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OLGU SUNUMU

Successful treatment of lichen amyloidosis with cryosurgery

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Summary

Lichen amyloidosis is a rare, chronic pruritic disorder characterized by amyloid deposition in the skin without evidence of visceral involvement. Clinically, lichen amyloidosis demonstrates discrete, intensely pruritic, hyperkeratotic papules that may coalesce into plaques, mainly located on the legs. Different treatment modalities including topical and intralesional corticosteroids, topical dimethyl sulfoxide, ultraviolet B, oral psoralen plus ultraviolet A, retinoids and dermabrasion have been described, but the results are generally disappointing. There are paucity of reports of liquid nitrogen cryosurgery as a treatment option in the management of lichen amyloidosis in the literature. In this article, we report a case of lichen amyloidosis, with pruritic hyperkeratotic papules on the left shin of 7 years' duration, successfully treated with cryosurgery.

Key words: Cryosurgery, lichen amyloidosis, treatment

Özet

Liken amiloidoz'un kriyocerrahiyle başarılı tedavisi

Liken amiloidoz, iç organ tutulumu olmaksızın deride amiloid birikimi ile seyreden, ender, kronik ve kaşıntılı bir cilt hastalığıdır. Liken amiloidoz, çoğunlukla bacaklarda yerleşim gösteren kaşıntılı, plak oluşumuna neden olabilen ayrı yerleşimli hiperkeratotik papüllerle klinik seyir gösterir. Tedavi seçeneği olarak topikal ve intralezyoner kortikosteroid, topikal dimetilsülfoksit, ultraviyole B, oral psoralen ve ultraviyole A, retinoik asidler ve dermabrazyon yer almakla birlikte, sonuçlar genellikle yüz güldürücü değildir. Literatürde Liken amiloidoz tedavisinde kriyoterapinin etkinliği ile ilgili çok az sayıda çalışma bulunmaktadır. Bu makalede, 7 yıldır devam eden, sol bacakta kaşıntılı hiperkeratotik papüllerin gözlendiği ve kriyoterapi ile başarıyla tedavi edilen bir liken amiloidoz olgumuzu sunuyoruz.

Anahtar kelimeler: Kriyoterapi, liken amiloidoz, tedavi

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Introduction

Lichen amyloidosis (LA) is an unusual, persistent pruritic papular eruption, characterized by cutaneous amyloid deposition without systemic involvement. The clinical features consist of discrete, intensely pruritic, hyperkeratotic skin colored or yellowish brown papules that may coalesce into plaques, mainly located on the lower extremities (1,2).

Although many treatment modalities including topical and intralesional corticosteroids, topical dimethyl sulfoxide (DMSO), ultraviolet B (UVB), oral psoralen plus ultraviolet A (PUVA), retinoids and dermabrasion have been described, the results are generally unsatisfactory (3-5). There is paucity of reports of liquid nitrogen cryosurgery as a treatment option in the management of LA in the literature. We hereby report a 24-year-old man diagnosed as LA, with pruritic hyperkeratotic papules on the left shin of 7 years' duration, successfully treated with cryosurgery.

Case Report

A 24-year-old man presented with a 7 year history of persistent, pruritic eruption on his left shin, unresponsive to topical corticosteroid treatment. The patient denied a history of trauma. He was otherwise healthy and had took no medication.

On physical examination we observed excoriated, firm, scaly hyperkeratotic papular lesions on his left shin (Figure 1). There were no other cutaneous or mucosal abnormalities. Routine laboratory investigations including complete blood count, renal, hepatic and thyroid function tests and urinary analysis were within normal limits.

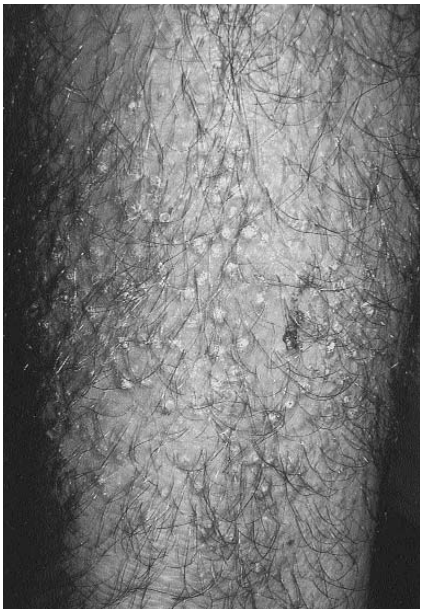

Figure 1. Discrete, scaly, hyperkeratotic papules on the shin

Ultrasonography of the thyroid and surrenal glands revealed no pathological findings.

A 4 mm punch biopsy specimen revealed orthokeratosis, hypergranulosis, irregular acanthosis, and a perivascular lymphohistiocytic inflammatory infiltrate and melanophages in the superficial dermis. In the papillary dermis, focal collections of amorphous eosinophilic-staining material that stained positive with crystal violet were present (Figure 2). A diagnosis of LA was established and

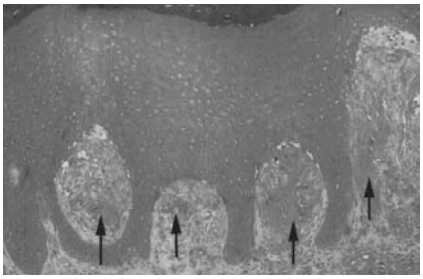

Figure 2. Arrows indicating focal collections of amorphous material in the papillary dermis consistent with lichen amyloidosis (crystal violet stain, X80)

the patient was treated with liquid nitrogen cryosurgery using a 15 second, single freeze thaw cycle. One month later there was prominent regression of the papular lesions, and a second session of cryosurgery was carried out. At the end of the second month, the patient was symptom free, and a near complete resolution of the lesions was achieved (Figure 3). A side effect of hypopigmentation was also observed in the treatment site. A control biopsy specimen of the previously involved skin showed clearance of amyloid deposits in the superficial dermis. No recurrence was noted during 6 months of follow-up.

Figure 3. Clearance of lesions with postinflammatory hypopigmentation after cryosurgery

Discussion

The term amyloidosis refers to the extracellular deposition of amyloid, which is normally a soluble autologous protein, in a characteristic abnormal form (6). Primary cutaneous amyloidosis is characterized by the deposition of amyloid in a previously apparently normal skin without internal organ involvement. Primary cutaneous amyloidosis has been classified into three types: macular, lichen (papular), and nodular (tumefactive form) amyloidosis (7). Unusual rare types, which include poikiloderma-like cutaneous amyloidosis, bullous amyloidosis and vitiliginous amyloidosis have been reported in the literature (7). LA is seen more frequently in South-east Asia, China and some South American countries (8). The course of LA is unknown, but genetic factors based on familial cases of LA, Epstein-Barr virus and chronic friction have been implicated in the etiology of the disease (1,9,10). According to the apoptosis theory, degenerated keratins from apoptotic keratinocytes are transformed into amyloid by dermal macrophages and fibroblasts (11). Yamagihara et al. considered that cutaneous amyloid deposits might also be secretory products of keratinocytes (12). The presence of amyloid K, an epidermal keratin derived amyloid suggests that the amyloid is derived from keratin peptides originating in the epidermis in LA (9).

The lesions of LA usually occur in a symmetrical distribution as multiple, scaly, closely set papules, some with a lichenified surface, mainly located on the lower legs, especially around the ankles, and, to a lesser extent, the trunk and the upper extremities. Atypical localization with involvement of the vulva, ears and buttocks and generalized cases have also been described (13-15). Histological features include eosinophilic globular masses of amorphous material in the papillary dermis that stain positively for amyloid with Congo red and crystal violet stains. Hyperkeratosis, hypergranulosis, slight degree of epidermal hyperplasia, colloid bodies, basal cell vacuolar degeneration, melanophages, and a mild superficial and perivascular lymphocytic infiltrate in the dermis may also be observed (6,9). The findings of excoriated, hard and hyperkeratotic papules on the anterior aspect of the shin as well as the deposition of amorphous amyloid material in the papillary dermis was consistent with a diagnosis of LA in our case.

LA is reported in association with several disorders including multiple endocrine neoplasia type 2A (Sipple syndrome), atopic dermatitis, lichen planus, mycosis fungoides, angiolymphoid hyperplasia with eosinophilia, Kimura's disease and Alagille syndrome (2,16-18) No associated dis-

ease has been observed in our patient despite detailed clinical and radiological examinations.

The treatment of LA is frequently disappointing. Topical corticosteroids have no effect on papular lesions, except for variable improvement in pruritus. Other treatment modalities including intralesional corticosteroid injection, topical tacrolimus, topical Capsaicin, oral and topical dimethylsulfoxide (DMSO), photo(chemo)therapy, retinoids, hydrocolloid dressings, dermabrasion, carbon dioxide laser ablation have been tried with mixed results (3-5,19,20). In LA, very limited data regarding effectiveness of cryosurgery exist. Hallel-Halevy et al. have reported a case of LA treated with topical corticosteroid preparation, retinoic acid 0.05% cream, cryosurgery, and systemic antihistamines with only minor effect, however the technique and freeze time of cryosurgery have not been described (12). In contrast, an open spray technique of liquid nitrogen cryosurgery with two sessions of a single 15 second, freeze thaw cycle resulted in complete resolution of pruritus and also clearance of amyloid deposits in the papillary dermis. The mode of action of cryosurgery in lichen amyloidosis should be explained by the destructive mechanism of this treatment modality (21). Cryosurgery is well-known to produce a destructive effect in the skin tissue depending on the depth of freeze, thus resulting in destruction of amyloid deposits located in the superfical dermis in lichen amyloidosis. The only side effect was postinflammatory hypopigmentation on the treatment site, which was neglected by the patient.

In conclusion, we consider that an open spray technique of cryosurgery may be considered in the therapy of localized LA resistant to other treatment options.

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