Speckled Acral Hypopigmentation: A New Pigmentary Disorder or an Unknown Presentation of a Known Disorder?

Sir,

Reticulate pigmentary disorders are a broad group of pigmentary disorders with significant overlap, often posing a diagnostic challenge due to variable phenotypic expression of similar gene defects. The acral reticulate pigmentary disorders described so far including reticulate acropigmentation of Kitamura, Dowling–Degos disease, and acropigmentation of Dohi are characterized by the presence of hyperpigmented macules with or without hypopigmented lesions. Here, we describe a patient presenting with only hypopigmented macules in a speckled pattern on acral sites.

A 17-year-old boy presented with a 2-year history of asymptomatic hypopigmented to depigmented macules on the dorsae of hands and feet. On examination, there were multiple, small (1–3mm), well-defined hypopigmented to depigmented macules symmetrically over the dorsae of hands and feet with prominent clustering over the sides. Lesions were mostly discrete, coalescing at few places, and present over a normal background skin, thus giving a speckled appearance [Figure 1a]. There were no hyperpigmented or atrophic macules, no palmar pits or break in dermatoglyphics, and no involvement of flexures. The patient was otherwise healthy with no significant medical history. He was born of nonconsanguineous marriage with no similar history in family.

Two biopsies done from the hypopigmented macules of both hands showed similar features. The number of melanocytes were normal but there were a striking number of macromelanosomes in melanocytes and keratinocytes [Figure 1b]. Fontana-Masson and HMB-45 staining showed similar findings [Figure 1c and d].

Reticulate pigmentary disorders are an evolving group of dermatoses with a recent report of unusual clinical presentation as "speckled acral hypopigmentation."The term familial was used by authors to describe a case similar to ours where a 14-year-old girl presented with speckled hypopigmentation on the sides of hands and feet bilaterally with a strong family history of similar lesions. Histopathological evaluation in this patient showed decreased number of melanocytes.[1]

Congenital symmetric acroleukopathy has been described in literature, but here depigmented macules are large and present over periungual areas.[2] Reticulate acropigmentation of Kitamura is characterized by the presence of well-defined atrophic hyperpigmented macules on the dorsum of hands and feet, palmar pits, and break in dermatoglyphics.[3] Rare cases of disseminated hypo- or depigmented macules and papules are described, but presence of only hypopigmented macules is not seen in this condition.[4] Dowling–Degos disease is

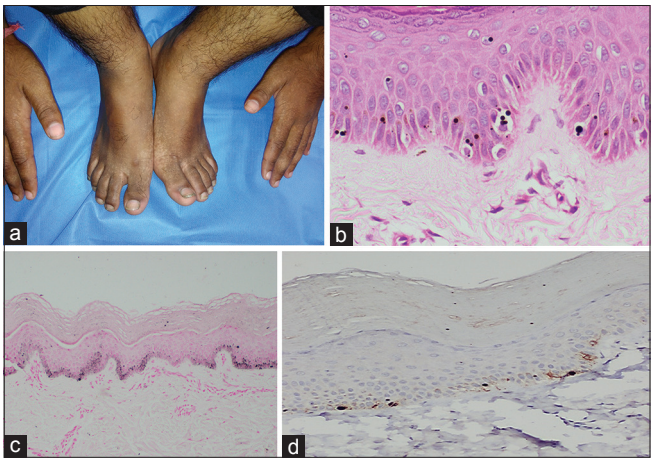

Figure 1: (a) Symmetric involvement of bilateral dorsae of hands and feet in form of multiple, small (1–3-mm sized), well-defined hypopigmented macules with prominent clustering over the sides. (b) Histopathology showing macromelanosomes in melanocytes as well as in keratinocytes (H and E, ×400). (c and d) Fontana-Masson and HMB-45 staining showing similar findings (×100, ×200)

characterized by the presence of hyperpigmented macules over the flexures along with comedonal papules on the back and neck.[3] Rarely, Dowling–Degos disease presenting as hypopigmented macules similar to our case has been described, but in this case lesions were widely distributed over the chest, back, axilla, upper arms, and inguinal folds and there was no clustering of lesions.[5] Acropigmentation of Dohi shows presence of both hyper and hypopigmented macules on the hands and feet, which may extend to the proximal extremities and face. Extragenital lichen sclerosus is a chronic inflammatory disorder characterized by the presence of small, porcelain white, mildly atrophic polygonal papules and plaques which commonly affects neck, shoulders, and upper portion of the trunk.[6] Confetti or guttate form of hypomelanotic macules seen in tuberous sclerosis patients present as 1–3 mm, numerous, small hypopigmented macules that typically occur symmetrically over the distal extremities. These hypopigmented macules usually develop during early childhood. Over time, these patients develop other characteristic cutaneous and systemic features of tuberous sclerosis.[7]

None of the above described differential diagnosis have macromelanosomes on histopathology. Histopathological differential diagnosis of macromelanosomes include Griscelli syndrome, Chediak Higashi disease, albinism, nevus spilus, neurofibromatosis, and xeroderma pigmentosum.[8,9] However, our patient did not have clinical features of any of the above described histopathological differentials.

Our patient has an unusual clinical presentation which does not fit into any of the well described reticulate pigmentary

disorders. It may be a hitherto unknown presentation of previously described disorders or it may be a unique disorder itself. Careful study of large number of patients and genomic analysis will be required to unfold this mystery.

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.

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Conflicts of interest

There are no conflicts of interest.

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