

Short Communication

Pigmented nevi within segmental nevus depigmentosus - a rare case

Sir, nevus is the Latin word for 'maternal impression' or 'birthmark' and indicates a circumscribed, non-neoplastic skin or mucosal lesion, usually present at or soon after birth, and fixed. Many, possibly all, nevi represent clones of genetically altered cells arising from mosaicism. Some of the nevi are commonly encountered like, acquired melanocytic nevi, Becker's nevi and verrucous epidermal nevi. Some of them are extremely rare like, apocrine nevi, proteoglycan nevi, nevoid ichthyosis hystrix, kissing nevus and pigmented nevi within nevus depigmentosus. Nevus spilus is comprised of a flat, macular component, subtly darker shade than the surrounding skin, resembling a café-au-lait spot. Within this pale background, there are lentigo-like lesions and also elevated darker-brown nevi. In our case, the background lesion is hypopigmented macule, resembling nevus depigmentosus. Interspersed within this hypopigmented macule, there are many hyperpigmented, lentigo-like macules.

An 18-year-old girl presented with asymptomatic, hypopigmented lesion over the right side of neck since five years of age. To start with, it was a small whitish dot. Gradually it increased in size. The lesion gradually extended to the right shoulder. Patient also developed small hyperpigmented dots over the existing hypopigmented lesion after one year. Since the past few years, the lesion was static. There was no history of receiving phototherapy. There was no other significant history. There was no family history of similar complaints.

General physical examination and systemic examinations were unremarkable. Cutaneous examination revealed a hypopigmented patch measuring about 25cm x 15cm situated over the right side of the lateral aspect of the neck extending to the right shoulder and right side of the upper back, along the C7-C8 dermatomes. There were multiple, brownish, round macules measuring 5 to 10 mm interspersed within the hypopigmented macule. The hyperpigmented macules are confined only within the hypopigmented macule and it was not present elsewhere (**Figure 1**). The hair in the hypopigmented patch was not depigmented. Wood's lamp examination revealed an off-white accentuation, in contrast to the chalky-white accentuation noted in vitiligo. Routine hematological and biochemical parameters were within normal limits. Histopathological examination of hypopigmented macule revealed normal melanocyte number with decreased melanin (**Figure 2**). Histopathological examination of the hyperpigmented macule revealed increased melanocytes (**Figure 3**). With this background, we made a diagnosis of lentiginos within segmental nevus depigmentosus.

In this case, first we thought of repigmenting vitiligo. But Histopathological examination of the hypopigmented area revealed normal numbers of melanocytes, ruling out the possibility of repigmenting vitiligo. Then we considered the possibility of pigmented nevi within the nevus depigmentosus. This was later confirmed by histopathological examination. The limitation of the lentiginos to the nevus depigmentosus was striking. Nevus depigmentosus is a congenital, unilateral, localized hypopigmented macule or patch that is stable in its size and distribution throughout life. It may be localized, segmental, or systematized.



Figure 1 Pigmented nevi within nevus depigmentosus.

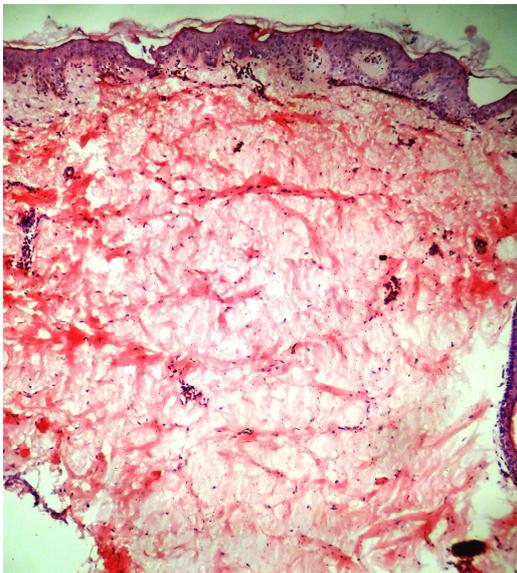


Figure 2 Histopathology of pigmented nevi within nevus depigmentosus.

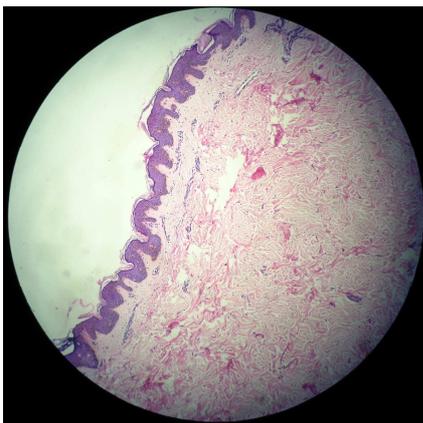


Figure 3 Histopathology of nevus depigmentosus

The development of lentigines within nevoid hypopigmentation has been reported only

rarely.^{1,2,3} There is a case report of development of lentigines within nevus depigmentosus as a result of NB-UVB therapy which was given as a result of mistaken diagnosis of nevus depigmentosus for segmental vitiligo.⁴ In some patients with hypomelanosis of Ito or piebaldism, acquired pigmented macules may occur. It has been reported that inactive cells might become functional to produce contraction of hypomelanotic areas. However, this is caused by a gradual “fading” in which the streaks acquire the same level of pigmentation as the surrounding skin. Somatic mosaicism is one genetic explanation for the presence of hypopigmented or hyperpigmented streaks. This mosaicism can involve a single gene, a cluster of genes, or an entire chromosome. In our case, the nevus depigmentosus underwent an unusual change, development of pigmented nevi. We report this case for its rarity.

References

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Pradeep Vittal Bhagwat, Chandramohan Kudligi, Suman Gurunathgouda Odugoudar, Sowmyashree Krishna

Department of Skin & STD, Karnataka Institute of Medical Sciences, Hubli, Karnataka

Address for correspondence

Dr. PV Bhagwat, Associate Professor,
Department of Skin and STD,
Karnataka Institute of Medical Sciences,
Hubli-580022, Karnataka
E-mail: sharadapbagwat@yahoo.com