A Localized Form of Darier's Disease

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We report the case of a 33-year-old woman who presented with skin manifestations that resulted in a differential diagnosis of either an epidermal nevus or a localized form of Darier's disease. We discuss the arguments for and against each possibility. The available data on localized forms of Darier's disease are also discussed.

Darier's disease was described simultaneously by Darier and White in 1989. Since then, many cases that some consider to be a localized form of the disease have been published. In a review, Starink[1] suggested that these forms could be epidermal nevi with peculiar characteristics. Recently, efforts have been directed at determining the gene responsible for the disease as well as clarifying the mechanism through which localized forms are produced. We report a case in which both differential diagnoses are considered.

Case Report

A 33-year-old woman presented with an increased number and extent of lesions that had appeared in the cervical region some 20 years previously. She reported that they initially affected her right shoulder and the right side of her neck; recently, they had spread to her back. There was no previous personal or family background of interest. She stated that the only symptoms were occasional pruritus and a possible increase in the lesions following exposure to sunlight.

Examination showed dispersed papulokeratotic lesions located at the base of her neck and right shoulder (Figure 1). On the lumbar region of her back, three or four papules with similar characteristics arranged along the midline were noted (Figure 2).

Differential diagnoses included Darier's disease or epidermal nevus. Biopsy specimens from both lesions showed acantholytic dyskeratosis, with the presence of *corps ronds* and grains (Figures 3 and 4).

Topical treatment was initiated with 0.025% tretinoin, leading to a discrete improvement.

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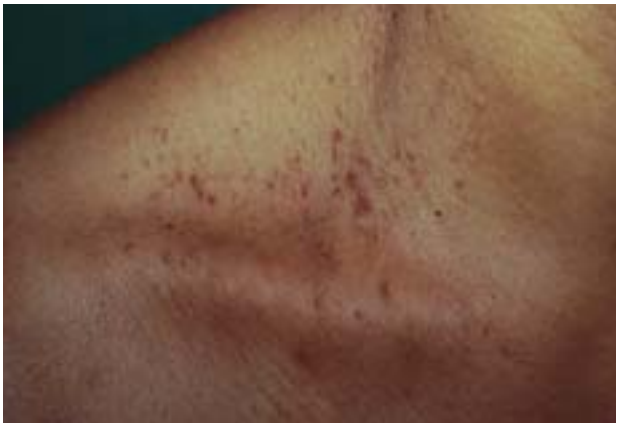

FIGURE 1. Disperse keratotic papules on the right shoulder.

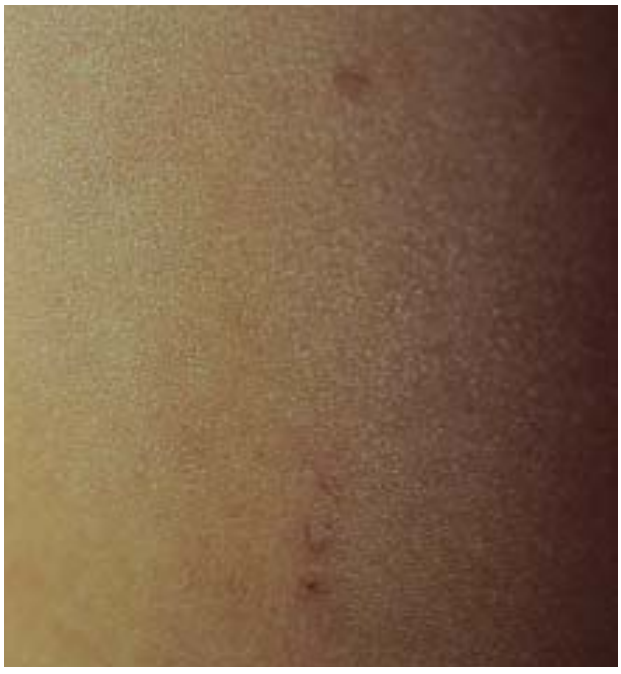

FIGURE 2. Linearly arranged keratotic papules on the mid-zone of the back.

Comments

Darier's disease is well-characterized clinically, but the histologic findings are not specific.[2] The first description of a localized form of Darier's disease was offered by Kreibich[3] in 1906. In these cases, the lesions follow

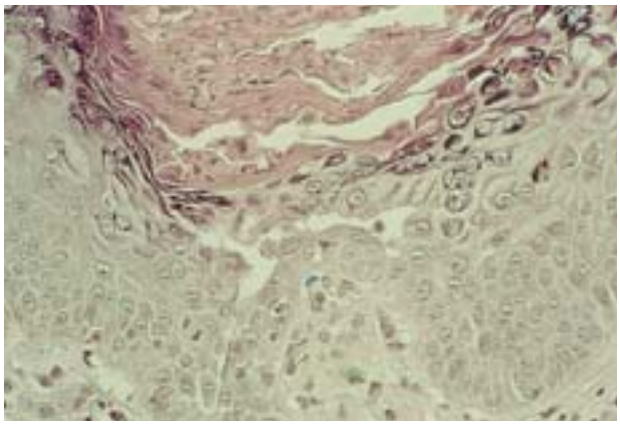

FIGURE 3. Biopsy shows hyperkeratosis with dyskeratosis and acantholytic clefts (H&E; original magnification, X 100).

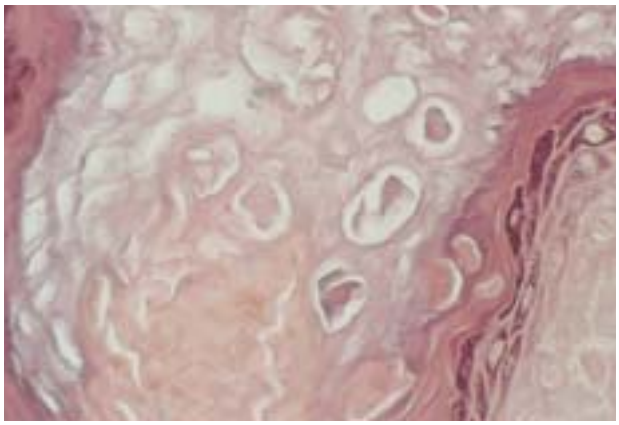

FIGURE 4. Biopsy shows corps ronds and grains (H&E; original magnification, X 400).

a linear pattern, similar to herpes zoster or the lines of Blaschko.[4] The affected area varies and a certain relationship is seen between the extent of the lesions and the involvement of other structures, such as the nails.[5]

There is still controversy about what some consider to be localized forms of Darier's disease and other special forms of epidermal nevus. The second diagnosis is supported by the absence of a hereditary pattern; however, in published cases of generalized Darier's disease, the possibility of undiagnosed localized forms in progenitors has not been ruled out, and neither has the follow-up of localized forms in descendants been specified. The appearance of the lesions at such a late age (onset occurred when the patient was 13) also supports the first diagnosis. There are also intermediate forms with uni- or bilateral extracutaneous involvement.[6-8]

In the cases published, both exposure to sunlight and sweating are cited as aggravating agents.[9] Forms associated with lymphomas[10] or triggered by psoralen/ultraviolet A radiation therapy have been described.[11] Response to treatment with vitamin A derivatives is satisfactory.

In view of the absence of a previous personal history of the disease, the present case suggested a differential diagnosis with epidermal nevus, but the clinical evolution and accompanying symptoms rather incline us toward a localized form of Darier's disease, in this case without extracutaneous affectation.

Darier's disease is a genetic disease with varying expression. According to recent studies, the gene responsible for the disease is located on chromosome 12, 12q23-24.[12] The possible mechanism of production of localized forms would be the presence of mosaics. The interest in determining whether these forms are accompanied by gonadal affectation would be due to the possibility of determining the degree of affectation of the descendants.

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