Acral Hemorrhagic Darier's Disease with Spontaneous Regression over time: A Case Report

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Abstract

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Title page

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"consent statement"

Written informed consent was obtained from the patient to publish this report in concordance with the journal's patient consent policy.

Acral Hemorrhagic Darier's Disease with Spontaneous Regression over time: A Case Report

Abstract:

we reported a-24-year-old woman, who was a known case of Darier Disease (DD) since the age of 5 years old. She was presented with hemorrhagic macules, and vesicles on the dorsum aspects of her hands, fingers, and palmar area with no triggers like trauma, and with spontaneous recovery.

KEYWORDS

darier disease, extremity, hemorrhagic skin macules

Introduction

Darier-White disease, also known as dyskeratosis follicularis or keratosis follicularis, is a rare autosomal dominant disorder that was firstly described by Darier and White about 132 years ago.[1]

The cause is a gene that encodes the sarco/endoplasmic reticulom Ca2+ ATP ase type 2, which causes calcium dyshomeostasis and endoplasmic reticulum stress including intercellular adhesion.[2,3,4]

Its incidence is 1 to 9 per 100,000 individuals. The disease usually begins before the third decade of life with a peak in puberty (11-15 years old) [5,6] ;due to sweating ,sebum exertion or change in bacterial flora.[7]

Diagnosis is confirmed by skin biopsy. Two prominent histologic findings in DD are supranasal acantholysis and dyskeratosis (corps ronds and grain) with hyperkeratosis. [8]

Classis DD is characterized by malodor keratotic, warty crusted papules and plaques, which are usually located in seborrheic areas, variable nail changes, whitish oral macules and papules(oral cobblestoning),and palmoplanta puctate pits that typically worsen in summer.[2,4]

Treatments for DD include:1- topical treatments such as:(retinoids, 5-fu, diclofenac, tacrolimus, tacalcitol, and steroids),2-oral treatments such as:(retinoids, cyclosporine, doxycycline, magnesium chloride, and naltrexone),3-physical approach such as:[dermabrasion, electrosurgical excision, surgical excision, lasers (Co2,pulse dye, Erbium),and radiotherapy].[9]

Acral hemorrhagic DD is a rare subtype with hemorrhagic macules and vesicles in extremities. Microscopic description showed suprabasal acantholytic cleft with intraepidermal hemorrhage above the cleft.[2] To date, fewer than 30 cases of acral hemorrhagic DD have been reported worldwide.

We reported the case of patient with DD who developed hemorrhagic macules and vesicles, with no symptoms that recovered spontaneously.

Case presentation:

A 24-year-old woman who was a known case of DD since the age of 5 years old (confirmed by skin biopsy from 15 years ago), came to our clinic with hemorrhagic macules and vesicles on her hands, fingers, and right palm (figure.1). Her symptoms worsened in summer. The lesions were completely asymptotic and their occurrence was not associated with trauma. Physical examination revealed hemorrhagic macules and vesicles on dorsum aspects of her hands and the palms (figure.2), as well as hyperkeratotic papules and plaques on face, neck, abdomen, and both forearms. No lesions were found on the mocuse. Moreover, the nails were completely asymptomatic and without any change.

Laboratory findings, including complete blood count(CBC), platelet count, international normalized ratio(INR), Prothrombin Time(PT), Partial Thromboplastin Time (PTT),high sensitivity C-reactive protein(CRP), antinuclear antibody(ANA), creatinine, glomerular filtration rate(GFR), and liver function tests(LFT), revealed no abnormalities in regard to her status.

The patient received several treatments, including oral corticosteroids, oral retinoids, and topical steroids and retinoids, during the course of her disease. Long-term use of oral corticosteroids has led to osteoporosis in the patient and on the other hand, she was resistant to oral retinoids and topical treatments. Therefore, she did not want to use topical and oral medicine. After 3 months, the hemorrhagic lesions spontaneously regressed without any treatment. Unfortunately, the patient was not satisfied with the skin biopsy.

Discussion:

Acral hemorrhagic DD is an uncommon skin disorder, which was firstly introduced by Jones et al. in 1964. They have presented 4 cases (including 3 women and one man), all of whom had hemorrhagic macules, vesicles, and papules on the dorsal aspects of their hands, palms (thenar and hypothenar), and feet. The lesions caused by trauma, were initially red and turned black over time. [1]Twenty-five years later, Coulson et al. have reported a 43-year-old woman with a diagnosis of acral hemorrhagic DD, which initially presented with red-purple blisters. Within a week, the blisters transformed into brownish-black macules, and finally disappeared in less than three weeks. Warm sunny weather and trauma were precipitating factors for causing hemorrhagic lesions.[10] A case with retinoid-induced hemorrhagic DD was reported for the first time by Gebauer et al., who was a-47-year-old woman treated with etretinate for approximately 10 years, and developed painless hemorrhagic bullae on the dorsal aspect of the hands without any association with trauma.[11] As well, in 2014, Zavattaro et al. have reported an 84-year-old woman suffering from hemorrhagic lesions on the palmoplantar surfaces due to acitretin consumption.[5]Burge and Wilkinson have also reported 9 of 163 patients (6%), Foresman et al. have reported 5 of 34 patients with darier diseases presented with hemorrhagic macules on palmoplantar areas.[6 ,12]Moreover, Jorg et al. have reported a76-year-old female patient, who also had acral hemorrhagic DD ,which presented in the summer.[13] Sanchez et al. have presented a 48-year-old man who was a new case of DD with developed blisters and small hemorrhagic black macules on the back of his hands, during the course of disease. The lesions improved relatively with the use of 5-fluorouracil 1% cream, and topical tazarotene .[14]Regazzini et al. have described three patients in two generations with acral hemorrhagic DD.[15]Vender et al. have presented a 65-year-old woman, with isolated acral hemorrhagic Darier's disease, nail changes, and bilateral keratotic papules on the dorsum and palms of her hands.[2] Flores-Terry et al. have reported three new cases of acral hemorrhagic DD. Accordingly, in two of them, summer and trauma were the triggering factors, but in one case, the lesions got worse in winter.[3]

In this study, our patient was a 24-year-old woman, known case of darier disease, without any positive family history, who developed hemorrhagic macules and vesicles on her hands, fingers, and right palm since last winter, which worsened in summer and regressed spontaneously in three months with no treatment. Our patient, like most reported patients, was female. Her lesions were similar to most previous cases, completely asymptomatic. Although her lesions started in the winter, they worsened in the summer, like most cases. No association was found between trauma and the incidence of these lesions. The patient has used no acitretin and retinoids for the past 7 years, and has taken retinoids for about two years before that, so her lesions were unrelated to retinoids. She refused to receive any type of treatment and the hemorrhagic lesions spontaneously regressed during three months.

In conclusion, we reported a rare variant of Darier-White disease, in order to increase cognition and awareness about this rare entity and favor early diagnosis, given the characteristic dermoscopic and clinical pictures presented by this rare variant of DD. We also showed spontaneous regression of this rare disease without any treatment.

Author contribution:

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Figures:

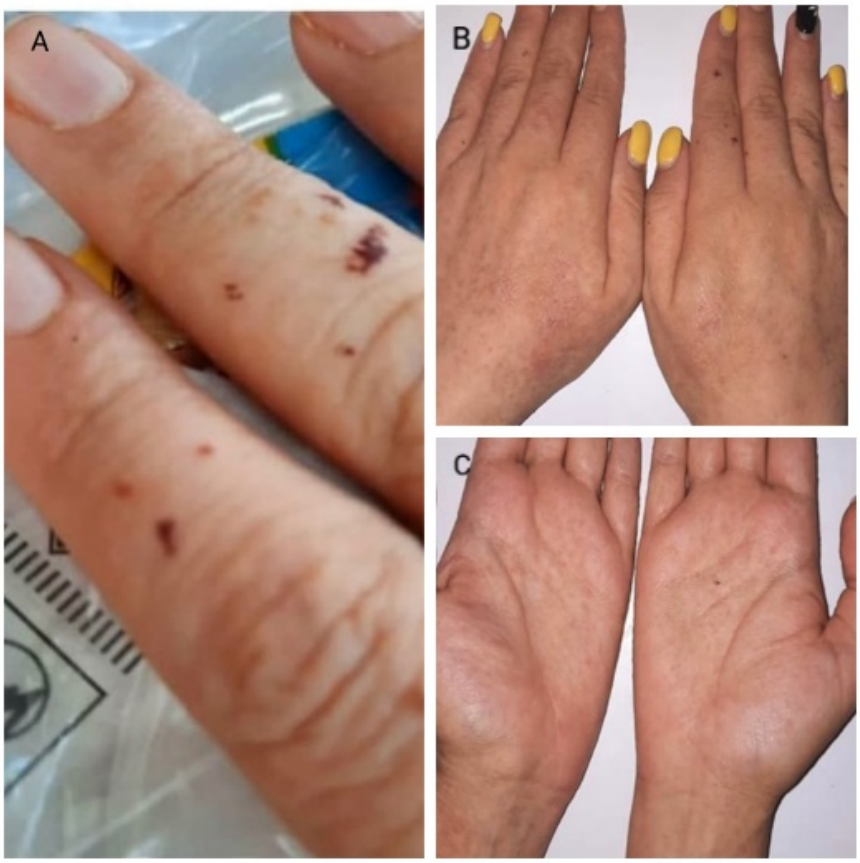

Figure 1.A, Hemorrhagic papules and vesicles on the dorsum of the fingers and.B, hands.C,palm

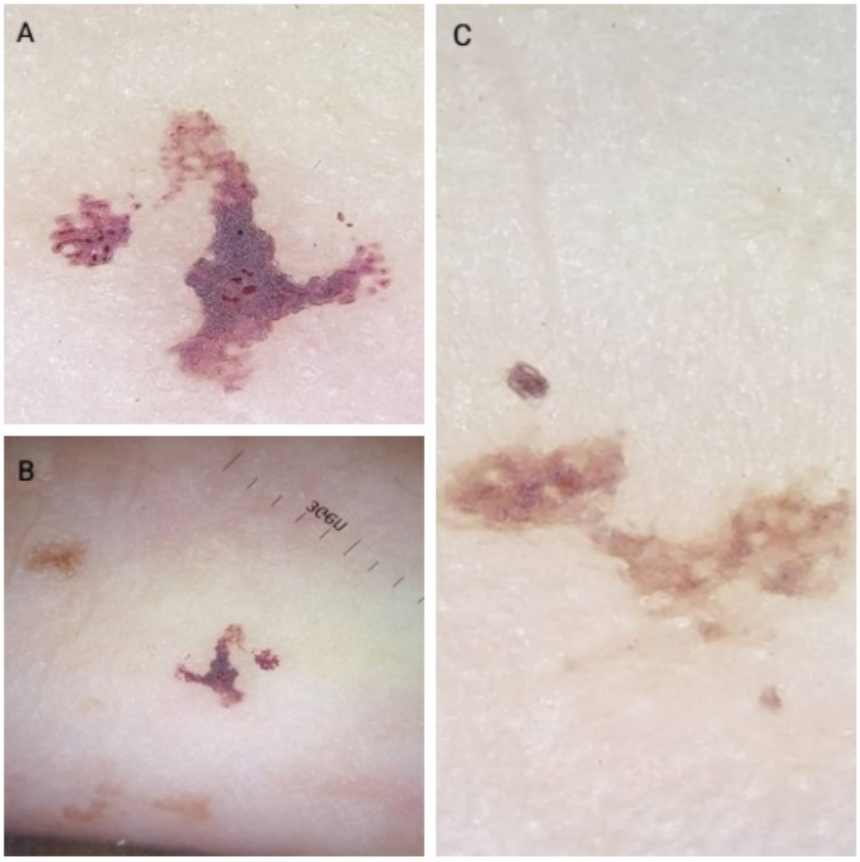

Figure 2.A and B, Dermoscopic image of a new lesion showing the signs of bleeding. C, Dermoscopic image of an old lesion with no sign of bleeding