Dermoscopy of Localized Darier's Disease in Fitzpatrick Type IV Skin

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A 25-year-old female presented with asymptomatic dark colored eruptions around the eyes, nose, and ears since 6 months. Clinical examination revealed multiple small skin-colored and hyperpigmented papules involving the eyelids and periorbital area, nasolabial folds, the area adjoining the nostrils, and the external ear. Papules ranged 1–3 mm in size and were discrete as well as coalesced to form greasy hyperpigmented plaques especially involving the eyelids and adjacent area, and around the nostrils [Figure 1]. The rest of the cutaneous examination including the nails and oral mucosa was unremarkable.

Noncontact dermoscopy of an area of confluent papules under polarized mode using DermLite™ DL3 (3 Gen, San Juan Capistrano, CA, USA) revealed multiple dark brown polygonal to round structures surrounded by grayish-white halo with superficial white scaling imparting a global honeycomb pattern. At places, these dark brown structures with whitish halo housed light brown follicular openings forming three zones – central light brown follicular opening, surrounding dark brown structure, and peripheral whitish halo. Focal areas with exaggerated pigment pseudo-network were also seen [Figure 2]. Histopathological analysis revealed hyperkeratosis, acanthosis, and focal suprabasal clefts containing dyskeratotic cells – corps ronds and corps grains [Figures 3 and 4]. The clinical and histological features were consistent with Darier's disease (localized form) and the patient responded satisfactorily to oral isotretinoin [Figure 5].

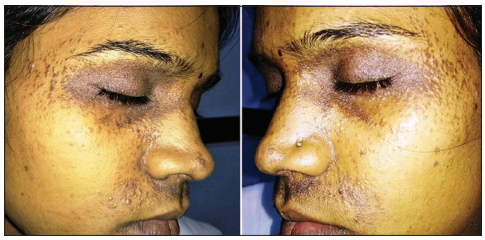

Figure 1: Multiple discrete and confluent greasy hyperpigmented papules involving the eyelids and periocular areas, perinasal area, nasolabial folds, and few papules on the temples, nose, and forehead

Darier's disease is an autosomal dominant disorder of keratinization due to mutations in *ATP2A2* gene, clinically characterized by symmetrically distributed multiple skin-colored to hyperpigmented keratotic papules and coalesced greasy plaques predominantly involving the seborrheic areas of the body. Other characteristic cutaneous features such as cobblestoning of hard palate, palmar pitting, and nail changes (V-shaped nicking at the distal edge of the nail, longitudinal white and red alternating streaks, subungual hyperkeratosis, splinter hemorrhages, and nail fragility) aid in clinical diagnosis.[1] Localized (segmental and nonsegmental) forms have also been described, possibly reflecting cutaneous mosaicism.[2,3]

Dermoscopy appears to be a useful ancillary tool in complementing the clinical diagnosis of Darier's disease as the features – brown polygonal or round structures (corresponding to the hyperkeratosis) and the surrounding whitish halo (corresponding to the acanthosis) appear to be consistent based on the available evidence.[4] The brown color of the round and polygonal structures is attributable to the melanin [Figure 3]. These findings have been described by Errichetti *et al*. as consistent features in 11 patients of biopsy-confirmed generalized Darier's disease.[5] In addition to these features, they also observed a pinkish

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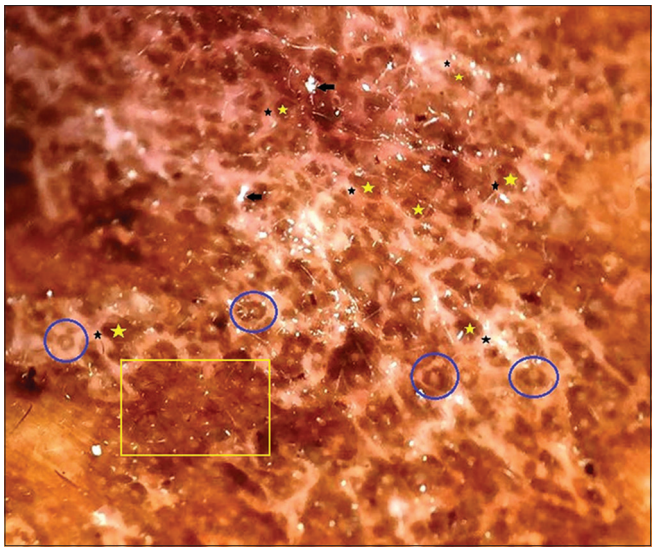

Figure 2: Noncontact dermoscopy of a plaque under polarized mode using DermLite™ DL3 showing multiple round to polygonal brown globules (yellow stars) with surrounding grayish-white halo (black stars). Some of these brown globules show central follicular light brown opening (blue circles) forming three zones (see text). Superficial white scales (black arrows) and focal areas with exaggerated pigment pseudo-network are also seen (yellow square)

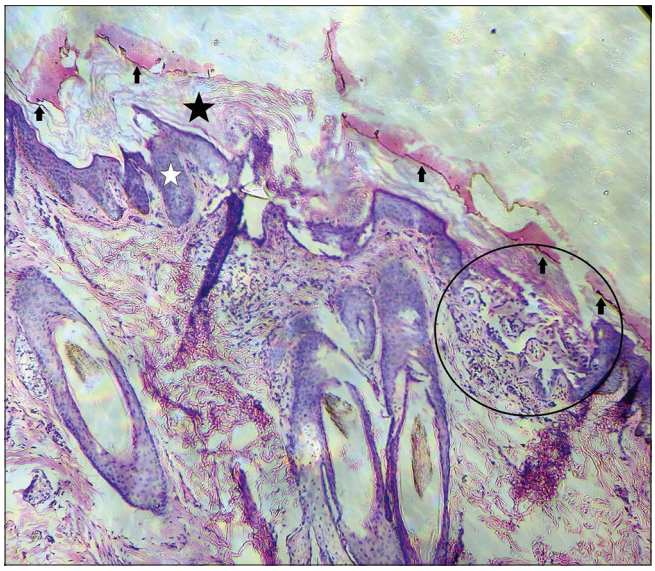

Figure 3: Photomicrograph showing hyperkeratosis (black star), acanthosis (white star), and focal suprabasal acantholysis (black circle) with the blister cavity showing dyskeratotic cells (see Figure 4). Also note the melanin in the hyperkeratotic stratum corneum (black arrows) [H and E, original magnification ×10]

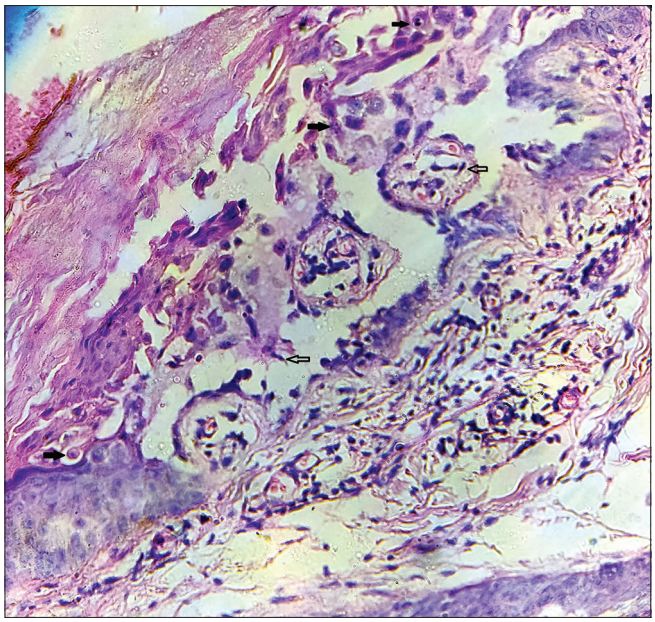

Figure 4: A suprabasal blister showing acantholysis and dyskeratotic cells – the corps ronds (black solid arrows) and corps grains (black hollow arrows) [H and E, original magnification ×40]

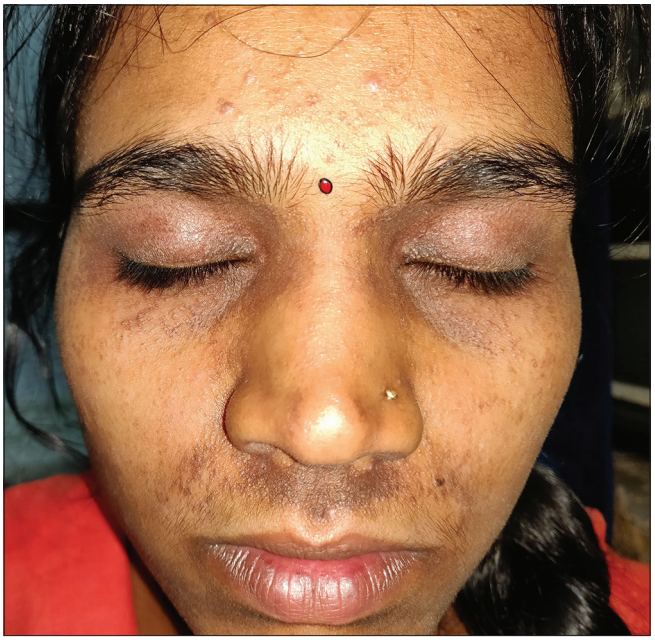

Figure 5: Satisfactory response after 4 weeks of treatment with oral isotretinoin

homogeneous background, vascular patterns, and whitish scales. They have reported similar features in a type 1 segmental form of the disease as well.[6] To the best of our knowledge, this is the first observation in an Indian patient (who had Fitzpatrick type IV skin). In comparison to the findings described by Errichetti *et al.*,[5] in the lighter skin type, vascular structures and erythematous background were not seen in our case and the "brown structures" were understandably darker in our case as opposed to yellowish or light brown color in the lighter colored skin. The "three zones" formed by the brown globules with central follicular opening and peripheral whitish halo was our another observation not described before.

Similar dermoscopic features have also been observed in other acantholytic dyskeratotic conditions such as acantholytic dyskeratotic acanthoma and Darier-like histological variant of Grover's disease because of the resembling microscopic features.[7,8] Hence, the dermoscopic features need to be correlated with the clinical picture. The findings nonetheless

assert the clinical diagnosis of Darier's disease, especially in such localized variants, and aid in differentiating from other clinically resembling disorders.[5] The available evidence, however, is still limited, and further observations are required in both in lighter and darker skin types to establish objective dermoscopic criteria for Darier's disease.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her names and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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