

## Hyperpigmented Stage of Incontinentia Pigmenti

Dear Editor,

A 2-month-old girl child presented with asymptomatic hyperpigmented skin lesions in a linear distribution on the extremities, since 7 days of her age. There was no history of vesicular or verrucous lesions. Her perinatal, developmental, and family history was not significant. On mucocutaneous examination, there was involvement of bilateral upper and lower limbs in the form of multiple hyperpigmented macules in a blaschkolinear arrangement with intervening skip areas [Figure 1]. These macules were arranged in multiple lines and were giving a “Chinese letter” appearance at places. Fine white scaling was also appreciable on close examination. Her oral cavity, hair, and nails were within normal limits. Systemic evaluation including ocular, skeletal, and neurological systems was unremarkable.

Dermoscopy of hyperpigmented lesions on the posterior calf using DermLite™ DL4 (3Gen, San Juan Capistrano, CA, USA) showed multiple irregular brownish dots and globules of varying shapes and sizes with background homogenous light brown pigmentation and overlying fine white scaling [Figure 2a]. These dots were arranged to form interrupted lines. A 3-mm punch biopsy was taken from the hyperpigmented macules on the left lower leg. Histopathological examination showed a few necrotic keratinocytes in the basal layer of epidermis with pigment incontinence in the superficial dermis, consistent with the diagnosis of hyperpigmented stage of incontinentia pigmenti [IP, Figure 2b].

IP (Bloch-Sulzberger syndrome) is an X-linked dominant genodermatosis characterized by loss of function mutations in the *IKBKG/NEMO* (inhibitor of kappa

polypeptide gene enhancer in B cells, kinase gamma/nuclear factor-kappa B essential modulator) gene. The disorder is lethal in males and is mostly seen in females, although a few boys with somatic mosaicism may survive. The cutaneous lesions of IP are distributed along the lines of Blaschko and progress through four stages namely: vesicular, verrucous, hyperpigmented, and hypopigmented.

The hyperpigmented stage is characterized by pigment incontinence in the superficial dermis resulting in bluish-grey dots on dermoscopy.<sup>[1,2]</sup> Dots and globules histopathologically correspond to melanin clumps, melanocytic aggregates, or melanophages which can be present at varying levels within the epidermis and superficial dermis, reflecting the resultant color. The presence of these pigmented structures at the dermo-epidermal junction gives a brownish hue, whereas their predominance in dermis manifests in a bluish coloration.<sup>[3]</sup> The interrupted lines of brown dots on dermoscopy could be a manifestation of the punctuated presence of melanin clumps along the dermo-epidermal junction and high up in the dermis. Various clinical mimickers of this stage are linear and whorled nevoid hypermelanosis (LWNH), epidermal nevus, progressive cribriform zosteriform hyperpigmentation, Becker's nevus, and X-linked dominant variant of chondrodysplasia punctata (Conradi-Hunerman-Happle syndrome) [Table 1].<sup>[4,5]</sup>

LWNH presents as a streaky configuration of hyperpigmented macules along the lines of Blaschko, mainly distributed on the trunk and extremities. Dermoscopic examination of such a whorled lesion shows partially curved and circular pigmented streaks arranged in a parallel fashion with focally distributed perifollicular hypopigmentation.<sup>[6]</sup>

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: WKHLRPMedknow\_reprints@wolterskluwer.com

**How to cite this article:** Ahuja R, Mustari AP, Bhari N. Hyperpigmented stage of incontinentia pigmenti. Indian Dermatol Online J 2021;12:371-3.

**Received:** 21-Dec-2019. **Revised:** 08-Jan-2020.  
**Accepted:** 06-Mar-2020. **Published:** 28-Sep-2020

**Rhea Ahuja,  
Akash P. Mustari,  
Neetu Bhari**

*Department of Dermatology and Venereology, All India Institute of Medical Sciences, New Delhi, India*

### Address for correspondence:

Dr. Neetu Bhari,  
Dermatology and Venereology,  
All India Institute of Medical  
Sciences, New Delhi, India.  
E-mail: drntbhari@gmail.com

### Access this article online

**Website:** www.idoj.in

**DOI:** 10.4103/idoj.IDOJ\_628\_19

### Quick Response Code:



**Table 1: Differential diagnosis of Blaschkolinear hyperpigmentation**

Diagnosis	Clinical presentations	Dermoscopic characteristics	Histopathological features
Incontinentia pigmenti, hyperpigmented stage	Four stages: vesicular, verrucous, hyperpigmented and hypopigmented. Hyperpigmented macules in linear streaks in a ‘Chinese letter pattern’	Bluish-grey dots	Necrotic keratinocytes, pigment incontinence
Linear and whorled nevoid hypermelanosis	Streaky configuration of hyperpigmented macules	Parallel arrangement of curved brownish streaks	Increase in basal layer pigmentation with elongation of rete ridges
Progressive cribriform zosteriform hyperpigmentation	Brownish macules in a sieve-like configuration	Reticulate pattern of brownish pigmentation with fingerprint like structures	Increase in basal layer pigmentation with elongation of rete ridges
Becker’s nevus	Geographical brownish patch with ‘splashed margins’ associated with hypertrichosis and acneiform eruptions	Structureless brownish hyperpigmentation with increased density of follicles	Acanthosis, increased basal layer melanization with squaring of rete ridges



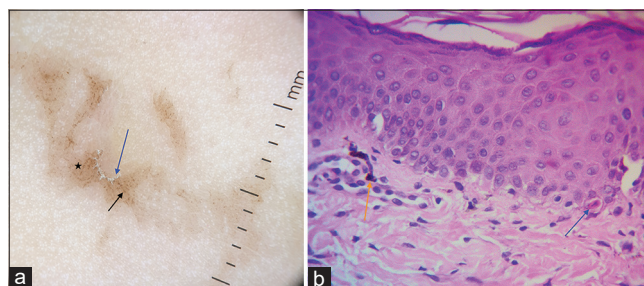
**Figure 1:** Hyperpigmented macules over the right lower leg distributing along the lines of Blaschko giving a “Chinese letter” appearance

Histopathology reveals an increase in basal layer melanization with occasional lentiginous elongation of rete ridges. Pigment incontinence is strikingly absent, thus explaining the absence of brown-grey dots and globules on dermoscopy as seen in our case.<sup>[4,7]</sup> Progressive cribriform zosteriform hyperpigmentation manifests as brownish macules in a sieve-like configuration in a Blaschkolinear pattern on the trunk.<sup>[5]</sup> Dermoscopy shows a diffuse or net-like brownish hyperpigmentation with occasional finger-print like structures or rosettes.<sup>[8]</sup> On histopathology, increase pigmentation of basal layer and elongation of rete ridges are seen. Dermal melanophages and interface dermatitis may be seen in some cases contributing to greyish-brown dots and globules similar to hyperpigmented IP, although the presence of background net-like brownish hyperpigmentation and fingerprint like structures help to differentiate it.

The significant difference in the mode of inheritance, frequency of systemic involvement and prognosis of these conditions necessitates the correct diagnosis. Thus, the dermoscopic evaluation may help in accurate early diagnosis and proper intervention.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have



**Figure 2:** (a) Dermoscopic evaluation (10×, polarizing, DermLite™ DL4, 3Gen, San Juan Capistrano, CA, USA) showing multiple irregular brownish dots and globules of varying shapes and sizes (black arrow) with background homogenous light brown pigmentation (star) and overlying fine white scaling (blue arrow). These dots were arranged to form interrupted lines. (b) Histopathological examination (H and E, 400×) showing a few necrotic keratinocytes in the basal layer of epidermis (blue arrow) with pigment incontinence in superficial dermis (yellow arrow)

given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

**References**

- Razmi M, Jogunoori S, Radotra BD, De D. Trichoscopy of whorled alopecia revealing “pigment incontinence” of incontinentia pigmenti. *Int J Dermatol* 2019;58:e156-8.
- Argenziano G, Soyer HP, De Giorgi V, editors. *Dermoscopy: A Tutorial*. Milan, Italy: EDRA Medical Publishing and New Media; 2000.
- Massi D, Giorgi V, Soyer HP. Histopathological correlates of dermoscopic criteria. *Dermatol Clin* 2001;19:259-68.
- Metta AK, Ramachandra S, Sadath N, Manupati S. Linear and whorled nevoid hypermelanosis in three successive generations. *Indian J Dermatol Venereol Leprol* 2011;77:403.
- Das A, Bandyopadhyaa D, Mishra V, Gharami RC. Progressive

- cribriform and zosteriform hyperpigmentation. Indian J Dermatol Venereol Leprol 2015;81:321-3.
6. Ertam I, Turk BG, Urkmez A, Kazandi A, Ozdemir F. Linear and whorled nevoid hypermelanosis: Dermatoscopic features. J Am Acad Dermatol 2009;60:328-31.
  7. Sinha P, Chatterjee M, Singh KK, Sood A. Linear and whorled nevoid hypermelanosis with depigmentation. Indian Dermatol Online J 2017;8:131-3.
  8. Malakar S, Mukherjee SS. Dermoscopic features of progressive cribriform and zosteriform hyperpigmentation. Our Dermatol Online 2018;9:343-5.