UNILATERAL DARIER'S DISEASE

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Unilateral Darier's disease was observed in an adult male involving one half of the body.

Key words : Darier's disease, Unilateral.

Darier's disease is a rare familial disease.[1] Although it is usually determined by an autosomal dominant gene, it may be the result of a mutation.[2] Its clinical variants include the hypertrophic, vesicular and the acute eruptive types.[3] Lesions occurring in a linear or a zosteriform pattern and which may involve one half of the body are noticed in 10% of the cases.[3] Still this manifestation is rare.

Case Report

Ten years ago, a 32-year-old man developed mildly itchy, papular lesions over the dorsum of the left foot which gradually progressed upwards and involved the left half of the body in a span of 5 years. Family history of similar skin lesions was absent. He was married and his two children were asymptomatic. He had numerous, closely grouped, hyperpigmented, keratotic, follicular and extra-follicular papules involving the left half of the body (Figs. 1 and 2), except the palm and sole, mucous membranes, genitalia, nails, scalp and face. On the trunk, the papules had a tendency for grouping at many places with a dermatomal distribution, while linear arrangement was the feature over the left forearm and left lower limb. A few lesions were also present over the back of left pinna. Multiple, discrete, 2-3 mm sized hypopigmented macules were present over the left half of the chest intermingled with the papules. Systemic examination was normal. Routine blood and urine analysis were within normal limits. Biopsy of one of the representative lesions showed typical histopathology of Darier's disease (Fig. 3).

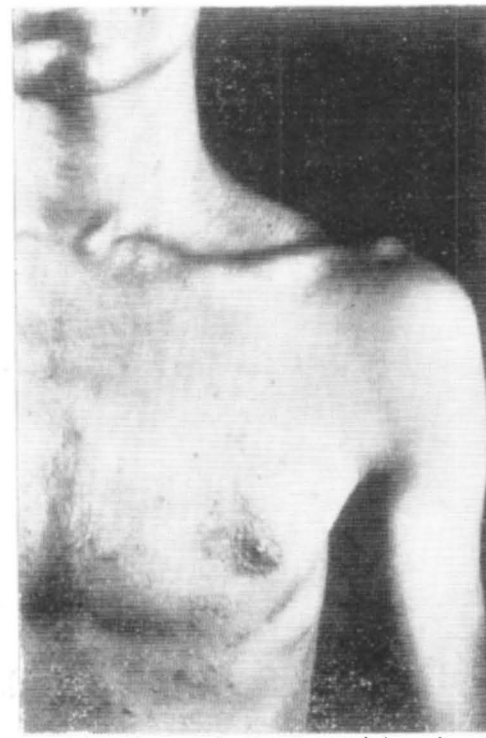

Fig. 1. Hypopigmented macules and hyperkeratotic papules over the front of chest confined to the left half of the body.

Comments

Unlike the classical type of Darier's disease, the unilateral variety is thought to be of non-genetic origin with a delayed onset.[4] A review of literature revealed that since 1933, only 32 cases of unilateral Darier's disease have been reported and of these the majority of cases had the onset in adult life.[5] These observations are consistent with the findings in our patient.

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Fig. 2. Zosteriform distribution of lesions on the back.

Fig. 3. Characteristic histopathology with a suprabasal cleft, a few acantholytic cells, villi and corps ronds (H & E x 189).

Cases of unilateral Darier's disease with both extensive and multiple lesions,[5-12] as well as those with localised lesions[5,13] have been described. The clinical presentation similar to our case has also been observed by Snyder[10] and Hesbacher.[11] The peculiar hypopigmented macules noted in our patient have also been observed by others.[3,15,16]

The histopathological findings of Darier's disease are also seen in warty dyskeratoma and rarely in epidermal naevi.[4] According to Ackerman,[14] actinic keratosis, transient acantholytic dermatosis and dermatofibroma can also exhibit focal histopathological changes of acantholytic dyskeratosis. Cases clinically typical of epidermal naevi but showing the histopathological picture of Darier's disease have been reported.[4,6] In the opinion of Demetree[4] some of the cases reported in the past as unilateral Darier's disease could have actually been epidermal naevi and he feels that as more cases of epidermal naevi undergo biopsy, the entity he labelled as unilateral, linear, zosteriform epidermal naevi with acantholytic dyskeratosis, will be better recognised. This view is supported by Starink and Woerdemann[5] who proposed the term acantholytic dyskeratotic epidermal naevus rather than unilateral Darier's disease to the 7 cases studied by them. The lack of family history, absence of other signs of Darier's disease and the peculiar distribution pattern of the lesions favours a naevoid origin. At the same time, the delayed onset of the lesions is not consistent with epidermal naevi which usually manifest at birth. In the light of these observations, classification of these cases as a variant of Darier's disease or as epidermal naevi is still a point of controversy. However, we consider the present case to be one of Darier's disease because of the delayed age of onset,

itching, typical lesions and the characteristic histopathology.

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