Darier's Disease with Guttate Leukoderma: Correlation of Dermoscopic and Histological Findings

A 15-year-old boy presented with multiple itchy hyperpigmented papules over face and trunk and hypopigmented macules over limbs for the last 8 years. The patient reported exacerbation of itching in the lesions during summers, especially after sweating. The hypopigmented macules were not preceded by any inflammatory lesions. Similar lesions were also noted in his mother. On examination, there were multiple skin-colored to hyperpigmented greasy papules and plaques present over the face, predominantly in the centrofacial area and the upper trunk. He also had multiple hypopigmented macules, 1-4 mm in size, on extensor surface of both upper limbs. Punctate depressions were seen on the palms [Figure 1]. Nails and oral cavity were normal.

Dermoscopic examination [DermLite[™] DL4 (3Gen, San Juan Capistrano, CA, USA)] of facial papules showed a central brownish area surrounded by a thin whitish halo and fine white scales,



Figure 1: (a) Discrete to confluent greasy papules over the nasolabial folds. (b) punctate depressions on the palms. (c) hypopigmented macules over the extensors of the forearm

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while dermoscopy of forearm showed multiple depigmented areas of variable size and shapes which were confluent at places [Figure 2a and b]. Histopathological examination of skin biopsy taken from papular lesion on chest showed hyperkeratosis, focal parakeratosis, acanthosis, necrotic keratinocytes, and suprabasal cleft formation, while that of hypopigmented macule did not reveal any abnormality [Figure 2c and d].

Darier's disease is an autosomal dominant disorder of keratinization caused by mutations in *ATP2A2* which encodes for the SERCA2 calcium pump.^[1] The disease



Figure 2: (a) Dermoscopy (10x, polarizing, DermLite™ DL4, 3Gen, San Juan Capistrano, CA, USA) of greasy papule showing central brownish area (blue arrow) with a thin whitish halo (oval shape) and fine white scales (green arrow). (b) Dermoscopy (10x) from forearm showing multiple depigmented areas (black arrow) of variable size and shapes, confluent at places (green arrow). (c) Skin biopsy (H and E, 100x) from greasy papule showing hyperkeratosis, parakeratosis (blue arrow), necrotic keratinocytes (black arrow), and suprabasal cleft (green arrow). (d) Skin biopsy (H and E, 100x) from hypopigmented macule was within normal limits

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Sachin Gupta, Nikhil Mehta, Shipra Agarwal¹, Neetu Bhari

Departments of Dermatology and Venereology and ¹Pathology, All India Institute of Medical Sciences, New Delhi, India

Address for correspondence: Dr. Neetu Bhari, Dermatology and Venereology, All India Institute of Medical Sciences, New Delhi, India. E-mail: drntbhari@gmail.com



is characterized by greasy, hyperkeratotic, skin-colored to hypopigmented papules predominantly in a seborrheic distribution at the scalp, face, trunk and flexures. Guttate leukoderma has been described as a rare manifestation of Darier's disease.^[2] Dermoscopic features of greasy papules in Darier's have recently been described.[3-5] Brownish areas and fine white scales on dermoscopy correspond to marked hyperkeratosis histopathologically and parakeratosis while a peripheral whitish halo correlates with marked acanthosis.^[3,6] The brownish areas have been referred as pseudocomedones by previous authors.^[7] Dermoscopy of guttate leukoderma lesions has not been reported previously. Thus, dermoscopic evaluation of greasy papules and guttate leukoderma may allow earlier diagnosis of this disease and may help in appropriate management.

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 b'e 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.

References

- 1. Takagi A, Kamijo M, Ikeda S. Darier disease. J Dermatol 2016;43:275-9.
- Harb J, Motaparthi K. Clinicopathologic findings of guttate leukoderma in Darier disease: A helpful diagnostic feature. JAAD Case Reports 2018;4:262-6.
- Lacarrubba F, Verzi AE, Errichetti E, Stinco G, Micali G. Darier disease: Dermoscopy, confocal microscopy, and histologic correlations. J Am Acad Dermatol 2015;73:e97-9.
- Errichetti E, Stinco G, Lacarrubba F, Micali G. Dermoscopy of Darier's disease. J Eur Acad Dermatol Venereol 2016;30:1392-4.
- Senhaji G, Jouari OE, Gallouj S, Lamouaffaq A, Mernissi FZ. Late onset Darier's disease: Clinical and dermoscopic features. J Dermat Cosmetol 2018;2:75-6.
- Errichetti E, De Francesco V, Pegolo E, Stinco G. Dermoscopy of Grover's disease: Variability according to histological subtype. J Dermatol 2016;43:937-9.
- Vázquez-López F, Lopez-Escobar M, Maldonado-Seral C, Perez-Oliva N, Marghoob AA. The handheld dermoscope improves the recognition of giant pseudocomedones in Darier's disease. J Am Acad Dermatol 2004;50:454-5.