Indonesian Journal of Medicine (2024), 09(04): 478-485
Masters Program in Public Health, Universitas Sebelas Maret

Case report
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The Effect of Combined Phototherapy and Topical Corticosteroid Treatment in a Rare Case: 57 Years Old Male Patient with Lichen Amyloidosis

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Received: May 23, 2023; Accepted: September 20, 2024; Available online: October 10, 2024

ABSTRACT

Background: Lichen Amyloidosis (LA) is one of the most common forms of primary localized cutaneous amyloidosis, with clinical manifestations of itchy blackish-brown hyperkeratotic papules. The most predilection site is the upper extensor of the legs. This case report aimed to improve the clinician's knowledge regarding clinical features and supporting examinations to the provision of appropriate therapy in LA.

Case Presentation: A 57-year-old farmer presented to the dermatology and venereology outpatient clinic of Dr. Moewardi General Hospital with itchy black spots nearly all over his body since 2 years ago. Dermatology examination obtained generalized papules and scaly hyperpigmented patches. Dermoscopy revealed a scar-like center with a whitish color in the center. Histopathological examination showed an amorphous eosinophilic (amyloid) in the dermis. Congo's red examination demonstrated a reddish-orange amyloid. We treated the patient with oral cetirizine 10 mg/day, desoximetasone 0.25% cream applied twice a day in the morning and in the evening, Carmed® cream 20% cream applied twice a day in the afternoon and night, phototherapy 350 MJ/cm2 twice a week. We observed for 14 weeks.

Results: Lichen amyloidosis results from amyloid deposits in the papillary dermis which are derived from the degradation of basal keratinocytes. The diagnosis is based on history taking, clinical examination, dermoscopy, and skin biopsy. The combination of phototherapy and topical corticosteroids can be an option for LA therapy, especially for the symptoms of pruritus.

Conclusion: Lichen Amyloidosis is the most common type of primary localized cutaneous amyloidosis, presenting as blackish-brown hyperkeratotic papules. The combined therapy of topical corticosteroids and phototherapy can significantly improve pruritus and skin lesions.

Keywords: Lichen amyloidosis, amyloid, congo red, phototherapy, topical corticosteroid

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Cite this as:
Sambodo SL, Mawardi P, Tansil I Dewi AK, Utama RFD, Octarica SG, Adjie SP (2024). Successful Combination Therapy with Phototherapy and Topical Corticosteroid in a Rare Case: 57 Years Old Male Patient with Lichen Amyloidosis. Indones J Med. 09(04): 478-485. https://doi.org/10.26911/theijmed.2024.09.04.08.

BACKGROUND

Lichen Amyloidosis (LA) is one of the most common types of primary localized cutaneous amyloidosis that caused by heterogeneous extracellular deposition of amyloid protein in the skin without any systemic

involvement (Weidner et al. 2017; Żychowska et al. 2021). Primary localized cutaneous amyloidosis consists of 3 main subtypes, there are lichen amyloidosis (LA), macular amyloidosis (MA) and nodular amyloidosis (NA). The etiology of LA is not fully understood. Several studies have explained the abnormalities signal through oncostatin M type II receptors and interleukin (IL)-31 receptors that cause keratinocyte apoptosis with amyloid deposition (Weidner et al. 2017).

The prevalence of LA is still rare, around 0.2% - 0.3% of the world's population and more common in individuals with darker skin phenotypes (Gunawan and Rahmawati, 2021). The specific epidemiological data on LA have not been recorded, but from several studies it was found that LA generally occurs in people with skin phenotypes III-IV from Asia, South America and the Middle of East (Aung, 2019). Lichen amyloidosis is also generally more common in people with the age of the 4th decade and above female (Aung, 2003; Weidner et al. 2017). Data from the previous study at the Department Dermatology and Venereology, Faculty of Medicine, University of Indonesia in 1983-1987 showed that there were 78 confirmed cases of LA, of which 9 patients were male and 71 patients were female. Sixty patients were aged between 20-50 years and 49 of them suffered from LA for less than 2 years (Djuanda et al. 1988).

The diagnosis of LA can be made through history taking, physical examination and supporting examination to exclude the differential diagnosis of LA (Ladizinski and Lee, 2014). Lichen amyloiddosis has clinical manifestations in the form of localized group hyperpigmented papules that are rarely generalized with a predilection for the inferior extremities, ankles and thighs characterized by very itchy symptoms. Persistent itching complaints and recurrent multiple hyperkeratotic papules in LA patients have a negative impact on the patient's quality of life (Wang et al. 2020). Over time, hyperkeratotic papules can form thickened plaques that are resistant to treatment. Dermoscopic examination of LA can be found a central hub and scar-like whitish color in the center and there are brownish dots in some places (Ouiam et al. 2018). Histopathological examination of LA gives a picture of amorphous eosinophilic (amyloid) in the dermis with hyperkeratosis and acanthosis of the epidermis (Harahap and Marwali, 1998).

The differential diagnosis of LA includes lichen planus and chronic lichen simplex (Ladizinski and Lee, 2014). Lichen planus is a T cell-mediated inflammation of the skin with manifestations of firmly circumscribed and flat polygonal papules (Mangold and Pittelkow, 2019). Dermoscopic examination can be found polymorphic pearly white structures with Wickham's striae and arboriform “fern leaf” projections (Friedman and Sabbahn, 2015). Histological examination of lichen planus shows a typical form of epidermal layer with hyperkeratosis, wedge-shaped hypergranulosis and rete ridges called sawtooth pattern. In the dermoepidermal layer can be found apoptotic cells and colloid-hyaline bodies (civatte bodies) (Ouiam et al. 2018; Ji, 2019). Chronic lichen simplex is a skin disorder with lichenification caused by chronic itching (Ji, 2019). We can found parakeratosis, orthokeratosis, hypergranulosis and psoriasiformis epidermal hyperplasia in histopathological examination of chronic lichen simplex. The papillary dermal showed collagen thickening and infiltration of inflammatory cell with superficial vascular plexus (Ouiam et al. 2018; Ji, 2019).

Patients with LA have several therapeutic options. There are topical or intralesional of steroid, antihistamin, ultraviolet (UV) light therapy, laser therapy, dermabrasion, scalpel scraping and retinoid agents (Ladizinski and Lee, 2014). The most commonly used therapy is steroid, because the pathogenesis of LA is related to the impaired of immune system (Shoen E et al. 2021). The challenge of the treatment of LA is usually associated with genetic factors and other concomitant diseases. A study in Bulgaria in 2020 described that LA associated with rheumatoid arthritis (RA). Lichen amyloidosis is also associated with dermatitis atopic, hepatitis C and lichen planus (Matsumura and Yamamoto, 2017). Although the pathogenesis of LA is unclear, it should be a concern for clinicians to sharpen the history taking and physical examination to diagnosis of LA (Tchernev, 2017). Based on these problems, the authors compiled this paper which discussed a case report of LA. This case report aims to improve the clinician's knowledge regarding clinical features and supporting examinations to determine the appropriate therapy in LA. Delayed diagnosis and therapy of LA may worsen the lesion and also impaired the quality of life of LA patients.

CASE PRESENTATION

A 57-year-old man came to the Dermatology and Venereology Outpatient Clinic at the Dr. Moewardi General Hospital Surakarta with chief complaint of black spots appearing on almost the entire body since 2 years ago. From the patient, since 2 years ago appeared nodules accompanied by itchy black spots on the body. Patient often scratch the skin, so that the skin becomes thick. The papules felt itchy, especially during the day and hot temperature.

There are no complaints of fever or muscle aches. The patient has not been treated for this complaint. The patient complained of black spots extending to both hands and feet 1.5 years later. The patient then checked himself into the Karanganyar Hospital and was given medicine by Dermatovenereologist but the patient can't recall the name of the medicine. Complaints felt slightly improve in term of itchiness after treatment at Karanganyar Hospital.

The complaint persisted. Itchy black spots almost all over the body, so it interferes with activities. The patient then went to Dr. Moewardi General Hospital Surakarta. Based on the past medical history, the patient had no history of hypertension, diabetes mellitus, atopy or chronic disease. The patient claimed that had food allergy to fish and shrimp which rarely recurs. Patient works as a farmer in the fields and rarely wore personal protective equipment from the sun such as hat, sunscreen or skin moisturizer. None of the patient's family members had previous similar complaints and there was no history of atopy, drug and food allergies or chronic diseases in the family.

RESULTS

Dermatology examination obtained generalized papules and scaly hyperpigmented patches (Figure. 1). Dermoscopy revealed a scar-like center with a whitish color in the center. Histopathological examination showed an amorphous eosinophilic (amyloid) in the dermis (Figure. 2). Histological examination with Hematoxylin and Eosin (H&E) staining in the epidermal layer showed parakeratosis, the dermis layer contained amorphous eosinophilic (amyloid) masses, lymphocytes and histiocytes (Figure. 3A). Specific examination using

Congo Red showed orange-reddish amyloid (Figure. 3B).

Based on the history, physical examination and supporting examinations, we diagnosed the patient with lichen amyloidosis. The patient received therapy with cetirizine tablet 10 mg a day, desoximetasone cream 0.25% and Carmed® cream 20% which were applied twice a day in the morning and evening, afternoon and night, respectively on affected area of the body and narrow band ultraviolet B (NB UVB) 350 phototherapy MJ/cm^2 twice a week. The patient showed good response of the therapy and revealed clinical improvement after 14 weeks.

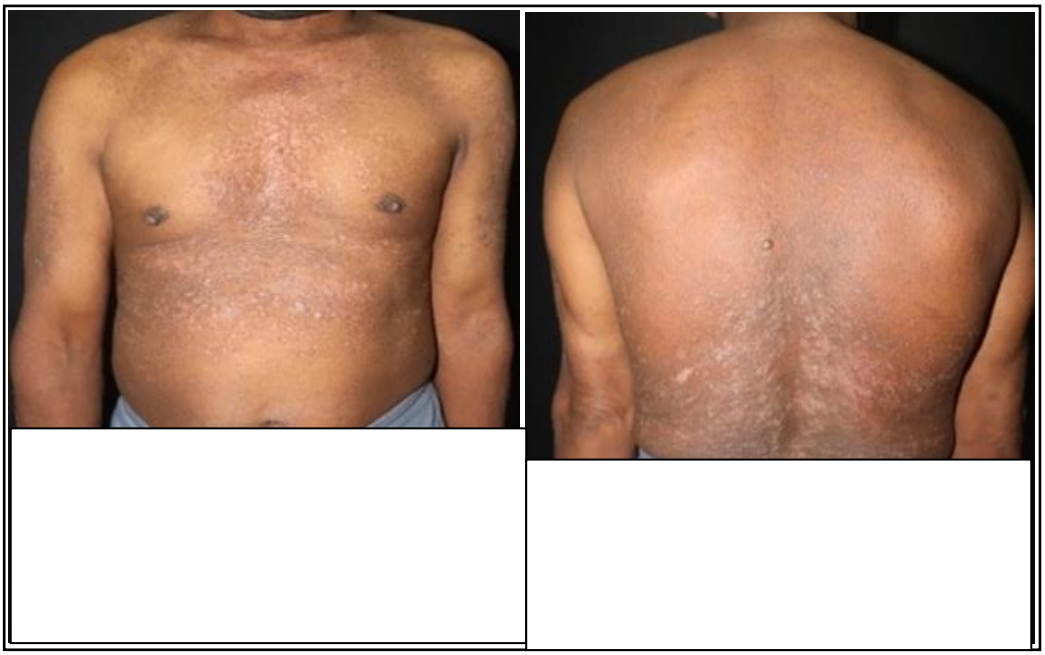

Figure 1. Clinical manifestation : papules, patches, multiple discrete hyperpigmented plaques, partly confluent, with overlying scales.

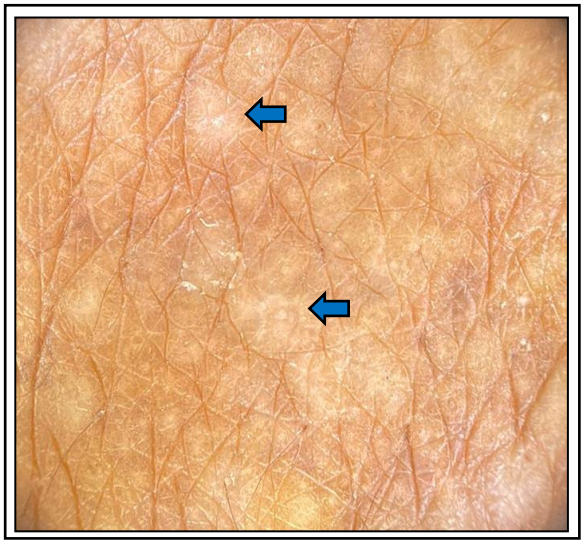

Figure 2. Dermoscopy findings : a scar-like center was seen with a whitish color in the center (blue arrow).

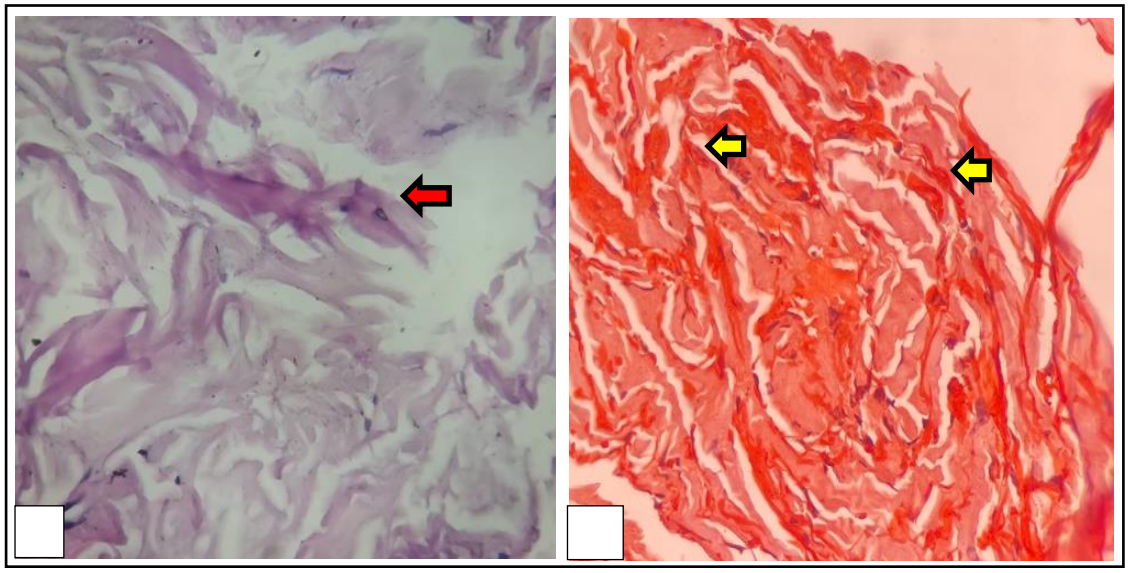

Figure 3. A. Histopathological findings : an amorphous eosinophilic (amyloid) mass (red arrow); B. Congo Red findings : reddish-orange amyloid (yellow arrow).

DISCUSSION

Lichen amyloidosis (LA) is the most common form of primary localized cutaneous amyloidosis with clinical manifestations of hyperkeratotic papules of brown-black color with a predilection most often on the upper extensor legs, but can also occur on the thighs, calves, forearms and back (Weidner et al. 2017).

The pathogenesis of LA is unknown, although it can be related to viral and genetic etiology. Lichen amyloidosis is caused by amyloid deposits in the papillary dermis resulting from degradation of basal cell keratinocytes (Humeda et al. 2020). Lichen amyloidosis is characterized by deposits of amyloid in the dermis without any systemic manifestations. Amyloid is a derivative of keratin where cytokeratin function as an amyloid precursor that affects the dermis (Ladizinski and Lee, 2014). The accumulation of amyloid causes an itchy process which can cause irritation to the skin (Guillet et al. 2022). Research by He A et al in 2016 explained that IL-31 may play a role in the itchy intensity associated with primary localized cutaneous amyloidosis (He et al. 2016).

The patient in this case is a 57 years old man with dark skin and the process of developing skin lesions also occurs slowly, from initially the lesions are just nodules accompanied by itchy black spots on the body until similar lesions appear in the affected area. Lichen amyloidosis is more common in males aged in the 5th and 6th decade. Lichen amyloidosis is a disorder that occurs chronically and progressively (Ladizinski and Lee, 2014). This patient has a job as a farmer and rarely uses personal protective equipment from the sun such as hat, sunscreen or skin moisturizer. Another study showed 70 patients with LA that 92% of patients had a history of using body scrubbers and 50% of patients had a history of exposure to sunlight (Chapalamadugu and Rao, 2020).

In the case of this patient, the clinical manifestations that appeared were papules, patches, hyperpigmented plaques accompanied by scales on them in the generalized region. According to Pramita et al in 2020,

the clinical manifestations of LA can be in the form of macules, papules and nodules. The papular type is characterized by multiple, discrete, hyperkeratotic papules which are very itchy and can confluent to form brown to red-brown plaques that feel rough and thick. Lesions often start unilaterally and can expand to be bilateral and symmetrical (Pramita et al. 2020).

Dermoscopy is a useful non-invasive diagnostic tool. The results of dermoscopy examination in this case showed a scar-like center with a whitish color in the middle. According to previous research, dermoscopy examination in LA cases will find two main, distinctive patterns, namely the central hub and scar-like in the form of a whitish color in the center and in several places there are brown spots (Ouiam, 2018).

Skin biopsy followed by histopathological examination is one of the diagnostic method with the best level of accuracy for diagnosing lichen amyloidosis and eliminating differential diagnoses (David, 2010). The patient underwent a biopsy examination of the lesion from the posterior truncus region and after examination with H&E staining the epidermal layer appeared parakeratosis, the dermis layer contained amorphous eosinophilic (amyloid) masses, instead of lymphocytes and histiocytes. Histopathological examination of LA will show pigmentary incontinence with melanophages as the main feature of lichen amyloidosis. Lichen Amiloidosis depicts eosinophilic amorphous deposits in the papillary dermis with hyperkeratosis and epidermal acanthosis (Harahap and Marwali, 1998). Amyloid deposits are confined to the papillary dermis and do not involve blood vessels or adnexal structures. Focal amyloid deposits are large enough to extend into the papillae and replace the elongated rete ridges (Ouiam, 2018; Ji, 2019). Amyloid deposits can be a marker of systemic disorders or chronic diseases including malignant skin processes such as basal cell carcinoma (Mawardi et al. 2019).

The patient was also examined by Congo Red and the results showed reddish-orange amyloid. The Congo Red examination is a standard examination to see amyloid in the skin (Chen et al. 2022). Congo Red examination has good sensitivity but must be carried out with the right technique because in some cases of lichen amyloidosis, either in clinical manifestations or systemically upright histopathologically but the Congo Red examination showed a negative result due to an error in the biopsy technique and method of staining the sample (Bowen et al. 2012).

Therapy for LA is aimed at reducing pruritic symptoms, avoiding secondary infections due to scratching of the skin, preventing secondary lesions in the form of lichenification and overcoming cosmetic complaints, however, LA lesions that form over time are often resistant to treatment (He et al. 2016). Until now, no therapy has been found. which are both curative and effective for lichen amyloidosis (Pramita et al. 2020). Therapeutic options for LA include topical or intralesional steroids, antihistamines, UV light therapy, laser therapy, dermabrasion, scraping with a scalpel and with retinoid agents (Ladizinski and Lee, 2014). Milder cases of LA can be given corticosteroids. Topically potentiated topical administration, occlusion or a combination of keratolytics such as urea and salicylic acid may be effective (Bowen et al. 2012; Groves RW, 2012).

Lichen amyloidosis is the most common type of primary localized cutaneous amyloidosis, clinically manifest as blackish brown hyperkeratotic papules. The combined therapy of topical corticosteroid and phototherapy results in significant

improvement of pruritis and skin lesions of LA.

AUTHOR CONTRIBUTION

Each author prepared the figures, authored the majority of the manuscript.

FINANCIAL SUPPORT AND SPONSORSHIP

The authors declare that this study had no funding resource.

ACKNOWLEDGEMENT

We thank to patient's family, all internists and surgeons who were involved in patients' care.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

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