



DARIERS DISEASE MASQUERADING AS PHOTODERMATITIS : A RARE CASE REPORT

Dermatology

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ABSTRACT

Darier's disease is an autosomal dominant genodermatosis characterized by chronic eruption of keratotic papules in the seborrheic areas of the skin. The disease usually begins during childhood but can manifest at any time. Both sexes are affected with equal frequency. Here we report a case of Darier's disease mimicking photodermatitis.

KEYWORDS

Darier's disease, Genodermatosis

INTRODUCTION:

Darier's disease or Darier- white disease, also known as keratosis follicularis, is an autosomal dominantly inherited genodermatosis characterized by hyperkeratotic warty papules in seborrheic regions with nail abnormalities and mucous membrane changes.² The onset is often insidious and slowly progressive. The disease tends to flare with heat and humidity.

CASE REPORT:

A 52-year-old male came to the Dermatology OPD with diffuse pigmentation and raised skin lesions for the past 6 months. The lesions started over the axilla and progressed to involve the nape of neck, back, scalp, forearms and legs. Later he developed similar lesions on the face. It was associated with itching. History of photosensitivity present. No history of similar lesions in the past. No similar complaints in the family.

On examination multiple warty papules were present on the scalp, forehead, nape of neck. Diffuse pigmentation of face, neck and extremities. Nails and oral cavity were normal.

Due to lesions in sun-exposed areas a diagnosis of photodermatitis was considered. Skin biopsy was done and it showed hyperkeratosis with follicular plugging, few vacuolated keratinocytes, focal basal cell degeneration and increased pigmentation in upper dermis. One part of epidermis showed suprabasal separation with evidence of corps ronds and grains. These features were consistent with Darier's disease.

DISCUSSION:

Darier's disease was first described independently by Darier and White in 1889.³ It is an inherited acantholytic dermatosis caused by mutation in the ATP2A2 gene on chromosome 12.⁴ It presents as chronic keratotic papules mostly in seborrheic areas. Other lesions include distinctive nail and palmoplantar abnormalities.^{5,6}

Darier's disease is prevalent worldwide and affects males and females equally. Rash usually starts during the early teenage but can present for first time in adult as well. Special variants include hypertrophic type, vesiculobullous type and linear type.

Histologically, supra-basal acantholysis is seen which leads to suprabasal clefts and lacunae. There will be hyperkeratosis and papillomatosis. In the epidermis there are rounded dyskeratotic cells with eosinophilic cytoplasm called corps ronds and small cells with shrunken cytoplasm called grains.

The close differentials are Seborrheic dermatitis, Hailey-Hailey disease, Dowling Degos disease and Acanthosis Nigricans. Mild disease can be managed with emollients and by avoidance of heat or UV radiation. Moderate to severe types are treated with topical mild to moderate potent corticosteroids and retinoids. Oral vitamin A in

combination with vitamin E has given excellent results.

CONCLUSION:

Darier's disease with an element of photosensitivity can be easily misdiagnosed as photosensitive dermatitis and this should be kept in mind when making a diagnosis of the latter.

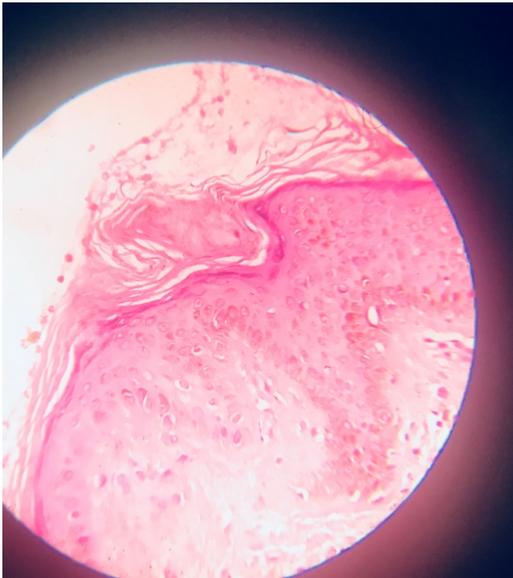
FIGURE 1-Clinical picture showing pigmented warty papules on the face, nape of neck and scalp.



FIGURE 2- Scanning view showing suprabasal detachment of epidermis at one end and hyperkeratosis along with follicular plugging on the other end



FIGURE 3- high power view showing follicular plugging along with corps ronds and grains.



ACKNOWLEDGMENT: None

CONFLICT OF INTEREST: The authors declare that they have no conflict of interest.

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