Case Report

Lychen Amyloidosis

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Abstract

Lychen amyloidosis is recognized by a persistent pruritic eruption of numerous separate hyperkeratotic papules, characterized by primary amyloid deposition in the skin. The cause is undetermined, but chronic irritation and cutaneous innervation have been suggested as etiological causes. Here, we report the clinicopathological findings of a patient with lychen amyloidosis.

ABBREVIATIONS

UVB: Ultraviolet B; YAG: Yttrium aluminum garnet.

INTRODUCTION

Lychen amyloidosis is a chronic pruritic skin illness characterized by the atypical extracellular tissue accumulation of amyloid without systemic damage. This malady is a primary cutaneous amyloidosis characterized by a persistent pruritic eruption of several hyperkeratotic papules that may join into plaques. This disease is identified most commonly in South America and Southeast Asia.

CASE PRESENTATION

A 33-year-old man presented with 5-month history of severe generalized pruritus and skin lesions. The patient informed no other healing troubles and denied any family history of skin problems.

Figure 1 Lichenified, erythematous and hyperkeratotic plaques located on the anterior forearm.

Figure 2 Amyloid deposits (A) – [In the papillary dermis (hematoxylin and eosin stain; scale bar 100 µm)] (B) - [Deposits exposed in the dermis using Congo red stain (scale bar 100 µm).] (C) – [Deposits stained with Congo red (amplification; scale bar 50 µm).] (D) - [An assemblage of irregularly intermixed fibrils is appreciated, consistent with amyloid (electron micrograph, scale bar 1 µm)].
illness. Results of physical examination exposed lichenified, erythematous and hyperkeratotic plaques, predominantly on the anterior forearms (Figure 1). The histological analyses showed slight globular deposits of an amorphous and unclearly eosinophilic material in the papillary dermis consist with amyloid (Figure 2A). Congo red stain revealed amyloid deposits (Figure 2B and 2C). These deposits were corroborated with electron microscopy analysis (Figure 2D). The patient received oral cyclosporine (4 mg/kg). Six weeks later, the size and number of the skin lesions were noticeably reduced. Eight months after beginning cyclosporine therapy, his skin lesions remained in remission and the circulating serum amyloid P values were within the normal range.

DISCUSSION

The etiopathogenesis of lichen amyloidosis is unknown, however, hereditary and viral causes have been identified as probable origins. Fibril proteins structure the principal constituent of amyloid; there are twenty-threedifferent forms of amyloid fibril (amyloid light chain (AL), transthyretin-associated (ATTR), amyloid associated (AA), etc.,). Other minor components are glycosaminoglycans and amyloid P.A particular precursor protein is correlated with each clinical condition; however in the skin the precursor protein has not totally identified, but it is considered as keratinocyte-derived. Lesions are located predominantly on the anterior legs, but can occur on the thighs, forearms, and back. The histological analysis shows amyloid accumulation in the papillary dermis and acanthosis of the epidermis. Amyloid deposits could be the result of epidermal trauma produced by long-term scratching or associated with nerve fiber density [1].

Mycosis fungoides, lichen planus, multiple endocrine neoplasia type 2A and atopic dermatitis are some clinical conditions, which have been correlated with lichen amyloidosis [2]. The clinical differential diagnosis included lichen simplex chronicus, hypertrophic lichen planus, lichen atrophicus and prurigo nodularis [3]. Current therapies for lichen amyloidosis are topical corticosteroids, etretinate, calcipotriene, UV-B phototherapy, YAG laser treatment, topical dimethyl sulfoxide, cyclophosphamide, cyclosporine and manual dermabrasion [4]. In this case the patient responded well to cyclosporine, without nephrotoxicity and hypertension, which are the two most usual and grave adverse effects encountered after using this drug.

REFERENCES