A Unique Duo Of Rare Morphologic Variants Of Darier's Disease

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Citation

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Abstract

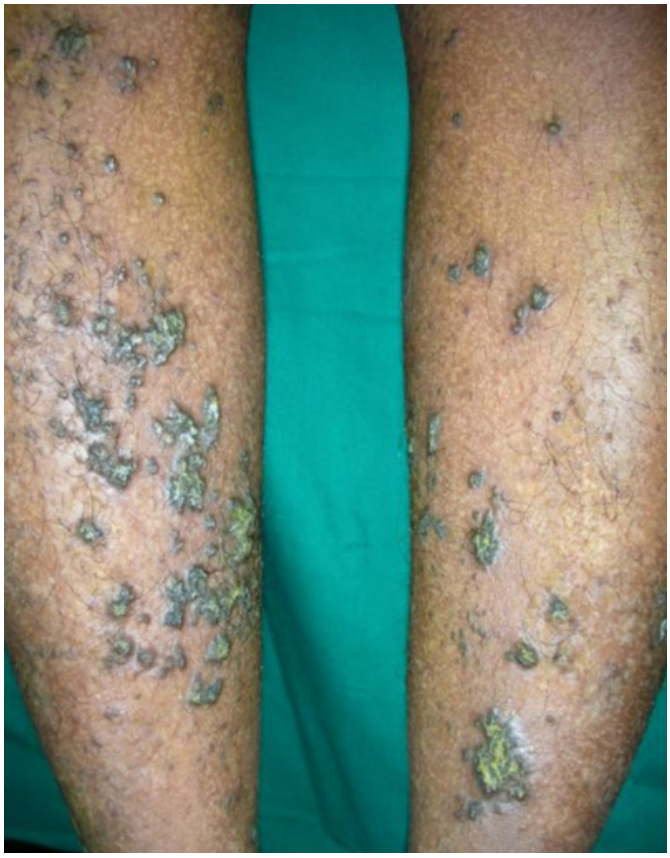
Darier's disease is an autosomal dominant disorder characterized clinically by presence of keratotic papules in a seborrheic distribution, nail involvement and mucosal lesions and histologically by the presence of focal areas of acantholysis in the suprabasal layer of epidermis, lacunae within the epidermis and an unusual dyskeratosis. We describe the case of a 38-year-old female came with asymptomatic brownish pigmented lesions over the face, chest and extremities of 20 years.

CASE REPORT

A 38-year-old female came with asymptomatic brownish pigmented lesions over the face, chest and extremities of 20 years. Initially, she noted the lesions over the face and retroauricular regions and subsequently over the chest, axillae and extremities in a span of 4 years. Other members of her family were not having similar disease. Physical examination revealed hyperpigmented keratotic papules distributed over seborrheic areas and unusually hyperkeratotic papules and plaques over the extremities, especially over the shins (Fig.1).

Figure 1

Figure 1: Cornifying as well as hypopigmented macules of Darier's disease over the legs (greenish tinge over the lesions is due to neem plant paste)



Hypopigmented macules 1-4 mm in size, discrete as well as coalescing were noted over the extremities. Dorsa of the hands showed typical dome shaped brown papules, with a few of the finger nails showing distal notching and red

longitudinal streaks. Palmar pits and typical oral mucosal lesions were also noted. Patient had been treated earlier with keratolytic agents in the past with minimal improvement. The lesions over the shins had been especially resistant to treatment. Skin biopsy from the representative keratotic lesion (shin) showed unusually prominent hyperkeratosis and papilomatosis in the epidermis, intraepidermal acantholysis, lacunae and dyskeratosis in the form of corps ronds and grains, features that were consistent with Darier's disease. Patient was put on topical salicylic acid ointment for extremities lesions and is under follow up.

Darier's disease is an autosomal dominant disorder characterized clinically by presence of keratotic papules in a seborrheic distribution, nail involvement and mucosal lesions and histologically by the presence of focal areas of acantholysis in the suprabasal layer of epidermis, lacunae within the epidermis and an unusual dyskeratosis.[1] Since 1889, when Dr James White described its typical clinical manifestations, a number of rare morphologic variants have been described including localized disease, painful cutaneous horns, isolated hemorrhagic acral lesions, hypertrophic intertriginous plaques, bullous lesions and leukodermic macules in black patients.[1,2] Clinically, the distinctive lesion is a firm, rather greasy, crusted papule that is skin-coloured or yellow–brown and the coalescence of these papules, produces irregular warty plaques or papillomatous masses, which, in the flexures, become hypertrophic, fissured and malodorous.[3] In our patient, in addition to these typical manifestations, hyperkeratotic papules and plaques were noted over the shins, which are features of the cornifying variant. Leukodermic perifollicular macules were also noted over the extremities and were seen predominantly over the legs. This unique combination of rare morphological variants viz. leukodermic macules with cornifying variant of Darier's disease has not been described earlier. Notable also in our patient was presence of other features of typical Darier's disease, which has seldom being noted in the cornifying variant.[2] Typically, these hypopigmented macules (guttate leukoderma) are localized to the trunk.[4] Originally regarded as postinflammatory hypopigmentation, these lesions were later recognized as an early or subclinical form of this acantholytic disorder and showed decreased melanin layer in the basal layer on histology.[4] Unusual feature in our patient was their localization to the legs.[4, 5]

The Darier's disease is refractory to treatment with most conventional keratolytic and anti-inflammatory agents. Topical corticosteroid/ antibiotic preparations may help itching but have no effect on the course of the disease.[1] Retinoids are of great value in treatment of Darier's disease; however the hypertrophic lesions respond poorly.[1] Erbium:YAG laser ablation effectively removes resistant Darier's disease and yields excellent long-term results in chronic, recalcitrant cases. This supercedes the earlier suggested modalities of selective surgical removal of the keratotic papules and circumscribed deeper ablation in follicular lesions, which are not feasible with dermabrasion.[6]

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