

# Hyperpigmented Macules in an Indian boy: is it EDP or LPP? - A Case Report

Sameeksha Chand<sup>1</sup>, Anuj Malhotra<sup>2</sup>

<sup>1</sup>Consultant Dermatologist, Graphic Era Institute of Medical Sciences, Dehradun - 248007, India.

<sup>2</sup>Medical Officer, The Hans Foundation, Jharkhand, India

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## Corresponding Author

Sameeksha Chand,  
Consultant Dermatologist,  
Graphic Era Institute of Medical Sciences,  
Dehradun- 248007,  
India.  
Email:  
sameeksha\_chand0902@yahoo.com

## Abstract

Erythema dyschromicum perstans (EDP) and Lichen planus pigmentosus (LPP) are both inflammatory disorders of pigmentation with unknown etiology and are difficult to differentiate clinically and histologically. There is no consensus on whether both entities are separate or belong to the same spectrum of cutaneous disorders. We present a 13 year old boy with mostly asymptomatic hyperpigmented lesions over body. The lesions were subsequently diagnosed as EDP after detailed examination and histopathological examination.

## Introduction

Erythema dyschromicum perstans (EDP) presents with asymptomatic hyperpigmented macules in sun-protected areas of the body. Similar lesions at the sun exposed sites are seen in lichen planus-pigmentosus (LPP). Flexural involvement is rare in either condition. Clinical and histological differentiation between the two is difficult. It remains unclear whether EDP is a separate entity or an abortive form of LPP.

## Case report

A 13 year old developmentally normal boy with no significant past and family history noticed asymptomatic pigmentation over the body since three months. The lesions started as pea sized redness over lower abdomen, gradually increasing in size and darkness. Subsequently, multiple lesions erupted over the face, neck, trunk, bilateral axillae, inguinal and popliteal regions. Unlike the initial lesion, most of these were not preceded by erythema. A recent lesion at the left upper eyelid had mild pruritis. There was no history of fluid filled lesions, medication use, trauma or family history of pigmentary disorders.

Examination revealed multiple, oval-irregular, dark-brown macules in generalized distribution. Left periorbital region, neck, upper back, abdomen, bilateral axillae, inguinal and popliteal regions were involved with size ranging from 2 X 2 mm to 5 X 2 mm as shown in Figures 1 and 2.

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**Figure 1.** Intertriginous location of hyperpigmented macules.

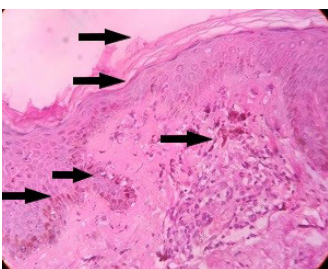


**Figure 2.** Hyperpigmented plaque with raised erythematous margins over upper eyelid.



An annular lesion was noticed over the nape of the neck. The surface was smooth and not shiny. There was no associated scaling or erythema. Two oval plaques at the left upper eyelid were shiny, violaceous with wrinkled centre and well-defined, elevated, partially blanchable, erythematous margin. Multiple hyperpigmented macules ranging in size from pinpoint to 3 mm were present over both palms since two months. The mucosae, nails and hair were normal. Histology of neck macule revealed basket weave hyperkeratosis, hypogranulosis, irregular epidermal atrophy, increased melanisation of the basal layer and vacuolated keratinocytes. Papillary dermis contained melanophages and multiple areas of mononuclear, perivascular inflammatory infiltrate. Reticular dermis showed focal area of inflammation with peri-adenexal involvement as demonstrated in Figure 3.

**Figure 3.** Histology from axillary macule



Blood count, urine-stool analysis, hepatitis serology, liver-

renal function, VDRL, vitamin B<sub>12</sub> tests were normal. Here, predominant sparing of the sun-exposed areas, centrifugal spread, infiltrated erythematous margins of the eyelid lesion and annular lesion over the neck suggest a clinical diagnosis of EDP. Typical flexural involvement in this case was striking and is rarely reported for EDP. Flexural involvement seen in our patient is a rare presentation of EDP.

## Discussion

EDP was reported in 1961 as an asymptomatic, slowly progressive ashy-gray hyperpigmentation of the trunk, extremities and face having raised erythematous borders (17.6%) that disappears after several months and may be difficult to perceive in dark skin.<sup>1,2</sup> Onset of EDP is between first and third decades of life with asymptomatic, symmetric, oval, polycyclic to irregular, ashy-grey macules of approximately 0.5 - 2 cm. Initially starting over the trunk, these spread to the face and extremities. Palms, soles, scalp, nails, and mucous membranes are spared.<sup>3</sup> Circinate and annular macules are typical of EDP. Our patient had a similar annular lesion over the nape of the neck. In children, EDP is rare and mostly reported in Caucasians (52%) than Asians (4%).<sup>3</sup> To the best of our knowledge, from India only two case reports of EDP or AD in children have been reported.<sup>4,5</sup> Unlike in adults, EDP in children can spontaneously resolve in 50% cases within two to three years.<sup>3</sup>

Differential diagnoses of EDP include LPP, multiple FDE, post-inflammatory pigmentation, late pinta, figurate erythemas, hemochromatosis, Addison's disease, melasma, leprosy, idiopathic macular eruptive pigmentation, macular amyloidosis, confluent and reticulated papillomatosis of Gougerot and Carteaud and acanthosis nigricans.<sup>6,7</sup>

The most frequent cause of confusion and controversy is LPP, characterized by occasionally pruritic, hyperpigmented macules predominantly in the exposed areas.<sup>8</sup> Bhutani et al first described LPP with symptoms similar to EDP. However, 1/3<sup>rd</sup> also had associated lichen planus (LP) clinically and histologically. Thus, they considered this a macular variant of LP.<sup>9</sup> Some authors consider EDP to be a variant of LP due to indistinguishable histology and immunofluorescence. However, Vega et al, presented differences between the two.<sup>10</sup> LPP is seen in older age group and has a typical 'actinic pattern'. In 30% cases, there can be LP lesions subsequently or simultaneously. Pigmentation in LPP does not have associated preceding inflammatory lesions. It is typically asymptomatic with occasional mild pruritus. Unlike

EDP, generalized hyperpigmentation does not develop in LPP. Involvement of flexures in LPP and EDP is rare.<sup>6,7</sup> Interestingly, our patient had palmar brown macules. This has not been described for either EDP or LPP. A biopsy from these lesions was refused by the parents.

The histology of EDP and LPP may be indistinguishable. Basal vacuolar degeneration and melanophages are seen in both disorders. Band like infiltrate at the dermo-epidermal junction (DEJ) is more likely in LPP while peri-adenexal involvement strongly suggests EDP. Late stages of LPP are typified by multiple melanophages in papillary dermis. While colloid bodies may occasionally be seen in EDP, Max-Joseph spaces are not. The immunopathology of EDP and LP are similar including populations of CD4+, CD8+ T cells and epidermal keratinocytes (HLA-DR+).<sup>5</sup>

Multiple agents, allergens and infections have been implicated in the etiology of both EDP and LPP like parasite infection, ammonium nitrite, cobalt, chlorothalonil, and barium sulphate. Systemic corticosteroids, Vitamin A, Q-switched Nd-YAG (1064 nm), 0.1% tacrolimus, and dapsone have been tried. In children, watchful waiting can be undertaken.<sup>8</sup> Our patient complained of new eruptions and was started on dapsone 50 mg with topical mometasone. He had no new lesions since dapsone was started but was lost to follow-up after two months.

## Conclusions

Although rare, hyperpigmented macules should also include EDP and LPP as differential diagnosis. Morphologically, EDP and LPP may be difficult to differentiate. Detailed history of onset and progression with histopathological examination may clinch the correct diagnosis.

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