

Lichen Planus Pigmentosus and Concomitant Frontal Fibrosing Alopecia in A Filipino Woman: A Case Report

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Abstract

Facial hyperpigmentation and pigmentary lesions are often difficult to distinguish in skin of color. Hence, correlation of clinical, dermoscopy and histopathology is necessary to establish definite diagnosis. Lichen planus pigmentosus is a condition of unknown etiology which presents with ashy pigmentation on the exposed areas. Frontal fibrosing alopecia is an uncommon variant of lichen planopilaris and presents with progressive recession of the fronto-temporal line. To date, coexistence of lichen planus pigmentosus and frontal fibrosing alopecia in Filipinos has never been reported.

Keywords: Lichen Planus Pigmentosus, Frontal fibrosing alopecia, Pigmentary disorders, Dermoscopy

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Received: June 7, 2023

Revision received: July 31, 2023

Accepted after revision: Aug 1, 2023

www.japa-edu.org

Lichen planus is a condition that is found worldwide, with its incidence varying from 0.22% to 1% depending on the geographic location [1]. It can affect various areas, including the skin and mucous membranes such as the oral, vulvovaginal, esophageal, laryngeal, and conjunctival mucosa. Different variants of lichen planus exist, characterized by variations in lesion morphology and site of occurrence. These subtypes include popular (classic), hypertrophic, vesiculo-bullous, actinic, annular, atrophic, linear, follicular, and lichen planus pigmentosus (LPP) [2]. Lichen planus pigmentosus is a pigmentation disorder characterized by the presence of dark-brown to grey macules, primarily appearing in sun-exposed areas. While it predominantly affects women, unlike frontal fibrosing alopecia (FFA), it is more frequently observed in individuals with higher skin phototypes [3]. FFA is a type of primary cicatricial alopecia characterized by lymphocytic infiltration and is considered a variant of lichen planopilaris [4] that is characterized by the gradual recession of the hairline in the frontotemporal area. While it predominantly affects postmenopausal women, there have been reports of its occurrence in premenopausal women as well as in males [5].

Case Presentation

We present the case of a 54-year-old Filipino woman with Fitzpatrick Skin type IV-V, housewife, who sought consultation for hyperpigmented patches on cheeks and extensor surface of upper extremities and one-year history of frontotemporal hair loss. History of hyperpigmentation started two years prior to consultation, when she noted the appearance of few hyperpigmented macules and papules on the extensor surface of the arm, and no topical medications were applied. Lesions increased in size and number coalescing into multiple hyperpigmented patches and plaques accompanied by pruritus, now involving the face and V-neck line. One year later, gradual

hair thinning on the frontotemporal area was noted. No history of chemical hair straighteners uses or illicit use of drugs or contact dermatitis. On examination, the patient had no medical illness and family history was also unremarkable. Upon dermatological examination, there was diffuse slate-gray pigmentation on forehead, cheeks, preauricular, perioral area (Figure 1a) and the neck (Figure 1b). Multiple well-defined irregularly shaped hyperpigmented plaques and flat-topped papules were noted on the extensor surface of upper extremities (Figure 1c). Examination of the scalp revealed focal thinning of hair on the frontotemporal area (Figure 1a).

Dermoscopy of the lesion on the extensor surface of arms showed dots and globules in a “hem-like pattern”, discrete bluish-gray deposits with occasional curvilinear distribution with sparing of follicular openings and accentuation of pigment around the follicular openings (Figure 2a) Dermoscopy of the scalp revealed irregularly distributed pinpoint white dots and absence of follicular ostia (Figure 2b) Skin punch biopsy was done on the face and the sections showed a basket woven stratum corneum with thinning of the epidermis and focal areas of vacuolar alteration of the basal cell layer. Some necrotic keratinocytes were seen. The dermis reveals civatte bodies and a mild superficial perivascular inflammatory infiltrate of lymphocytes (Figure 2c). Histologic examination of the scalp revealed a basket woven stratum corneum, thinning of the epidermis with focal areas of vacuolar alteration in the basal cell layer with necrotic keratinocytes. The dermis revealed fibrosis, numerous-pigment laden macrophages and perivascular and perifollicular inflammatory lymphohistiocytic infiltrate. (Figure 2d) Dermoscopy and histopathology findings were consistent with LPP and FFA respectively.



Fig.1 (a) Diffuse slate-gray pigmentation on forehead, cheeks, pre-auricular, perioral area Fig. (b) Diffuse slate-gray pigmentation on the neck Fig. (c) multiple well-defined irregularly shaped hyperpigmented plaques and flat-topped papules were noted on extensor surface of upper extremities and posterior surface of the thigh

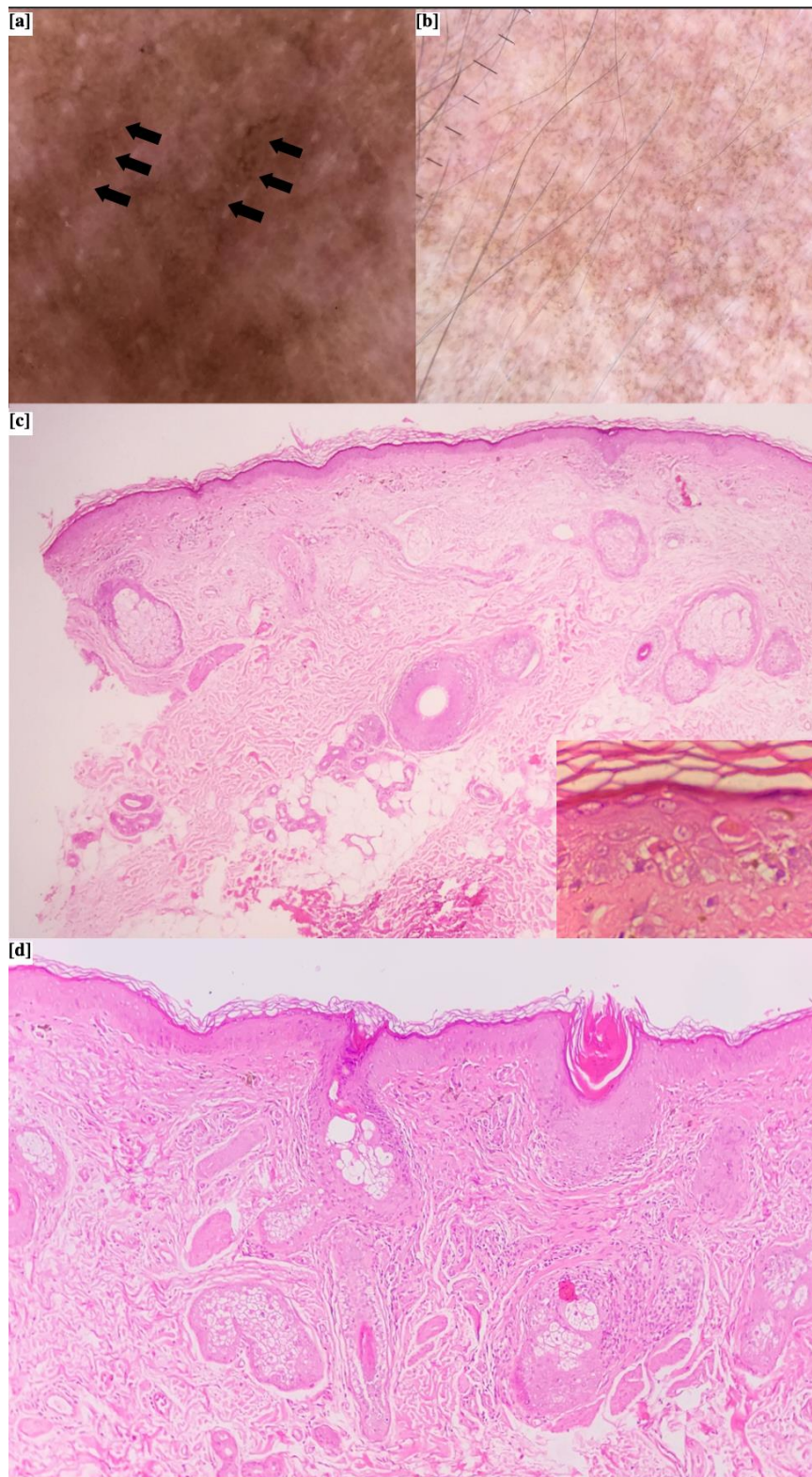


Fig. 2 (a) Dermoscopy of the lesion on the extensor surface of arms showed dots and globules in a “hem-like pattern” (black arrows) (b) Dermoscopy of the scalp revealed irregularly distributed pinpoint white dots and absence of follicular ostia. (c) Histopathological examination (HPE) of the skin from the face showed thinning of the epidermis and focal areas of basal cell layer vacuolar alteration. The dermis reveals Civatte bodies and a mild superficial perivascular inflammatory infiltrate of lymphocytes. (d) HPE of the scalp revealed thinning of the epidermis with focal areas of basal cell layer vacuolar alteration with necrotic keratinocytes. The dermis revealed fibrosis, numerous-pigment laden macrophages and perivascular and perifollicular inflammatory lymphohistiocytic infiltrate (a,b Dermlite DL2x10 ; c, H&E, 10x; inset, 40x; d, H&E, 20x).

Discussion

The uniqueness of our case report lies in the rare coexistence of LPP and FFA in a 54-year-old Filipino woman with Fitzpatrick Skin type IV-V. LPP and FFA are two distinct clinical variants that fall within the broader spectrum of lichen planus [6]. The exact cause of the LPP is uncertain, and it is thought that immunological processes and exposure to certain chemical and physical agents, including sunlight, mustard oil, henna hair dye, and amla oil, may contribute significantly to the development of the condition [7].

Dots and globules in a “hem-like pattern”, discrete bluish-gray deposits with occasional curvilinear distribution with sparing of follicular openings and accentuation of pigment around the follicular openings were the observed dermoscopic findings. It is difficult to differentiate using dermoscopy alone with its differential diagnosis such as ashy dermatosis and erythema dyschromicum perstans. In the global consensus statement by Kumarasinghe et al8, ashy dermatosis, erythema dyschromicum perstans, and LPP are in the spectrum of the condition called acquired macular pigmentation of uncertain etiology (MPUE). There should also be an erythematous border to definitely label a condition as erythema dyschromicum perstans which is not present in our patient. In addition, the consensus also stated that if there is any history of pruritus and clinical features of papules and plaques associated with pigmented lesions, the condition is unlikely to be ashy dermatosis. In contrast with other conditions, LPP lesions commonly affect the head and neck and exposed and non-sun-exposed areas [8]. The findings of dots and globules in a linear/hem pattern in our dermoscopy were similar to the findings of Devanda et al9. in their case series of 27 patients with LPP. While these dermoscopic findings are not specific to LPP alone, it can be a diagnostic clue in diagnosing this condition.

Skin punch biopsy was done on the face and the sections showed a basket woven stratum corneum with thinning of the epidermis and focal areas of vacuolar alteration of the basal cell layer. Some necrotic keratinocytes were seen. The dermis reveals civatte bodies, melanophages and a mild superficial perivascular inflammatory infiltrate of lymphocytes. (Figure 2d). Based on the consensus by Kumarasinghe et al8, histopathology of ashy dermatosis, erythema dyschromicum perstans and LPP may appear similar during some periods of the disease and that the melanophages in the dermis causes the ashy pigmentation in ashy dermatosis, erythema dyschromicum perstans, and LPP. Our findings were similar to that of the frequent histological findings of LPP described in the literature including perifollicular hyperkeratosis, atrophy with hydropic or vacuolar degeneration of the basal layer of the epidermis, and sparse lymphohistiocytic or lichenoid infiltrates along the dermis, along with pigmentary incontinence and the presence of melanophages [10].

Dermoscopy of the scalp in our patient revealed irregularly distributed pinpoint white dots which resemble empty follicles and absence of follicular ostia. It is generally accepted that FFA initially presents as a non-scarring condition, and early intervention may help restore the function of damaged hair follicles. Distinguishing this condition from androgenetic alopecia, traction alopecia, or alopecia areata solely based on scalp dermoscopy can be challenging. Based on the algorithm by Shim et al., the hair shaft abnormality should be checked first [11]. Our patient did not present with dermoscopic findings of tapering hairs also known as exclamation hairs, yellow dots and black dots which are diagnostic of alopecia areata. Also, alopecia would present with a small round to oval hairless patch which is not present in our patient hence the group ruled this entity out. Another differential diagnosis would be

traction alopecia. Based on our patient's history, there was no past history of using tight ponytails, bun or braids or use of chemicals or heat on her hair. In addition, dermoscopic findings of traction alopecia would reveal broken hair or black dots which are not present in our patient. The closest differential would be female pattern androgenetic alopecia which can be differentiated clinically as this would present with a "Christmas tree" pattern of diffuse hair loss at the middle of the hairline as compared with FFA which would present with symmetric and band-like hair loss involving the frontal hairline. The findings from the multicenter study conducted by the International Dermoscopy Society, which examined FFA using clinical and dermoscopic parameters, indicated that 93.6% (176 out of 188) of participants showed empty follicles, and 92% (173 out of 188) exhibited a lack of follicular ostia which is similar in our study [12]. It is challenging to differentiate androgenetic alopecia and FFA using a dermoscope due to the similarities in their dermoscopic findings hence performing a scalp biopsy to confirm the diagnosis in patients with pattern hair loss.

The histological examination of the scalp is demonstrated on Figure 2c. Histopathological confirmation is necessary for the diagnosis of FFA. Our study was similar with the findings of Rao et al. where the histopathological findings of their group revealed marked perifollicular fibrosis, lymphocytic inflammation in a lichenoid pattern around the infundibulum, isthmus, and follicle bulge, and a reduction in the number of follicles replaced by fibrous tracts [13].

The treatment of both LPP and FFA remains to be challenging. Managing LPP typically involves the application of topical medications such as steroids, immunomodulators, keratolytics, hydroquinone (with or without retinoic acid), azelaic acid, kojic acid, glycolic acid, vitamin A, and a 10% dimethyl-sulfoxide aqueous solution, among other options. However, the

effectiveness of these treatments can vary significantly [7]. Our patient was prescribed topical steroids and topical calcineurin inhibitors. Patient was then lost to follow-up.

The treatment approach for FFA encompasses various strategies. These include the administration of chloroquine at a dosage of 200mg twice daily for a duration of 6-9 months, or the use of doxycycline as a systemic anti-inflammatory agent. Additionally, potent topical steroids, intralesional steroids (at a concentration of 4-8mg/ml), topical minoxidil, and topical calcineurin inhibitors are commonly employed. Some authors also recommend short courses of systemic steroids as part of the management plan [14]. In a retrospective cohort study by Panchaprateep et al [15], up to 90% of their patients reported improved or stable FFA after receiving antiandrogen (finasteride or dutasteride) or antimalarial along with topical treatment. In patients unresponsive to steroids, antiandrogen and antimalarial, systemic immunosuppressive agents such as cyclosporine and mycophenolate mofetil are also possible treatment options for this condition [15]. Baseline laboratories prior to starting of an anti-malarial and topical minoxidil were advised to the patient but she was eventually lost to follow-up.

In a retrospective descriptive analytical study conducted by de Brito et al. [16], involving 104 patients with FFA associated with LPP, the researchers concluded that LPP could serve as an indicator or warning sign for the development of FFA. Our own findings align with their study, as we also observed the presence of LPP preceded the onset of FFA.

In recent years, there has been a significant rise in the number of reported cases of FFA globally. The coexistence of LPP and FFA indicates that the underlying inflammatory process may have systemic implications beyond the scalp [17]. Furthermore, FFA can have a detrimental effect on patients' quality of life, leading to psychological distress. The

presence of LPP and/or other facial changes in association with FFA may further exacerbate its impact [18].

Conclusion

LPP, a rare variant of Lichen planus, is frequently associated with FFA. Dermoscopy is essential in diagnosing LPP and FFA especially in skin of color. Histopathology remains to be the gold standard in the diagnosis.

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