Lichen planus pigmentosus with frontal fibrosing alopecia: A case report

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ABSTRACT:

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A 62-year-old woman presented with diffuse net-like hyperpigmentation for 1 year. Dermoscopic examination showed a pseudonetwork pattern of dark brown-gray dots which is characteristic feature of lichen planus pigmentosus. There is confirmed diagnosis by histopathological findings. In this patient, the pigmented lesions are predominantly confined to intertriginous and flexural areas called lichen planus pigmentosus inversus (LPPI). She also had a total loss of eyebrows and band-like scarring alopecia in the frontotemporal hairline which was consistent with frontal fibrosing alopecia (FFA). Here we report a patient presented with LPPI coexisting with FFA.

Key words: Lichen planus pigmentosus, lichen planus pigmentosus inversus, frontal fibrosing alopecia

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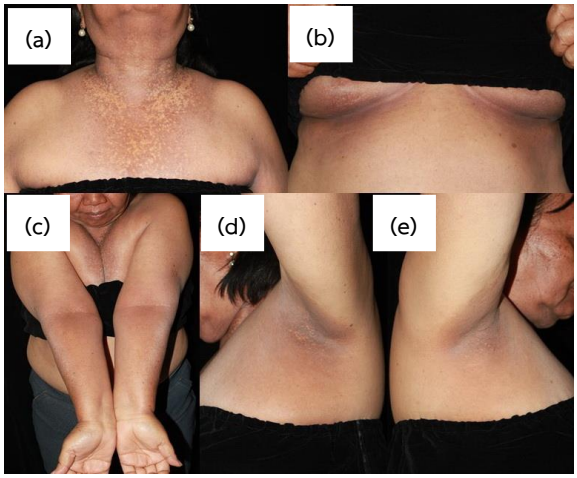
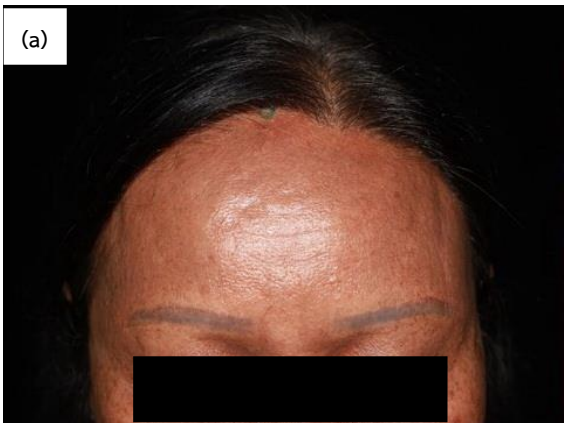


Figure 1 Net-like hyperpigmentation and hypopigmentation on the (a) neck, (b) inframammary folds, (c) antecubital fossa and (d,e) axillae of a patient with lichen planus pigmentosus.

Case report

A 62-year-old woman presented with itchy reticulate hyperpigmented patches on the neck and arms for one year. The pigmented patches later spread to both underarms and inframammary folds. The itching was worse with heavy sweating. She also had a total loss of eyebrows. She also observed that her face had been darkened for more than ten years after total abdominal hysterectomy.

Clinical examination revealed multiple, reticulated, dark brown-gray and hypopigmented macules and patches located symmetrically on the neck, axillae, antecubital fossa and inframammary folds (Figure 1). Generalized hyperpigmentation of the face was also noted. In addition, a band-like scarring alopecia with perifollicular scales was observed in the frontotemporal area (Figure 2a). The eyebrows were also loss with tattoo, while eyelashes and other body hairs were intact. The oral mucosa, palms, soles and nails were normal.



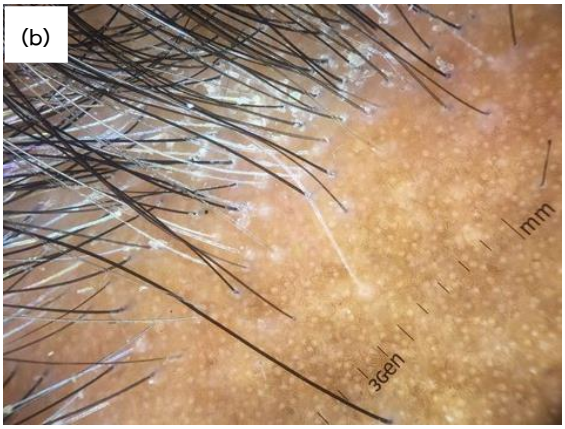


Figure 2 (a) Band-like scarring alopecia with perifollicular scales in the frontotemporal hairline. (b) The non-polarized light dermoscopy showed perifollicular scales, erythema and white halo of patient with frontal fibrosing alopecia (10x).

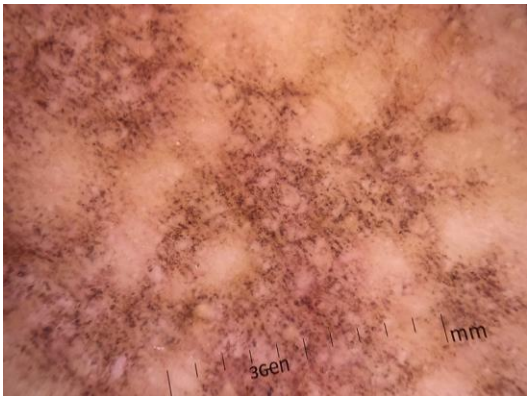


Figure 3 Appearance of lichen planus pigmentosus showed irregular dark brown-gray dots in a pseudonetwork pattern under non-polarized light dermoscopy (40x).

The dermoscopic examination of the reticulated patch showed irregular dark brown-gray dots in a pseudonetwork pattern (Figure 3). The frontal hairline showed perifollicular scales, erythema and white halo (Figure 2b).

Skin biopsy from the hyperpigmented patch demonstrated typical lichen planus pigmentosus (LP pigmentosus) features including epidermal atrophy with hypergranulosis, band-like lymphocytic infiltrates with necrotic keratinocytes, and marked pigment incontinence in the upper dermis (Figure 4a, 4b). Interestingly hypopigmented patch also revealed band-like lymphocytic infiltrates and pigment incontinence in the upper dermis. In addition, histopathology from the frontal hairline showed lichenoid lymphohistiocytic infiltrates and fibrosis at the perifollicular area which was consistent with lichen planopilaris (Figure 4c). Therefore, the clinical correlation helped establish the diagnosis of frontal fibrosing alopecia (FFA).

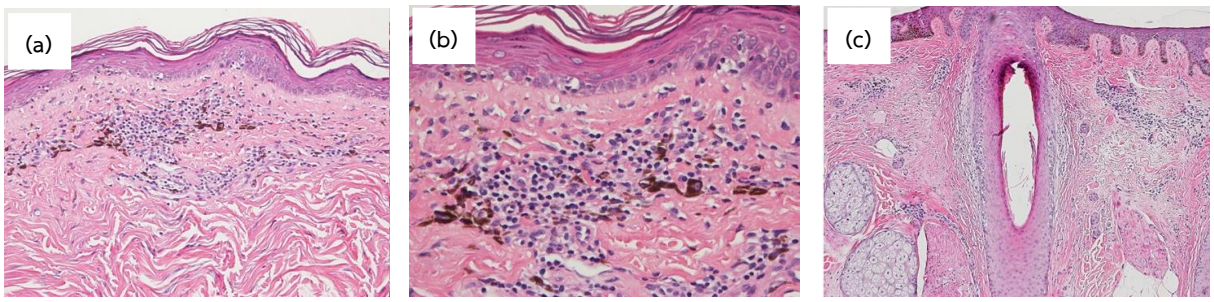


Figure 4 (a,b) Skin biopsy from hyperpigmented patch of the neck showed band-like lymphocytic infiltrates and pigment incontinence in the upper dermis which consistent with lichen planus pigmentosus (H&E, 200x and 400x, respectively). (c) Skin biopsy from hair follicles with scales in frontal hairline showed lichenoid lymphohistiocytic infiltrates and fibrosis at the perifollicular area which consistent with lichen planopilaris (H&E, 100x).

The diagnosis of this patient was coexistence of LP pigmentosus and FFA. Oral corticosteroid (prednisolone 40 mg/day) was initiated, together with 0.1% tacrolimus ointment on the neck and

0.25% desoximetasone lotion on the scalp. Prednisolone was tapered off, then topical medications for long term treatment. The pigmented lesions became lighter and lesser pruritus after two months. Mild improvement of the perifollicular scales on the scalp was also noted.

Discussion

Our patient presented with pruritic dark brown-gray macules and patches on sun-exposed areas, especially the face and the neck. The differential diagnosis includes LP pigmentosus, ashy dermatosis, Riehl's melanosis, drug-induced hyperpigmentation and dyschromic amyloidosis.[1,2] In this patient, LP pigmentosus is most likely due to the dermoscopic characteristics of irregular dark brown-gray dots in a pseudonetwork pattern. In addition, marginal band-like scarring alopecia with trichoscopic characteristics of perifollicular scales in the frontotemporal hairline was consistent with frontal fibrosing alopecia (FFA). A diagnosis required the correlation between comprehensive clinical evaluations and histopathologic findings.[3,4]

Nowadays, dermoscopy is a very useful tool in the diagnosis of various skin and hair disorders. Many hyperpigmented maculopapular diseases can be initially differentiated by dermoscopic characteristics including lichen planus pigmentosus, confluent and reticulated papillomatosis, erythema ab igne, friction melanosis, primary cutaneous amyloidosis and Dowling-Degos disease.[5] The main dermoscopic patterns of LP pigmentosus are pseudonetwork pattern, dotted pattern, speckled blue-gray dots and blue-gray dots arranged in circles.[1] In our patient, dermoscopic examination showed irregular dark brown-gray dots in a pseudonetwork pattern which was consistent with the reticulate hyperpigmentation of LP pigmentosus.[1,2] Furthermore, in frontotemporal hairline, perifollicular scales and white halo were observed. These findings are dermoscopic features of FFA, which are helpful to differentiate from other marginal alopecias.[6]

LP pigmentosus is a pigmented variant of lichen planus presenting clinically as a diffuse, reticulated, blotchy, and linear or perifollicular brownish to gray-black pigmentation mainly on sun-exposed areas, the face and the neck. LP pigmentosus is common in young to middle-aged adults with skin phototypes III-IV. However, there is LP pigmentosus subtype in which pigmented lesions are confined to intertriginous and flexural areas, and skin folds called lichen planus pigmentosus inversus (LPPI).[4] LPPI have been reported in Asians and the axilla is most consistently involved.[7,8] The etiology and pathogenesis of LPPI are still unknown. It is postulated that Koebner phenomenon such as

friction and tight clothing can be the trigger factors. In addition, the hormonal factors may involve, which LPPI tends to occur in menopausal women. Similar to LP pigmentosus, it is evidenced to associate with hepatitis c viral infection.[9,10]

There is a significant increase in the number of coexistence of LP pigmentosus and FFA reports. The two diseases share similar underlying mechanisms of lymphocyte and Langerhans cell-mediated conditions. FFA is a primary lymphocytic cicatricial alopecia characterized by progressive hair loss along the frontotemporal hairline and the eyebrows. Like LP pigmentosus, FFA predominantly affects menopausal women. The clinical progression and histopathology suggest that LP pigmentosus and FFA are on the same spectrum of disease. Although FFA occurs mainly in Caucasian women, the coexistence of FFA and LP pigmentosus has been reported mostly in dark-skin patients.[1] Several reports revealed that LP pigmentosus often preceded the onset of FFA by several months to years.[3] In our patient, the precise onset of LP pigmentosus and FFA cannot be precisely identified. However, here we reported the patient presenting with FFA coexisting with LPPI.

Treatment of LP pigmentosus and FFA are challenging. Therapeutic recommendation for LP pigmentosus includes topical and systemic corticosteroids, 0.1% tacrolimus ointment and systemic retinoids. Recently, significant improvement in pruritus and violaceous plaques of LP pigmentosus has been reported after administration of prednisolone (40mg) for one month.[2] Pigment specific lasers, dapsone combined with the oral immunomodulator, topical tacrolimus combined with photoprotection are also effective in some patients.[4] In addition, a treatment of FFA is also difficult. Current treatment options including oral antibiotics, oral antimalarial drugs, topical, intralesional and systemic corticosteroids, 5α reductase inhibitors, and isotretinoin, demonstrate a fair clinical response. However, giving antimalarial drugs to FFA coexisting with LP pigmentosus should be cautious due to the drugs can bind to melanin and might worsen LP pigmentosus lesions.[1]

In conclusion, the association between LP pigmentosus and FFA seems not uncommon. Searching for the evidence of FFA in the patients with LP pigmentosus is strongly, especially in menopausal women.

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