

Lichen planus pigmentosus in a blaschkoid distribution

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Abstract

Lichen planus pigmentosus is a pigmentary disorder of unknown etiology, with diffuse hyperpigmentation of sun-exposed areas, more commonly seen in some ethnic and racial groups. We report an unusual case of lichen planus pigmentosus in a 40-year-old man with Fitzpatrick type III skin that was present in a blaschkoid distribution on the trunk, a distribution that has been rarely reported. This unique presentation of lichen planus pigmentosus may contribute to better understanding of the etiology, as the blaschkoid distribution may reflect underlying cutaneous mosaicism that renders those cells more susceptible to an insult that results in lichen planus pigmentosus. This disorder should be considered in the differential diagnosis of macular hyperpigmentation, especially in those from more commonly affected ethnic and racial groups, even when the distribution is atypical and in the absence of history of sun exposure.

Keywords: lichen planus pigmentosus, lichen planus, blaschkoid

Introduction

Lichen planus pigmentosus is a condition of unknown etiology characterized by the insidious onset of macular hyperpigmentation. The hyperpigmentation is usually bilateral and symmetric on sun-exposed areas and flexural folds in a diffuse pattern [1]. A retrospective study of 124 patients with lichen planus pigmentosus found that nearly 90% had involvement of the face and neck, suggesting that sun exposure plays a role in the pathogenesis [1].

There are reports of atypical patterns of lichen planus pigmentosus including linear, zosteriform, and segmental [2-7]. Lichen planus pigmentosus in a blaschkoid distribution is rarely encountered, with only two previous case reports [8, 9]. We present a 40-year-old man with an unusual manifestation of lichen planus pigmentosus presenting on the trunk in a blaschkoid pattern without history of significant sun exposure.

Case Synopsis

A 40-year-old man with Fitzpatrick type III skin tone presented with hyperpigmented, violaceous macules and patches on his left trunk that had appeared four months previously (**Figure 1**). The hyperpigmented areas were occasionally pruritic, but were otherwise asymptomatic. The hyperpigmentation extended from the left lower back to the left lower abdomen. The remainder of the body, including the head and neck, were spared. There was no history of preceding rash, trauma, application of topical treatments, or herpes zoster infection.

Histopathologic examination of a macule on the left lower abdomen revealed a lichenoid inflammatory infiltrate with prominent pigment incontinence, consistent with a diagnosis of lichen planus pigmentosus (**Figure 2**). The patient was prescribed mometasone 0.1% cream to be applied twice daily for the associated pruritus. He was subsequently lost to follow up.

Case Discussion

Lichen planus pigmentosus is a chronic pigmentary disorder of unknown etiology first described by



Figure 1. Lichen planus pigmentosus with characteristic hyperpigmented macules and patches following lines of Blaschko.

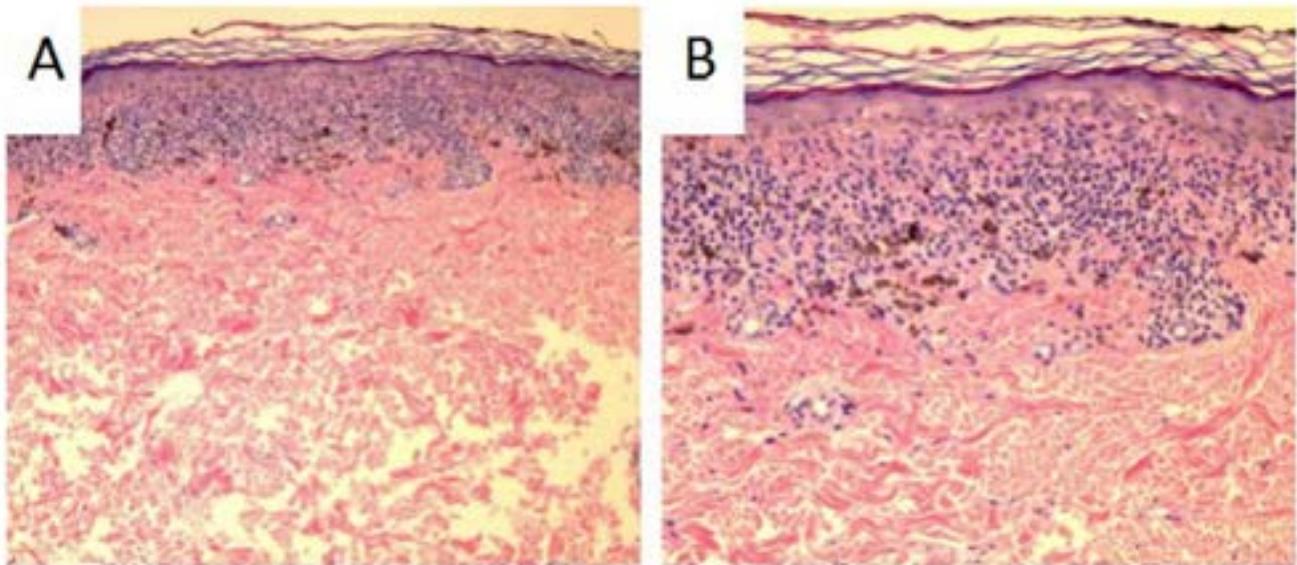


Figure 2. H&E Histopathological examination of a punch biopsy taken from a macular lesion on the left lower abdomen demonstrates a lichenoid inflammatory infiltrate with prominent pigment incontinence, as is typical of lichen planus pigmentosus (A) 10x, (B) 20x.

Bhutani et al. [10]. It was first reported in India, where it is seen fairly frequently, but it also occurs in other racial and ethnic groups [10]. Onset is typically in the third or fourth decade of life and it has a chronic and persistent course. Clinically, it is characterized by macular hyperpigmentation that later forms confluent patches [10, 11]. The color may range from blue-black to slate grey to dark brownish-black [1]. There is no preceding or surrounding erythema [1, 10-12]. About 30% of patients experience pruritus [1]. Although lichen planus pigmentosus has a

variable distribution [1, 10, 11], it is usually bilateral and symmetric on sun-exposed areas and flexural folds, with involvement of the face and neck in almost 90% of cases [1]. It most commonly begins in the preauricular region [1]. Proposed inciting agents include UV exposure, topically applied mustard oil, which contains a potential photosensitizer, and amla oil [1]

Histologically, lichen planus pigmentosus is similar to lichen planus (LP), though it also shows epidermal

atrophy and marked pigment incontinence. Lichen planus pigmentosus is considered to be a variant of LP given these similarities [10, 11], even though lichen planus pigmentosus spares the palms, soles, and nails and has a more chronic course [1]. There is no known effective treatment for lichen planus pigmentosus, although a range of therapeutic options including topical tacrolimus, topical and systemic corticosteroids, high doses of vitamin A, and laser treatments have been attempted with variable and limited efficacy [12, 13]. A recent clinical trial showed some improvement with low-dose oral isotretinoin therapy [13].

Our patient presented with an unusual blaschkoid distribution of hyperpigmentation. His presentation was also unique in the sparing of sun-exposed areas, including the head and neck. A blaschkoid distribution of lichen planus pigmentosus has been described in only two previous cases [8, 9]; in both cases the skin lesions also occurred on the trunk. Previous cases have highlighted the potential overlap between linear, zosteriform, and blaschkoid lichen planus pigmentosus [6]. In contrast to the rarely reported blaschkoid lichen planus pigmentosus, blaschkoid LP has been more commonly reported [14-16]. Pregnancy has been associated with blaschkoid LP [17].

These atypical distribution patterns in both LP and lichen planus pigmentosus may offer insight into the pathogenesis of these separate but related conditions. Although LP is thought to be an immunologically mediated disorder in which CD8+ T cells first recognize an antigen associated with MHC class II on keratinocytes and then lyse the keratinocytes [12], the etiology of lichen planus pigmentosus is less well understood. A blaschkoid distribution of lichen planus pigmentosus may reflect underlying cutaneous antigenic mosaicism, resulting in distinct properties that may render them more susceptible to potential triggers. Because the vast majority of cases of lichen planus pigmentosus involve photo-exposed regions, it is possible that a mutation in cells affected by the condition renders them more likely to become hyperpigmented with UV exposure. In some instances, as in our patient, there is no history of significant sun exposure, other trauma, or topical application to the affected region, highlighting our

limited understanding of the pathogenesis of lichen planus pigmentosus.

Conclusion

Lichen planus pigmentosus should be considered in the differential diagnosis of macular hyperpigmentation, especially in those from more commonly affected ethnic and racial groups, even when the distribution is atypical and in the absence of history of proposed environmental triggers including sun exposure and topical applications.

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