

# A unilateral linear pattern of lichen planus pigmentosus

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## Abstract

Lichen planus pigmentosus (LPP) is a rare chronic variant of lichen planus. LPP distribution tends to be symmetrical and is frequently seen on the face, neck, and trunk. Atypical clinical patterns of LPP, including segmental, zosteriform, and linear, were reported in the literature. Herein, we report a rare presentation of LPP in a 16-year-old female with a unilateral linear pattern along the Blascko lines.

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## Introduction

Lichen planus (LP) is a chronic inflammatory disease that involves the skin, hair, nails, and/or mucous membranes.<sup>1</sup> LP is a cell-mediated autoimmune disease with T cells attacking basal keratinocytes with an unknown etiology, that some have linked to drugs and infections.<sup>1</sup> The classic presentation of LP is characterized by pruritic, polygonal, shiny, and purplish papules and plaques.<sup>1</sup> It may also involve mucosal membranes, such as the genitalia, but mainly it involves the oral mucosa.<sup>1</sup> LP has several variants that have been reported in the literature, including hypertrophic, atrophic, follicular, annular, linear, guttate, actinic, bullous, and pigmentosus.<sup>1</sup> Lichen planus pigmentosus (LPP) is a rare chronic variant of LP.<sup>1</sup> It usually affects middle-aged adults with darker skin types, such as Middle Easterners and Southeast Asians.<sup>1</sup> The clinical presentation of LPP is reticulated hyperpigmentation, with dark brown or gray macules/patches, and its distribution tends to be symmetrical and frequently seen on the face, neck, and trunk.<sup>1</sup> Moreover, rarer LPP variants can present in a linear or segmental distribution along Blaschko lines, with only a few cases reported in the literature.<sup>2,3</sup> Herein, we report a rare presentation of LPP in a 16-year-old female with a unilateral linear pattern along the Blascko lines.

## Case Report

An otherwise healthy 16-year-old girl presented with unilateral asymptomatic hyperpigmentation for 8 months. The patient reported no inflammation or erythema preceding the hyperpigmentation. There was no recent history of drug intake or previous trauma. There were no similar complaints in the family. Of note, the parents are not consanguineous; the father is known to have vitiligo, and the mother is known to have oral LP, hypothyroidism, and dyslipidemia. On examination, there were unilateral whorled ill-defined hyperpigmented macules and patches over the left side of the neck, arm, axilla, chest, abdomen, and inner thigh in a blaschkoid distribution associated with nail changes mainly pitting (Figure 1). Histopathology findings showed superficial melanin incontinence with a very mild perivascular lymphocytic infiltrate and no lichenoid interface reaction (Figure 2). Dermoscopy demonstrated pigmentation with brown dots consistent with melanophages (Figure 3). The diagnosis of LPP was established, and treatment with tacrolimus 0.1% ointment BID and hydroquinone 4% was initiated. The patient was stable with no new lesions at 6 months of follow-up.

## Discussion and Conclusions

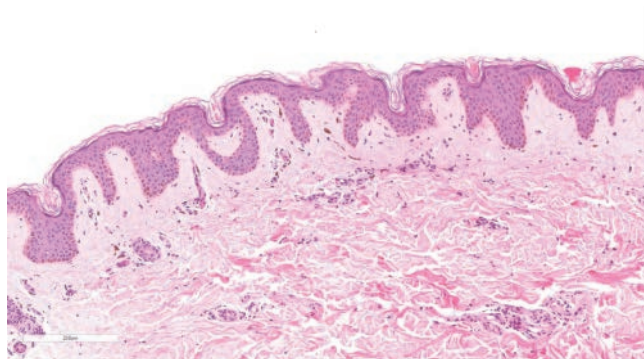
LPP is a chronic idiopathic pigmentary disease of unknown etiology affecting middle-aged adults with racial and ethnic backgrounds.<sup>1</sup> Unlike the violaceous papules and plaques seen classically in LP, LPP patients present with bilateral diffuse symmetrical macu-

lar hyperpigmentation on sun-exposed sites such as the face and neck with no involvement of the scalp, mucosa, and nails, as well as flexural folds such as the axillary, submammary, and inguinal areas.<sup>1</sup> Histologically, LP and LPP are quite similar, but melanin incontinence is more predominant in LPP.<sup>3</sup> Atypical clinical patterns of LPP, including segmental, zosteriform, and linear, were reported in the literature.<sup>2-5</sup> However, only a few cases reported a unilateral linear pattern along the Blascko lines, as seen in our case.<sup>3,4,6</sup> The main clinical characteristics reported in these cases were the presence of hyperpigmented macules and patches mainly localized over the trunk, thigh, pectoral, and submammary areas following the Blascko

lines. Currently, there is no approved treatment for LPP. However, multiple treatment modalities were tried with variable success, such as topical tacrolimus, topical and systemic corticosteroids, high doses of vitamin A, and laser treatments.<sup>7,8</sup> Successful use of topical tacrolimus was seen in a few cases. Moreover, a recent study found the use of oral isotretinoin at low dosages to be beneficial in stabilizing and reducing hyperpigmentation.<sup>8</sup> In conclusion, LPP can present in different atypical patterns. LPP in a blaschkoid distribution is rarely encountered and should be included in the differential diagnosis of unilateral hyperpigmentation along the Blascko lines, particularly in darker skin types.



**Figure 1.** Patient at initial presentation. Examination revealed unilateral whorled, ill-defined hyperpigmented macules and patches over the left side of the neck, arm, axilla, chest, abdomen, and inner thigh in a blaschkoid distribution.



**Figure 2.** Histological examination of a skin punch biopsy shows focal hydropic changes on the basement membrane with melanophage deposition (hematoxylin-eosin stain, magnification  $\times 10$ ).



**Figure 3.** Dermoscopy showing multiple brown dots on the background of a light brown structure-less area.

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