

Lichen Amyloidosis Without Itching Indicates That it is Not Secondary to Chronic Scratching

Aquilina Ramírez-Santos, Javier Labandeira*, Benigno Monteagudo and Jaime Toribio

Department of Dermatology, Complejo Hospitalario Universitario, Faculty of Medicine, C/ San Francisco, s/n ES-15782 Santiago de Compostela, Spain.

*E-mail: jlabandeira@meditex.es

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Sir,

The origin of lichen amyloidosis (LA) is controversial. It is a form of primary cutaneous amyloidosis in which fibrillar proteinaceous material is deposited solely in the skin, without associated systemic manifestations. LA is typically highly pruritic, and the deposition of fibrillar material has generally been considered to be secondary to the scratching induced by other subjacent pruritogenic processes. However, our literature search revealed one previous report of LA without pruritus and a small-series study in which pruritus preceded lesions in most but not all patients. Here we present a case of widespread lichen amyloidosis with lesions on the trunk and extremities first noted 7 years previously by a young man, who at no stage reported pruritus. We conclude that, contrary to the actual view, pruritus and scratching are merely possible symptoms of this dermatitis, not requisite causal factors.

CASE REPORT

An 18-year-old man without noteworthy personal or family antecedents consulted for asymptomatic lesions first noted 4 years previously, initiating on the anterior surface of the legs and subsequently extending to arms and trunk. Dermatological examination revealed multiple hyperkeratotic papular lesions 2 mm in diameter with reddish-brown colouring, present bilaterally and symmetrically in the pre-tibial regions (Fig. 1), anterior surface of thighs, extensor regions of upper arms and forearms, abdominal wall and lumbar region. Neither mucosal involvement nor other cutaneous manifestations were detected, and the physical examination

was otherwise normal. Routine blood and urinalyses, serum protein electrophoresis, and serum levels of anti-nuclear antibodies (ANA), extractable nuclear antigens (ENA), immunoglobulins G, A and M, and complement fractions C3 and C4 were all normal.

Biopsy of the pre-tibial lesions showed orthokeratotic hyperkeratosis, acanthosis and, in dermis, nodular deposits of eosinophilic acellular material, perivascular inflammatory infiltrate, and pigmentary incontinence, without involvement of vascular or adnexal structures. The dermal deposits showed staining with Congo red and greenish fluorescence with thioflavin T (Fig. 2).

Following diagnosis of lichen amyloidosis, treatment was commenced with oral valacyclovir (1 g every 8 h for 7 days), and clobetasol propionate in pre-tibial areas (for 4 weeks), without improvement. Treatment with oral acitretin (0.43 mg/kg body weight/day) was thus commenced, with improvement of the lesions on the arms noted within 4 weeks, and subsequently on thighs and trunk, but without modification of the pre-tibial lesions after 7.5 months of treatment. The acitretin treatment induced formation of periungual granulation tissue on the big toe of each foot, and generalized skin xerosis, which gave rise to pruritus and scratching despite associated emollient treatment. After termination of the acitretin treatment, the xerosis and pruritus disappeared. Three years later the lesions are stable and the patient remains without pruritus.

DISCUSSION

LA manifests clinically as hyperkeratotic papules of variable colour (yellowish to reddish brown), generally



Fig. 1. Yellowish-brown hyperkeratotic papules in pre-tibial regions.

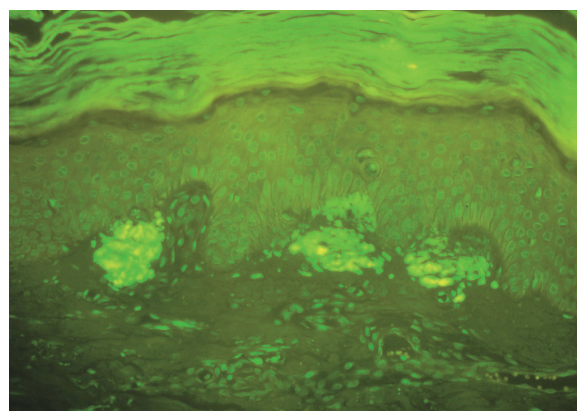


Fig. 2. Deposits of amyloid material in the papillary dermis, showing greenish fluorescence (thioflavin T, $\times 200$).

highly pruritic, above all in the initial stages. Hence LA is widely considered as a variant of lichen simplex chronicus, and chronic scratching as the cause and not as a result of the amyloid deposition (1).

However, in addition to the patient described here, there has been at least one previous report of LA without pruritus (2), and a small-series study found that pruritus preceded lesions in most but not all patients (1). Chronic scratching likewise cannot explain cases in which generalized lesions appear over the entire tegument (3, 4), thus other factors must be involved. LA is generally sporadic, but familial presentation with dominant autosomal inheritance has been reported, and it has recently been suggested that a locus located on the short arm of chromosome 1 may be involved (5). LA has also been reported in association with diverse diseases, including multiple endocrine neoplasia (MEN) 2A (6), atopic dermatitis (7), mycosis fungoides (8) and chronic urticaria (3). Epstein-Barr virus has been implicated in its aetiology (9), but the case in question responded to antiviral treatment, unlike in the present study.

The co-occurrence of macular amyloid lesions and amyloid lichen, denominated mixed amyloidosis (10), is frequent, and some authors have argued that the way in which amyloid is deposited in the skin depends on location, with macular deposits in the back vs. palpable deposits in pre-tibial regions and extremities (11). However, in our patient we did not observe pigmented maculae indicative of mixed amyloidosis; by contrast, we did observe LA lesions in the back.

In conclusion, since the lesions were first detected 7 years ago, our patient has not reported pruritus or shown signs of scratching. Thus the present case can be classified as non-itchy LA, as far as we know the second case of this type

reported. In our opinion, the available data indicate that LA should not be considered secondary to chronic scratching.

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