Progressive Cribriform and Zosteriform Hyperpigmentation

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To the Editor:

Progressive cribriform and zosteriform hyperpigmentation (PCZH) was first described by Rower et al[1] in 1978. The diagnostic criteria included the following: (1) uniformly tan cribriform macular pigmentation in a zosteriform distribution; (2) a histologic pattern that consisted of a mild increase in melanin pigment in the basal cell layer and complete absence of nevus cells; (3) no history of rash, injury, or inflammation to suggest postinflammatory hyperpigmentation; (4) onset occurring well after birth with gradual extension; and (5) lack of other associated cutaneous or internal abnormalities.[1]

Many pigmentary disorders occurring along the Blaschko lines are included in differential diagnosis of PCZH such as incontinentia pigmenti (IP), progressive zosteriform macular pigmented lesion (PZMPL), and linear and whorled nevoid hypermelanosis (LWNH). However, PCZH is considered to be the localized variant (the late onset) of LWNH.[2] We report a case of PCZH, a segmented and delayed form of LWNH.

A 25-year-old woman presented with asymptomatic progressive multiple brownish macular eruptions arranged in a zosteriform pattern on the left arm and thigh of 3 months' duration. There was no history of injury or any prior cutaneous changes. There was no personal or family history of similar eruptions and she was otherwise in good health. She was not taking any medications. Physical examination showed linear, uniformly tanned, cribriform hyperpigmentation along the Blaschko lines on the left arm and thigh (Figure 1). Routine laboratory tests, including complete blood cell count with differential, were normal. Assuming a diagnosis of PCZH or PZMPL, we performed a punch biopsy on the left upper arm. The histopathologic findings showed increased pigmentation of the basal layer. There were a few

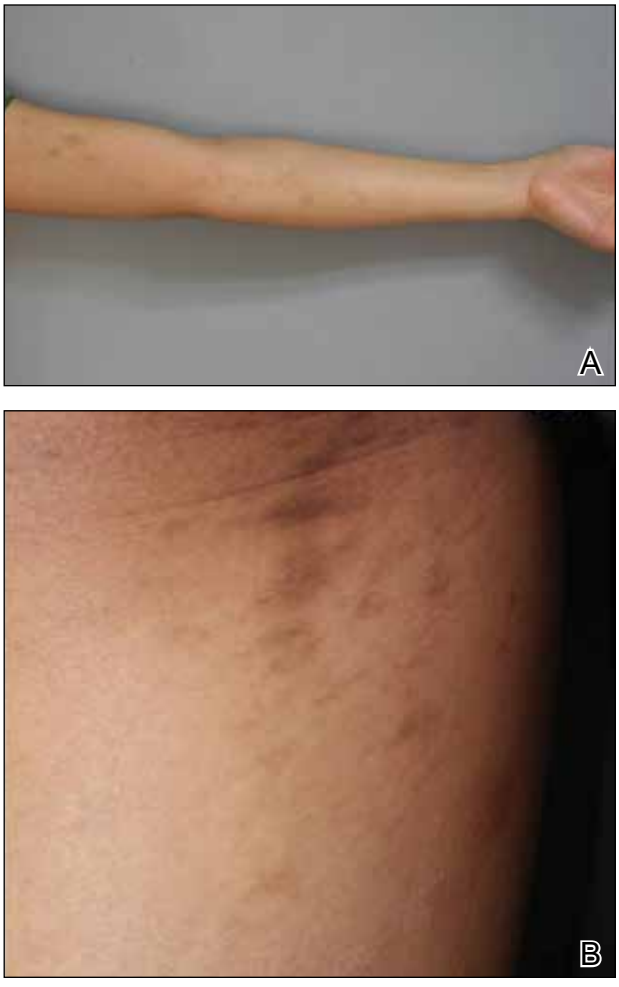

Figure 1. Asymptomatic linear, uniformly tanned, cribriform hyperpigmentation along the Blaschko lines on the left arm (A) and thigh (B).

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The authors report no conflict of interest.

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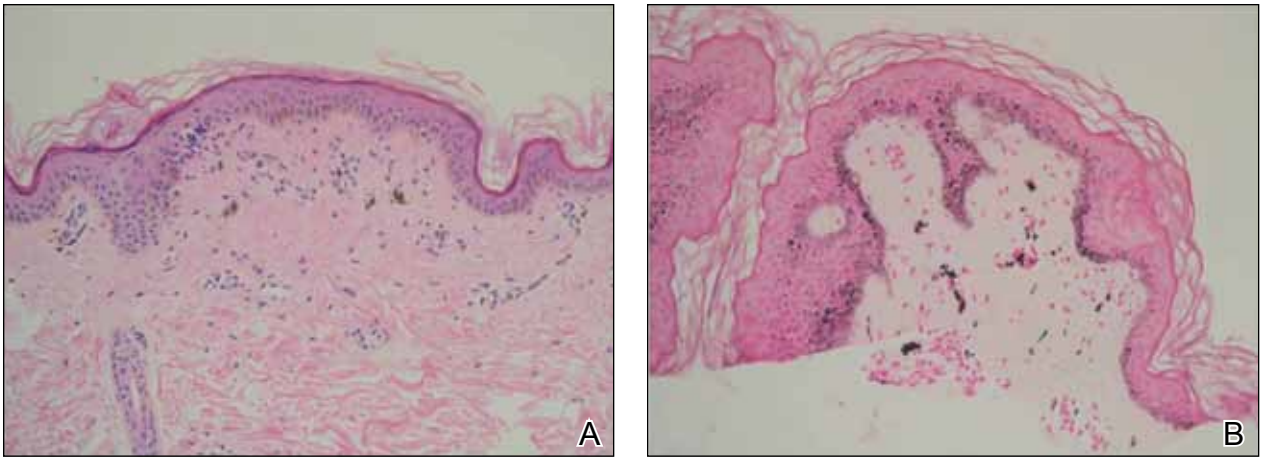

Figure 2. Histopathologic findings showed increased pigmentation of the basal layer with a few dermal melanophages. No nevus cells were present (A)(H&E, original magnification ×100). Fontana-Masson stain showed an increase in melanin in the basal layer (B)(original magnification ×100).

dermal melanophages and no nevus cells present (Figure 2A). Fontana-Masson stain showed an increase in melanin in the basal layer (Figure 2B). On the basis of these clinical and histological findings, a diagnosis of PCZH was made. She was observed without treatment for 6 months showing no change.

Progressive cribriform and zosteriform hyperpigmentation is a disorder of pigmentation along the Blaschko lines. The trunk is the most common site of involvement.[3] In the differential diagnosis, other pigmentary disorders along the Blaschko lines must be excluded, including the pigmentary stage of IP, PZMPL, and LWNH. In IP, characteristic inflammatory vesicular and verrucous stages usually precede the whorled pigmentation.[4] In approximately 80% of cases, IP is associated with various congenital abnormalities, particularly of the central nervous system, eyes, and teeth.[5] Progressive zosteriform macular pigmented lesion is a chronic pigmentary dermatosis similar to PCZH but is characteristically accompanied by pruritus as a prodromal symptom. It is usually preceded by multiple pruritic macular pigmentation in part of the dermatome for a period of time. Then the size and number of the pigmented macules abruptly increases and coalesces into patches.[6] Linear and whorled nevoid hypermelanosis was first described by Kalter et al[7] in 1988. It is characterized by swirls and whorls of hyperpigmented macules without preceding bullae or verrucae along Blaschko lines, usually occurring within the first 2 years of life. The lesions are stable in some patients but can spread in others, stabilizing by 2 to 3 years of age.[7-10] It has been referred to as zosteriform lentiginous nevus, zebralike hyperpigmentation, and reticulate hyperpigmentation distributed in a zosteriform fashion.[2,9]

Linear and whorled nevoid hypermelanosis can be distinguished from PCZH by a diffuse or localized pattern and an association of congenital anomalies.[3] However, neurologic and skeletal anomalies also can be observed in PCZH.[11] Additionally, not all LWNH cases show a diffuse type.[2] Therefore, LWNH has been used to encompass a wide spectrum of clinical entities, ranging from the congenital or perinatal form described by Kalter et al[7] to the segmented and delayed form described by Rower et al[1] for which there is a tendency to use the term *progressive cribriform and zosteriform hyperpigmentation*.[2,10,11] There are no clinical and histologic differences between PCZH and LWNH, other than a later onset.[2] Although some authors reported that PCZH and LWNH have increased hyperpigmentation of the basal layer and prominent melanocytes without incontinence of pigment on histopathology,[2,7,8] other reports have demonstrated that both could show pigment incontinence,[3,10,12-14] such as in our case.

Progressive cribriform and zosteriform hyperpigmentation is considered to be the localized variant as well as the late onset of LWNH.[2] We report a case of PCZH, a segmented and delayed form of LWNH without systemic abnormalities.

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