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# Case of Lichen Planus Pigmentosus with Atypical Presentation: A Case Report

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**ABSTRACT**

Lichen planus pigmentosus (LPP) is a chronic pigmentary disorder that shows diffuse or reticulated hyperpigmented, dark brown macules and patches on the sun-exposed areas such as the face, neck and other flexural folds. Clinically, it is different from classical lichen planus because LPP has a longer clinical course and it manifests with dark brown macules. In case of LPP, involvement of the scalp, nail or mucosal area is rare. The histopathological findings of the lesions show an atrophic epidermis, the presence of melanophages and a vacuolar alteration of the basal cell layer with a sparse lymphohistiocytic lichenoid infiltration. Here we report a case of lichen planus pigmentosus over sun protected areas.

**Keywords:** Lichen Planus Pigmentosus; Pigment Incontinence; Sun protected areas.

## INTRODUCTION

Lichen planus pigmentosus (LPP) was first described by Bhutani *et al.*[1] The lesions are small, brown, oval macules with diffuse borders. Later, they merge to form pigmented areas which are grey or brown. The pigmentation may be diffuse, reticulate, blotchy, or perifollicular. The patches are usually symmetrical in distribution but may be found in a segmental, zosteriform, or blaschkoid pattern.[2]

## CASE REPORT

A 21 year old male, waiter by occupation, hailing from Assam presented to the dermatology OPD with complaints of a symptomatic black discoloration mainly over back, neck, arms, thighs since 4 months.

Patient first noticed brown to black a symptomatic lesion over upper left scapular region which gradually spread to involve the entire back, abdomen, bilateral shoulders, lateral aspect of upper arm, buttocks, thighs and lower legs. There was no history of preceding erythema or erythematous borders over the lesions which is generally seen in Erythema Dyschronicum Perstans.

There was history of application of mustard oil all over body everyday in the night since 3 years

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of age. Patient denies any history of long duration of sun exposure or outdoor activities as the lesions were mainly present over the sun protected areas sparing the sun exposed areas and flexural folds.

On examination multiple ill defined diffuse hyper pigmentation tending towards symmetry over back, bilateral shoulders, arms, buttocks, thighs and knees was seen. (Fig. a-f)

**Fig. A:** Absence of lesions over face, neck and hands.

**Fig. B:** Absence of lesions over face, neck and hands.

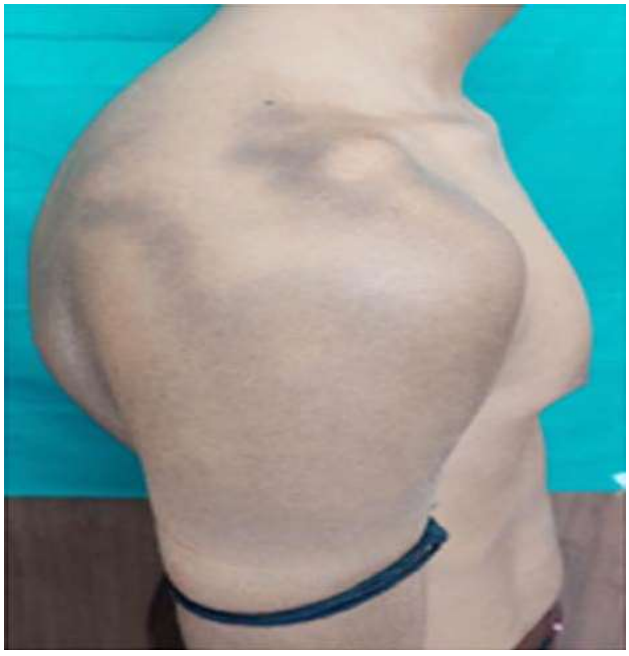


**Fig. C:** Depicts the presence of diffuse hyperpigmentation over arms, abdomen and back

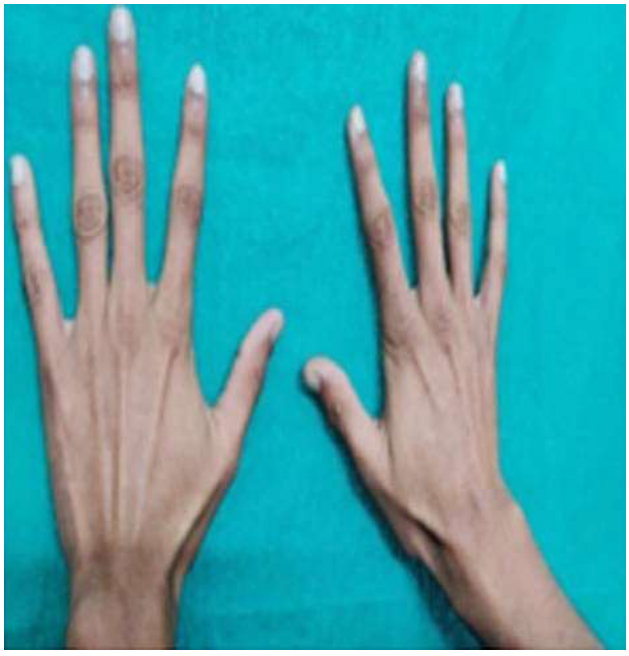


**Fig. D:** Absence of lesions over face, neck and hands.

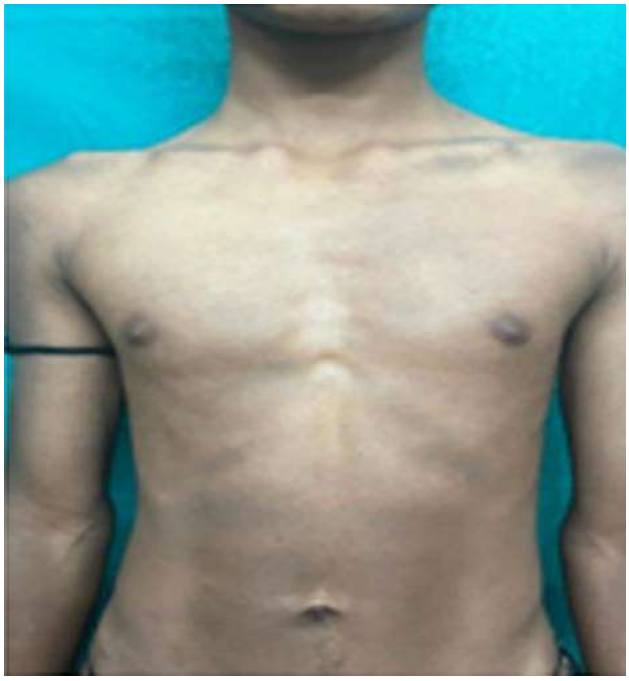


**Fig. E:** Depicts the presence of diffuse hyperpigmentation over arms, abdomen and back

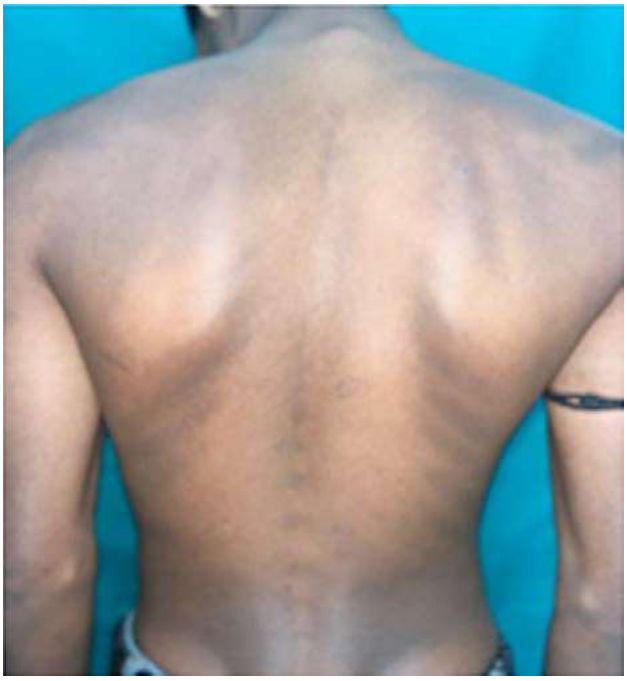
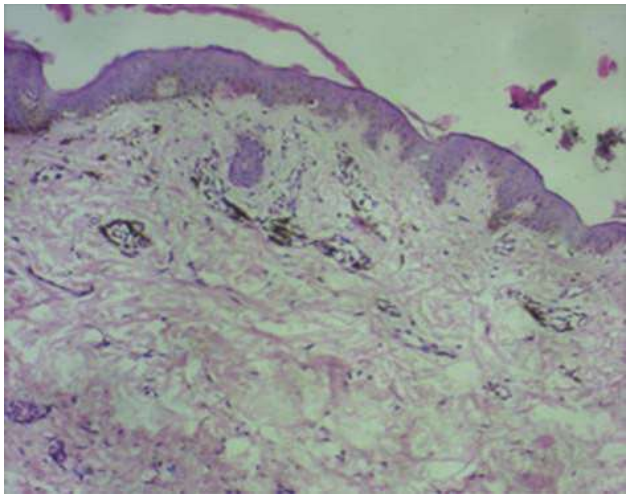


**Fig. F:** Depicts the presence of diffuse hyperpigmentation over arms, abdomen and back



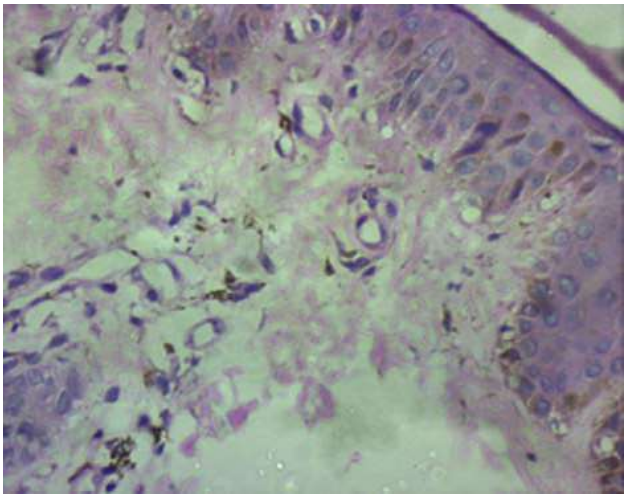


**Fig. G & H:** Histopathology of the skin lesions showed thinning of the epidermis, a superficial band like lymphocytic infiltrate with basal layer vacuolization and pigmentation incontinence in superficial dermis. Fig (g) HE 40x, Fig (h) HE 100x

There was complete sparing of face, intertriginous areas, forearm, legs, hands, feet, nails, oral and genital mucosa.

Histopathological examination shows sparse superficial perivascular lymphocytic infiltrate with numerous melanophages within the papillary dermis. The papillary dermis is slightly thickened and shows delicate fibroplasia and mucin. Overlying epidermis shows focal vacuolar change in the basal layer and infiltration of interface by lymphocytes. The epidermis is flattened at places. The findings were suggestive of Lichen Planus Pigmentosus.

## DISCUSSION

Lichen planus pigmentosus is a chronic inflammatory pigmentary disorder.[1] LPP is essentially a disease of the adult, starting insidiously after the age of 30. It occurs in both sexes but shows a female preponderance. It has been reported to occur predominantly in people with darker skin. Although the etiology is essentially unknown, a number of agents have been reported to act as predisposing factors. The occurrence in exposed areas in many patients has led to the proposition

that sunlight may be a principal etiological agent mustard oil which contains the potential photosensitizer allyl thiocyanate, amla oil where photosensitivity may be caused by fragrances, cosmetic agents such as kumkum, hair dyes, etc. Abnormalities in T lymphocyte functions have also been implicated.[2-6]

Morphological variants described in order of frequency from highest to lowest are diffuse, reticular, blotchy, perifollicular, segmental, zosteriform and linear.[8-11] Another variant is LPP inversus which was described in 2001 by Pock et al., defining it as a variant of LPP limited to intertriginous and flexural regions, sparing sun exposed areas.[12, 13]

Dermoscopy shows with discrete brownish to bluish grey dots, globules, blotches, rods and white lines against a diffuse brownish background. Brown indicates epidermal pigmentation while the grey and blue dots indicate melanin incontinence in papillary and reticular dermis, respectively.[14,15]

The histologic findings of LPP show hyperkeratosis and atrophy of the epidermis with vacuolar degeneration of the basal layer. A perivascular lymphohistiocytic infiltration and pigmentary incontinence in the dermis are also noted.[3,9]

The most common differential diagnosis of LPP include Idiopathic eruptive macular pigmentation, Erythema Dyschromicum Perstans, Riehl's melanosis, Ochronosis, Hori nevus, Fixed drug reaction and Post-inflammatory Hyper pigmentation.[16,17]

**Table 1:** Depicts few differentiating features of the mentioned differential diagnosis.

| Differential Diagnosis | Comment |
|---|---|
| Idiopathic eruptive macular pigmentation | Occurs in younger patients. Brown to grey macules start in the middle area of the trunk and then spread to proximal areas of the limbs. Histopathology reveals pigmentation of the basal layer with mild perivascular inflammatory infiltrate. It is an epidermal hypermelanosis and does not show significant melanophages in the papillary dermis. [16,17,18] |
| Ashy dermatosis/erythema dyschromicum perstans | EDP presents with blue-grey, regularly shaped, hyperpigmented macules compared with dark brown, irregularly shaped, and ill-defined hyperpigmented macules in LPP with erythematous raised active borders. Histopathological evaluation reveals superficial dermal melanin and melanophages in LPP, whereas EDP usually has deep dermal melanophages, giving rise to the characteristic brown–grey color in LPP and the bluish-grey hue in EDP.[16, 19,20] |
| Riehl's melanosis/pigmented contact dermatitis | Characterized by facial hyperpigmentation, most pronounced on the forehead and in the zygomatic and/or temporal region. A correlation with clinical history of contactants and positive patch testing is necessary.[16,17] |
| Ochronosis | History of hydroquinone use at high concentration for a prolonged period, most commonly on the face. Usually does not affect neck and flexural areas.[16] |
| Hori nevus | Bluish-grey or dark brown 2–5 mm macules caused by dermal melanocytes affecting the cheeks, temples, or forehead. Biopsy confirms diagnosis.[16] |
| Fixed drug reaction | Round, initially erythematous macules with history of medication intake prior to onset.[16] |
| Post inflammatory pigmentation | Previous history of dermatosis which leaves pigmentation as it subsides.[16] |

## CONCLUSION

This case exhibited an atypical clinical variety of LPP, including a wide region of grey brown patches occurring on the photo protected sites sparing the sun exposed and intertriginous areas showing characteristic histologic features of Lichen Planus Pigmentosus.

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