

Lichen Amyloidosis: A Bullous Variant

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Abstract

Introduction: Lichen amyloidosis is a common skin disease seen among Asian patients. **Clinical Picture:** Typical features range from macular hyperpigmentation to pruritic, lichenified, hyperpigmented papules. However, in this rare bullous variant of lichen amyloidosis, bullae and vesicles are present. **Histopathologically,** deposits of amyloid were seen in the papillary dermis, associated with an intraepidermal or subepidermal blister. **Treatment and Outcome:** No good treatment so far, but pruritus can be relieved by topical steroid. **Conclusion:** It is important to screen for systemic amyloidosis with the relevant investigations as it can present similarly with blistering eruptions, in which the prognosis would be grave.

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Key words: Asians, Blisters, Lichen amyloidosis, No systemic involvement

Introduction

Systemic amyloidosis, first reported in 1886,¹ is characterised by deposits of amyloid in various organs including the skin, which may manifest as skin purpura, papules, nodules and occasionally bullae (termed bullous amyloidosis in the literature), predominantly distributed over the periorbital and intertriginous areas.^{2,3} In contrast, lichen amyloidosis is solely a cutaneous amyloidosis problem and is not associated with bullae formation except in two rare cases, and now we are reporting the third such case.

Case Report

A 21-year-old Chinese man presented with an itchy rash on the face and limbs since childhood, consistent with the diagnosis of endogenous eczema. However, there were some tense blisters on the legs that healed with scarring. There was no other medical history of note. His family members were unaffected.

Physical examination revealed an extensive, uneven, rippled hyperpigmentation over the trunk and limbs, consistent with the diagnosis of lichen amyloidosis. However, some unusual blisters were noted over the legs (Fig. 1) and the right groin.

A punch biopsy specimen taken from a blister on the left calf revealed an old subepidermal bulla with re-epithelialisation. The blister had a necrotic roof and contained neutrophils. The dermis showed superficial perivascular

infiltrates of lymphocytes and histiocytes (Fig. 2). Amyloid deposits were visible in the dermal papillae on Congo red staining. Under electron microscope, the skin showed normal appearing keratinocytes. There was no degenerative cytologic changes in the basal cells. The hemidesmosomes and anchoring fibrils were normal in morphology and number. Globules of amyloid deposits were identified in the dermis (Fig. 3). Other tests including the direct immunofluorescence test, antibodies to epidermolysis bullosa acquisita antigen, anti-nuclear antibodies, blood and urine porphyrins, serum immunoglobulins and Bence Jones protein were all normal or negative.

Discussion

Lichen amyloidosis (LA) is a persistent, pruritic eruption consisting of multiple discrete hyperkeratotic and hyperpigmented papules, which sometime coalesce to form plaques. The sites of predilection are the shins and extensor aspect of the arms. The aetiology of LA remains unknown. It is more common among the Chinese⁴⁻⁶ than other ethnic groups.

In 1967, Chow and Burns⁷ reported a 55-year-old man with localised cutaneous amyloidosis, presenting with both haemorrhagic and serous bullous eruptions on the lower extremities. There were no features of systemic involvement or myeloma after 10 years of follow up.

In 1985, Ruzicka et al⁸ described a 45-year-old woman

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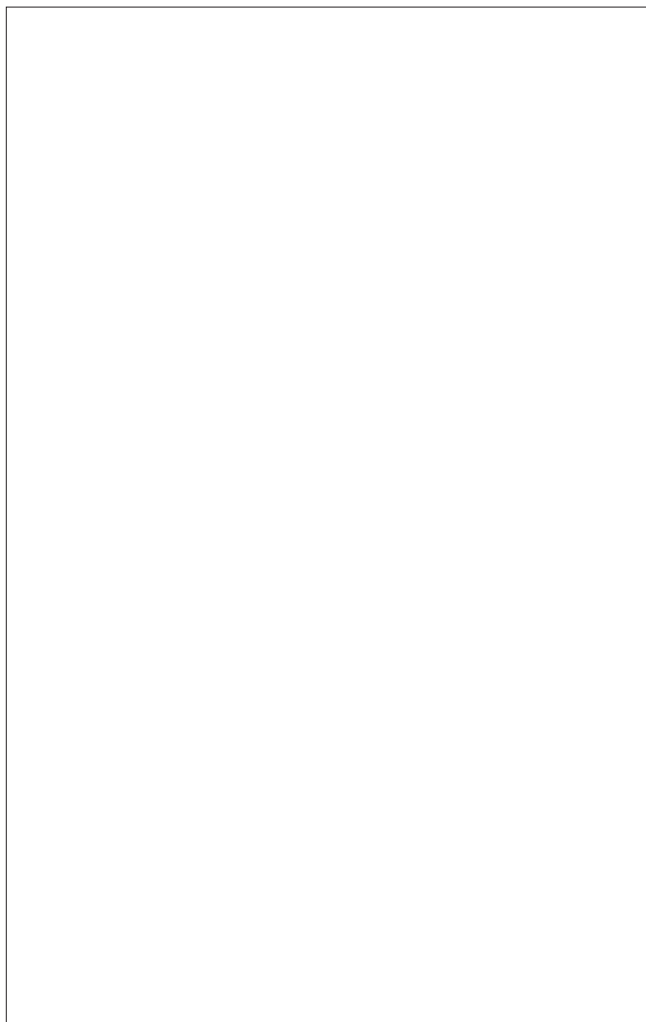


Fig. 1. A close-up of the left calf showing hyperpigmentation, white scars and hyperkeratotic blisters (arrows).

presenting with a 12-year history of relapsing generalised bullous dermatosis. Histological examination showed subepidermal blister and amyloid deposits in the upper dermis. Ultrastructurally, blister formation was seen at the lamina lucida level. Further investigations including serum and urine electrophoresis, immunoelectrophoresis, direct and indirect immunofluorescence tests were all negative.

Besides the typical features of lichen amyloidosis present in our patient, the blisters containing clear fluid on the lower limbs were indeed unusual. Therefore the clinical differential diagnoses initially considered were epidermolysis bullosa dystrophica, epidermolysis bullosa acquisita and the porphyrias. Our investigations however had adequately excluded these conditions.

The bullae seen in bullous amyloidosis can be either intradermal or subepidermal and are often induced by trauma.⁹ The exact mechanism in which bullae are formed remains unknown, although shearing forces breaking up the amyloid deposits have been postulated.¹⁰

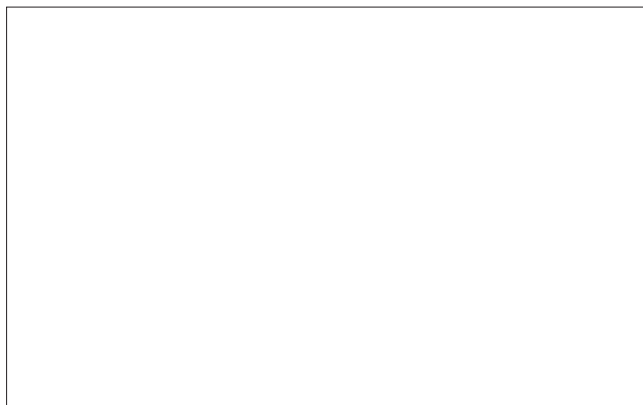


Fig. 2. Lesional skin biopsy specimen showing an old subepidermal vesicle with re-epithelisation containing few neutrophils. (H&E stain; original magnification x40)

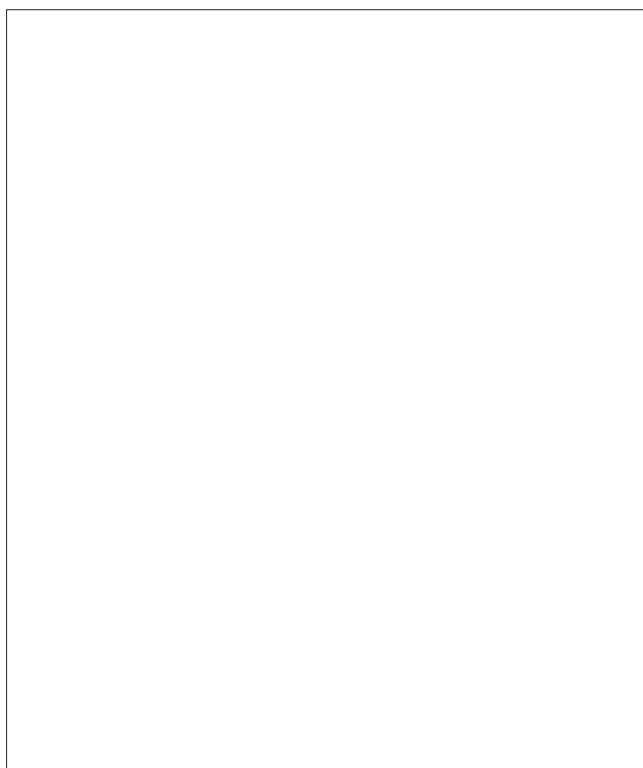


Fig. 3 Electronmicrograph showing basal keratinocytes of epidermis with normal hemidesmosomes and anchoring fibrils. There are deposits of amyloid (A) in the dermis (original magnification x 21, 375). (Inset) Higher magnification of the amyloid fibrils (original magnification x74, 250).

In conclusion, our patient has bullous variant of lichen amyloidosis, similar to the two earlier reported cases. Although it is unlikely that he will go on to develop the systemic form of the disease, close follow up is warranted.

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