Response to oral acitretin in lichen amyloidosis

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ABSTRACT

We report the therapeutically challenging case of a patient with severe and extensive lichen amyloidosis (LA) who responded to oral acitretin and topical corticosteroids. Colloid milia and terra firma-forme dermatoses were noted post healing of the lesions of LA. There has been no recurrence of lesions post 8 months of follow-up. We recommend that acitretin should be used more often in severe and recalcitrant cases of LA.

Key words: Acitretin, lichen amyloidosis, retinoids

INTRODUCTION

Lichen amyloidosis (LA) is either a primary disease which is characterized by the deposition of amyloid in previously normal skin without any association with other cutaneous or systemic disorders or it can be a response to chronic scratching secondary to atopic dermatitis, stasis dermatitis or an interface dermatitis. The treatment of generalized LA is difficult. There are few reports documenting response of oral retinoids in this condition. We report a patient with extensive LA who responded very well to treatment with oral acitretin.

CASE REPORT

A 59-year-old female presented with a 17 year history of intense pruritus with progressive appearance of multiple hyperkeratotic papules over the lower extremities and abdomen. There was severe pruritus with an imperative need to scratch. The past history was not significant for any pre-existing cutaneous or systemic disorder. She gave a history of use of topical corticosteroids with salicylic acid combination off and on with no relief. Upon dermatological examination, there were bilaterally symmetrical hyperkeratotic, hyperpigmented, dome shaped papules over the anterior and posterior aspects of the legs, lateral aspects of the thighs and the abdomen [Figure 1]. Laboratory investigations including the complete blood count, erythrocyte sedimentation rate, renal, hepatic, lipid and thyroid profiles, urine analysis and IgE levels were normal. The clinical diagnosis of LA was confirmed on histopathology by the presence of globular eosinophilic deposits throughout the papillary dermis that stained positive with Congo red [Figure 2].

The patient was started on oral acitretin 25 mg once daily with topical 0.05% halobetasol propionate with 3% salicylic acid lotion at bedtime. A month later, there was 90% reduction in itching and the lesions of LA started to flatten out. On completion of two months of daily dosing, acitretin was decreased to 25 mg every alternate day and the topical steroid-salicylic combination was stopped. There was complete flattening of the lesions with the disappearance of itching [Figure 3]. The patient was further maintained on oral acitretin 25 mg twice a week for two months, after which acitretin was stopped. The lesions of LA healed with colloid milia formation and the patient had terra firma-forme dermatosis on the posterior aspect of the legs post-treatment [Figure 4]. The patient did not experience any subjective adverse effects related to acitretin. Hepatic and lipid profiles repeated every two months were found to be normal. The results were maintained at eight months follow-up.

DISCUSSION

The treatment of LA is unsatisfactory particularly when the disease is severe and extensive. Multiple treatments have been advocated but none are uniformly effective. Among the topical treatments, potent topical corticosteroids, calcipotriol, keratolytics, dimethylsulfoxide and tacrolimus

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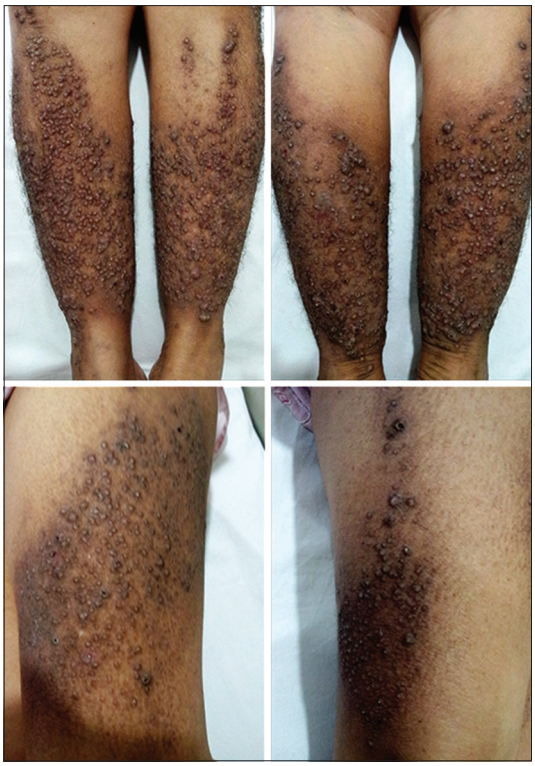

Figure 1: Pre-treatment photograph – Multiple hyperkeratotic, hyperpigmented papules over the legs, calves and the medial aspect of thighs

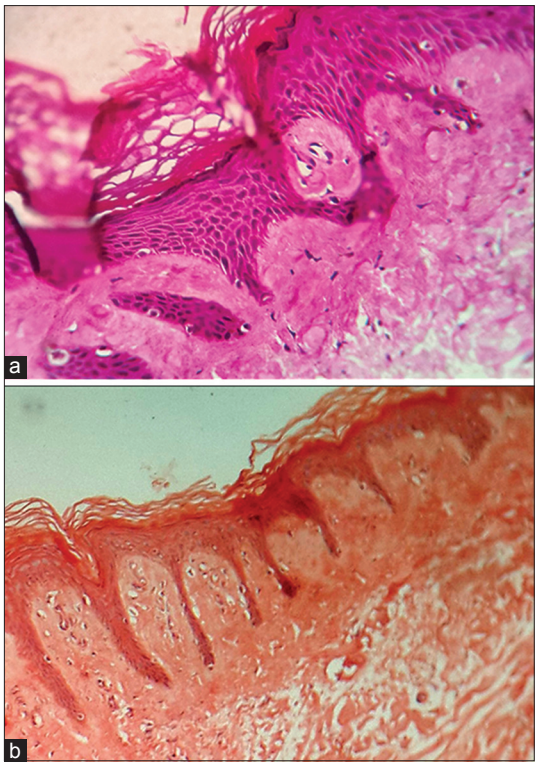

Figure 2: (a) Eosinophilic globular deposits throughout the papillary dermis (H and E, ×10) (b) Deposits confirmed to be amyloid (Congo red stain, ×10)

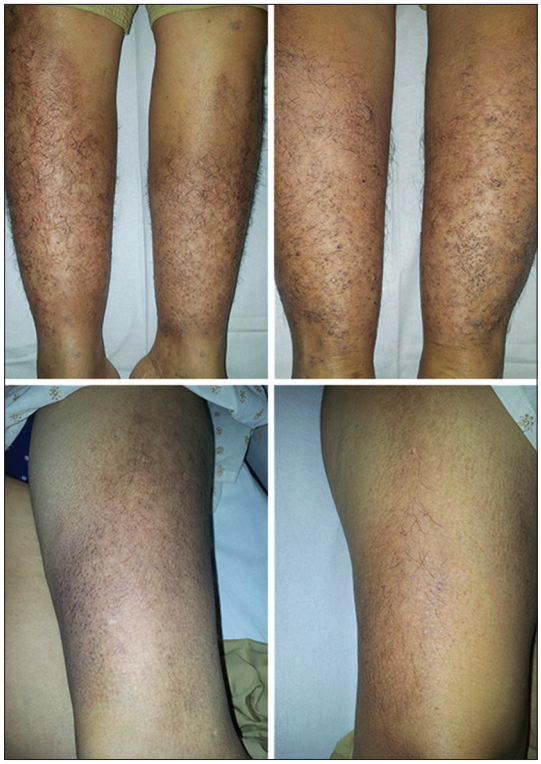

Figure 3: Complete flattening of the lesions of lichen amyloidosis after 4 months treatment with oral acitretin

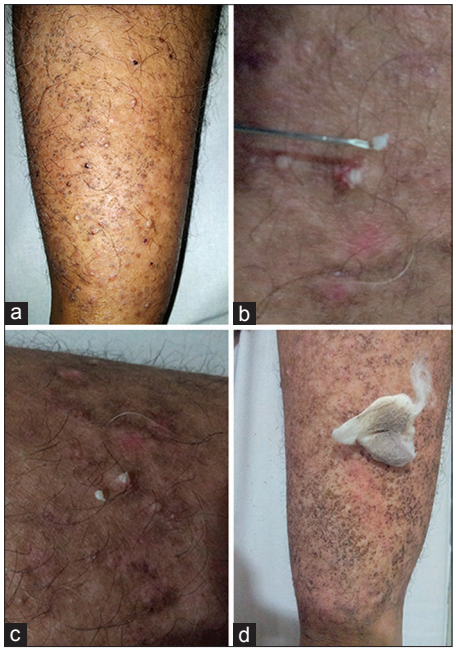

Figure 4: (a) Lesions of lichen amyloidosis healed with the formation of colloid milia (b) Confirmation of colloid milia by needling of one lesion (c) Extrusion of whitish keratinous material confirms the diagnosis of colloid milia (d) Complete removal of the dirty black appearing material on the calf using an alcohol soaked cotton swab confirms the diagnosis of terra firma-forme dermatoses over the posterior aspect of legs

have been tried. Oral treatments that have been used include colchicine, cyclosporine, low dose cyclophosphamide and oral retinoids with or without photochemotherapy. Procedural treatments that have been tried include deep peels such as trichloroacetic acid and phenol, dermabrasion, lasers such as erbium-yttrium aluminum garnet and CO_2 Lasers.

There have been varied responses with the use of oral retinoids in LA. There have been many reports demonstrating the usefulness of oral etretinate [1,2] acitretin[3,4] and isotretinoin.[5] Some describe a rapid response within 1-2 weeks with complete clearance of pruritus and papules and long lasting remissions when retinoids are ceased. [1,3-6] Others have a positive but incomplete response to treatment with a relapse after discontinuation.[2] There has also been a report of no response.[7] The response of LA to retinoids possibly is dependent upon the genetic constitution of the patient and the type of LA.

The apoptotic basal keratinocytes are thought to be the source of amyloid K implicated in the causation of LA. Repeated mechanical trauma induced by chronic friction is one of the proposed mechanisms for keratinocyte degeneration. Acitretin has marked anti-inflammatory and anti-proliferative action that inhibits epidermal acanthosis and further inflammatory injury to the basal keratinocytes. It also promotes keratinocyte differentiation and prevents amyloidosis by correcting keratin 5:14 imbalance.[8] Regulation of the levels of heat shock protein prevent the aberrant folding of keratin[9,10] and reduction in the apolipoprotein E by the retinoids suppress the production of amyloid K.[4] Furthermore, retinoids are potent inducers of apoptosis and may stimulate macrophages to phagocytose and remove the amyloid K deposits from the dermis. These two effects could down regulate amyloid K formation.

An excellent response to acitretin was observed in the treatment of extensive LA in our patient. No adverse effect or relapse till date was observed. Thus, acitretin should be considered more often in the treatment of extensive, recalcitrant cases of LA.

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