Pruritic Eruptions on the Limbs

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A 55-year-old male of Asian ancestry presents to the office with an intensely pruritic eruption of 10 years duration. He has tried numerous topical steroid medications, without significant improvement. Often he will scratch at the lesions until there is bleeding.

Examination reveals numerous red-brown hyperkeratotic papules which form large plaques on the arms, trunk and legs (Figures 1 and 2). They have a symmetrical distribution, with relative sparing of the scapular regions.

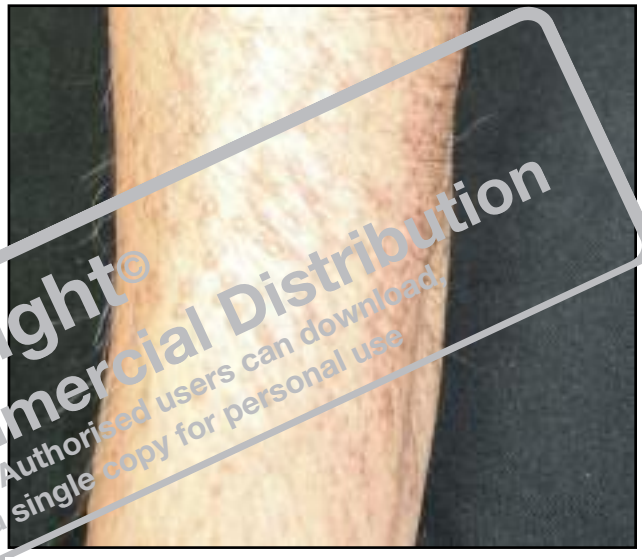

Figure 1: Plaques on patient's arm

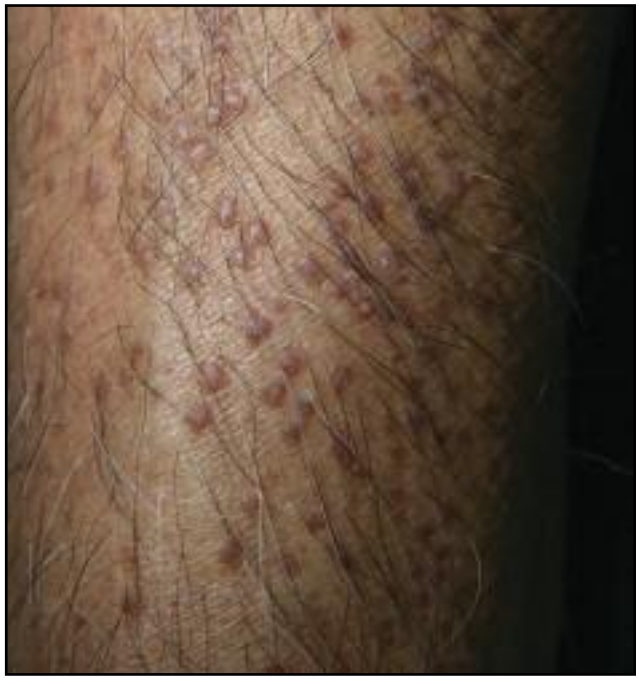

Figure 2: Plaques on patient's leg

What is your diagnosis?

a. Scabies infestation
b. Lichen amyloidosis
c. Molluscum contagiosum
d. Bullous pemphigoid

Answer: Lichen Amyloidosis (LA)

About LA

The term amyloidosis describes a heterogenous group of disorders characterized by the deposition of biochemically-unrelated proteins in the extracellular tissue of the body. However, in all instances, the deposited amyloid protein fibrils all demonstrate an affinity for Congo red staining on histology.

In systemic amyloidosis (SA), the amyloid deposition occurs in multiple organ systems such as the CNS, heart and gastrointestinal tract. SA has been

implicated in neurodegenerative disorders (*e.g.*, Alzheimer's disease) as well as malignancies (*e.g.*, multiple myeloma) and a variety of chronic inflammatory conditions, such as rheumatoid arthritis.

In contrast, localized amyloidosis only occurs in a single, tissue-specific site (*e.g.*, skin). Primary cutaneous amyloidosis is classified into three groups depending on the location of amyloid deposits in the skin: macular, lichen (papular) and nodular type. LA refers to keratin-derived amyloid deposits that develop in the papillary dermis.

LA is much more common in the Asian population. It is rare in Caucasians. Patients typically present with a longstanding history of pruritic red-brown papules located on the back and extremities. Often, the discrete papules coalesce to form large plaques. The eruption has a symmetrical distribution. Often there is a history of self rubbing the skin with a towel or brush (friction amyloidosis). A definitive diagnosis for LA can be established by taking a skin biopsy and then treating the tissue with a special stain (Congo red stain) to identify amyloid deposits under polarized light.

Familial case reports suggest the importance of genetic factors. LA has also been identified as a dermatological manifestation of multiple endocrine neoplasia type 2 (MEN2), a condition which can also cause thyroid carcinoma, pheochromocytoma or parathyroid disease.

Treatment

LA is a chronic condition and response to treatment is often unsatisfactory. Topical potent steroid creams, calcineurin inhibitors and keratolytic agents may be used. Systemic antihistamines may provide relief of pruritus and control excoriation habit. Intralesional steroid injection, cryotherapy, dermabrasion, surgical excision and laser ablation may be attempted for recalcitrant lesions. However, lesions are prone to recurrence. Some patients with widespread involvement have demonstrated significant improvement and even clearing of lesions with narrowband ultraviolet light (UVB) therapy (personal observations, S.L.). Dx

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