Coexistence of Linear and Inversus Variants of Lichen Planus Pigmentosus: A Rare Occurrence

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Abstract
Lichen planus pigmentosus (LPP) is a rare disorder affecting primarily the people with ethnic skin. It results in hyperpigmentation of sun-exposed sites along with the involvement of flexures. In accordance with clinical patterns, different variants such as diffuse, blotchy, reticulate, and perifollicular have been described. Rare variants such as flexural (inversus), mucosal, linear, and zosteriform have been reported scarcely. Inversus LPP refers to the involvement of major flexures such as inframammary area, axillae, and groins in the absence of involvement of photo-exposed sites. We present an unusual case presenting with coexisting linear and inversus variants of LPP.

Key Words: Inversus lichen planus pigmentosus, lichen planus pigmentosus, linear lichen planus pigmentosus

Introduction
Lichen planus pigmentosus (LPP) is a disorder of hyperpigmentation that usually affects the people with ethnic skin. It usually affects the photo-exposed areas and flexures simultaneously. Apart from the diffuse, blotchy, reticulate, and perifollicular variants, unusual variants such as linear, flexural (inversus), mucosal, and zosteriform have been described in LPP. We describe a case having both inversus and linear variants of LPP.

Case Report
A 55-year-old woman, with skin phototype III presented with a 1-year history of asymptomatic, slate gray-brownish hyperpigmented patches on her inframammary area. She also complained of a linear hyperpigmented patch on her trunk for 8 months. There was no history of itchy, inflammatory lesions before the appearance of hyperpigmented lesions. After the development of inframammary hyperpigmentation, the patient developed herpes zoster affecting left thoracic (T5) dermatome, for which she was treated with valacyclovir 1 g three times a day for 10 days and lesions healed leaving behind hypopigmented atrophic scars. She was otherwise systemically well.

Clinical examination revealed large, ill-defined, slate gray-colored patches of size around 15 cm × 15 cm over the inframammary area [Figure 1a]. A linear streak of bluish-blackish hyperpigmentation following blaschkoid pattern was observed inferior to the left inframammary patch, partly covering the same. The linear streak was noticeably darker [Figure 1a and b] than the inframammary patches. Hypopigmented, circular to serpiginous, slightly atrophic macules of healed herpes zoster were present on left inframammary patch. There was no evidence of oral, nail, scalp, or follicular lichen planus.

She was investigated with punch skin biopsies, which were taken from areas of above-mentioned pigmentary changes. Histopathology demonstrated atrophied epidermis, necrotic keratinocytes, focal basal cell vacuolization, mild perivascular infiltrate, and minimal melanin incontinence from larger patches [Figure 2a] whereas that from linear patch revealed almost normal epidermis, dense melanin incontinence, and perivascular lymphocytic infiltrate [Figure 2b]. Epidermal pigmentation was normal at all sites and band-like infiltrate was not observed.

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A diagnosis of coexisting linear and inversus LPP was made, and the patient was started on tacrolimus 0.1% ointment. She had minimal improvement at a month follow-up.

**Discussion**

LPP is a disorder of hyperpigmentation characterized by the presence of slate gray or brownish macules and patches on photo-exposed sites and flexures. LPP as a terminology has undergone a lot of debate lately, owing to its considerable clinicopathological overlap with erythema dyschromicum perstans and pigmented contact dermatitis. The presence of blaschkoid/linear LPP evaded this confusion in our case. Exact pathomechanism of LPP is elusive; however, characteristic bluish-gray color to these patches is imparted by dermal melanin incontinence resulting from basal cell damage.

Pock et al. described seven patients of LPP inversus affecting only non-sun-exposed intertriginous areas. They hypothesized that rapid hydropic degeneration of epidermal basal layer combined with nonappearance of acanthosis results in the formation of characteristic brownish macules and patches instead of papules and plaques in LPP. They described the presence of melanophages with the absence of active basal cell degeneration as regressive LPP.

Blaschko’s lines represent the paths followed by keratinocytes during embryogenesis, and disorders following these lines are characterized by somatic mosaicism, which reflects a clone of cells harboring a mutation that puts them at risk of developing a certain disease. The presence of linear LPP over LPP inversus might represent type-2 mosaicism, wherein, an individual having a generalized disorder (LPP inversus) presents with more severe manifestation of the same disorder in one dermatome (in this case, linear LPP patch that was much more darker than the LPP inversus).

We observed changes of active basal cell degeneration in LPP inversus patches, whereas changes suggestive of regression were observed in that from linear LPP. Isolated involvement of inframammary areas in the absence of involvement of sun-exposed sites, that is face and neck, rendered a diagnosis of LPP inversus in our case. The rapid development of patches in the absence of pruritic papules and plaques of classical lichen planus, classic distribution, morphology, and histopathology substantiated the diagnosis of coexisting inversus and linear variants of LPP in the index case, both of which are rare themselves, even if present alone.

Although a few reports are present in the literature regarding atypical variants of LPP, the overall incidence is still rare. We report this case for its classical clinical appearance and rarity, with the dual presence of two unusual variants of LPP (flexural/inversus LPP and blaschkoid/linear LPP) together in the same subject.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.
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