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Title
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Journal
Dermatology Online Journal, 27(4)

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Publication Date
2021

DOI
10.5070/D3274053159

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Linear lichen planus pigmentosus of the face with histological findings of lichen planopilaris—an uncommon variant of lichen planus

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Abstract

Lichen planus pigmentosus and lichen planopilaris are two clinically and histologically distinct forms of lichen planus. Lichen planus pigmentosus presents with sudden onset hyperpigmented macules and patches, predominantly in darker skin phototypes. On the other hand, lichen planopilaris is a scarring follicular variant of lichen planus that presents with progressive, permanent patches of alopecia. It is not uncommon for different variants of lichen planus to clinically coexist with each other. However, to our knowledge, there has been no previous reporting of linear lichen planus pigmentosus of the face with histological features of lichen planopilaris. We herein present a hybrid case of these two entities.

Keywords: face, lichen planus, linear, pigmentosus, planopilaris

Introduction

Lichen planus pigmentosus (LPPig) is a subtype of lichen planus (LP) with several described configurations. Linear LPPig has rarely been described on the face. Lichen planopilaris is a scarring follicular variant of LP with three recognized presentations: classic, frontal fibrosing alopecia (FFA), and Graham-Little-Piccardi-Lasseur syndrome. Histological features of LPPig demonstrate an interface dermatitis of the basement membrane with pigment incontinence and melanophages in the papillary dermis. On the other hand, lichen planopilaris presents with lymphocytic destruction of the follicular epithelium. A clinical link between LPPig, FFA, and less commonly classic lichen planopilaris has been reported in darker skin phototypes [1-5]. To date, no associations have been described between linear LPPig of the face with histologic findings of lichen planopilaris. For this hybrid entity we propose an encompassing term, linear lichen planopilaris pigmentosus.

Case Synopsis

A healthy 27-year-old man presented for evaluation of hyperpigmented macules and small patches on his face. The lesions first started on the forehead with gradual spreading to the left side of the face over two months. He denied any associated pruritus, tenderness, blistering, or scaling. There was no preceding trauma, no new medications, excessive sun exposure, or any topical application.

Skin examination was notable for linearly arranged brown to violaceous macules and small confluent patches on the left medial forehead extending to the left nasal sidewall, left infraorbital cheek, and left lateral upper cutaneous lip (**Figure 1**). No atrophy, follicular plugging, or perifollicular erythema were noted. There were no oral mucosal lesions, nail dystrophy, or any patches of hair loss on the scalp. There was no loss of facial hair or eyebrow thinning.

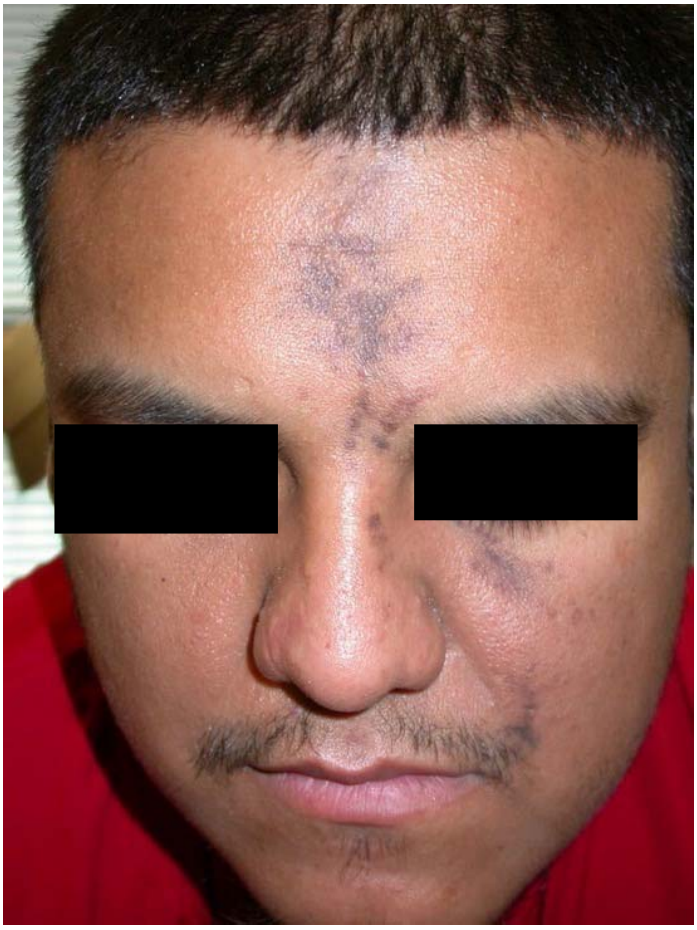

Figure 1. *Linear brown-violaceous macules and small confluent patches on the left medial forehead extending to the left nasal sidewall and left medial cheek.*

A three mm punch biopsy from the forehead was performed. Histologic examination demonstrated a superficial perivascular lymphocytic infiltrate and extensive perifollicular and perieccrine inflammation (**Figure 2A)**. Closer sections revealed a vacuolar and focally lichenoid interface dermatitis of the basement membrane with melanin pigment incontinence in the papillary dermis (**Figure 2B)**. A dense lichenoid infiltrate with destruction of the follicular infundibulum and isthmus were also noted, along with early perifollicular scarring (**Figure 3A, Figure 3B)**. Although the clinical findings were consistent with linear LPPig, the combined histologic features of lichen planopilaris favor an overlap diagnosis between these two entities.

The patient received therapy with hydroxychloroquine 200mg twice a day, topical hydrocortisone 2.5% cream, and diligent photoprotection. After one month of treatment there were no new lesions.

Case Discussion

Lichen planus pigmentosus is a rare acquired variant of LP predominantly affecting phototypes III to VI [6]. The disorder has a slight female predilection and is more common in middle-aged patients of Indian, Latin American, African, and Asian heritage [6-8]. Unlike other LP subtypes, LPPig often spares the scalp, mucosa, and the nails [6-9]. Lichen planus pigmentosus presents with bilateral, symmetric

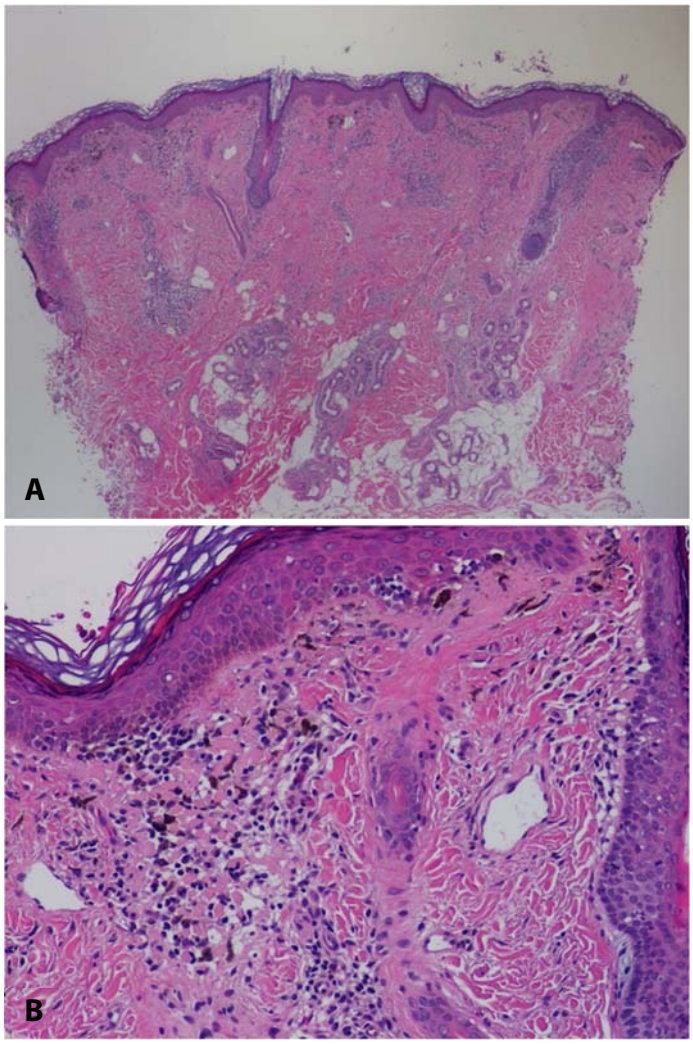

Figure 2. ***A)*** *Lower magnification showing a superficial perivascular infiltrate with perifollicular and perieccrine inflammation. Also noted is paucity of sebaceous glands. H&E, 40×.* ***B)*** *Epidermal atrophy with vacuolar and focally lichenoid interface dermatitis. Melanin pigment incontinence and melanophages are seen in the papillary dermis. H&E, 200×.*

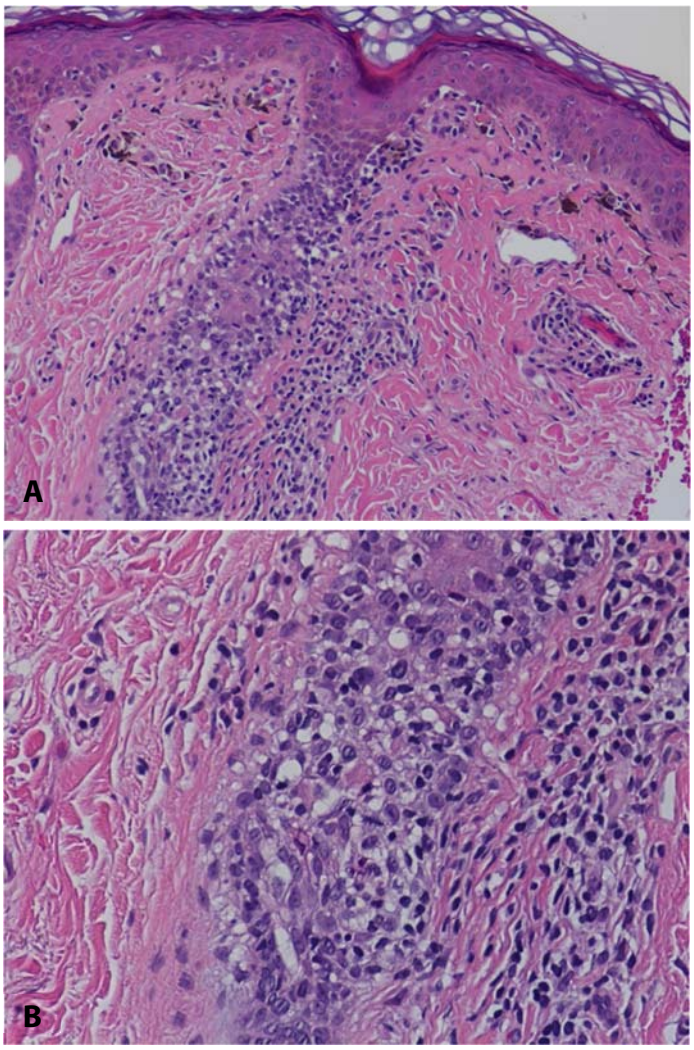

Figure 3. ***A)*** *Dense lichenoid infiltrate centered around the follicular infundibulum and isthmus. H&E, 200×.* ***B)*** *Follicular basal layer inflammation with early fibrosis. H&E, 400×.*

blue-gray macules and coalescing patches that more commonly affect sun-exposed areas of the head and neck [10,11].

Lichen planus pigmentosus can demonstrate significant clinical overlap with erythema dyschromicum perstans (EDP), another acquired pigmentary disorder considered as a principal entity in the differential diagnosis of LPPig [10]. Some dermatologists consider EDP and LPPig analogous to each other, whereas others, based on clinical and histopathological differences, consider them to be distinct [10]. Zaynoun et al. in 2008 attempted to categorize EDP and LPPig within the spectrum of ashy dermatoses [10-12]. This classification includes ashy dermatosis, EDP, and simulators such as LPPig [10-12]. Erythema dyschromicum perstans can be considered synonymous with ashy dermatosis, the latter lacking an erythematous rim more typical of active EDP [10,11].

A more recent global consensus statement from 2018 considers EDP and LPPig as distinct pathologies of acquired macular pigmentation of uncertain etiology [10,11]. Despite the substantial overlap, clinical differences allow for differentiation between the two. Erythema dyschromicum perstans presents with insidious-onset ashy-gray hyperpigmented macules and patches, characteristically larger than 5cm [11]. Unlike LPPig, EDP favors sun-protected areas of the trunk and extremities [10,11]. Additionally, evolving lesions of EDP are characterized by an erythematous and sometimes raised border, which is not seen in LPPig [10,11].

Several morphologies of LPPig have been described, including diffuse, reticular, blotchy, annular, perifollicular, and inverse [6]. These are not mutually exclusive and may coexist in the same patient. Linear LPPig is an uncommon unilateral variant that has been reported in a segmental, zosteriform, or Blaschkoid distribution [8,9,13,14]. The Blaschkoid and segmental configurations more frequently affect the trunk. Linear LPPig of the face is very rare and to the best of our knowledge, only few cases have been described in the literature [9]. The linearity of the lesions suggests in part a predetermined genetic mosaicism [13,14]. It is postulated that the affected areas are populated by a group of cells with distinct immunologic and antigenic profiles. Following exposure to an as-of-yet unknown trigger, a mosaic T cell response likely accounts for the presentation in this variant [9,13,14].

Although the exact etiology of LPPig is yet to be determined, possible precipitators include ultraviolet radiation, hepatitis C infection, and exposure to contact allergens such as nickel, amla oil, mustard oil, and para-phenylenediamine in hair dyes and henna [6-9]. Other reported triggers include photosensitizing fragrances such as bergamot oil, musk ambrette, and sandalwood oil [9]. The immunopathogenesis of LPPig results from cytotoxic T lymphocytes that recognize and subsequently attack the epidermal keratinocytes [6,15]. Histologically, this manifests with degeneration of

the basal layer keratinocytes, pigment incontinence, and melanophages in the papillary dermis [6]. The interface dermatitis in LPPig ranges from a scant vacuolar inflammation to a denser lichenoid infiltrate in earlier lesions [6,10,15,16].

The histopathological findings of LPPig are by themselves not specific. Vacuolar degeneration of the basal layer, pigment incontinence with dermal melanophages, and a superficial perivascular infiltrate can be seen in both LPPig and EDP or ashy dermatosis [16]. Lichen planus pigmentosus tends to demonstrate more epidermal change, including hyperkeratosis and hypergranulosis, greater degrees of perifollicular involvement, and pigmentary incontinence [16]. Similar to early lesions of LPPig, active EDP, which clinically manifests with an erythematous border, displays a more robust lichenoid infiltrate [11,17]. Centrally, the lesions have a more subtle basal layer vacuolar degeneration and a more scant superficial dermal infiltrate [10,17]. In contrast to LPPig, the melanin deposits in EDP tend to be localized deeper in the dermis, accounting clinically for a darker blue-gray hue related to the Tyndall effect [10]. The histopathological differences are subtle and may not always be present, thus the diagnosis of LPPig relies on clinical correlation including distribution, color, and border of the lesions [10].

Lichen planus pigmentosus has been described in association with other variants of lichen planus in approximately one-third of the cases [11]. An association of particular interest is that between LPPig and subtypes of lichen planopilaris [1-5]. Lichen planopilaris is a primary lymphocytic cicatricial alopecia where reactive T lymphocytes attack follicular antigens [8,18]. It more commonly affects Caucasian and Indian females on the vertex and parietal scalp [8,18]. Clinical findings reveal perifollicular erythema with scale and scarred patches of alopecia with loss of follicular ostia. Three presentations of lichen planopilaris are recognized: classic, FFA, and Graham-Little-Piccardi-Lisseur syndrome [8,18]. Although classic lichen planopilaris can be multifocal, FFA only affects the frontotemporal scalp [18]. Histologically, in lichen planopilaris and FFA a lichenoid interface dermatitis is seen around the isthmus and infundibulum of new or active lesions [8,18]. With time, fibrosis and scarring replace the follicular basement membrane, sebaceous glands, and root sheaths [8,18].

In 2012, a retrospective review of 24 patients in South Africa reported that FFA was preceded by LLPig by an average of 14 months, suggesting that LPPig could herald the onset of this variant [2]. This postulate has been reinforced by few other subsequent reports of FFA coexisting with LPPig [3-5]. Lichen planus pigmentosus in association with classic lichen planopilaris of the scalp has been reported in one case [1]. However, even histological specimens of LPPig with or without clinical association with FFA have demonstrated a higher incidence of perifollicular inflammation and interface changes of the acrosyringeal epithelium [15,19]. Although further investigations are needed to establish a definitive link and a temporal association, it is reasonable to suspect that LPPig could precede lichen planopilaris or its FFA variant.

Given no clinical findings of lichen planopilaris or FFA, the presence of infundibular basal layer degeneration in our case was unexpected. To our knowledge, there have been no reports describing overlapping linear LPPig with histological lichen planopilaris. The significance of this finding is yet to be determined. However, we propose that patients with LPPig and any of its variants should be carefully evaluated and clinically monitored for evidence of hair loss.

This is important as advanced lichen planopilaris is irreversible. Additionally, both lichen planopilaris and LPPig lead to significant psychological morbidity. There is no cure and management focuses first on halting active inflammation [18,20]. Depending on disease severity, more common treatment modalities for lichen planopilaris include topical and intralesional corticosteroids, oral hydroxychloroquine, and immunomodulating agents such as cyclosporine and mycophenolate mofetil [18]. For LPPig, options to alleviate disease activity include topical corticosteroids, tacrolimus ointment, and systemic hydroxychloroquine [20]. The residual post inflammatory hyperpigmentation can then be addressed with lightening modalities.

Conclusion

We report a clinical case of linear lichen planus pigmentosus of the face with histological features of lichen planopilaris. The patient's biopsy was notable for epidermal and infundibular basal layer degeneration along with early perifollicular fibrosis. These histological findings were in the absence of any clinical evidence of perifollicular erythema, scalp, or facial hair loss. This is likely an uncommon presentation as no other cases have been reported to our knowledge. We propose the term linear lichen planopilaris pigmentosus to describe this hybrid entity.

Potential conflicts of interest

The authors declare no conflicts of interests.

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