

CASE AND RESEARCH LETTER

Satisfactory Response to Narrowband UV-B Therapy in Generalized Lichen Amyloidosis[☆]

Liquen amiloide generalizado con respuesta satisfactoria a ultravioleta B de banda estrecha

To the Editor:

Lichen amyloidosis (LA) is a form of primary localized cutaneous amyloidosis that is associated with pruritic skin-colored or hyperpigmented hyperkeratotic papules^{1,2}; these papules can coalesce to form plaques that often display a wavy pattern. The lesions are found mainly in the pretibial region, and generalized involvement is uncommon.^{3,4} Numerous approaches have been used to treat LA, although none has achieved a complete cure. We present a case of generalized LA that responded satisfactorily to narrowband UV-B therapy.

The patient was a 69-year-old man with Fitzpatrick skin type III and a past history of insulin-dependent diabetes mellitus, arterial hypertension, and ischemic heart disease. He consulted for pruritic lesions on the trunk and extremities that had first appeared 4 years earlier and had been treated unsuccessfully with topical corticosteroids. A series of laboratory and additional tests were performed to screen for potential causes of the pruritus, and the patient did not present an underlying eczematous condition. Physical examination revealed slightly desquamating plaques formed of multiple hyperpigmented hemispheric papules, many of which had been eroded by scratching. The lesions affected the anterior and posterior surfaces of the arms and legs, abdomen, back, and buttocks, with a symmetrical bilateral distribution (Fig. 1). Histopathology revealed compact orthokeratotic hyperkeratosis, irregular acanthosis, and deposits in the papillary dermis that caused widening of the papillae with lateral shift of the epidermal ridges. The deposits were composed of acellular, amorphous eosinophilic material with abundant fissures caused by retraction; the material fluoresced green with thioflavin T (Fig. 2). We thus confirmed the diagnosis of



Figure 1 Extent of the skin condition before treatment. Inset: Detailed image of the lesions on the thigh.

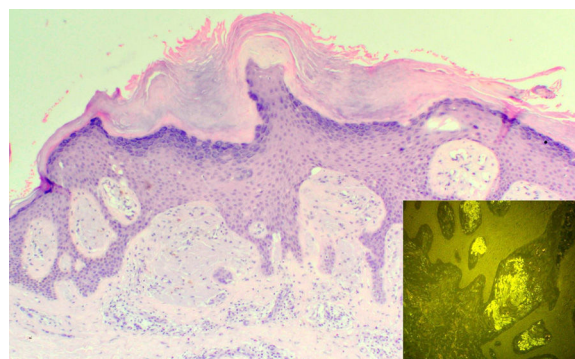


Figure 2 Compact orthokeratotic hyperkeratosis, irregular acanthosis, and deposits in the papillary dermis that caused widening of the papillae with lateral shift of the epidermal ridges (hematoxylin-eosin, original magnification x100). Inset: Deposits of amyloid material in the papillary dermis showing green fluorescence (thioflavin T, original magnification x200).

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LA. Phototherapy with narrowband UV-B was started (3 times weekly). The initial dose was 0.35 J/cm² (determined according to phototype), which was increased by 10% to 20% at each session. A significant reduction in the pruritus and progressive flattening of the papules in all the affected areas was observed from the initial sessions. Complete resolution of the lesions on the trunk and arms was achieved after 57 sessions and a cumulative dose of 109.27 J/cm² (maximum dose, 2.33 J/cm²) (Fig. 3). The physical and symptomatic improvement persisted at the time of writing, after 4 months of follow-up, with no further treatment.

Treatment of LA is somewhat unsatisfactory. The different approaches tried include topical or intralesional corticosteroids, oral and topical dimethylsulfoxide, calcineurin inhibitors, oral retinoids, cyclophosphamide, dermabrasion, and neodymium:yttrium-aluminum-garnet laser therapy, as well as various phototherapy modalities.^{1,5}

Only 3 cases of LA treated satisfactorily with narrowband UV-B therapy have been reported to date.⁵⁻⁷ In 2 of those cases, LA was associated with refractory atopic dermatitis, and narrowband UV-B was combined with other treatments to improve the clinical condition.^{5,6} The third case involved a man with generalized LA whose lesions characteristically affected areas of the body with lower skin temperatures, sparing areas with higher temperatures. The patient's response to narrowband UV-B monotherapy was excellent.⁷

The amyloid substance in LA is thought to originate from necrosis of the epidermal keratinocytes of the basal layers of the epidermis.⁸ Narrowband UV-B therapy diminishes

basal cell activity, thus reducing the production of amyloid.⁵ Furthermore, narrowband UV-B has proven effective for the treatment of pruritus by suppressing the proliferation of keratinocytes and reducing apoptosis of keratinocytes and T cells and inflammation. In this way, it is possible to diminish or eliminate one of the factors that very probably induce and/or worsen LA. Other authors consider that the effectiveness of narrowband UV-B therapy is due to the marked elevation of the temperature in the booth, with the consequent increase in skin temperature and suppression of the heat-dependent synthesis of amyloid.⁷

Grimmer et al.⁹ recently reported 2 cases of LA treated with a combination of psoralen-UV-A therapy and oral acitretin. The authors concluded that this modality was effective and practical and that its effects were maintained over time. Finally, a study that compared the efficacy of topical corticosteroids with UV-B phototherapy and topical psoralen-UV-A in patients with LA found better results, namely, diminished pruritus and a decrease in the number of skin lesions, in the areas treated with phototherapy.¹⁰

To our knowledge, we present the second case of LA not associated with other skin conditions and that responded satisfactorily to narrowband UV-B in monotherapy. Although further studies are necessary to define the exact mechanism of action and level of efficacy, we believe that narrowband UV-B therapy could represent a safe and effective alternative in the management patients with generalized LA lesions.

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Figure 3 Improvement in lesions after completing treatment with narrowband UV-B therapy (57 sessions). Inset: Detail of the lesions on the thigh.

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Vogt-Koyanagi-Harada Disease: A Disorder Unfamiliar to Dermatologists[☆]

Enfermedad de Vogt-Koyanagi-Harada. Una entidad poco conocida para el dermatólogo

To the Editor:

Vogt-Koyanagi-Harada disease is a rare condition that is characterized by bilateral uveitis, meningitis, auditory symptoms, and skin disorders such as vitiligo, poliosis, and alopecia.¹ This report aims to draw attention to a condition that is widely discussed in the literature yet unfamiliar to dermatologists.

We present the case of a 37-year-old man from Bolivia who was seen in the emergency department with vertigo, headache, eye pain, and loss of vision in both eyes that had begun a week earlier. Three months later he was referred to the dermatology department with rapid-onset alopecia of the scalp. Physical examination revealed horizontal rotatory nystagmus at rest and on looking upward and to the right. Ophthalmological examination revealed a marked loss of visual acuity in both eyes; anterior pole biomicroscopy revealed Tyndall phenomenon with posterior synechiae (Fig. 1). Nonscarring diffuse alopecia was also observed on the scalp, together with poliosis of the right eyebrow (Fig. 2, A and B). The results of the blood workup—complete blood count, biochemistry, clotting, protein analysis, antibodies to antinuclear antigen and extractable nuclear antigen, serology for syphilis and viruses (hepatitis A, B, and C viruses; human immunodeficiency virus; Epstein-Barr virus; cytomegalovirus), and thyroid hormones—were negative or normal. Cerebral magnetic resonance imaging ruled out meningeal or encephalic disease. Histocompatibility testing was positive for HLA-DR4 and HLA-DR53. These findings enabled a diagnosis of Vogt-Koyanagi-Harada disease to be confirmed. Treatment with intravenous methylprednisolone (1 g) was prescribed for 3 days, with subsequent oral administration tapered over 12 weeks. The patient's clinical condition improved considerably, not only in terms of systemic manifestations, but also with complete resolution of the alopecia. However, the new hair remained whitish-gray in color (Fig. 3).

Vogt-Koyanagi-Harada disease, which was first reported in 1951, is a rare systemic inflammatory disorder mediated

by T lymphocytes acting against pigmented cells in the uvea, skin, inner ear, and leptomeninges.¹

It has an incidence of 6.5 cases per million and is more common in Hispanic and Native American individuals and Asian women aged between 20 and 50 years. Although its etiology is unknown, immunogenetic analysis suggests a strong association with some HLA antigens (DR4, DRB1, and DR53) and genes of the tyrosinase family that cause susceptibility to the disease.² It is associated with various infectious diseases and other disorders whose pathogenic mechanisms are of immunological origin, such as autoimmune thyroid disease, psoriasis, linear immunoglobulin A bullous dermatosis, and inflammatory vitiligo as a consequence of the destruction of melanocytes by cytotoxic CD8⁺ T lymphocytes.^{2–4}

Diagnosis is based essentially on clinical manifestations, as there are no specific confirmatory tests, and the diagnostic criteria include dermatologic findings (alopecia, poliosis, and vitiligo) (Table 1). Typical extracutaneous clinical manifestations include disorders of the eyes (bilateral granulomatous uveitis, retinal detachment, and loss of vision) and neurological and auditory conditions (headache, aseptic meningitis, vertigo, nystagmus, and hypoacusis).⁵

Alopecia can be seen in 70% of cases within weeks or months after the onset of ocular symptoms. It resolves completely after a variable period. Alopecia and the poliosis traditionally reported by ophthalmologists were recently considered to be alopecia areata with subsequent growth of white hair.⁶ Vitiligo affects almost half of all cases; it is often symmetrical and the pigmentary changes may be permanent.

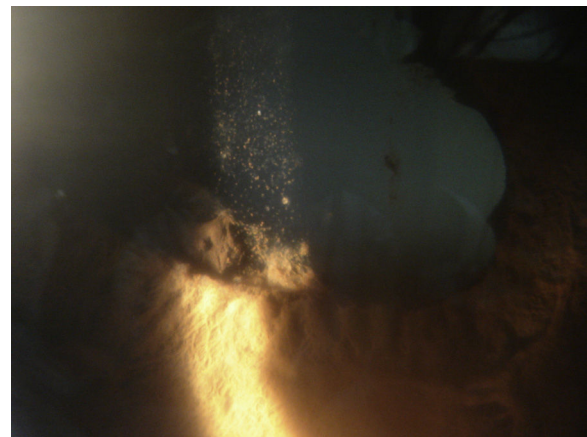


Figure 1 Tyndall phenomenon with the presence of posterior synechiae characteristic of anterior uveitis.

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