

12-30-2023

Lichen amyloidosis treated with topical combination therapy and narrowband ultraviolet B phototherapy

Ulfah Hasna Hasibah

Department of Dermatology and Venereology, Dr. Sitanala Hospital, Tangerang, Banten, Indonesia

Siti Aisha Nabila Ferina

Department of Dermatology and Venereology, Dr. Sitanala Hospital, Tangerang, Banten, Indonesia

Eka Komarasari

Department of Dermatology and Venereology, Dr. Sitanala Hospital, Tangerang, Banten, Indonesia

See next page for additional authors

Follow this and additional works at: <https://scholarhub.ui.ac.id/jdvi>



Part of the [Dermatology Commons](#)

Recommended Citation

Hasibah, Ulfah Hasna; Ferina, Siti Aisha Nabila; Komarasari, Eka; Esti, Prima Kartika; and Angin, Lenti Perangin (2023) "Lichen amyloidosis treated with topical combination therapy and narrowband ultraviolet B phototherapy," *Journal of General - Procedural Dermatology & Venereology Indonesia*: Vol. 7: Iss. 2, Article 7.

DOI: 10.7454/jdvi.v7i2.1153

Available at: <https://scholarhub.ui.ac.id/jdvi/vol7/iss2/7>

This Article is brought to you for free and open access by the Faculty of Medicine at UI Scholars Hub. It has been accepted for inclusion in Journal of General - Procedural Dermatology & Venereology Indonesia by an authorized editor of UI Scholars Hub.

Lichen amyloidosis treated with topical combination therapy and narrowband ultraviolet B phototherapy

Cover Page Footnote

We would like to extend our gratitude to IMERI Writing Centre for the proofreading process.

Authors

- Ulfah Hasna Hasibah
Department of Dermatology and Venereology, Dr. Sitanala Hospital, Tangerang, Banten, Indonesia
- Siti Aisha Nabila Ferina
Department of Dermatology and Venereology, Dr. Sitanala Hospital, Tangerang, Banten, Indonesia
- Eka Komarasari
Department of Dermatology and Venereology, Dr. Sitanala Hospital, Tangerang, Banten, Indonesia
- Prima Kartika Esti
Department of Dermatology and Venereology, Dr. Sitanala Hospital, Tangerang, Banten, Indonesia
- Lenti Perangin Angin
Department of Pathology Anatomy, Dr. Sitanala Hospital, Tangerang, Banten, Indonesia

Case Report

Lichen amyloidosis treated with topical combination therapy and narrowband ultraviolet B phototherapy

Ulfah Hasna Hasibah¹, Siti Aisha Nabila Ferina¹, Eka Komarasari¹, Prima Kartika Esti¹, Lenti Perangin Angin²

1. Department of Dermatology and Venereology, 2. Department of Pathology Anatomy, Dr. Sitanala Hospital, Tangerang, Banten, Indonesia

Email: hasnahasibah97@gmail.com

Abstract

Background: Lichen amyloidosis (LA) is characterized by linear rows of firm, pigmented, grouped, hyperkeratotic papules that can form a plaque, usually occurring on the shins and forearms, with intense itch as the prominent symptom.

Case Illustration: A 57-year-old female complained of brown spots on her shins and arms that gradually thickened six years ago. The lesions were brown, multiple, discrete, slightly scaly papules forming hyperpigmented plaques. The result of the histopathology examination showed an acanthotic epidermis with hyperkeratotic foci and eosinophilic amorphous mass deposits in the papillary dermis with brown pigments. The working diagnosis was lichen amyloidosis. Topical treatments were ointment consisting of 6% salicylic acid mixed with clobetasol propionate ointment 0.05% used once daily and emollient used twice daily. Narrowband ultraviolet B (NB-UVB) was administered at a dose of 300 mJ/cm² once a week and increased by 10-20% on the next episodes. After six weeks of treatment, there were no new brown spots, the lesions became thinner and less erythematous, and the itch decreased significantly.

Discussion: The factors that induce and worsen LA are pruritus and scratching. Topical combination therapy of salicylic acid and corticosteroid can increase the effectiveness of treatment on thick, scaly plaque lesions. NB-UVB was found to reduce pruritus.

Conclusion: Topical combination therapy of keratolytic agents and potent corticosteroids can be used as a non-invasive therapy to improve skin lesions by thinning these lesions in LA patients. NB-UVB phototherapy has also been significantly shown to relieve a patient's severe itch.

Keywords: *lichen amyloidosis, phototherapy, topical combination therapy*

Background

Amyloidosis is a rare and heterogeneous group of disorders characterized by the deposition of abnormally folded proteins in tissues. Amyloid deposits are formed from globular, soluble proteins, which undergo misfolding and aggregate into insoluble fibrils, which leads to progressive organ damage. Amyloidosis can be classified as systemic, localized, and hereditary familial types. The deposit of amyloid can be localized with deposits present in a single organ in the former case and affecting various organs and tissues throughout the body in the latter; hence, it can become systemic. Exclusively localized amyloid

deposits have been associated with at least 19 protein types, while at least 14 protein types (and many more variants) appear to be consistently associated with systemic amyloidosis.¹ Primary localized cutaneous amyloidosis (PLCA) may have various manifestations, depending on the site of amyloid deposition in the skin and without systemic involvement. There are three subtypes of it, namely lichen amyloidosis (LA), macular amyloidosis (MA), and nodular amyloidosis (NA). LA is the most common form of PLCA.^{2,3} LA is a rare skin disease. The prevalence of cutaneous LA is 0.2-0.3%. LA is more common in Southeast Asia and some South American Countries.⁴ Djuanda A, et al. reported 78 cases of LA in Jakarta, Indonesia during 1983-

1987.⁵ LA is usually present in the fifth and sixth decade and is more common in men and patients with higher Fitzpatrick skin types. An early symptom of LA is intense pruritus that improves with sun exposure and worsens during stress. The patient scratches due to pruritus, leading to a hyperpigmented lesion. Clinically, LA is characterized by linear rows of firm pigmented grouped hyperkeratotic papules that can form a large plaque, which usually occurs on the shins and forearms bilaterally. The upper back can also be involved.²

The diagnosis is often made clinically, but histopathological assessment is performed as confirmatory.⁶ Because of the intense pruritus, the epidermis will be acanthotic, papillomatous with compact horn, hyperkeratotic, basal keratinocytes undergo hyperpigmentation, and the rete ridges become elongated. In the papillary dermis, there are small collections of amphophilic material surrounded by melanophages.² In this paper, we will discuss a case of lichen amyloidosis, a cutaneous or localized variant of the disease.

Case Illustration

A 57-year-old female patient complained of brown spots on both shins that gradually thickened since six years ago. At first, it showed up as red spots with a reddish base that slowly spread and thickened; this symptom also affected her arms. The lesions on her skin feel itchy, sometimes hindering her activities. She had tried multiple oral and topical treatments without progress. Complaints other than her skin lesion were denied. History of diabetes mellitus, atopic dermatitis, and other diseases were also denied.

The physical examination of both extremities' antibrachial region and shin showed multiple brown, discrete, slightly scaly papules forming moniliform hyperpigmented plaques. The shin site was thicker than the antibrachial, and all regions were firm.

The differential diagnoses for these patients were lichen amyloidosis, lichen nitidus, lichen planus, and lichen chronic simplex. We performed a skin biopsy for histopathological examination. The biopsy showed that the tissue was lined with hyperplastic stratified squamous epithelium, elongated psoriasiform rete ridges with acanthotic epidermis with hyperkeratotic foci. Eosinophilic amorphous mass deposits were seen in the papillary dermis, with mild chronic inflammatory cells in between. There were also bits of chronic inflammatory cells around the blood vessels and

locally visible cells with brown pigment (Figure 1). To eliminate the possibility of systemic involvement, we performed laboratory tests such as complete blood count and blood glucose. All laboratory results were within normal limits.

The working diagnosis for this patient was lichen amyloidosis. Observation was done for six weeks. For the treatment, in the first two weeks, the patient was given cetirizine 1x10 mg orally and an ointment made up of 4% salicylic acid and 10 % anhydrous lanolin in 60 g white vaseline applied twice daily. In the third week, we added narrowband ultraviolet B (NB-UVB) 300 mJ/cm² once a week.

Topical treatment was changed to emollient (10 % anhydrous lanolin in white vaseline) twice daily and an ointment made of 6 % salicylic acid mixed with clobetasol propionate 0.05% once daily. In the fourth week, NB-UVB intensity was increased to 350 mJ/cm² and increased to 380 mJ/cm² for the next week. Lastly, in the sixth week, the NB-UVB intensity was increased to 390 mJ/cm².

Information, education, and communication (IEC) were provided to the patient about her current disease, factors that can cause the lesion to thicken, which is mainly due to pruritus and scratching, treatments, the length of therapy to be given, possible side effects of the treatment and the possibility of recurrence after the treatment. The patient had informed consent for the examination and the treatment.

After six weeks of treatment, there were no new brown spots. The lesions on the antibrachial region were thinner and less erythematous (Figure 2). The lesions on her shins became slightly thinner, (Figure 3). The itching sensation decreased considerably on both regions.

Discussion

LA is the most common form of PLCA, caused by the deposition of heterogenic amyloid proteins in the skin without systemic involvement. The systemic involvement in amyloidosis should be excluded by laboratory testing.³ LA has been associated with diabetes mellitus and autoimmune connective tissue disease.⁷ In this patient, a complete blood count and blood glucose level were performed, and the result was normal. Thus, diabetes mellitus was not an associated disease in the patient. Moreover, the patient did not complain of any other symptoms except the skin lesions. Therefore, systemic involvement in amyloidosis could be excluded.

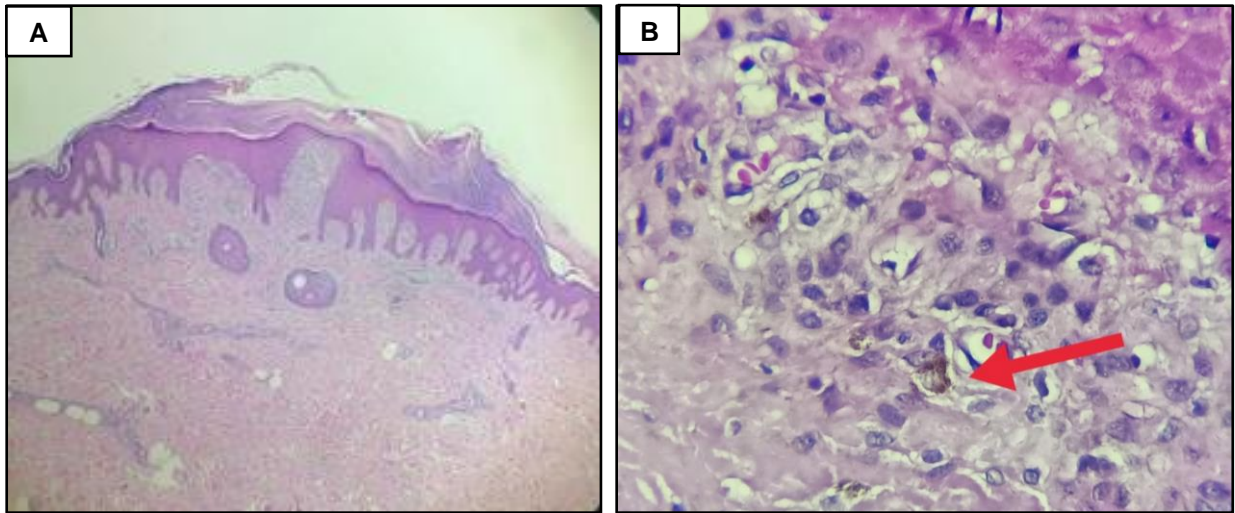


Figure 1. A. Histopathology Using Haematoxylin & Eosin Stain.

A. Image in 40x magnification shows that the tissue lined with hyperplastic stratified squamous epithelium, elongated psoriasiform rete ridges with acanthotic epidermis with hyperkeratotic foci. B. In 400x magnification shows bits of chronic inflammatory cells around the blood vessels and locally visible cells with brown pigment. The red arrow shows pigment incontinence.

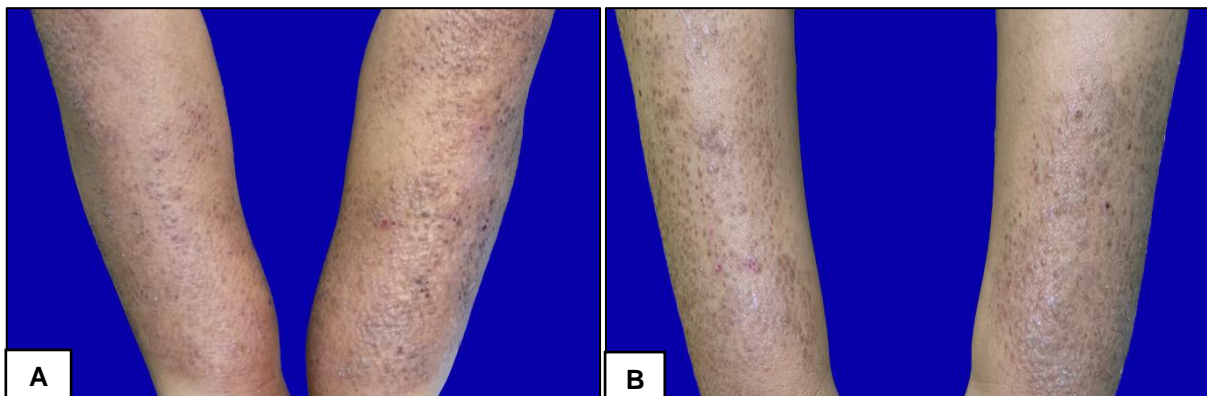


Figure 2. Lesion on the Bilateral Antebrachial were Thinner and Less Erythematous.

A. Before treatment. B. After six weeks of treatment.

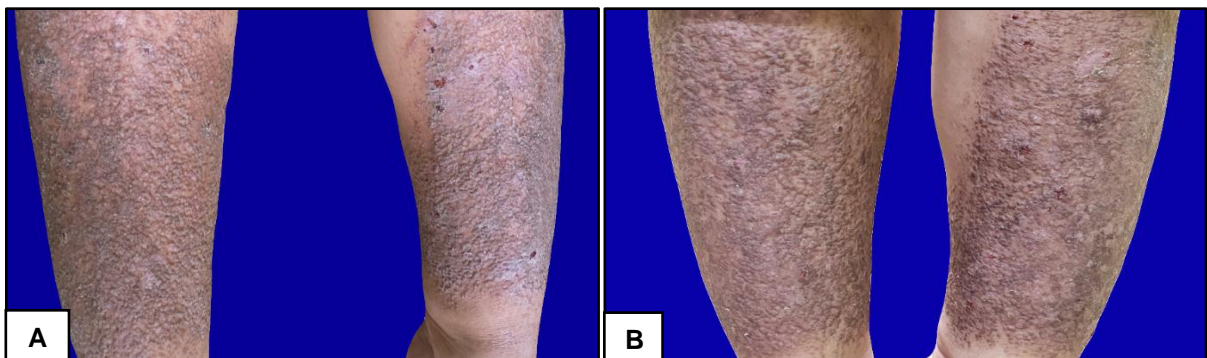


Figure 3. Lesion on Both Shins were Thinner.

A. Before treatment. B. After six weeks of treatment.

The exact aetiology and pathogenesis of LA are not well understood. LA is thought to be associated with other skin disorders that cause scratching, such as atopic dermatitis (AD).⁸ This patient had no history of AD. Other etiologic factors that have been proposed are genetic predisposition, Epstein-Barr virus, and environmental factors.⁶ The factors that induce and/or worsen LA are pruritus and scratching.⁹ Scratching can lead to inappropriate apoptosis of keratinocytes, which degenerate into keratin peptides. Furthermore, dermal macrophages convert the degenerated keratin peptides into amyloid fibrils, which are deposited in the papillary dermis.^{8,10}

Diagnosis of LA is based on clinical features and histopathological examination. Clinically, it is characterized by discrete pruritic hyperpigmented hyperkeratotic papules, which can become confluent to form plaques, usually beginning unilaterally and then extending symmetrically bilaterally.^{3,11} LA is primarily localized in the anterior aspect of the shins, back, forearms, dorsum of the feet and thighs.^{3,6,8} While dermatological status in this patient was suggestive of several diagnoses such as lichen amyloidosis, lichen nitidus, lichen nitidus, lichen planus, and lichen chronic simplex, thus, histological examination should be performed to confirm the diagnosis.

Histopathology of LA using haematoxylin & eosin (H&E) stain shows acanthosis and a hyperkeratotic epidermis with elongated rete ridges and eosinophilic amorphous deposits in the papillary dermis.^{6,10,12} Histopathological examination of the patient shows tissue covered by hyperplastic stratified squamous epithelium, elongated psoriasiform-rete ridges with acanthotic and hyperkeratotic epidermis. Eosinophilic amorphous masses were found in the papillary dermis with mild chronic inflammatory cells in between. Brown pigment incontinence was also found in the dermis with chronic inflammatory cells lined around blood vessels. Therefore, the histopathological features of the patient were following the diagnosis of LA.

Treatment of LA remains challenging since there is no standard treatment, and treatment is usually unsatisfactory due to the probability of recurrence.^{6,13} The main aim of LA treatment is to control pruritus, thus decreasing scratching frequency and hyperkeratosis.¹² Treatment options are reported, including topical and systemic agents, such as topical and intralesional steroids, topical calcineurin inhibitors, oral retinoid, oral cyclophosphamide, and cyclosporine.⁹ Topical therapy with occlusion is important to increase the

potency of therapy and give protection against trauma. Other modalities are dermabrasion, phototherapy with NB-UVB, or a combination of psoralen with ultraviolet A (PUVA) and acitretin and laser therapy.¹³

Salicylic acid is a topical keratolytic agent. In the concentration of 3% to 6%, it causes the shedding of scales by softening the stratum corneum, loosening connections between corneocytes, and dissolving the intracellular matrix, thereby increasing the absorption of other topical agents into the skin.¹⁴ Topical corticosteroids bind to glucocorticoid receptors and inhibit DNA synthesis and mitosis. As an antiproliferative agent, it has been known to reduce the proliferation and size of keratinocytes and inhibit fibroblast activity and collagen formation.¹³ Therefore, topical combination therapy of salicylic acid and corticosteroid can increase the effectiveness of therapy on thick, scaly plaque lesions.^{12,13} Two case reports showed the satisfactory effect of thinning the lesions and significant improvement in severe itching in LA treated by desoximetasone cream and salicylic acid.^{11,12} In another case report, no improvement in the lesions was reported; however, the patient reported reduced itching. Hence, a combination of therapeutic modalities may be required.¹³

Phototherapy is a safe treatment option for LA but requires several control periods. The latest phototherapy study with NB-UVB found that NB-UVB therapy alleviates the activity of basal cells, thus decreasing amyloid production and effectively reducing pruritus, a factor that worsens LA.⁹ The antipruritic effects of phototherapy occur through several pathways. Cutaneous cells such as keratinocytes, Langerhans cells, mast cells, eosinophils, and infiltrating lymphocytes release cytokines and inflammatory factors as essential mediators in chronic pruritus. These cells can be reached and are affected by UVB. Moreover, UV light also reaches and interacts with a network of cutaneous sensory nerves in the epidermis and upper dermis.

Phototherapy may induce "soluble antipruritic factor" (sAPF) and indirectly inhibit the itch signal pathway.¹⁵ Phototherapy may alleviate itching and abolish an essential factor that induces and worsens LA. Furthermore, phototherapy reduced topical treatment and decreased antihistamine use, further reinforcing the use of NB-UVB in skin diseases associated with pruritus.¹⁶

In this case, the patient was given a topical combination therapy of salicylic acid 6% as the

keratolytic agent and clobetasol propionate 0,05% as the corticosteroid, and narrowband UVB phototherapy once a week with an initial dose of 300 mJ/cm² and increased by 10-20% on the next episodes. After being observed for six weeks, the lesions on the patient's arms showed improvement, while the lesions on her shin showed no significant improvement. However, this combination therapy has been shown to relieve itching significantly. The patient was also educated about the recurrence of the disease, mainly due to the cycle of pruritus, scratching, and hyperkeratosis.

Conclusion

Topical combination therapy of keratolytic agents and potent corticosteroids is a non-invasive therapy that is able to improve lesions in LA patients. NB-UVB phototherapy can significantly relieve severe itching in LA patients. Eliminating the main risk factors, such as pruritus and scratching, can reduce the severity of LA. However, a longer duration of observation is needed to explore the effectiveness of this combination therapy further.

Acknowledgments

We would like to extend our gratitude to IMERI Writing Centre for the proofreading process.

Author Contributions

UHH and SANF are joint first authors of this manuscript. EK and PKE are the supervisors and reviewers of this manuscript. LP participated in the histopathology data analysis and reviewed this manuscript.

Conflict of Interest

No conflict of interest.

References

1. Picken MM. The pathology of amyloidosis in classification: A review. *Acta Haematol.* 2020;143(4):322–34.
2. Gorevic PD, Phelps RG. Amyloidosis. In: Kang S, Amagai M, Bruckner A, et al. editors. *Fitzpatrick's dermatology in general medicine.* 9th ed. New York: McGraw Hill Education; 2019. p.2258–70.
3. Weidner T, Illing T, Elsner P. Primary localized cutaneous amyloidosis: A systematic treatment review. *Am J Clin Dermatol.* 2017;18(5):629–42.
4. Mehrotra K, Dewan R, Kumar JV, Dewan A. Primary cutaneous amyloidosis: A clinical, histopathological and immunofluorescence study. *J Clin Diagn Res.* 2017;11(8):WC01–5.
5. Carolin A, Saraswati PDA. Lichen amiloidosis - diagnosis dan terapi. *Cermin Dunia Kedokteran.* 2023;50(6):306–9. Indonesian.
6. Kim Y, Ioffreda MD, Chung CG. Lichen amyloidosis of the scalp and forehead. *Dermatol Online J.* 2017;23(11):1-3.
7. George AE, Rajan RS, Nandakumar G. A clinical and histopathological study of cutaneous amyloidosis. *J Evol Med Dent Sci.* 2017;6(71):5050–4.
8. Chu H, Shin JU, Lee J, Park CO, Lee KH. Successful treatment of lichen amyloidosis accompanied by atopic dermatitis by fractional CO₂ laser. *J Cosmet Laser Ther.* 2017;19(6):345–6.
9. Alonso-Gonzales J, Rodríguez-Granados MT, Toribio J. Satisfactory response to narrowband UVB therapy in generalized lichen amyloidosis. *Actas Dermosifiliogr.* 2013;104(6):527–9.
10. Humeda Y, Beasley J, Calder K. Clinical resolution of generalized lichen amyloidosis with dupilumab: A new alternative therapy. *Dermatol Online J.* 2020;26(12):4–7.
11. Pramita N, Adiguna M, Suryawati N. Lichen amiloidosis dengan terapi topikal kombinasi. *Media Dermato-Venereologica Indonesiana.* 2020;47:206–10. Indonesian.
12. Indah MS, Made N, Puspawati D, Saputra H. Lichen amyloidosis with combined topical therapy: A case report. *Bali Dermatology and Venereology Journal (BDV).* 2019;2(2):25–7.
13. Gunawan H, Rakhmawati FA. Penatalaksanaan lichen amiloidosis dengan desoksimeson dan asam salisilat topikal: Laporan kasus. *Intisari Sains Medis.* 2021;12(1):379–84. Indonesian.
14. Kwartia SG, Loss M. Other topical medications. In: Kang S, Amagai M, Bruckner A, et al., editors. *Fitzpatrick's dermatology in general medicine.* 9th ed. New York: McGraw-Hill Education; 2019. p.3619.
15. Legat FJ. The antipruritic effect of phototherapy. *Front Med.* 2018;5(11):1–9.
16. Foerster J, Boswell K, West J, et al. Narrowband UVB treatment is highly effective and causes a strong reduction in the use of steroid and other creams in psoriasis patients in clinical practice. *PLoS One.* 2017;12(8):1–14.