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Case Report

Extragenital Lichen Sclerosis: Defined to Hairline, Trichoscopic and Histopathologic Description

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Abstract

Lichen sclerosis is an idiopathic inflammatory disease. It is a disease that most commonly affects the anogenital area with extragenital disease in 15-20% of patients. We report a case of a 25-year-old man who presented hypopigmented lesions that are confirmed to be hairline. Trichoscopy and histopathology of the affected area confirm the diagnosis of extragenital lichen sclerosis.

Keywords: Lichen; Hypopigmented lesions; Sclerosis; Hairline; Trichoscopy.

Introduction

Lichen sclerosis is an idiopathic inflammatory disease first described by Hallopeau in 1887 [1]. The etiology of lichen sclerosis is a mystery, with evidence suggestive of autoimmune activity with association with HLA class II antigens and comorbidity with autoimmune diseases, especially thyroid disease, in patients with genetic background.

It is a disease that most commonly affects the anogenital area with extragenital disease in 15-20% of patients [2-4]. Extragenital disease is characterized clinically by white, polygonal papules that coalesce into plaques, located on the neck and shoulders and are usually asymptomatic [2]. The rate of isolated extragenital lichen sclerosis (ELS) is 2.5% [4].

Case Report

A 25-year-old male, skin type 5, presents to the outpatient dermatology clinic with a complaint of asymptomatic hypopigmentation along the hairlines for three weeks only. Not proceeded by itching or burning. The clinical examination showed a well-demarcated hypopigmented linear patch on the border of the frontal hairline with a solitary polygonal porcline white macule (Figures 1 and 2). Oral and genital mucosae were normal. Wood's light examination showed no accentuation.

Figure 1: Well demarcated linear hypopigmented patch along the left frontal hairline.

Figure 2: Well-demarcated linear hypopigmented patch along the right frontal hairline.

Dermoscopy

Showed structureless whitish, homogenous areas, with erythematous halo and peripilar white halo. (Figures 3 and 4).

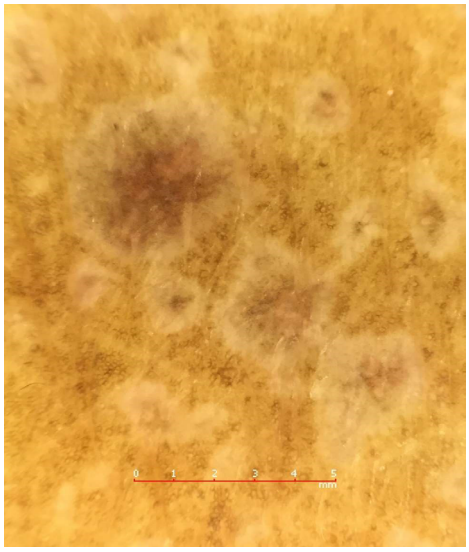

Figure 3: Dermoscopy showing white structureless areas and telangiectasia.

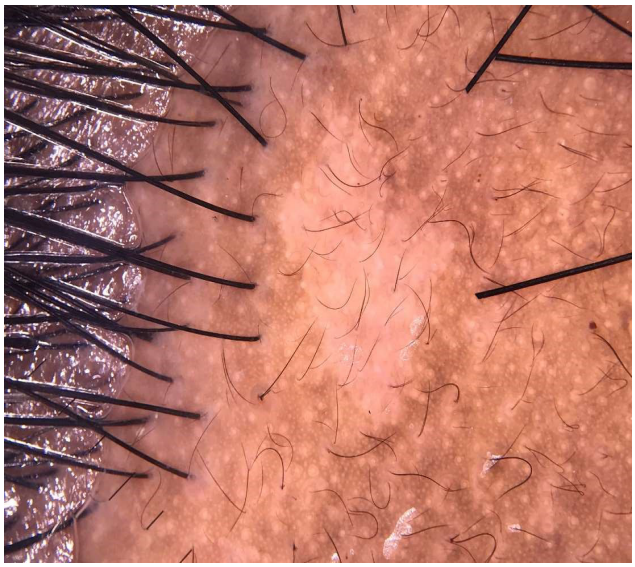

Figure 4: Dermoscopy showing the white structureless area. Notice the reservation of vellus hair.

Complete blood count, thyroid function test, testosterone and antinuclear antibody (ANA) were within normal limits

Histopathology

A 3 mm punch biopsy was performed and demonstrated a hyperkeratosis, irregular acanthosis, mild spongiosis, and focal vacuolar degeneration of the basal cell layer (figure 3). The dermis showed focal sclerosis with Occasional melanophages and very few perivascular and periadnexal lymphohistiocytic infiltrate (figure 5).

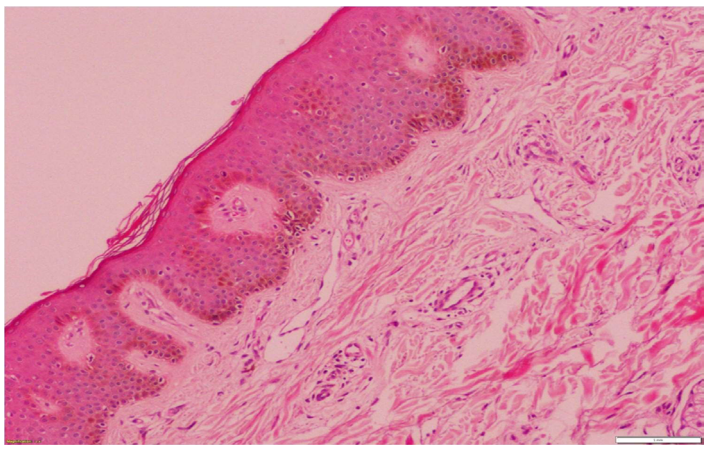

Figure 5: Hyperkeratosis, acanthosis, mild vacuolar interface changes, and minimal perivascular lymphocytic infiltrate. Note melanocytes on the basal epidermal layer.

The diagnosis of extra-genital lichen was retained. The patient was treated with topical clobetasole 0.05% with great response on 2 months follow-up.

Discussion

Lichen sclerosis is a chronic relapsing skin condition characterized by inflammation followed by scarring and atrophy. The typical presentation of extragenital lichen sclerosis is polygonal bluish-white papules that coalesce over time into erythematous plaques with an increasingly atrophic and wrinkled appearance. Advanced features include follicular plugging Unusual distribution of extragenital lesions have been reported, blaschkolineal and linear clinical forms [5].

Few cases of extragenital lichen sclerosis have been present with Scalp involvement, to our knowledge this is the first case present with restricted hairline involvement.

Disease	Clinical	Trichoscopy	Pathology
Vitiligo	asymptomatic depigmented macules and patches, Leukotrichia are often present in lesional skin	-perifollicular depigmentation -leukotrichia -erythema and telangiectasia	complete loss of melanin pigment in the epidermis and absence of melanocytes, with occasional lymphocytes at the advancing border of the lesions
Frontal fibrosing alopecia	frontotemporal loss of both terminal and vellus hairs. Hair loss occurs in a band-like distribution	-Loss of follicular openings -Peripilar erythema and cast -Loss of villous hairs	perivascular and Perifollicular Lymphocytic infiltrate concentrated around the hair bulge area, dermal fibrosis, loss of sebaceous glands and follicular dropout

Table 1: The differential diagnoses are summarized in the following table.

The typical dermoscopic features of extragenital lichen sclerosis are bright white/white-yellowish patches, and yellowish-white keratotic follicular plugs [6]. Not affecting the hair color.

Treatment of extragenital lichen sclerosis is typically with ultra-potent topical corticosteroids or even intralesional. There are many other treatment options with variable response including retinoids and, phototherapy [7], estrogen, and topical tacrolimus.

In our case, excellent improvement was noted with topical clobetasol ointment, used once daily for 4 months then every other day for another 4 months, and then twice weekly on the remaining lesion.

On 12-month follow up the lesions totally disappear apart from 0.5*0.2 scar on frontal scalp. No genital or skin lesions are noted.

The natural history of extragenital lichen sclerosis has been poorly characterized. Periodic follow-up is recommended given the unpredictable periods of exacerbation and stability known for this disease.

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