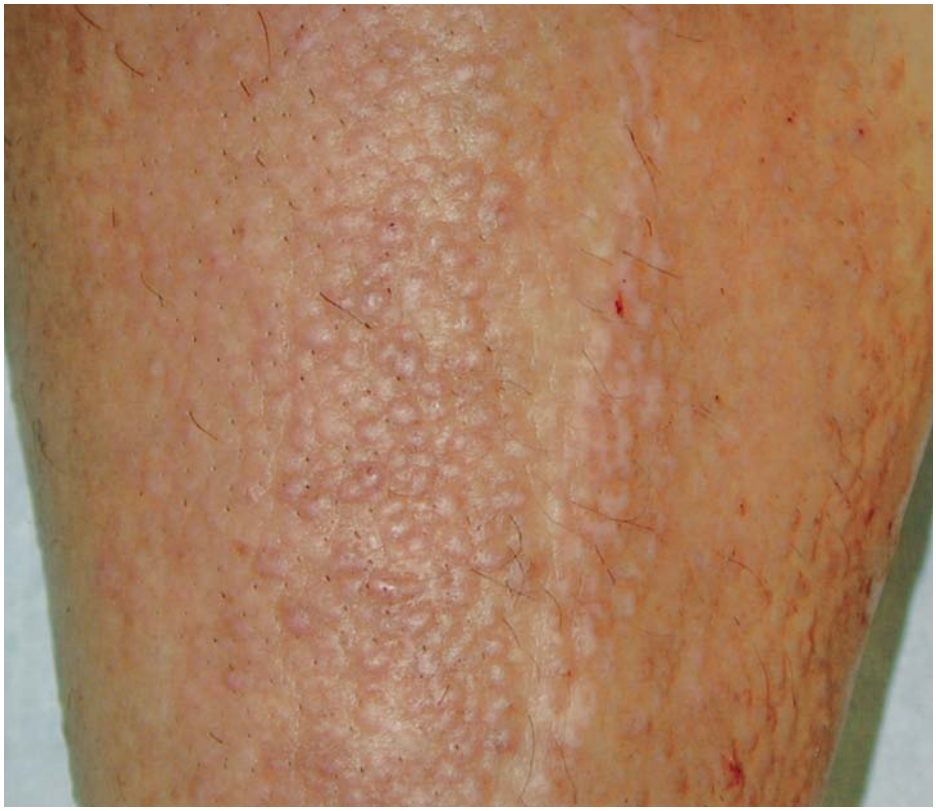
What Is Your Diagnosis?



A 63-year-old Asian woman presented with a pruritic rash on the extensor surfaces of her arms and right leg. The rash did not resolve with antihistamines, topical steroids, and a moisturizer.

PLEASE TURN TO PAGE 301 FOR DISCUSSION

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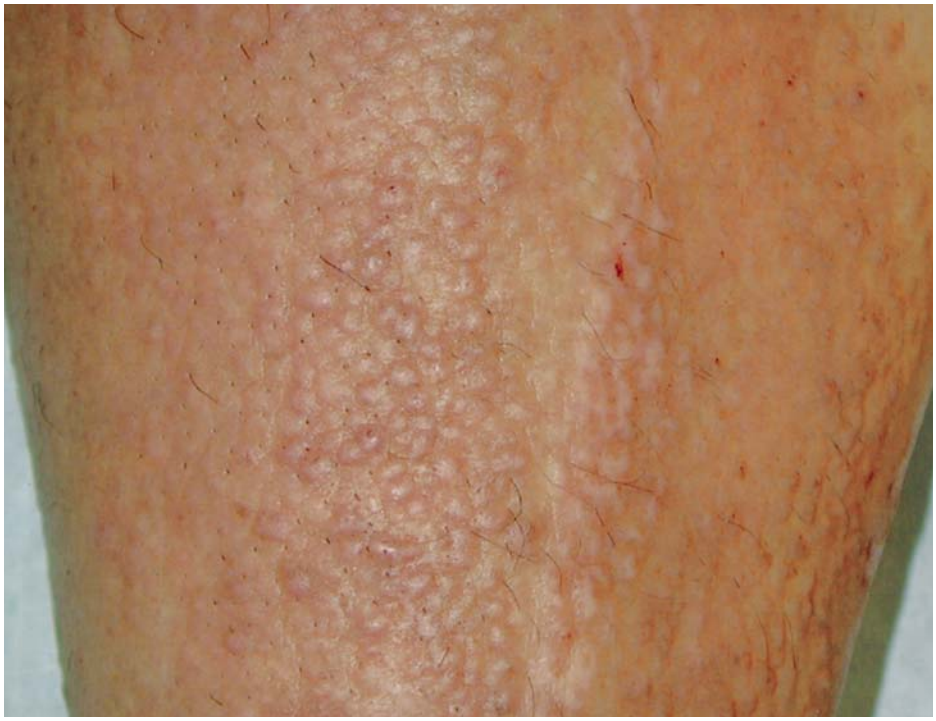
The authors report no conflict of interest.

The Diagnosis: Lichen Amyloidosis



Amyloidosis refers to a group of conditions consisting of one of a family of biochemically unrelated proteins sharing characteristic staining properties within one or more tissues.[1-3] The apple green birefringence of Bennhold Congo red–stained preparations under polarizing light is characteristic.[1,3] Amyloid deposition may occur throughout many organs of the body or may be restricted to a single tissue site.[2] The biochemical composition of amyloid fibrils varies according to the type of amyloidosis and accumulates because of different pathogenic mechanisms.[2] Under electron microscopy, all amyloid deposits are composed of linear, nonbranching, aggregated fibrils arranged in a loose meshwork.[1] The fibrils have an antiparallel β-pleated sheet configuration with polypeptide chains arranged perpendicular to the long axis of the fibrils.[1,3]

Lichen amyloidosis is the most common form of localized cutaneous amyloidosis.[3,4] The current theory is that focal epidermal damage induced by long-term scratching and rubbing causes filamentous degeneration of keratinocytes, followed by apoptosis and conversion of colloid bodies into amyloid material in the papillary dermis.[2,3,5] This theory is supported by the fact that dermal amyloid deposits cross-react immunohistochemically with keratin.[2,5] Lichen amyloidosis clinically presents as a persistent pruritic eruption of multiple discrete red-brown hyperkeratotic papules that may coalesce to form plaques.[3,5,6] This eruption principally is distributed on the pretibial surfaces but rarely may spread to the calves, ankles, dorsa of feet and thighs, and even extensor aspects of the arms.[3,5,6]

Lichen amyloidosis affects middle-aged adults and is more common among the Chinese.[1,5] In addition, lichen amyloidosis occasionally has been reported in association with connective tissue diseases, such as systemic sclerosis, primary biliary cirrhosis, and systemic lupus erythematosus.[2,5] Lichen amyloidosis also has been reported in association with multiple endocrine neoplasia, type 2, also known as Sipple syndrome, an autosomal dominant triad consisting of medullary thyroid carcinoma, pheochromocytoma, and hyperparathyroidism.[1]

The diagnosis of lichen amyloidosis depends on the histochemical, immunohistochemical, or ultrastructural demonstration of amyloid material in a

skin biopsy specimen.[2] Histologically, amyloid deposits can be demonstrated with many stains. The best known is the Bennhold Congo red stain, but lichen amyloidosis actually stains better with Ehrlich aniline crystal violet and thioflavine T. Other stains include the periodic acid-Schiff; methyl violet; various cotton dyes such as pagoda red and Sirius red, which are more commonly used in systemic forms of amyloidosis; Phorwhite BBU; and fluorescent dyes.[1,4] Sections of lichen amyloidosis usually are confined to the papillary dermis and do not involve blood vessels or adnexal structures.[1,2,5] A characteristic reticulated pattern is seen because of pigment incontinence between amyloid deposits. In addition, hyperplasia of the papillary epidermis with focal hyperkeratotic scale is seen.[1,2,5] Angioplasia also may be present. Small amorphous globules within the papillae coalesce and expand the papillae and displace the rete ridges laterally.[2,5]

Because pruritus is believed to be the primary trigger for the deposition of amyloid, most treatment modalities are directed toward the relief of this symptom. Antihistamines are moderately effective, and topical and intralesional steroids are beneficial if combined with other modalities.[1] More aggressive treatments such as surgical excision, dermabrasion, or treatment with the CO_2 laser have been used; however, lesions and pruritis tend to recur.[3,6] Anecdotal reports of response to topical dimethyl sulfoxide and etretinate therapy also have been reported.[2,6]

REFERENCES

1. Al-khenaizan S. Lichen amyloidosis [eMedicine Web site]. September 22, 2005. Available at: www.emedicine.com /derm/topic16.htm. Accessed May 15, 2002.
2. Breathnach SM. Amyloidosis of the Skin. In: Fitzpatrick TB, Eisen AZ, Wolff K, et al, eds. *Dermatology in General Medicine*. Vol 2. New York, NY: McGraw-Hill, Inc; 1993:1845-1852.
3. Finkel LJ, Headington JT. Cutaneous mucinosis and amyloidosis. In: Moschella SL, Hurley HJ, eds. *Dermatology*. Vol 2. Philadelphia, Pa: WB Saunders Co; 1992:1589-1604.
4. Wang WJ. Clinical and histopathological characteristics of primary cutaneous amyloidosis in 794 Chinese patients. *Chin Med J*. 2001;64:101-107.
5. Black MM. Amyloid and the amyloidoses of the skin. In: Champion RH, Burton JL, Ebling FJG, eds. *Textbook of Dermatology*. Vol 4. Boston, Mass: Blackwell Scientific Publications; 1992:2333-2344.
6. Behr FD, Levine N, Bangert J. Lichen amyloidosis associated with atopic dermatitis: clinical resolution with cyclosporine. *Arch Dermatol*. 2001;137:553-555.