Acantholytic dyskeratotic epidermal nevus which mimic as a darier's disease

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Abstract
Darier's disease is characterized by multiple hyperkeratotic lesions and crusted papules or plaques involving seborrheic areas along with palmoplantar pits, and nail changes. Epidermal naevus may show acantholytic histology similar to darier's disease. Presenting a case of acantholytic dyskeratotic epidermal nevus mimicking darier's disease. Negative family history, no significant signs of darier's disease with limited distribution of lesions favour a nevoid origin.

Keywords: Darier's disease, Epidermal naevus, Retinoids.

Introduction
Epidermal nevi are localized malformed epidermis consisting of verrucoid scaly papules or plaques presented along the lines of Blaschko's. Rarely, epidermal naevi show acantholytic histology resembling darier's disease, a dominantly inherited skin condition characterized by widespread warty papules. The prevalence of the disease is estimated to be 1 in 50000 people. Phenotypic expression of darier's disease (DD) is variable. In 1906 the first localised pattern of DD was reported.[1] Other variants to be recognised since then are unilateral, linear, segmental or zosteriform DD.[2] Here we present a case of acantholytic dyskeratotic epidermal nevus mimicking darier's disease.

Case Report
A 31-year-old male presented with a 7-year history of skin lesions over body which were progressive in nature. Patient also had associated complaint of itching and burning over the affected sites. Patient did not give history of worsening of lesions in summer or after sun exposure. No other family member had similar illness. Patient was being treated for the same by a private doctor in the form of oral methotraxate and laser therapy before he presented to our department. Cutaneous examination showed multiple hyperpigmented, greasy, warty papules and plaques with adherent crusts and scales over both legs and feet with multiple discrete erythematous keratotic papules over palms, bilateral dorsum of hands, abdomen, back and soles (Fig. 1 a,b,c). There were no other mucosal lesions, nail changes or systemic complaints. Biopsy from the scaly papules showed hyperkeratosis, parakeratosis, moderate acanthosis, orthokeratosis with papillomatosis of epidermis and dyskeratosis (in the form of corps ronds and grains) in the epidermis (Fig. 3). Routine laboratory tests were within normal limits. We gave patient oral steroids and isotretinoin with vitamin A supplements and topical retinoids which showed marked improvement within a year (Fig. 2 a,b,c).

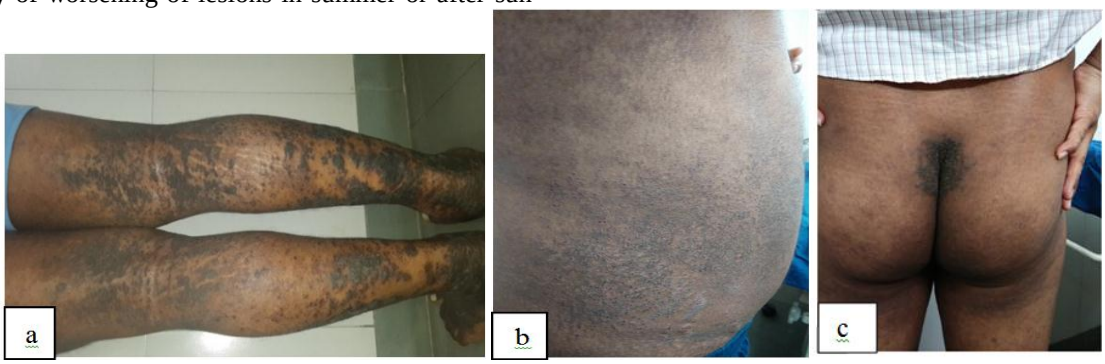

Fig.1: pre-treatment photograph (a) Multiple hyperpigmented, greasy, warty papules and plaques with adherent crusts and scales over both legs (b) abdomen (c) back

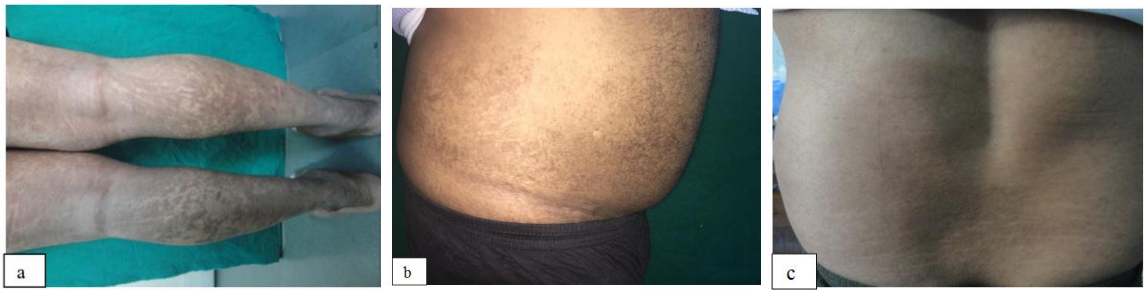

Fig. 2: Post-treatment photographs (a) legs (b) abdomen (c) back

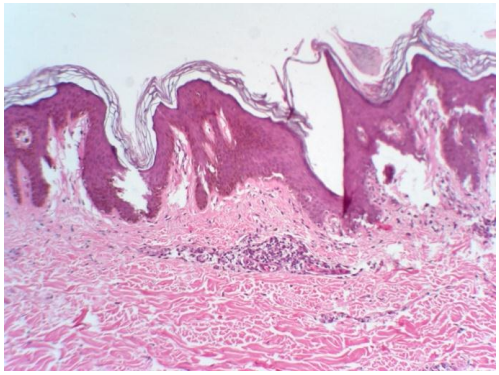

Fig. 3: Hyperkeratosis, parakeratosis, moderate acanthosis, orthokeratosis with papillomatosis of epidermis and dyskeratosis (in the form of corps ronds and grains) in the epidermis

Discussion

Darier's disease, Lutz-Darier-White disease, keratosis follicularis; is a skin disorder characterized by multiple hyperkeratotic lesions and crusted papules or plaques involving seborrheic areas along with palmoplantar pits, and nail changes.[3] The mutation in the ATP2A2 gene at chromosome 12q24.1, encoding the sarco endoplasmic reticulum calcium pumping ATPase type 2 (SERCA2) is the major responsible factor in occurrence of DD. The penetrance is complete and the expressivity is variable in adults.[4] Clinically, the disease is identified by keratotic papules or plaques mainly at the seborrheic areas with occasional nail or mucosal involvement. The disease exacerbate during summers and sun-exposure as well as during heat and humid weather. In 1906, an uncommon linear variant of this condition was first reported which lacks the classical features of Darier disease, and only shows localized lesions in a linear fashion. Usually there is no family history. Two subtypes are described, type 1 is commoner, presents along the lines of Blaschko, and is suggested to result from post zygotic somatic mutations. Type 2 being rarer, severe is mainly characterized by linear streaks and resulting from a heterozygous germline mutation including somatic loss of heterozygosity of the wildtype allele in a segmental area.[5] The localized variants classically ivolves a limited area and other significant features associated with classical DD are usually absent. The differential diagnoses of localized DD include Herpes zoster, lichen striatus, Grover's disease, lichen planus and linear nevoid disorder. Negative family history, no significant signs of darier's disease with limited distribution of lesions favour a nevoid origin.[6] Segmental (localized, linear or zosteriform) forms of DD may be clinically indistinguishable from Hailey-Hailey disease, seborrheic dermatitis and transient acantholytic dermatosis. Histologically, loss of cohesion between suprabasal epidermal cells (acantholysis) results in suprabasal clefting with papillomatosis and dyskeratosis. Systemic retinoids are the drug of choice in darier's disease. Sunscreen, cool cotton clothing, and avoidance of hot environments prevent flares. Moisturizers with urea or lactic acid can reduce scaling and hyperkeratosis. A low-or mid-potency topical steroid is useful for inflammation. Topical retinoids (adapalene, tazarotene gel 0.01%, tretinoin) can reduce hyperkeratosis. Topical 5-fluorouracil (5-FU) has been used effectively in some patients.

Conclusion

A rare case of acantholytic dyskeratotic epidermal nevus mimicking darier's disease who was given oral steroids and isotretinoin with Vitamin A supplements and topical retinoids and the improvement was found to be 80% within a year.

Reference

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