Speckled Pigmentation in Nevus Depigmentosus-An Unfamiliar Concurrence

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Nevus depigmentosus (ND) is a relatively rare congenital disorder presenting with hypopigmented macules and patches as a result of functionally defective melanocytes and abnormal melanosomes. Clinically, three variants have been described. A localized variant has a single localized hypopigmented macule or patch with serrated borders. The segmental variant has a block-like distribution of hypopigmented lesions without crossing the midline. Systematized/linear/whorled variant is associated with seizures, hemihypertrophy, mental retardation, yellow hair, or atopic dermatitis.[1] Herein, we report a rare presentation of ND with speckled pigmentation in the entire patch.

A 16-year-old female patient presented with an asymptomatic hypopigmented patch over the face and neck since childhood. There was no history of systemic abnormality or family history of similar lesions. Clinical examination revealed a well-defined hypopigmented patch over the neck extending from the chin to the upper chest measuring 10 × 4 cm with irregular serrated borders. Interestingly, specks of hyperpigmentation of various sizes were present throughout the patch [Figure 1a]. Diascopy excluded nevus anemicus as a differential diagnosis. Dermoscopy showed a hypopigmented patch having an altered pigment network and feathery margins. Specks of hyperpigmentation showing an accentuated pigment network inside the patches were also seen [Figure 1b].

Histopathology examination of both hypopigmented and hyperpigmented regions showed the presence of melanin pigment within epidermal basal keratinocytes. Immunostaining with HMB 45 showed a reduction in the number of melanocytes in the hypopigmented lesion in comparison to the hyperpigmented lesion [Figure 2a and b]. As a part of the treatment plan, dermabrasion followed by autologous non-cultured epidermal cell suspension procedure was performed on a test patch measuring 4 × 4 cm with minimal response.

Nevus depigmentosus is a congenital pigmentary disorder described by Lesser in 1884 and characterized by hypopigmented macules and patches.[1] It is a manifestation of cutaneous mosaicism, wherein a clone of melanocytes is unable to produce melanin.[2] The clinical diagnostic criteria of ND are i) leukoderma present at birth or of an early onset, ii) no alteration in the distribution of leukoderma throughout life, iii) no alteration in texture or change in sensation in the affected area, and iv) absence of hyperpigmented border. Dermoscopy of ND shows a hypopigmented patch with an irregular and serrated border and a faint reticular network. The borders show a pseudopod pattern protruding into the normal skin. Hairs within the patches are of normal color without peripheral hyperpigmentation. Specks of pigmentations are found at the margins in a few cases.

Similar to our case, ND with lentigines[3] and acquired melanocytic nevi[4] have been described in the literature. In our case, there were specks of hyperpigmentation throughout the lesion. The following hypotheses could be put forward to explain the same: 1) reversal of mutation in melanocytes as an escape mechanism from mosaicism, 2) excessive melanogenesis to overcome defective melanin synthesis, 3) release of mosaicism due to neural

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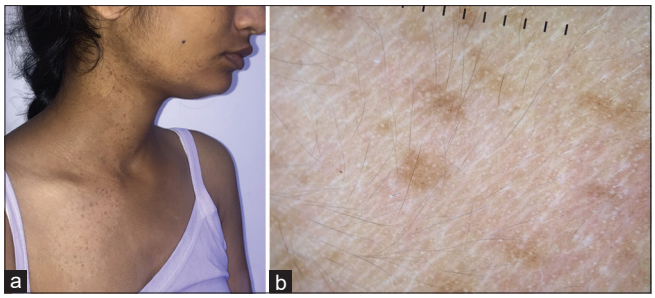

Figure 1: (a) A case of nevus depigmentosus showing a well-defined hypopigmented patch with irregular borders over the neck extending from the chin to the upper chest measuring 10 × 4 cm. Specks of hyperpigmented macules uniformly distributed within the patch can be appreciated. (b) Dermoscopy image showing a hypopigmented patch having an altered pigment network and feathery margins and specks of hyperpigmentation with an accentuated pigment network inside the patches

mechanisms such as an increase in αMSH receptors, and 4) twin spotting.

Although repigmentation in ND is possible by grafting techniques, the results are inconsistent among various reports.[5] Deviations from normal clinical presentations are rare, our report exemplifies one such clinical presentation.

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.

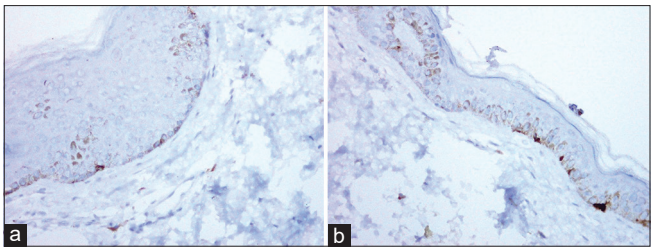

Figure 2: HMB45 immune staining shows a reduction in the number of melanocytes in lesion biopsy (a, 400×) as compared to the non-lesion skin (b, 400×)

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Conflicts of interest

There are no conflicts of interest.

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