VESICULAR DARIER'S DISEASE: Report of a case occurring in a linear distribution

M. Z. MANI * G. PHILIPS † M. MATHEW ‡

Summary

A case believed to be linear vesicular Darier's disease occurring in a 5 year old male is reported. Features which suggested vesicular Darier's disease in preference to Benign familial chronic pemphigus (B. F. C. P.) were the linear distribution, the absence of a family history and also the pronounced dyskeratosis seen histologically.

Darier's disease or keratosis follicularis is a rare but well known heriditary disorder of keratinization. Localized linear and zosteriform variants of Darier's disease are still rarer[1-6]. The lesions in the localized form of Darier's disease may mimic naevus unius lateris[2,6] or lichen striatus[5]. In view of this clinical resemblance, Zelligman and Pomeranz[7] have emphasized that all linear or zosteriform verrucous lesions should be examined histologically.

Darier's disease is also known to occur in a vesicular form[8,9]. Controversy has raged over the relationship of vesicular Darier's disease to benign familial chronic pemphigus (Hailey-Hailey). Some authors[8,9,10] are of the opinion that benign familial chronic pemphigus (B. F. C. P.) and vesicular Darier's disease are the same condition. Points advanced in favour of this view are:

1. The occurrence of both diseases in the same patient[9,10,11].
2. The fact that "corps ronds", which are due to dyskeratosis, have also been observed in B.F.C.P.[12]

On the other hand, points suggesting that these two diseases are different entities are:—

1. On more critical analysis it has been seen that the histological features of vesicular Darier's disease are different from those of B.F.C.P.[13]
2. Genetic studies have shown independent inheritance for the two diseases, without overlap[14].

The controversy has not yet been settled. Lever[15] has stated that until more is known about the aetiology of the two diseases it may be best to regard Darier's disease and B.F.C.P. as independent entities.

Even though several cases of vesicular Darier's disease have been reported[8,9] to our knowledge, no case of

* Lecturer, Dermatology Section, Department of Medicine.
† House Physician, Dermatology Section, Department of Medicine.
‡ Professor and Head, Department of Medicine.
Christian Medical College and Brown Memorial Hospital, Ludhiana 141008 Punjab, India.

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vesicular Darier's disease occurring in a linear distribution has been reported so far.

Case Report

A five year old male was brought to the dermatology clinic of our hospital on 11th October 1977. He had been developing recurrent episodes of itchy vesicular lesions on the right upper extremity and the right scapular region for four years prior to the hospital visit. There was no history of lesions on other sites. The lesions used to become erythematous and later there would be pus discharge. Each episode used to improve with systemic antibiotics and various local applications, but the disease used to relapse frequently and was always aggravated in summer.

Nobody in the patient's family was suffering from a similar disease. The general physical examination and systemic examination did not reveal any abnormality. The patient was apparently of normal intelligence.

On cutaneous examination, the lesions were seen to be confined to the right upper extremity and the right scapular region. Four plaques were present in a linear configuration and of these one plaque was on the dorsum of the hand, two were on the extensor aspect of the forearm and one plaque was present on the right scapular region. The plaques were approximately 2–3 cm in width, were erythematous in colour and were covered with crusts and scales. A seropurulent discharge was present and some pustules were also seen on the plaques. The plaque on the right scapular region was less crusted than the others and consisted of hyperkeratotic non-follicular papules.

Hypopigmented, non atrophic macules were seen extending in a linear distribution along the right upper extremity, in intervening areas of the skin which were not covered by the plaques. They preserved the continuity of the linear distribution. These hypopigmented macules presumably represented previously healed lesions.

There were no lesions elsewhere on the skin, mucosa, palms or soles. Hair and nails were normal. A skin biopsy was performed from a plaque on the left forearm and also from the plaque on the left scapular region. The patient was sent home on wet dressings and local application of a cream containing betamethasone and neomycin. He returned after two weeks of treatment with marked improvement. The plaques showed some residual erythema and crusting. There was a prominent area of diffuse hypopigmentation surrounding the plaques, at the sites where the plaques had healed (Fig. 1). Biopsies were taken from the forearm and also the scapular region.

Biopsy Report :

Both sections show suprabasal clefts, villi formation shrivelled, dyskeratotic acantholytic cells, "corps ronds" and few "grains" (Fig. 2a). The section from the biopsy of the plaque on the right

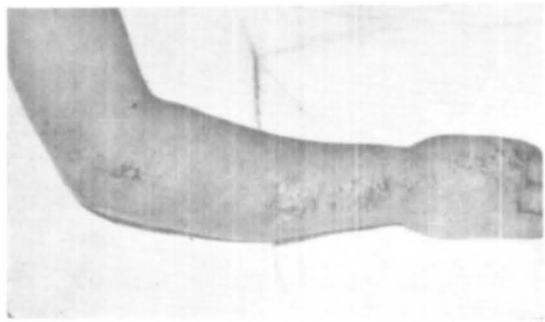

Fig. 1 Clinical picture after use of steroid-antibiotic combination. Three partially healed plaques are seen on the hand and forearm. The plaques consist of erythema and crusting. Hypopigmented macules are seen around and in between the plaques.

forearm shows in addition, an intra-epidermal vesicle, by the side of which there is the dilated ostium of a sweat duct filled with keratin (Fig. 2b).

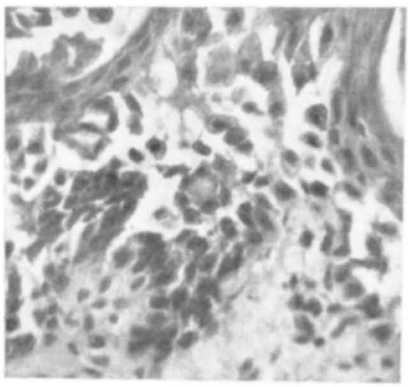

Fig. 2A Biopsy from scapular region (x 400) The suprabasal clefts and the shrivelled dyskeratotic acantholytic cells are well seen. Some of the acantholytic cells appear like "grains".

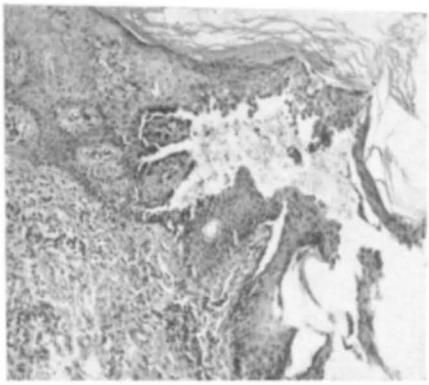

Fig. 2B Biopsy from the forearm. There is hyperkeratosis and an intraepidermal bulla containing acantholytic cells and RBC'c. 2 "villi" are seen on the lefi. "Corps ronds" are present within the superficial part of the stratum spinosum. A dilated ostium of sweat duct is seen above the bulla.

Impression :

The features are suggestive of Darier's disease.

Discussion

Localized Darier's disease presenting in a linear or zosteriform configuration has only rarely been reported[1-6]. In most of these cases, the lesions have consisted of hyperkeratotic papules and in two cases they resembled epidermal naevus[2,6]. So far, only one case has been reported in which the biopsy showed an intraepidermal vesicle[3]. However, even in this case, the clinical features did not suggest the vesicular type of Darier's disease. The lesions consisted of umbilicated papulo-nodules and no material could be obtained on incision with a scalpel.

In our patient, even though there was a clinical resemblance to the lesions of benign familial chronic pemphigus (B.F.C.P.) in our opinion the latter condition was ruled out histologically. There were marked dyskeratotic changes in several of the acantholytic cells, many of which were elongated, and some of them looked like "grains". Acantholysis was not very extensive and there were many "corps ronds" present in the superficial part of the stratum spinosum and also in the stratum granulosum.

Atypical forms of B.F.C.P. have been described[16], but none of these cases had lesions in a linear or zosteriform distribution. In our patient there was linear configuration of the lesion which have already been described in Darier's disease. Absence of a family history in our case could also be interpreted in favour of a localized form of Darier's disease[4].

The relapsing and remitting course, the favourable response to antibiotics and the aggravation of the disease in summer are features that may be interpreted in favour of either vesicular Darier's disease[8,9] or of B.F.C.P.[18]

Noteworthy features in our patient were the occurrence of the vesicular

form of Darier's disease in a linear distribution and the absence of any associated zosteriform lesions. Another feature was the presence of the hypopigmented macules situated around and between the plaques and which helped to complete the linear configuration of the whole lesion. They probably represented sites of previously healed lesions.

The very early age of onset at one year is intriguing. The lesions of the ordinary type of Darier's disease usually appear between the ages of 8 and 16 years[17]. The localized form of Darier's disease usually appears at an even later age[1-6]. In the case of B.F.C.P. the age of onset is usually in adolescence or early adult life[18].

We would say in conclusion that it may be open to question whether the case we have reported was a linear form of vesicular Darier's disease or of B.F.C.P. However, due to the reasons we have already enumerated, we are of the opinion that this was a case of localized vesicular Darier's disease.

References:

1. Robinson HM : Keratosis Follicularis. Report of a case with linear arrangement of lesions limited to the Rt. lower extremity, Arch Dermatol, 62 : 137, 1950.
2. Snyder W : Unilateral Keratosis Follicularis. Report of a case clinically resembling Nevus Unius Lateris, Arch Dermatol, 78 : 95, 1958.
3. Kellum RE and Haserick TR : Localized linear keratosis follicularis. Response to intralesional Vit-A and simultaneous occurrence of warty dyskeratoma, Arch Dermatol, 86 : 450, 1962.
4. Leeming JAL : Acquired linear nevus showing histolcgical features of keratosis follicularis, Br J Dermatol, 81 : 128, 1969.
5. Hesbacher EN : Zosteriform keratosis follicularis treated topically with Tretinoin, Arch Dermatol, 102 : 209, 1970.
6. Anderson NP : Darier's Disease (Unilateral), Arch Dermatol Syph, 58 : 581, 1948.
7. Zeligman I and Pomeranz J : Variations of congenital ichthyosiform erythroderma, Arch Dermatol, 91 : 120, 1965.
8. Ellis FA : Vesicular Darier's disease (so called benign familial pemphigus), Arch Dermatol Syph, 61 : 715, 1950.
9. Finnerud CW and Szymanski FJ : Chronic benign familial pemphigus, a possible vesicular variant of keratosis follicularis, Arch Dermatol Syph, 61 : 737, 1950.
10. Niordson AM and Sylvest B : Bullous dyskeratosis follicularis and acrokeratosis verruciformis, Arch Dermatol, 92 : 166, 1965.
11. Canor S and Sagher F : Keratosis follicularis (Darier) and familial benign chronic pemphigus (Hailey-Hailey) in the same patient, Brit J Dermatol, 77 : 24, 1965.
12. Herzberg JJ : Pemphigus Gongerot/Hailey-Hailey, Arch Klin Exp Derm, 202 : 21, 1955.
13. Pierard J et al : Archs Belg Derm Syph, 24 : 381, 1968.
14. Raaschou-Nielsen W and Reymann F : Acta Derm Venereol, 39 : 280, 1959.
15. Lever WF : Familial Benign Pemphigus, Dermatology in General Medicine, Edited by Fitzpatrick TB, Mc Graw-Hill, New York, 1971, p 661.
16. Lyles TW, Knox JM and Richardson JB : Atypical features in familial benign chronic pemphigus, Arch Dermatol, 78 : 446, 1958.
17. Ebling FJ and Rook A : Darier's Disease, Textbook of Dermatology, Vol. 2, 2nd Edition, Edited by Rook A, Wilkinson DS and Ebling, Blackwell Scientific Publications, Oxford, 1972, p 1170.
18. Sneddon IB : Benign Familial Chronic Pemphigus, Textbook of Dermatology, Vol. 2, 2nd Edition, Edited by Rook A, Wilkinson DS aad Ebling FJ (Eds), Blackwell Scientific Publications, Oxford, 1972, p 1304.