

# Lichen amyloidosis with combined topical therapy: a case report



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## ABSTRACT

**Introduction:** Lichen amyloidosis (LA) is a rare case, characterized by circumscribed, highly pruritic, hyperkeratotic, and hyperpigmented papules occurring typically over the shins, outer aspects of upper arms, and on the upper back with amyloid deposits in the papillary dermis. Several therapeutic strategies, including topical steroids, oral antihistamines, cyclosporine, retinoids, laser, phototherapy, cryosurgery, and surgical interventions, have been reported as treatment options for patients with LA, but no standardized treatment has been established.

**Case report:** A 58-year-old man came to the Dermatovenereology Outpatient Department complaints of itchy blackish-brown papules on the shins. Dermatology examination found discrete multiple

hyperpigmentation papules and plaque covered with white scale. A scar-like center surrounded by brownish circles or white edges was found from the dermoscopic examination. The histopathological examination found thickened keratin with compact orthokeratosis and hyaline materials in the papillary dermis with dendritic melanophages. The patient diagnosed with LA and treated by combining desoximetasone cream 0.25% with 3% salicylic acid. The papules on the legs had flattened in the patient, with a significant improvement in the severe itching after three weeks.

**Conclusion:** Combination therapy of potent corticosteroids and keratolytic seems to be an appropriate modality and well-tolerated by LA patients. Skin lesion becomes thinner, and pruritus is reduced.

**Keywords:** lichen amyloidosis, topical corticosteroid, keratolytic

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## INTRODUCTION

LA is the most common form of primary localized cutaneous amyloidosis (PLCA).<sup>1</sup> Presents as persistent, pruritic plaques on the shins or other extensor surfaces. Initial lesions are discrete, firm, scaly, skin-coloured, or hyperpigmented papules, which later coalesce into plaques that often have a rippled or ridged pattern.<sup>5</sup> It is a challenge to lichen amyloidosis therapy, primarily aims to control the pruritus, recurring scratching, and lichenification. Herein, we describe a case of LA was improved after treatment with a combination of topical corticosteroid with keratolytic.

## CASE REPORT

A 58-year-old man came to the Dermatovenereology Outpatient Department complaints of blackish-brown spots on both lower limbs for one year accompanied by pruritus, intense scratching. He has a history of hypertension for two years, which treated with propranolol 10 mg every 24 hours. The patient was treated five months ago for the same complaints. He was given topical cream twice daily on the limbs; he forgot the name of the drugs. He felt there is no improvement. The application

of traditional ingredients to the skin was denied. Similar history in the family was denied.

In physical examination, we found discrete multiple hyperpigmentation papules, round shape with diameters varying from 0.2 to 0.3 cm on both lower extremities. Some papules are confluent and configure hyperpigmentation plaque with geographical shape, 2x3 – 4x6 cm, covered with a white scale. These lesions are hard on palpation (**Fig. 1a-e**). Dermoscopic examination revealed a scar-like centre surrounded by brownish circles or white edges (**Fig. 2a**). Histopathological examination with hematoxylin and eosin staining found thickened keratin with compact orthokeratosis. The epidermis appears normal. There were hyaline materials in the papillary dermis with dendritic melanophages in between (**Fig. 3a-c**).

The patient diagnosed with LA and was treated with cetirizine 10 mg tablets orally every 24 hours, desoximetasone cream 0.25% combined with 3% salicylic acid, which was applied every 12 hours in thick lesions on both lower extremities. Significant improvement was noted after three weeks of therapy. The lesion becomes thinner, and pruritus was reduced. We continue therapy with combination hydrocortisone 2.5% cream and urea 20% cream.

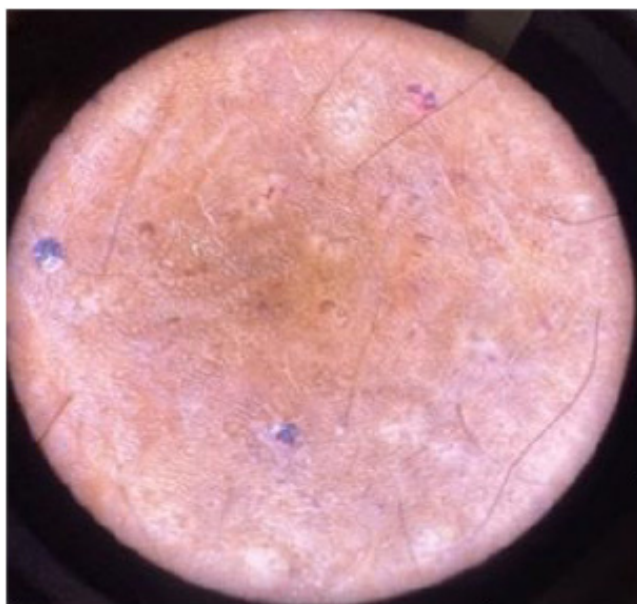
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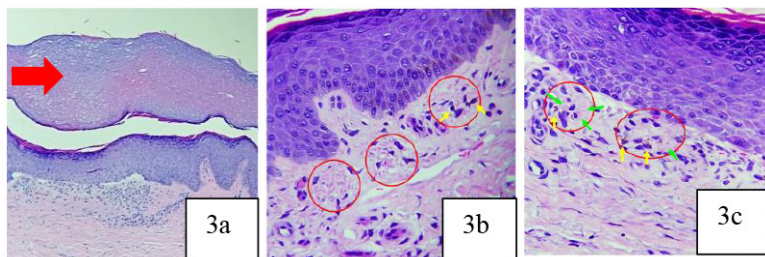
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**Figure 1.** (a-e) Multiple hyperpigmented papules covered by white scales. Symmetrically distributed in lower extremities and hard palpated



**Figure 2.** Dermoscopic examination revealed a scar-like center surrounded by a brown circle or white edges. There are brown dots in some spots



**Figure 3.** Histopathological examination from right crus region. (a) red arrow: thickened keratin with compact orthokeratosis. (b) & (c) green arrows: hyaline deposition materials; yellow arrows: dendritic melanophages

## DISCUSSION

LA is the most common form of PLCA.<sup>1</sup> Primary localized cutaneous amyloidosis is characterized by the presence of amyloid deposits in the skin, without any deposits in internal organs.<sup>7</sup> Predominance found in males, age of 50 and 60, and Fitzpatrick skin type III and IV.<sup>2</sup> Possible causes are high friction or scratching, genetic predisposition, Epstein-Barr virus infection, and environmental factors.<sup>3</sup> The previous study reported mutations in the oncostatin M receptor (OSMR) gene, which encodes the oncostatin M-specific receptor beta (OSMR $\beta$ ). OSMR $\beta$  is a component of type II oncostatin M (OSM) receptor and IL-31 receptor; signalling OSM and IL-31 plays a role in proliferation, differentiation, apoptosis, and inflammation of keratinocytes.<sup>5</sup> IL-31 shows a vital role in the condition of pruritic skin.<sup>7</sup>

The initial symptom of this disorder is intense pruritus that may increase with sun exposure. Hyperpigmented lesions are presumed to be secondary to scratching.<sup>2</sup> Usually presents as persistent, pruritic plaques on the shins or other extensor surfaces, e.g. anterior thighs or forearms. Lesions often begin unilaterally but can extend to be bilateral and appear symmetrical.<sup>5</sup>

The differential diagnosis is chronic lichen simplex chronicus and lichen planus. Chronic lichen simplex (LSC) gives rise to lichenification. There is a typical scale known as Wickham's striae.<sup>8</sup> Differences can be seen from histopathology. In hematoxylin and eosin staining techniques, focal amyloid deposits are large enough to extend into papillae and replace rete ridges that undergo lateral elongation in lichen amyloidosis. The epidermis above will experience acanthosis and hyperkeratosis. Several melanophages and amyloid deposits found near the epidermal basal layer. A rare lymphohistiocytic infiltrate usually found close the amyloid deposits.<sup>5</sup> It is necessary to do the immunohistochemical examination to determine the fibril protein.<sup>2</sup> The histopathology of the lichen simplex will reveal hyperplasia and epidermal hyperkeratosis, spongiosis and parakeratosis area.<sup>7</sup> That histopathology pictures of the lichen planus appear due to damage of basal epidermis and lichenoid lymphocytic reaction. In the papillary dermis found eosinophilic colloid bodies and lymphocyte infiltrates which looks like a ribbon.<sup>2</sup>

Therapeutic targets are to stop the pruritus-scratching cycle with topical or intralesional corticosteroid combined with the keratolytic agent, topical dimethylsulfoxide (DMSO), oral retinoid, calcipotriene, cyclophosphamide, laser, phototherapy, cryosurgery, and surgical interventions.<sup>4,6</sup> Administration of topical

corticosteroids, in mild cases, combined by occlusion or in combination with keratolytic agents such as salicylic acid, can increase the effectiveness of treatment.<sup>5</sup> Glucocorticoid receptors can inhibit T cell proliferation and cause cell apoptosis by inhibiting T cell growth factor, interleukin (IL)-2. Mitosis and DNA synthesis inhibition is mediated by topical corticosteroids, and they are known to decrease proliferation and keratinocyte size.<sup>10</sup> Salicylic acid widely used in almost any base with 0.5% to 60% concentrations. Shedding of scales by softening the stratum corneum, dissolving the intracellular matrix, and loosening connections between corneocytes were the functions of salicylic acid in 3% to 6% concentrations.<sup>11</sup>

In a study that compared the efficacy of topical corticosteroids versus either UVB or topical PUVA phototherapy for the treatment of LA, there was greater improvement in the pruritus and roughness scores on the sides treated with either form of phototherapy, with PUVA being marginally better in reducing pruritus.<sup>5</sup> Small series have reported successes with carbon dioxide resurfacing surgical laser, YAG laser, tocoretinate (a synthetic esterified compound of tocopherol and retinoic acid), narrowband ultraviolet B light therapy, and a combination of psoralen and ultraviolet A with acitretin.<sup>12</sup> Reported surgical procedures performed on lichen amyloidosis such as electrodesiccation, scraping technique with a scalpel, and dermabrasion healed and without recurrence. However, prolonged wound healing, superficial scarring, hypopigmentation, and edema.<sup>9</sup> This patient is treated with cetirizine 10 mg oral tablets every 24 hours to suppress pruritus and prevent scratching and friction is thought to play an essential role as a predisposing factor to lichen amyloidosis. Topical corticosteroids desoximetasone 0.25% combined with keratolytic, salicylic acid 3% cream are applied every 12 hours on thick lesions in lower extremities. Three weeks after therapy, there was good clinical improvement in which papules and hyperpigmented plaques become thinner.

## CONCLUSION

Combination therapy of potent corticosteroids and keratolytic appears to be an appropriate modality and well-tolerated by LA patients. This non-invasive and low-cost management has resulted in a meaningful improvement in the quality of life for these subjects. A similar approach is a reasonable option to consider other similar presenting

patients. More extensive controlled studies are needed further to establish the efficacy of this new treatment modality.

## CONFLICT OF INTEREST

Authors stated no conflict of interest related to the case publication.

## ETHICS IN PUBLICATION

The patient had received signed informed consent regarding the publication of their respective photograph in the journal article.

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