Pigmented Papules and Patches of the Folds: A Case Series of Lichen Planus Pigmentosus Inversus with Review of the Literature

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Abstract

We report nine cases of Lichen Planus Pigmentosus (LPP) inversus in Caucasian patients. They were 5 women and 4 men aged from 29 to 77 years with violaceous hyper pigmented oval or annular macules and patches mostly located in axillary folds but also groin, submammary folds, popliteal and genital regions. Histological examinations showed a band-like lymphocytic infiltrate in the upper dermis and melanophages in the papillary dermis. LPP inversus is a rare variant of LPP appearing in non-sun exposed areas. In contrast to LPP which occurs almost exclusively in dark skinned individuals, a comprehensive review of the literature revealed that about half of the cases reported of LPP inversus affects fair skinned individuals including Caucasian patients.

Keywords: Lichen planus pigmentosus; Hyper pigmented; Lymphocytic infiltrate; Papillary dermis

Introduction

Lichen Planus Pigmentosus (LPP) inversus is a rare variant of LPP occurring in flexural regions with asymptomatic to mildly pruritic violaceous and dark-brownish macules and/or patches [1]. Usually mucosae, scalp and palmoplantar area are not affected [2]. LPP is described almost exclusively in dark skinned individuals. We report here nine cases of LPP inversus in nine Caucasian patients, and an accurate review of the literature about the published cases. Including ours, 48 cases of LPP inversus have been reported, a half of which in Caucasian individuals.

Case Report

Nine Caucasian patients, five women and four men, with asymptomatic to mildly pruritic, sharply demarcated violaceous and hyperpigmented macules and patches located at different folds, mainly axillae and groins, were included in the study. Clinical and histopathological characteristics are shown on (Table 1). The age of onset of our patients range from 29-77 years, with a mean age of 44.7 years and a female:male ratio of 1.25:1. No association with systemic diseases, viral hepatitis and drug exposure was documented in any case. Lesions appeared from 4-18 months before diagnosis. A 4 mm punch biopsy was performed in all patients except patients 3 and 5. Histological changes were similar in all cases with a slight to moderate inflammatory infiltrate in the upper dermis with a band like pattern. Vacular alterations of the basal epidermal layers were observed in all patients, but only in four patients were prominent. Melanophages in the upper dermis were present in all cases. In case no. 8, immunohistochemistry showed the presence of CD8 positive and granzyme positive lymphocytes in close contact with damaged keratinocytes of the basal epidermal layers. Only three patients were treated with topical steroids without any significant improvement except for one that showed a complete resolution. After a follow up of 6-24 months, most patients showed persistent asymptomatic lesions (Figures 1 and 2).

Discussion

LPP is a rare lichenoid dermatitis characterized by the presence of hyper pigmented, dark-brown macules and patches in sun exposed areas occurring mainly in dark skinned individuals such as those from India and the Middle East [3,4]. In 2001, Pock et al. [1] described seven...
Figure 1: Clinical appearance of Lichen planus pigmentosus inversus: multiple hyperpigmented lesions with smooth surface. (A, B) Early erythematous-violaceous lesions at sacral area (A) and left axillary fold (B), (C) Sub-mammary and (D) axillary fold lesions resulting with the appearance of the late phase (brownish).

Figure 2: Histologic aspects of lichen planus pigmentosus inversus: basal vacuolar changes, a band-like infiltrate of lymphocytes in the upper dermis and incontinence of pigment (A). Numerous CD8+ cells in the basal epidermal layers (B), granzyme + cells in close apposition to damaged basal keratinocytes (C).

<table>
<thead>
<tr>
<th>References</th>
<th>Age</th>
<th>Sex</th>
<th>Location</th>
<th>Pruritus</th>
<th>Duration</th>
<th>Histology</th>
<th>Race</th>
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<tr>
<td>Pock et al. 2001</td>
<td>66</td>
<td>M</td>
<td>axillae, back</td>
<td>mild</td>
<td>2 months</td>
<td>A,B</td>
<td>caucasian</td>
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<td></td>
<td>54</td>
<td>F</td>
<td>axillae, shins</td>
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<td>6 months</td>
<td>A,B</td>
<td>caucasian</td>
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<td></td>
<td>68</td>
<td>F</td>
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<td>5 months</td>
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<td>71</td>
<td>F</td>
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<td>60</td>
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<td>M</td>
<td>axillae, groin, wrists</td>
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<td>46</td>
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<td></td>
<td>62</td>
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<td>Bennassar et al. 2009</td>
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<td></td>
<td>59</td>
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<td></td>
<td>54</td>
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<td>no</td>
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<td></td>
<td>25</td>
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<td>Jung et al. 2011</td>
<td>31</td>
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for those patients with more widespread lesions, post-inflammatory hyperpigmentation, figurate erythema, fixed drug eruption and LPP [2]. The clinical and the histological features are usually sufficient in making the diagnosis. A case report described the dermoscopic features of LPP inversus, with diffuse brown patches containing multiple granular gray-brown dots and an overlying scale [6].

LPP inversus belongs to the lichenoid dermatosis which includes lichen planus, LPP, lichenoid drug eruptions, annular lichenoid dermatitis of youth, graft versus host diseases [7]. LPP inversus microscopically resembles late phase lichen planus lesions, with epidermic atrophy, irregular hydropic degeneration of the basal layer and absence of epidermic hyperplasia. There is an exuberant pigmentary incontinence and a lichenoid inflammatory infiltrate with lymphocytes and histiocytes [2]. The pathogenesis appears to be related to a CD8+ T lymphocyte-mediated cytotoxic activity against basal keratinocytes [2] as suggested by immunohistochemical features performed in some patients, including our [3,8,9]. Pock et al. proposed that in LPP inversus the lichenoid reaction occurs within a short period of time with dramatically intensive hydropic degeneration of basal keratinocytes, with no time for compensatory increased proliferation of keratinocytes, as it occurs in typical lichen planus such that the papules transform quickly into brown macules [1,10]. No drugs, infections or systemic co-morbidities have been documented in LPP inversus cases neither in literature nor in our cases. External stimuli, such as friction [Koebner phenomenon], may be a triggering factor [9], but this has been postulated as an explanation for the isolated or exclusive distribution of lesions in intertriginous area [11], and it has not been confirmed.

Treatment of LPP-inversus is not well established. Some cases undergo spontaneous remission within months to years [2]. Topical treatment with high potency corticosteroids [9,12,13] or topical tacrolimus have been used to accelerate the process [12,14], but with minimal improvement. A case report showed only a slight lightening of the lesion after oral deflazacort 45 mg tapered gradually over a period of 2 months [15], and two cases reported improvement of lesions after the discontinuation of wearing tight underclothes [13]. Three of our cases were treated with topical steroids, with no significant improvement except for one that showed a complete resolution, five patients were lost at follow up and the last one had no treatment with spontaneous resolution of pruritus but permanence of pigmentation [16,17].

References

Table 2: Summary of LPP-inversus cases reported in literature.


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