Linear and whorled nevoid hypermelanosis versus incontinentia pigmenti: Pigmentary incontinence is not a differential histological feature

Dear Editor,

Linear and whorled nevoid hypermelanosis (LWNH) is characterized by hyperpigmented macules in swirls and streaky configuration along the lines of Blaschko without preceding inflammation.1–3 LWNH, first defined by Dr. Kalter in 1988, is considered a pigmentary disorder with genetic mosaicism, which results in clonal migration and proliferation of embryonic melanocyte precursors.2,4 By contrast, incontinentia pigmenti (IP), another segmental disorder, usually proceeds through four cutaneous stages: (1) vesicular stage; (2) verrucous stage; (3) hyperpigmented stage; and (4) hypopigmentation stage, along with pigmentary incontinence microscopically. Herein, we describe a typical case of LWNH despite moderate pigmentary incontinence. We argue that pigmentary incontinence may not be a differential histological feature between LWNH and IP.

An 8-month-old girl developed macular hyperpigmentation with linear and whorled configuration over her face, trunk, and extremities since birth (Figure 1). There were no preceding vesicles, verrucous lesions, or popular eruptions. The birth history was uneventful with normal developmental milestones. Her mother recalled a localized linear and whorled hyperpigmentation over her flank area since infancy. Under the tentative diagnosis of LWNH, skin biopsies were performed in a hyperpigmented area and a hypopigmented area on the lower back. Microscopically, there was increased basal pigmentation in the hyperpigmented area. However, moderate melanin incontinence was also present in the hyperpigmented area (B1 and B2). The Melan-A (B3 and B4) and Fontana-Masson (B5 and B6, 100×) stains showed slightly enhanced basal pigmentation and significantly increased numbers of melanocytes in the hyperpigmented area, respectively. The melanin incontinence in the hyperpigmented area was further demonstrated ultrastructurally within the dermal macrophage. Based on the clinical history and pathological findings, the final diagnosis of LWNH was rendered. The patient was asked for regular checkups in our clinics in 3–6 month intervals.

The typical histopathology of LWNH showed increased pigmentation of the basal layer and prominent melanocytes. The pigment incontinence is usually, but not always, absent. There has been no estimation on the frequency of pigment incontinence in LWNH because only a few cases have been reported.3,5 Albeit the pigment incontinence was considered a pathological marker of IP by the interface changes, the presence of pigment incontinence may not totally preclude the diagnosis of LWNH. Delaporte et al5 reported a case of LWNH and suggested that pigment incontinence may not be taken as a specific marker to distinguish between IP and LWNH. In our case, the clinical hyperpigmentation was not preceded by inflammation, bullae, or verrucae; the mucosa, eyes, palms, or soles were not involved, and no other congenital anomalies were associated. All these evidences pointed to the diagnosis of LWNH.

In summary, we describe a patient with LWNH who demonstrated both basal pigmentation and pigment incontinence microscopically. We conclude that pigmentary incontinence may not be a differential microscopic feature between IP and LWNH.
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Figure 1 (A) There was linear and whorled configuration of macular hyperpigmentation over the face, trunk, and extremities. Two sites of skin biopsy were performed in a hyperpigmented area (white arrow) and a hypopigmented area (black arrow). Microscopically, there was moderate pigmentation in the basal keratinocytes in both hyperpigmented and hypopigmented areas. However, (B) there was significant melanin incontinence in the hyperpigmented area (B1 vs. B2, H&E, 100 ×). The Melan-A (B3 vs. B4, 100 ×) and Fontana-Masson (B5 vs. B6, 100 ×) stains showed slightly enhanced basal pigmentation and significantly increased numbers of melanocytes over the hyperpigmented area. H&E = hematoxylin and eosin.