COEXISTENT BULLOUS DARIER'S DISEASE WITH XERODERMA PIGMENTOSUM

Radha Mittal and Samarjit Sandhu

A case of bullous Darier's disease had typical brown follicular papules, greasy crusted papules, discrete keratotic papules and vesiculo-bullous lesions on the dorsum of the hands and feet. In addition, characteristic lesions of xeroderma pigmentosum as extensive freckles intermingled with atrophic white spots were present on the face, trunk and limbs. This combination of bullous Darier's disease with xeroderma pigmentosum has not been reported earlier.

Key words : Darier's disease, Bullous, Xeroderma pigmentosum.

Darier's disease and xeroderma pigmentosum are two rare distinct diseases. Darier's disease is determined by autosomal dominant gene or it may manifest due to gene mutation.[1] Vesiculo-bullous lesions as a bullous variant were described by Jablonska and Chorzelski in 1958.[2] In some cases of Darier's disease, there are keratotic papules on the dorsa of hands and feet that resemble clinically those seen in acrokeratosis verruciformis of Hopf.[3] Histopathologically, these papules may show dyskeratosis and lacunae as seen in Darier's disease or changes characteristic of acrokeratosis verruciformis of Hopf.[4] Acrokeratosis verruciformis of Hopf is an independent entity as it is distinct both genetically and histopathologically from Darier's disease, suprabasal clefting being found only in the Darier type papules.[3,5]

Case Report

A 20-year male had photosensitivity, giddiness and weakness on exposure to sunlight for the last 6 years. Five years back, the patient developed greasy, crusted, follicular papules and scales on the scalp and a few crusted, yellow-brown papules on the seborrhoeic sites such as axillae, groins, naso-labial folds, ears, retro-auricular regions, buttocks, natal cleft and lumbo-sacral region. Simultaneously, he also developed discrete, scaly, 1 to 1.5 cm diameter papules on the dorsum of hands and feet and 1 to 3 mm freckles on the face which spread to the upper limbs, lower limbs and trunk. Four-and-a-half years back, atrophic white spots also appeared. The disease spread more rapidly during summers when vesicles and bullae with secondary infection also appeared mainly on the dorsa of hands and feet. Skin of the palms and soles was thick and waxy. One 2.5 cm diameter cafe au lait spot was present near the umbilicus. The face and neck had diffuse pigmentation. Both eyes had pterygium and left eye had one bitot spot. Hair and nails were normal.

Biopsy from a freckle on the face revealed hyperkeratosis, melanin accumulation in the basal cell layer and upper dermis and mononuclear infiltrate in the dermis. Second biopsy from a follicular papule in the axilla revealed hyperkeratosis, papillomatosis, numerous lacunae with acantholytic cells, corps ronds and grains. Dermis showed oedema and mononuclear cell infiltrate (Fig. 1). Third biopsy from a papule on the hand showed marked hyperkeratosis, grains, irregular hypergranulosis, acanthosis, lacunae, dyskeratosis, corps ronds, mid-epidermal and suprabasilar cleft. Suprabasal cleft, lacunae, corps ronds and dyskeratosis were seen under high power in the section

From the Departments of Dermatology and Pathology, Government Medical College and Rajendra Hospital, Patiala-147 001.

Address correspondence to : Dr R R Mittal.

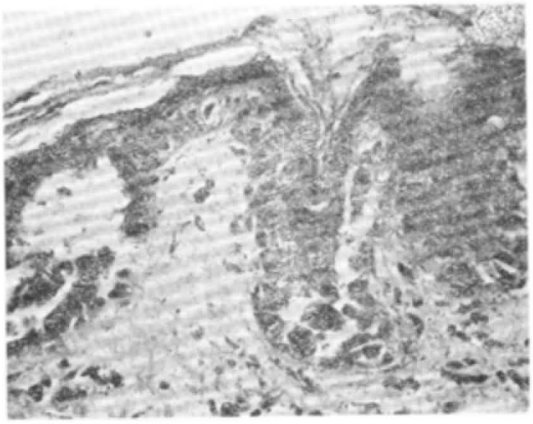

Fig. 1. Hyperkeratosis, lacunae with acantholytic cells, corps ronds, grains and dermal cellular infiltrate (Hematoxylin and Eosin stain × 200).

from the third biopsy. Thus, the first biopsy was consistent with xeroderma pigmentosum, second with Darier's disease and third showed typical histopathological changes of bullous Darier's disease.

Comments

A possible relationship between acrokeratosis verruciformis and Darier's disease has been repeatedly discussed. Typical histopathological changes of Darier's disease in keratotic papules clinically resembling acrokeratosis verruciformis of Hopf show that these are manifestations of Darier's disease itself as already discussed by Panja.[3] Onset of Darier's disease mostly occurs between the ages of 10 to 20 years and our patient developed it at the age of 15 years. Minor seasonal fluctuations and spontaneous remissions can occur and our patient noticed aggravations during summers with appearance of vesiculo-bullous lesions and secondary infection. It also progressed more rapidly in summers.

Xeroderma pigmentosum is a group of autosomal recessive disorders due to DNA repair defects that cause photosensitivity. Heterozygotes may show freckling.[6] Photosensitivity and susceptibility to infection is seen both in Darier's disease and xeroderma pigmentosum. Family history was negative for both Darier's disease and xeroderma pigmentosum and this combination in our patient can be due to gene mutation.

References

1. Lever WF and Schaumburg-Lever G : Congenital diseases (Genodermatoses), in : Histopathology of Skin, Sixth ed, JB Lippincott Company, Philadelphia, 1983; p 70.
2. Jablonska S and Chorzelski T : Zur Klassfikation des Pemphigus Hailey-Hailey, Dermatologica, 1958; 117 : 24-38.
3. Panja RK : Acrokeratosis verruciformis (Hopf) A clinical entity, Brit J Dermatol, 1977; 96 : 643-652.
4. Wisman M : Verruciform manifestations of keratosis follicularis, Arch Dermatol, 1960; 81 : 1-14.
5. Ebling FJG, Marks R and Rook A : Disorders of keratinization, in : Textbook of Dermatology, Fourth ed, Editors, Rook A, Wilkinson DS, Ebling FJG et al : Oxford University Press, Bombay, 1987; p 1440.
6. Burton JL and Rook A : Genetics in dermatology, in : Textbook of Dermatology, Fourth ed, Editors, Rook A, Wilkinson DS, Ebling FJG et al : Oxford University Press, Bombay, 1987; p 144.