

Symmetric Lichen Amyloidosis: An Atypical Location on the Bilateral Extensor Surfaces of the Arms

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PRACTICE POINTS

- Lichen amyloidosis (LA) classically presents as a pruritic and papular eruption localized to the pretibial surface of the legs.
- Nonpruritic and generalized variants are rare.
- This case represents a pruritic and nongeneralized case located on the arms; LA should be considered for any localized and pruritic eruption on the arms.

To the Editor:

Lichen amyloidosis (LA) classically presents as a pruritic, hyperkeratotic, papular eruption localized to the pretibial surface of the legs.¹ Nonpruritic and generalized variants have been reported but are rare.² Although it is the most common subtype of primary localized cutaneous amyloidosis, LA is a benign condition but is difficult to eradicate.¹ The precise pathophysiology is poorly understood, but chronic frictional irritation is closely associated with the eruption. We present a nongeneralized case of LA in an atypical location.

A healthy 30-year-old woman presented with an intermittent itchy rash on the elbows and knees of 2 years' duration. The patient was first diagnosed with lichen simplex chronicus (LSC) and initially responded well to treatment with fluocinonide ointment 0.05%. Nearly 2 years after the initial presentation, she developed recurrent symptoms and sought further treatment. She reported frequent scratching in association with episodes of anxiety. Examination revealed numerous 1- to 3-mm, flesh-colored to light brown, monomorphic, dome-shaped papules over the extensor surfaces of the bilateral arms and left pretibial surface (Figure 1).

Although in an atypical location, LA was clinically suspected due to the morphology, and a biopsy was performed given the evolving nature of the lesions. The differential diagnosis included LSC, hypertrophic lichen planus, papular mucinosis, prurigo nodularis, and pretibial myxedema. Pathology revealed small eosinophilic globules in the papillary dermis (Figure 2), and cytokeratin 5/6 immunostaining showed amorphous papillary dermal deposits consistent with keratin-derived amyloid deposition (Figure 3). The deposits stained positive for Congo red and displayed apple green birefringence under polarized light. Thus, the diagnosis of LA was confirmed. After limited success with triamcinolone ointment 0.1%, the patient was transitioned to clobetasol cream 0.05% with notable physical and symptomatic improvement.

Amyloidosis is histopathologically characterized by extracellular deposits of amyloid, a polypeptide that



FIGURE 1. Flesh-colored to light brown, monomorphic, dome-shaped papules (1–3 mm) over the extensor surfaces of the bilateral arms.

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The authors report no conflict of interest.

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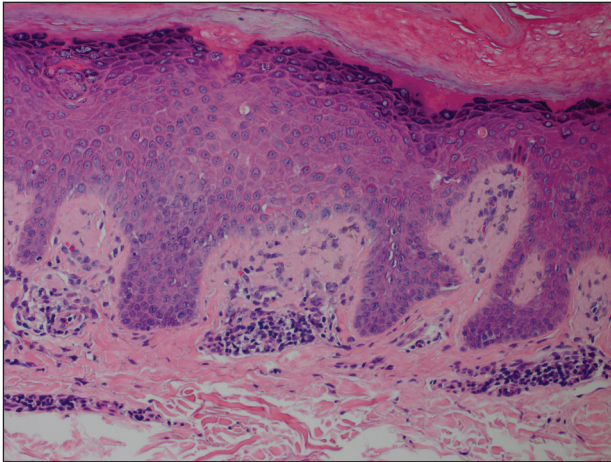


FIGURE 2. Eosinophilic globules in the papillary dermis (H&E, original magnification $\times 20$).

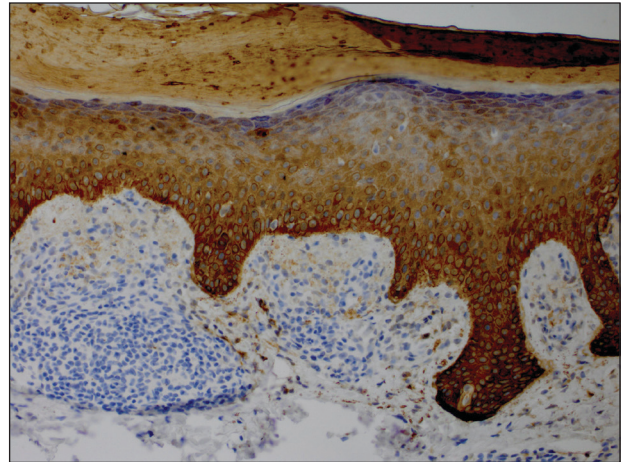


FIGURE 3. Cytokeratin 5/6 immunostaining showed amorphous papillary dermal deposits (original magnification $\times 20$).

polymerizes to form cross- β sheets.³ It is believed that the deposits seen in localized amyloidosis result from local production of amyloid, as opposed to the deposition of circulating light chains that is characteristic of systemic amyloidosis.³ Lichen amyloidosis is the most common subtype of primary localized cutaneous amyloidosis.¹ The amyloid in this condition has been found to react immunohistochemically with antikeratin antibody, leading to the conclusion that the amyloid is formed by degeneration of keratinocytes locally due to chronic rubbing and scratching.⁴⁻⁶

The possibility remains that this patient first presented with LSC 2 years prior and secondarily developed LA due to chronic trauma. Indeed, LA has been proposed as a variant of LSC. In both conditions, scratching seems to be the most important factor in the development of lesions. It has been proposed that treatment should primarily focus on the amelioration of pruritus.⁵

Five percent to 10% of cases of LA have been found to have some form of upper extremity involvement.⁷ However, these cases typically are associated with a generalized presentation involving the trunk and arms.^{2,7}

Our patient had no evidence of disease elsewhere. When evaluating a localized, pruritic, monomorphic, papular eruption on the extensor surfaces of the arms, LA may be an important consideration.

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