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Lichen amyloidosis of the scalp and forehead

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Abstract

Lichen amyloidosis is a subtype of primary localized cutaneous amyloidosis (PLCA), which presents as discrete, firm, closely-set 1-3mm, dome-shaped brown papules commonly involving the anterior aspect of shins and extensor surfaces of forearms. We present a case of an otherwise healthy man in his 30s with solitary facial involvement of lichen amyloidosis, which is very uncommon.

Keywords: amyloid, depositional disorders

Introduction

Lichen amyloidosis, a subtype of primary localized cutaneous amyloidosis (PLCA), commonly involves the anterior aspects of shins and extensor surfaces of forearms. The diagnosis of lichen amyloidosis is often made clinically but histopathological assessment can be confirmatory.

Case Synopsis

A man in his 30s presented with a three-year history of monomorphous, tan-colored scaly papules over the left frontal hairline, extending onto the forehead (**Figure 1**). The lesions had gradually increased in number over a period of two years, and were reportedly asymptomatic. There was no improvement with the use of moisturizers or topical antibiotics. A biopsy of a representative lesion was obtained for histopathological assessment, which demonstrated epidermal collarettes surrounding pink hyalinized and amorphous material that filled and expanded



Figure 1. Multiple monomorphous, tan-colored scaly papules over the left frontal hairline, extending onto the forehead.

the papillary dermis (**Figure 2**). Congo red stain was performed, and the material demonstrated apple green birefringence with polarized light microscopy (**Figure 3**). These findings were consistent with a diagnosis of lichen amyloidosis in the frontal hairline and forehead.

Case Discussion

Amyloidosis is a heterogenic group of diseases defined by extracellular deposits of misfolded proteins with features of apple-green birefringence with polarized light microscopy of Congo red-stained tissue. In the skin, amyloidosis may be primary localized or secondary to systemic disease. Primary localized cutaneous amyloidosis (PLCA) is characterized by amyloid deposits in the upper dermis. Amyloid in PLCA is thought to be derived from inappropriate apoptosis of keratinocytes with filamentous degeneration and are usually confined to the papillary dermis [1]. There are three

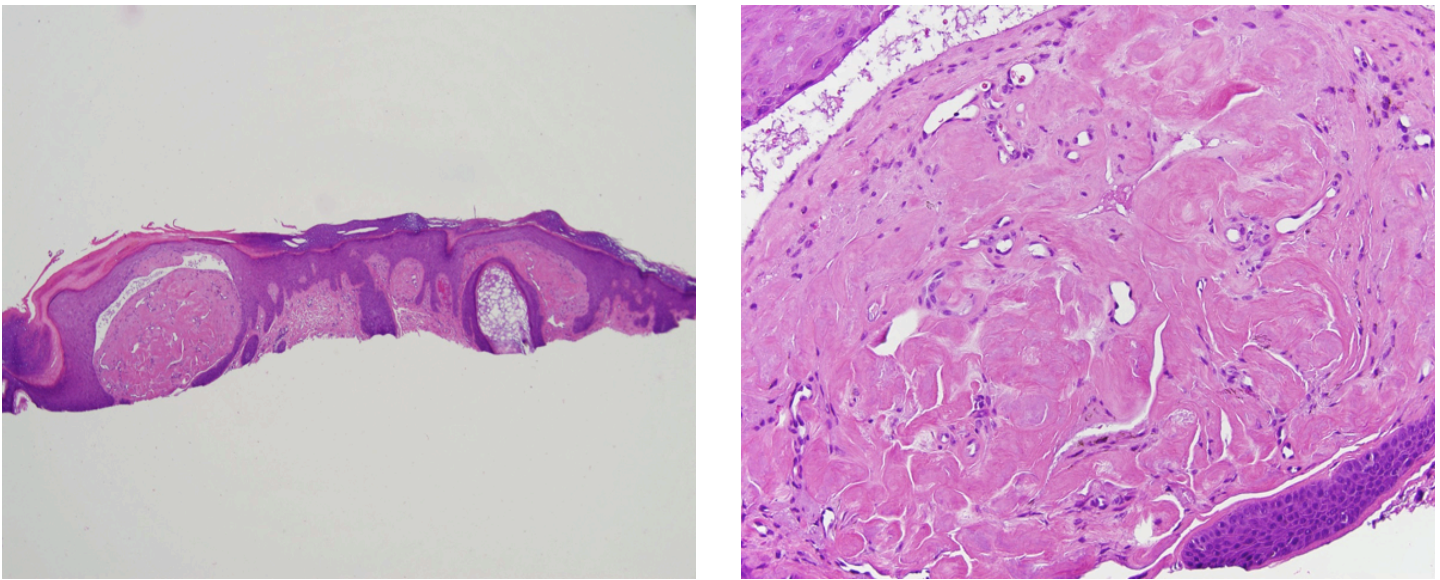


Figure 2. A biopsy specimen of the papule was obtained, and histopathology revealed epidermal collarettes surrounding pink hyalinized and amorphous material that filled and expanded the papillary dermis. H&E, A) 40%; B) 200%.

main forms of PLCA: macular, lichen, and nodular [2]. The most common subtypes are macular and lichen amyloidosis. Macular amyloidosis presents as pruritic brown patches in a rippled pattern. Lichen amyloidosis presents as discrete, firm, closely-set 1-3mm, dome-shaped, brown papules [3].

PLCA is a relatively common skin disorder among Asians and South Americans [3]. Oftentimes, patients confirm prior trauma, rubbing, and scratching of the skin [4]. The exact etiology is not yet fully understood but genetic predisposition, Epstein-Barr virus, and environmental factors have all been proposed as possible etiologic factors [5]. It has been described

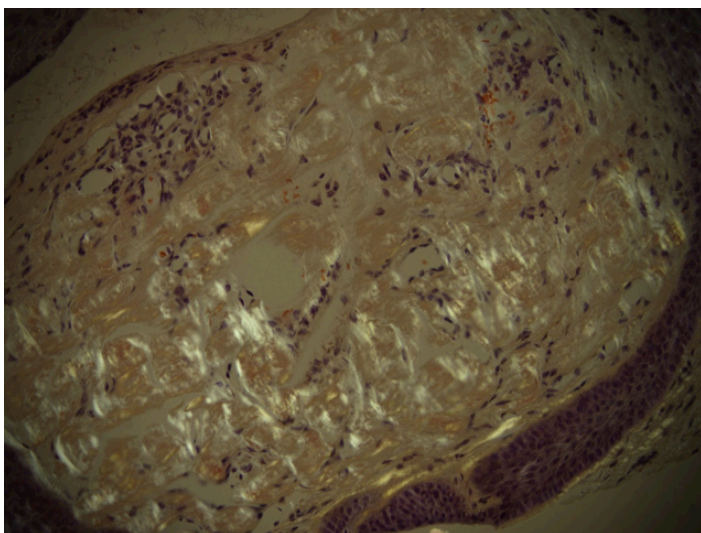


Figure 3. Congo red stain was performed, and the material demonstrated apple green birefringence with polarized light microscopy, 200%.

in patients with atopic dermatitis, sarcoidosis, and psoriatic patients receiving psoralen plus UVA [6]. An estimated 10% cases of PLCA appear to be familial with autosomal dominant inheritance [7].

Lichen amyloidosis is commonly seen on the anterior aspect of shins and extensor surfaces of forearms [8]. Clinically, the lesions may be pruritic or asymptomatic [8]. The differential diagnosis of lichen amyloidosis includes lichen simplex chronicus, post-inflammatory hyperpigmentation, prurigo nodularis, and colloid milium [3]. To our knowledge there have been only rare case reports of lichen amyloidosis with facial involvement. One was of a patient with generalized lichen amyloidosis on the face, neck, shoulders, and back for three years [9]. Another was of a Chinese patient with lichen amyloidosis on the upper lip and nasolabial folds [10].

Although the diagnosis of PLCA is often made clinically, histopathologic evaluation can be confirmatory. Histopathologic changes include hyperkeratosis, hypergranulosis, basal layer vacuolar degeneration, and keratinocytes in various stages of degeneration found throughout the epidermis [8]. Elongated rete ridges, some of which assume a claw-shape configuration around dermal papillae, are also present [8]. The dermal papillae contain confluent, homogenous eosinophilic masses [8].

Many therapeutic modalities are available but none

are considered to be curative or satisfactory [6]. These include: potent topical corticosteroids, topical calcineurin inhibitors, topical dimethyl sulfoxide (DMSO), PUVA, UVB phototherapy, systemic retinoids, cyclosporine, electrodesiccation, dermabrasion, pulsed dye laser, and YAG laser [6].

Conclusion

Lichen amyloidosis should be considered in a differential diagnosis of discrete, firm, closely-set, dome-shaped brownish papules especially in patients of Asian or South American origin. Suspicion for lichen amyloidosis should not be excluded based on facial involvement because it can infrequently present on the face. Familiarity with clinical and histopathologic features of lichen amyloidosis can facilitate definitive diagnosis.

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