Lichen Planus Pigmentosus and Lichen Planopilaris Coexistence

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Abstract

Lichen planus (LP) is a known chronic inflammatory condition affecting skin and mucosal surfaces. It has different variants. Many of these variants can coexist together. Lichen planus pigmentosus (LPPigm) is one of these variants known in the literature to coexist or precede frontal fibrosing alopecia (FFA). However, only a few cases reported, for LPPigm to develop or precede the classical lichen planopilaris (LPP). We reported a case of 48 years old woman who presented with LPPigm and then developed classic LPP with a good response to treatment in her LPPigm.

***Keywords*:** frontal fibrosing alopecia (FFA), lichen planopilaris (LPP), lichen planus pigmentosus (LLPigm).

Introduction

Lichen planus is a common inflammatory condition affecting skin, hair, nails, and mucous membranes.[1] It has multiple clinical variants based on the site involved and the morphology of the lesions.[2] LPPigm and LPP are uncommon variants of LP. LPPigm manifests clinically as greyish blue to brown color macules and patches with a diffuse, reticular, linear, or perifollicular pattern on the exposed parts of the body mainly the face and the neck.[3] LPP; a follicular form of LP, is classified as one of the primary lymphocytic cicatricial alopecia's. It typically affects the vertex and parietal scalp. It presents with white, atrophic, or scattered patches of alopecia associated with perifollicular erythema and scales, keratotic follicular papules, and loss of follicular ostia.[1,4] LPPigm can present concomitantly with other variants of LPP such as frontal fibrosing alopecia (FFA) which has been reported in the literature, but the coexistence of LPPigm and classic LPP like in our case here is extremely rare.[1]To the best of our knowledge there is only few cases reported worldwide with LPPigm association with LPP and no cases reported in Oman.

Case Report

A 48-year-old female presented to dermatology clinic with a few months history of hyperpigmentation on both sides of the face, which involved the whole face later on. She was initially diagnosed as melasma and was put on Kligman's formula then azeliac acid without significant improvement. Lesions were asymptomatic, but the condition was cosmetically concerning her. There was no history of new medications used including contraceptives. She was not pregnant and she denied excessive sun exposure. Examination showed diffused bilateral brown to gray-colored hyperpigmentation only on the face [Figure 1] and the rest of the body was not involved. Skin biopsy was taken and showed; focal basal layer vacuolization with few civatte bodies and focal lymphocytic exocytosis associated with

marked melanin incontinence and peri-vascular lymphohistiocytic infiltrate [Figure 2]. Based on both the clinical and the histological findings, the suggested diagnosis was LPPigm. She was started on topical tacrolimus 0.1% twice daily and tranexamic acid 250 mg twice daily added after a few weeks along with strict sun avoidance and sunblock use for 6 months with mild improvement. Then acetritin 10 mg was added which was increased to 25mg after 3 months. She showed good response over time in term of the pigmentation. After 1 year, she experienced excessive hair shedding from her frontal scalp, which then, involved her lateral eyebrows with no other hairy areas affected. Examination showed a 6 cm ill-defined patch of reduced hair density in mid- frontal scalp with maintenance of the frontal hairline, positive hair pull test, and semi complete loss of lateral portions of both eyebrows (figure 3). Add to the above, skin-colored small honeycomb-like papules were seen on the affected eyebrows, on the areas lateral, and above them on both sides [Figure 3]. A dermoscopic examination of the scalp showed a honeycomb pigment pattern with a perifollicular white halo and scaling. Two punch biopsies were performed. The first one was taken from the affected area in the frontal scalp, which showed epidermal orthokeratosis, focal lymphocytic exocytosis, and basal layer hyperpigmentation. Dermis showed superficial and deep perivascular lymphohistiocytic infiltrate with characteristic perifollicular lichenoid infiltrate with interface changes, Civatte bodies and fibrosis with no mucin [Figure 4]. The other one was taken from the described honeycomb-like skin colored papules and showed: focal interface dermatitis in follicular epithelium, moderate superficial perivascular and interstitial lymphohistiocytic infiltrate with few plasma cells and pigment incontinence. The patient was diagnosed as a case of LPPigm and LPP. We started her on minoxidil 5% and betamethasone solution. The patient showed some hair regrowth with new baby hairs but the condition was stabilized and small new patches appeared requiring more intervention. We initiated hydroxychloroquine and now she is on it with no further progression.

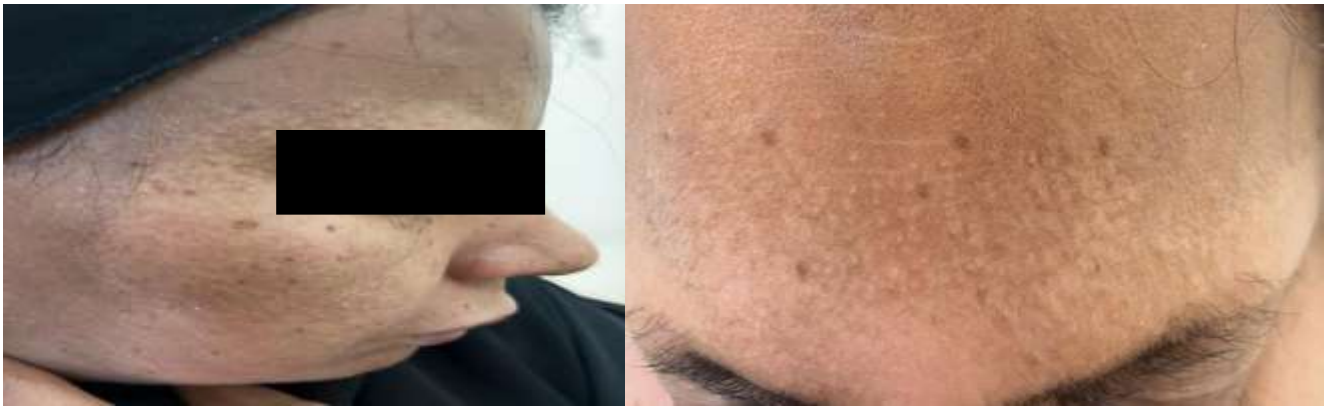

Figure 1: diffused blue to gray colored hyperpigmentation on bilateral temple and forehead

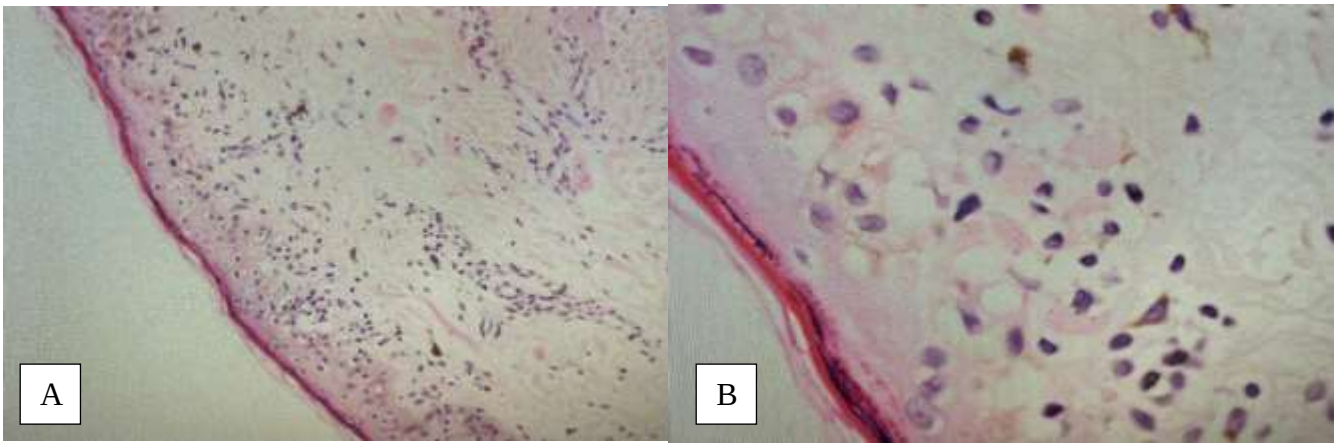

Figure 2: Histopathology of face pigmentation shows:: focal layer vacuolization with few civatte body and focal lymphohistocytic exocytosis associated with melanin incontinence and peri-vascular-histocytic infiltrate

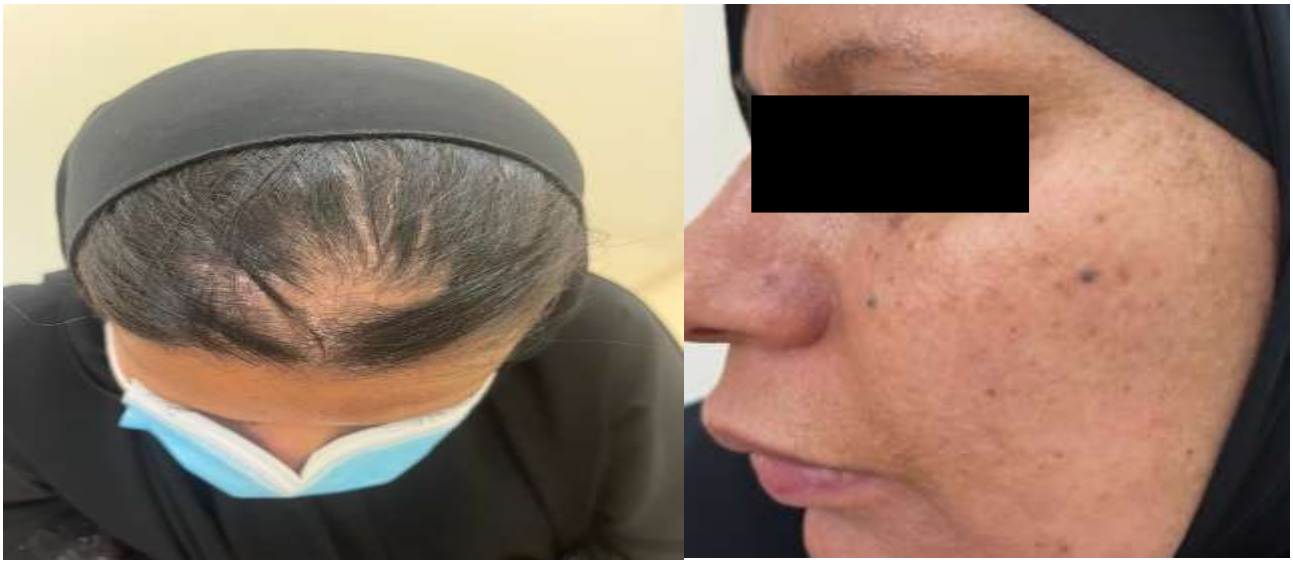

Figure 3: well demarcated patch of reduced hair density on mid-scalp with maintenance of frontal hairline and loss of lateral eyebrow hair and the skin colored small honeycomb like papules lateral and above the affected eyebrow.

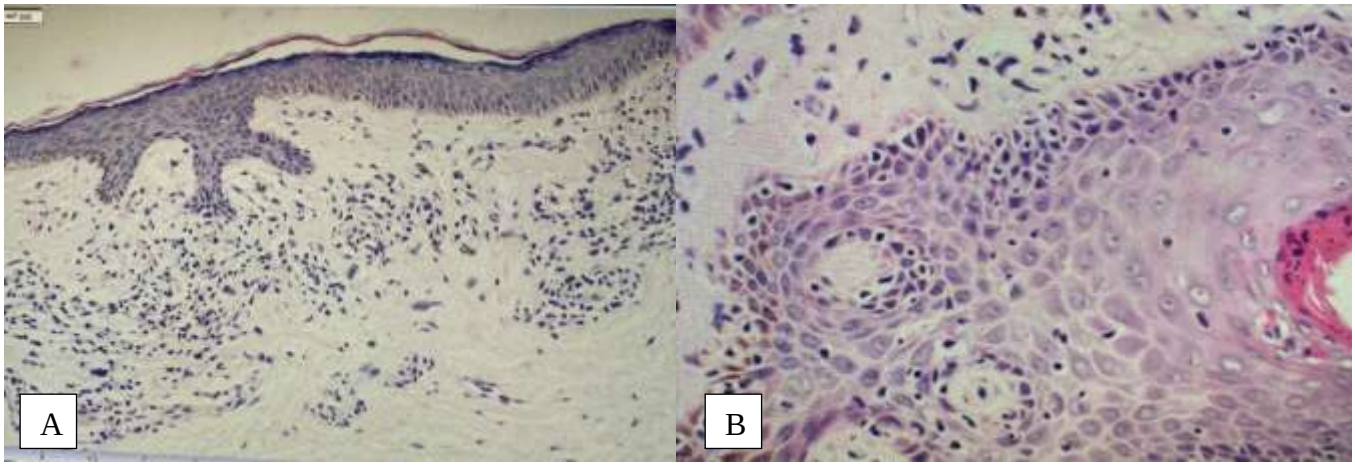

Figure 4: Basal layer hyperpigmentation. Dermis showed, superficial and deep perivascular lymphohistiocytic infiltrate with perifollicular lichenoid infiltrate with interface changes.

Discussion

LPPigm is an uncommon variant of LP that affects patients of skin types III to V, mainly type IV.[5] It mostly affects females, with a peak age of onset between 30-39 years.[1] It presents with an insidious onset of of ill-defined brown to gray macules that develop over sun-exposed areas most commonly the face and the neck, without any features of inflammation or preceding rashes. It can progress into confluent hyperpigmented brown patches. Typically, the disease is asymptomatic although occasionally can have mild pruritus.[1,6] The common pigmentation patterns are diffuse, reticular, blotchy, and perifollicular.[7] Our patient had a typical clinical presentation of diffuse LPPigm but her age was higher than the average at onset. Histopathologically, the classical findings show hyperkeratosis, thinning and basal cell vacuolar change of the epidermis, perivascular and/or perifollicular infiltrate, melanophages, and a band-like infiltrate in the upper dermis, which are the same in our patient. Erythema dyschromicum perstans "ashy dermatosis"

share the same findings in its well-established patches mainly interface dermatitis and pigmentary incontinence but clinically no clues toward the latter condition.[8]

Recently the disease gained a lot of interest and even its dermoscopic features were described. These include pseudo-network patterns, rhomboids with asymmetric pigmented follicular openings, a dotted pattern, and loss of facial vellus hair.[9]

LPP is a form of primary scarring alopecia that is characterized by inflammation and fibrosis of the follicular unit causing irreversible hair loss if untreated.[10] It mainly affects middle-aged Caucasian and Indian populations with slight female predominance (1.8:1.3). Clinically, it presents with white, atrophic, or scarred patches of alopecia on the vertex and parietal scalp occasionally associated with keratotic follicular papules and positive hair pull test. Patients can experience itching, burning, scaling, tenderness, and increased hair-shedding.[1]

LPP is classified into three different variants: classic LPP, FFA, and Graham-Little-Piccardi-Lassueur syndrome. FFA is also another scarring type of alopecia that is present with band-like loss of hair mostly in the frontotemporal area of the scalp that may later on affect eyebrows, axillae, and even the pubic area. For the purpose of differentiation between this disease and LPP, updated diagnostic criteria were suggested (Table 1).[11] The last variant of LPP, the one that we will not focus on in this report is Graham-Little-Piccardi-Lassueur syndrome. It is considered the rarest form of LPP. It is characterized by 3 features; scalp scarring patchy hair loss, nonscarring hair loss from axillae and groin, and hyperkeratotic follicular papules on trunk and extremities that usually proceed the previous two features.[12,13]

Table 1: Updated diagnostic criteria for frontal fibrosing alopecia.

Major
1.Cicatricial alopecia of the frontal, temporal, or frontotemporal scalp on examination, in the absence of follicular keratotic papules on the body
2.Diffuse bilateral eyebrow alopecia
Minor
1.Typical trichoscopic features: perifollicular erythema, follicular hyperkeratosis, or both
2.Histopathologic features of cicatricial alopecia in the pattern of FFA and LPP on biopsy
3.Involvement (hair loss or perifollicular erythema) of additional FFA sites: occipital area, facial hair, sideburns, or body hair
4.Noninflammatory facial papules

In the last few years, trichoscopy has gained more interest and with time, LPP and FFA features were described. Again, both conditions share the same features that were initially described in LPP which are, perifollicular erythema, perifollicular desquamation, and loss of follicular orifices.[1] In FFA, even their prevalence in patients was mentioned as follows; tube-like perifollicular scaling was seen in 89% of the cases, perifollicular erythema in 77% and the lonely hairs in 67.9%.[14]

In histopathology, LPP is characterized by perifollicular lymphohistiocytic infiltrate, sometimes with a lichenoid pattern that is more prominent in the upper portion (isthmus and infundibulum regions), vacuolar degeneration of basal cells, necrotic keratinocytes. Over time, there is a reduction and loss of sebaceous glands and the destruction of the entire hair follicle, which is replaced by connective tissue.[15]

The same histological findings are present in FFA with less inflammatory infiltrate and more apoptotic cells.[16-18]

Although it is one disease with different variants and the same dermoscopic and histologic features, some of these variants commonly coexist together in the same patient. The coexistence of LPPigm and FFA is well established in the literature compared to the coexistence of classic LPPigm and LPP, which is extremely rare. We only found a few cases reported in the literature.[1,4,7] Al Marek et al reported a postmenopausal Fitzpatrick V female who presented with dark pigmentation on the sides of her neck and was diagnosed as LPPigm. The pigmentation was preceded by scarring hair loss 20 years back, affecting her vertex scalp, later on, was diagnosed as LPP.[1] Cobos et al. reported a

premenopausal African American woman who presented with dark pigmentation on her arms, neck, and face that was diagnosed as LPPigm. The pigmentation was preceded by LPP affecting her frontal scalp, a few portions of her occipital scalp, and eyebrow for few years.[4] Our patient and the reported ones were all females. The lag time between LPPigm and LPP ranges from one to twenty years. Our patient presented initially with LPPigm followed LPP unlike the other two reported patients who showed the opposite. Chen et al reported a case series of six patients with follicular LPPigm on slate grey colored macules over the neck, upper limbs, and trunk with the LPP noticed in the scalp of two of them (one male and one female).[10] Thus, for patients who present with either LPPigm or LPP, it is useful to evaluate them for the other condition periodically.

With the application of updated FFA criteria, our patient can be labeled as a case of FFA but clinically her frontal hairline was preserved and it is known that LPP could also affect eyebrows and rest of body hairs.[11]

As the two conditions, share many clinical descriptions, same dermoscopic and histopathologic features. Along with the lag time that can reach up to 20 years between the two diagnoses; when there is a coexistence, we think that the three variants of LPP could be just different degrees of severity for the same condition with different literature views and descriptions according to the time of the patient presentation. That is why we think that our patient is in transition state between LPP and FFA.

Conclusion

Lichen planus is a chronic inflammatory dermatological disorder with multiple clinical and morphological variants. Many of these variants commonly coexist together and most dermatologists are aware of them. However, the rare coexistence of some variants should always be considered in non-classical cases. With the above, early diagnosis and treatment would help in the avoidance of possible sequelae of most of these scarring and psychologically distressing conditions.

Disclosure

The authors declared no conflicts of interest. Consent was obtained from the patient.

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