be divided into several clinical variants. Among them, hypertrophic LP should be differentiated from LA. It is typified by very itchy, elevated, hyperkeratotic, shiny, papuloplaques to nodules with a purplish hue that is usually not observed in LA. Topical corticosteroids are the first-line therapy in LP.

Lichen simplex chronicus is an intensely itchy skin condition that is usually caused by chronic scratching or rubbing because of pruritus.[6] Clinically, it is characterized by thickened, scaly plaques with accentuated skin markings. They are most commonly observed on the scalp, nape of the neck, ankles, extensor aspects of the limbs, and anogenital areas. The goal of treatment is identification of the systemic causes of itch and interruption of the vicious itch-scratch cycle. Sedating antihistamines and potent topical steroids are first-line measures for these patients.

Prurigo nodularis, somewhat similar in pathogenesis to LSC, is a skin condition caused by picking and scratching resulting mostly, but not always, from underlying itch.[6] Prurigo nodularis typically presents as severely itchy, firm, 0.5- to 3-cm hyperkeratotic nodules, usually with overlying excoriation, preferentially on the extensor aspects of limbs. Because prurigo nodularis usually appears in concert with underlying itch, efforts should be made to look for any systemic causes, such as hepatobiliary tract disease, renal dysfunction, hypothyroidism or hyperthyroidism, or occult malignancy, besides the primary skin disease.[7] Treatment is similar to that for LSC, with potent topical steroids being the first-line therapy. With patients suffering from widespread disease, broad- and narrow-band UVB phototherapy, or topical or oral psoralen and UVA phototherapy, can be considered.[6-8]

Pretibial myxedema is a skin condition that is most commonly associated with hyperthyroidism and usually appears as a late manifestation of Graves disease.[9] The clinical presentation ranges from asymptomatic, nonpitting, waxy nodules or plaques to confluent induration with prominence of the follicles giving rise to a peau d'orange appearance. Although lesions are usually located on the pretibial areas, they might affect other sites of the body as well. Therefore, some authors prefer the term *thyroid dermopathy*. Pretibial myxedema usually runs a chronic course and some patients have persistent skin lesions even after thyroid disease is effectively treated. Reported successful therapies include topical or systemic steroids, compression therapy, intravenous immunoglobulin, intralesional octreotide, and plasmapheresis.[10]

Management

Treatment of LA is usually unsatisfactory and complete clinical regression is seldom achieved. Sedating antihistamines are moderately effective in relieving the associated pruritus of LA. Several other treatment modalities have been reported with variable success, and they include topical and intralesional corticosteroids, topical dimethyl sulfoxide, calcineurin inhibitors, topical psoralen and UVA phototherapy, narrow-band UVB phototherapy, dermabrasion, hydrocolloid dressings, and pulsed-dye laser therapy.[11] Systemic retinoids can provide both cosmetic and symptomatic improvement for patients with LA resistant to topical corticosteroids. Prevention of behaviour causing trauma should be addressed with patients whose LA most likely resulted from chronic friction or trauma.

Dr J.F. Chen is a senior resident and **Dr Y.F. Chen** is a dermatologist in the Department of Dermatology at the Tri-Service General Hospital in Taipei, Taiwan.

Competing interests
None declared

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