Incontinentia Pigmenti-Dermoscopy Features of Early Evolving Disease

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A 6-day-old female neonate, born to nonconsanguineous parents, presented with multiple fluid-filled and crusted lesions over the trunk and extremities since birth. The mother gave a history of early fetal death at 6 months of conception, previously. Cutaneous examination revealed multiple papulo-vesicles, vesicles on an erythematous base, crusted erosions, and minimally crusted erosions on a hyperpigmented base, in a linear pattern, following Blaschko lines over the trunk and bilateral upper and lower limbs [Figure 1]. Neurological and ophthalmological examination revealed no abnormalities. Skin biopsy could not be performed because of lack of parent's consent. As our patient presented with papulo-vesicles, clear vesicles, and crusted erosions, secondary bacterial infection was not suspected, and hence smear for Gram's stain was not performed. Dermoscopic examination of the papulo-vesicles and clear vesicular lesions showed yellow oil-drop-like globules surrounded by an erythematous halo [Figure 2a] and early crusted erosions showed yellow blotches, whitish scales, and hyperpigmented base [Figure 2b]. The evolved crusted lesions showed greyish-black patches in a linear distribution with multiple black dots within. The sweat gland orifices usually seen as white dots and hair follicles were absent in the pigmented patches [Figure 2c]. The clinical and dermoscopic findings confirmed the diagnosis of incontinentia pigmenti (IP).

IP is a rare X-linked dominant disorder presenting at birth or shortly after in four cutaneous stages-vesicular, verrucous, hyperpigmented, and atrophic/hypopigmented. There may be ocular, dental, and neurological involvement also. IP is caused by a mutation of the NEMO gene/IKBKG gene which is essential for protection against tumor necrosis factor-induced apoptosis.[1,2] Typical clinical findings along with family history, skin biopsy, and IKBKG gene analysis as additional criteria are used to diagnose IP.[1]

The differentials for our case in the vesicular stage include congenital herpes simplex, bacterial infections, varicella, epidermolysis bullosa, and bullous pemphigoid and a close differential in the hyperpigmented stage is linear nevoid hyper melanosis.[1] The Dermoscopy findings of all four stages of IP and correlation with the histopathological changes collated from previous case reports are summarized in Table 1.[1-4] Clinically our case had papulo-vesicles, clear vesicles, crusted erosions, and subsiding hyperpigmented erosions with less crusting. The dermoscopy of papulo-vesicles and vesicles revealed yellow spots with an erythematous halo. This is due to eosinophilic spongiosis and corresponds to the early vesicular stage.[1,2] The crusted erosions showed yellow blotches, peripheral white scaling, and underlying hyperpigmentation. The above dermoscopic findings are similar to that reported by Minic *et al*.[1] and Arora *et al*.[2] in the vesicular stage of IP. The evolved crusted lesions showed hyperpigmented patches with black dots within, due to pigment incontinence and the absence of white dots (sweat pores) and hairs which have been described in the later hyperpigmented stage of IP.[3] In linear nevoid hyper melanosis, dermoscopy reveals brownish streak pigmentation over the whorled areas in a linear or circular arrangement showing follicular white dots and geographical edges, whereas in IP the follicular white dots are absent in the hyper and hypo-pigmented patches.[3,5]

IP, being a rare disease, only a few cases with dermoscopy findings have been reported, and fewer in the early stages of the disease.[1] Our findings of absent white dots (sweat pores) and hairs in the black patches helps to differentiate from linear

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How to cite this article: Vijaikumar M, Saravanan Y, Christin Armel JS. Incontinentia pigmenti-dermoscopy features of early evolving disease. Indian Dermatol Online J 2025;16:212-4.

Received: 27-Feb-2024. **Revised:** 02-Jun-2024. **Accepted:** 03-Jul-2024. **Published:** 26-Nov-2024.

Access this article online

Website: https://journals.lww.com/idoj

DOI: 10.4103/idoj.idoj_179_24

Quick Response Code:

Table 1: Correlation of histopathology and Dermoscopy in the clinical stages of Incontinentia pigmenti

Stages of IP	Histopathologic findings	Dermoscopic findings
Stage 1 Vesicular stage	Eosinophilic spongiosis, intraepidermal vesicles containing eosinophils and many apoptotic keratinocytes in epidermis.[1,4]	Oil-drop such as yellowish structureless areas in a linear arrangement and yellow serocrusts on an erythematous skin.[1,2]
Stage 2 Verrucous stage	Papillomatosis, hyperkeratosis, acanthosis and apoptotic cells forming squamous eddies in epidermis. Dilated blood vessels in the dermal papillae, peri-vascular lymphocytes and eosinophils and major melanin incontinence.[1,4]	Early verrucous lesions are star-shaped, yellowish or whitish on an erythematous and a slightly pigmented skin. Well-developed lesions have dotted vessels surrounding a central keratotic part, or can be distributed on the lesion, with thrombosed capillaries, resembling a viral wart.[1]
Stage 3 Hyperpigmented stage	No more hyperplasia with only scattered apoptotic keratinocytes in epidermis. Proliferation of capillaries in the papillary and superficial reticular dermis with eosinophils. Marked melanin incontinence and numerous melanophages in dermis.[1,4]	Linear hyperpigmented streaks show exaggerated normal reticulate pigmentation and few grey dots suggesting pigment incontinence These streaks characteristically lack sweat gland orifices and hair follicles.[1,3]
Stage 4 Atrophic/ hypopigmented stage	Atrophy and massive reduction of melanin in the basal layer in epidermis. Homogenization of collagen and dilated capillary vessels in the dermal papillae. Complete absence of pilosebaceous units and eccrine gland.[1,4]	Discrete, ill-defined white areas with loss of sudomotor appendages. Numerous very small dotted vessels present on the surrounding hypo- and normally pigmented skin. Perifollicular depigmentation in the surrounding normal skin.[1]

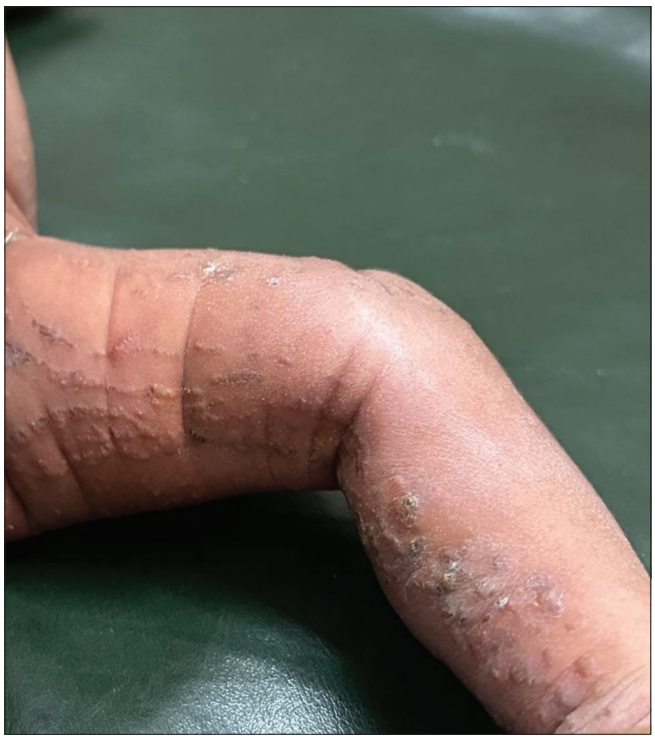

Figure 1: A 6-day-old neonate with incontinentia pigmenti showing vesicles and crusted erosions in a linear distribution along blaschko lines on the left leg

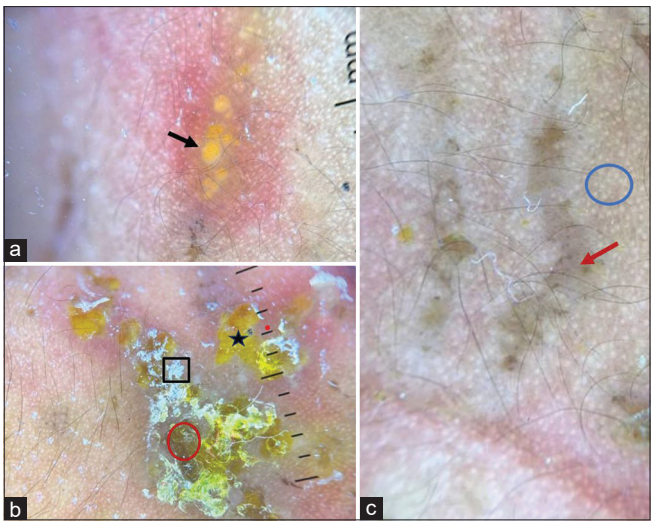

Figure 2: Dermoscopy showing (a) yellow dots with erythematous halo (black arrow) over the sero papules and vesicles (b) yellow blotches (star), whitish scales (square) and hyperpigmented base (red circle) over early crusted erosions and (c) linear hyperpigmented patches with black dots within, corresponding to pigment incontinence (red arrow) over the evolved hyperpigmented erosions. White dots (blue circle) corresponding to sweat orifices and hair were absent (Dermlite DL4 . polarized light, 10x)

nevoid hypermelanosis and confirms the earlier reports.[1,3,5] Patients of IP may have lesions of different stages as the lesions evolve and dermoscopy as a noninvasive, versatile tool may be used to demonstrate this. In our case of a 6-day-old neonate, dermoscopic findings of the early vesicular stage, crusted erosions, and subsiding erosions evolving to hyperpigmented stage of IP were demonstrated simultaneously and helped in excluding the above differentials when a skin biopsy was not possible.

Declaration of patient consent

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

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