

LICHEN AMILOYDOSIS – CASE REPORT

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Summary

Lichen amyloidosis is the most common form of primary cutaneous amyloidosis. It is characterized by the presence of hyperkeratotic, pruritic and persistent papules, which may coalesce forming red brownish plaques, found mostly on the extensor surface of the extremities. Histopathological findings are represented by the deposits of insoluble amyloid fibres in the papillary dermis.

We describe the case of a 47 year-old female patient, who was admitted to our clinic for the presence of infiltrated patches and plaques, composed of light brown, grouped papules, involving the anterior and posterior trunk, arms and thighs, of 15 years duration. The diagnosis of lichen amyloidosis was established based on clinical and histopathological examination.

We aim to discuss the main clinical, paraclinical and treatment aspects in lichen amyloidosis.

Key words: primary cutaneous amyloidosis, Lichen amyloidosis.

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Introduction

Amyloidoses are a group of rare diseases, characterized by the deposition of insoluble fibers of amyloid in the extracellular tissue. Amyloid is an amorphous, hyaline, eosinophilic, material, consisting of plasma proteins with low molecular weight, grouped in insoluble fibrillar structures [1, 5].

Lichen amyloidosis is more common in the Asian and South American population. It preponderantly affects men and usually develops after the age of 50 [4].

The classic presentation of lichen amyloidosis consists of multiple red-brownish, hyperkeratotic papules, which may coalesce forming intensely pruritic plaques, frequently resistant to treatment. The most common sites of distribution are the limbs and trunk [5] Initially, the lesions are unilateral, but as the disease progresses the

lesions develop bilaterally in a symmetrical manner. Verrucous changes may develop on the surface of the plaques resembling hypertrophic lichen planus. The lesions are intensely pruritic and some authors believe that chronic scratching leads to the destruction of keratinocytes and the deposition of amyloid [6, 7].

On histopathologic examination, amyloid deposits are found in the dermal papillae. The upper epidermis may exhibit vacuolar degeneration of the basal layer. As in lichen simplex chronicus, hyperkeratosis and acanthosis may be observed. On Haematoxylin & eosin staining, the amorphous material becomes intensively eosinophilic in the papillary dermis. Amyloid stains red-orange with Congo Red, and in polarized light has a green appearance. Immunohistochemical studies may be positive for the keratinocyte marker Cytokeratin 5 [9, 10, 11].

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Lichen amyloidosis is a benign, chronic disease. However, it can be associated with several systemic disorders. Among them is , an autosomal dominant syndrome. In these patients, the lesions of lichen amyloidosis are localized interscapular.

The treatment consists mainly in topical applications of corticosteroids.

Case report

A 47 year-old female patient was admitted to our clinic for the presence of infiltrated patches and plaques, composed of light brown, grouped papules, involving the anterior and posterior trunk, arms and thighs, of 15 years duration (fig. 1). Paraclinical investigations revealed polycythemia, slightly increased urinary proteins and mixed dyslipidemia. Abdominal ultrasound revealed first grade hepatic steatosis and bilateral renal lithiasis.

The histopathological examination showed hyperorthokeratosis, mild hypergranulosis, acanthosis and irregular papillomatosis. The dermal papillae were enlarged due to eosinophilic deposits. Capillary dilatations were seen in the whole dermis, as well as endothelial swelling. Perivascular inflammatory lymphocytic infiltration could also be observed. Red-orange deposits were identified in the dermal papillae in Congo Red staining (fig. 2). The histopatho-

logical findings were, therefore, compatible with the diagnosis of lichen amyloidosis.

Treatment consisted of emollients and topical corticosteroids, which lead to clinical improvement of lesions.

Discussions

Cutaneous lesions can be identified in primary skin amyloidosis or in systemic forms of the disease [1]. Primary cutaneous amyloidosis includes three types: macular, nodular and lichenoid forms [2]. Amyloid is a homogeneous, amorphous compound consisting of abnormally folded plasma proteins with low molecular weight, grouped into insoluble fibrillar structures. [1, 6]. It forms intradermal deposits of eosinophilic material, highly resistant to proteolytic enzymes [1]. The origin of amyloid, in lichen amyloidosis and macular amyloidosis, appears to be the deterioration of epidermal keratin filaments that accumulate into the dermis [8]. The etiology of the disease is unknown. Genetic predisposition, Ebstein-Barr virus or environmental factors have been incriminated in the occurrence of the disease.

The diagnosis of lichen amyloidosis is based on the clinical appearance and localization of skin lesions (especially on the shins). The histopathological examination is needed to confirm the clinical suspicion lichen amyloidosis.



Fig. 1. Infiltrated patches and plaques, composed of light brown, grouped papules

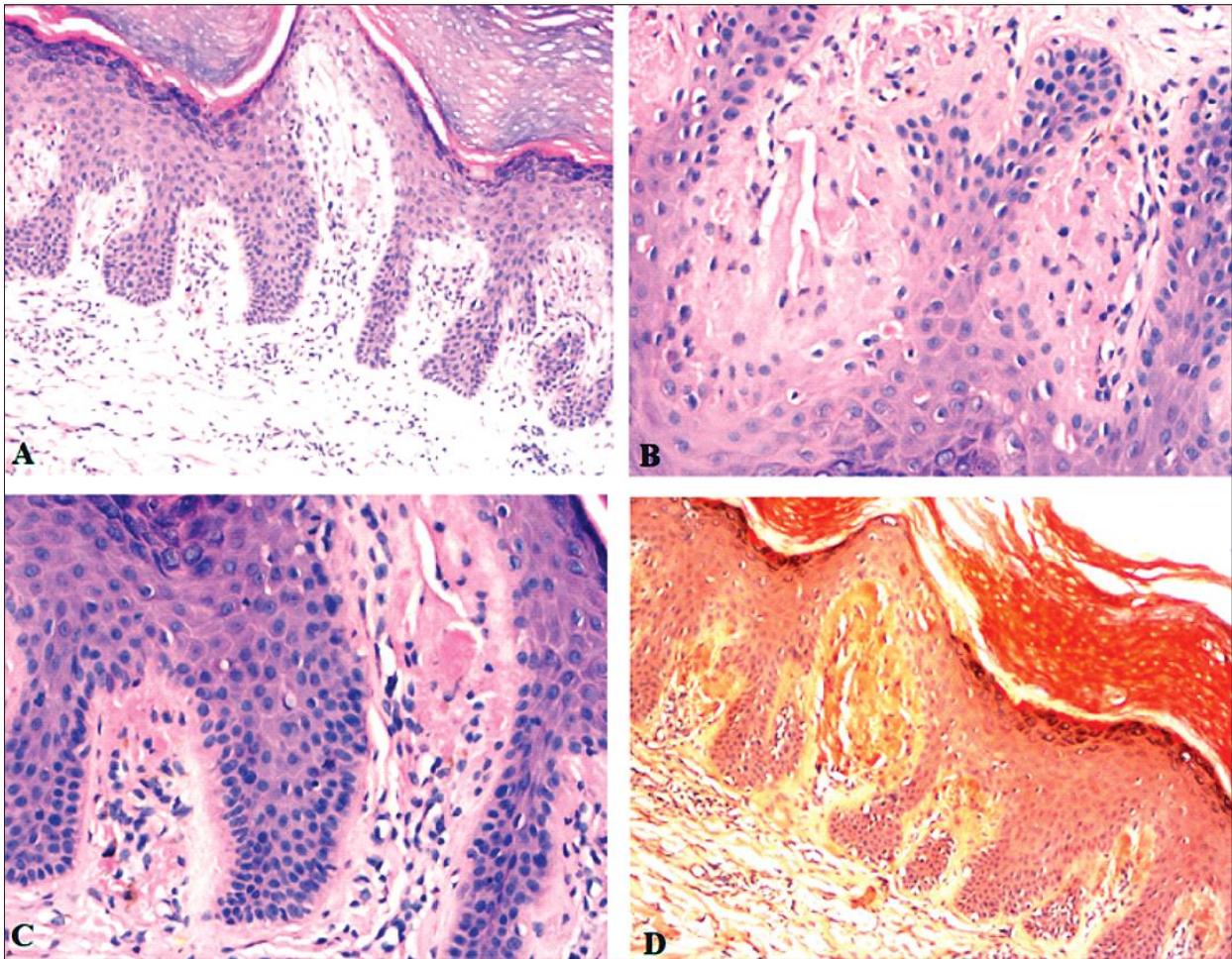


Fig. 2. Haematoxylin & eosin stain (A) magnification x 40, (B,C) magnification x 200, (B,C) showing hyperorthokeratosis, mild hypergranulosis, acanthosis and irregular papillomatosis. The dermal papillae were enlarged due to eosinophilic deposits. Capillary dilatations of the whole dermis and endothelial swelling and perivascular inflammatory lymphocytic infiltration are also observed. (D)- Red-orange deposits in the dermal papillae in Congo Red staining

Lichen amyloidosis is a chronic, benign condition. There is no curative treatment available. Alleviation of pruritus can be achieved by the administration of H1 antihistamines. The aesthetic appearance can be improved through the use of topical or intralesional glucocorticoids, cauterization or cryotherapy, as well as dermabrasion. Phototherapy and laser treatment may represent therapeutic options in case of refractory lichen amyloidosis [12].

Conclusions

Lichen amyloidosis is a rare condition limited to the skin. The diagnosis is based on the clinical aspect of skin lesions and their localization. Therapeutic options are limited and none is curative.

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Conflict of interest
NONE DECLARED

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