UNILATERAL DARIER'S DISEASE

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Various variations of Darier's disease have been mentioned in the literature. Here we describe a young male with unilateral involvement with clinical and histopathological features typical of Darier's disease.

Key Words : Follicular papules, Keratosis follicularis, Palmar pits

Introduction

Darier's disease is a disorder of keratinization determined by an autosomal dominant gene of variable penetrance or occurring as new mutation. It is characterized by persistent eruption of hyperkeratotic papules particularly in the seborrhoeic distribution.[1] The lesions in the flexures are likely to become hypertrophic and malodorous.[2] Various other variations such as vesicobullous and linear or zosteriform types have been described in the literature. Minute pits when present on palms and/or soles are considered to be pathognomonic of Darier's disease.[3] However, there is great variablity in the extent of involvement ranging from Darier's disease of nails only to generalized involvement of the whole body.

Case Report

A 16-year-old male presented to us with itchy, erythematous papules and dry, hyperkeratotic plaques which were present in the left axilla for the last 10 years. The lesions had started when has was about 6 years old and were non-itchy and discrete to begin with. However, they slowly progressed in size and number and then coalesced to give rise to the larger hyperkeratotic plaques (Fig. 1). For the last 5 years, the left half of the trunk, neck and left upper limb also developed similar small, skin-coloured and erythematous, greasy, crusted papules which turned out to be follicular on close examination. These follicular papules were in patches on the trunk and neck but in peculiar broad, linear spiralling bands on the left upper limb starting from the wrist and going up to the axilla and shoulder.

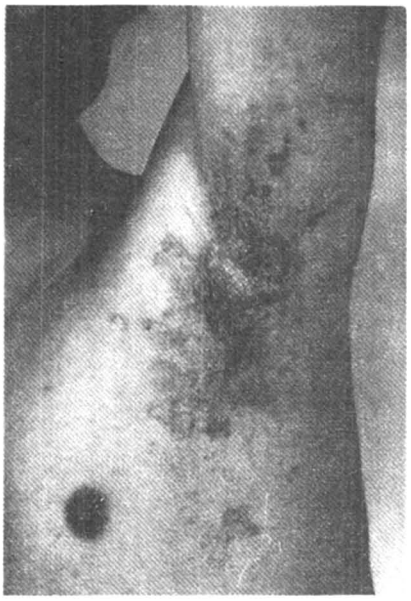

Fig. 1. Hypertrophic lesions of Darier's disease in the axilla.

In addition, the left palm and the ventral aspect of the third and fourth fingers showed punctate and plaque-like keratoses with overlying as well as separate minute pits. There was no pigmentary disturbance and the

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examination of the rest of the body, hair, nails and mucous membranes did not reveal any abnormality.

There was a history of worsening of the lesions and the itch in hot and humid weather. There was no associated systemic complaint or history of bullous eruption or any significant family history. The blood counts, sugar, urea, creatinine and electrolyte levels were all within normal ranges.

The histopathological examination of the biopsy specimen taken from axilla revealed hyperketatosis, acanthosis, superabasal clefting with acantholysis and chronic inflammatory infiltrate in the dermis together with downgrowth of epidermal cells, thus supporting the diagnosis of Darier's disease.

Discussion

The early onset of the disease, the typical follicular crusted papules, characteristic keratoses on the palms and the pathognomonic minute pits gave a strong clinical suggestion of Darier's disease which was supported by the histopathological findings. This conglomeration of features is enough to differentiate unilateral Darier's disease from acantholytic dyskeratotic epidermal naevi with which it could have been confused as there was no family history.[4] To our knowledge, this is the first authentic report of unilateral Darier's disease from India though it has been reported from other countries.

References

1. Phillips SB, Baden HP. Darier-White disease and miscellaneous hyperkeratotic disorders. In: Fitzpatrick TB, Eisen AZ, Wolff K, et al, editors. Dermatology in general medicine. New York : McGraw Hill, 1993: 547-52.
2. Elsbach EM, Nater JP. The hypertrophic forms of Darier's disease. Dermatologica 1960; 120: 93-4.
3. Griffiths WAD, Leigh IM, Marks R. Disorders of keratinization. In : Champion RH, Burton JL, Ebling FJG, editors. Textbook of dermatology. London : Blackwell, 1992: 1325-90.
4. Leeming JAL. Acquired linear naevus showing histologic features of keratosis follicularis. Br J Dermatol 1969; 81 : 128-31.