

# Linear Darier disease after COVID-19 infection



Aaron Burch, BS,<sup>a</sup> Tyler Long, DO,<sup>b</sup> and Craig Garofola, DO<sup>b</sup>

**Key words:** acantholysis; COVID-19; COVID-19 vaccine; cutaneous; Darier disease; dermatology; dyskeratosis; genodermatosis; linear Darier disease.

## INTRODUCTION

Darier disease, also named Darier-White disease and keratosis follicularis, is an inherited skin disease that commonly presents in childhood but can present later in life. Clinically, disease is characterized by multiple brown keratotic papules coalescing into greasy plaques in the seborrheic distribution/areas of the body. Lesions are histologically characterized as acantholytic dyskeratosis with associated hyperkeratosis.<sup>1</sup> Most cases first appear between ages 6 and 20 years old and clinically present in a generalized and symmetric distribution, but approximately 10% of patients present with localized disease described as segmental, unilateral, linear, or zosteriform.<sup>2</sup> Here, we discuss a case of a 74-year-old Caucasian male with a linear truncal rash after hospitalization for COVID-19 infection. While COVID-19 vaccination has been reported to precipitate Darier disease, Darier disease after natural COVID-19 infection has not been previously published.<sup>3</sup>

## CASE PRESENTATION

A 74-year-old Caucasian male with no past medical history presented with a raised, scaly erythematous rash located on the trunk for the past 5 months. The patient reported no pain but complained of moderate pruritus. Five months prior, the patient was hospitalized for COVID-19 infection and the rash appeared shortly after discharge. The patient did not receive a COVID vaccination prior to this infection nor after the admission. Current medications included aspirin, and the

patient completed a short course of oral steroids for COVID pneumonia symptoms. The patient denied any associated symptoms or history of any similar skin findings. The patient also denied fever, chills, cough, blisters, recent sore throat, diarrhea, or joint aches. He reported no household contacts with a similar rash. Patient had no recent changes in medications and reports no new personal care products. Examination revealed brown vesicular and keratotic papules in a linear distribution on the left lateral abdomen (Fig 1). Differential diagnoses included linear Darier disease, Grover disease, and acantholytic dyskeratotic epidermal nevus. Saucerization biopsy showed acantholytic dyskeratosis, confirming a diagnosis of linear Darier disease (Fig 2). Topical tazarotene 0.05% treatment was initiated with moderate improvement at 3 months.

## DISCUSSION

We are reporting a case of linear Darier disease after COVID-19 infection. Linear Darier disease is a rare, localized variant that occurs in approximately 10% of patients with this autosomal dominant disease.<sup>2</sup> Lesions commonly follow Blaschko's lines due to the underlying genetic mosaicism from postzygotic mutations in genes encoding SERCA2 pump.<sup>4</sup> Histopathology demonstrates acantholysis and dyskeratosis represented by the hallmark findings of corps ronds and grains. Disease can be triggered and further exacerbated by heat, stress, medications, or infection.<sup>5</sup> It is possible that this case represents a

From the Edward Via College of Osteopathic Medicine, Auburn, Alabama<sup>a</sup>; and Department of Dermatology, LewisGale Hospital Montgomery, Blacksburg, Virginia.<sup>b</sup>

Funding sources: This research was supported (in whole or in part) by HCA Healthcare and/or an HCA Healthcare affiliated entity. The views expressed in this publication represent those of the author(s) and do not necessarily represent the official views of HCA Healthcare or any of its affiliated entities.

IRB approval status: Exempted.

Patient consent: Consent for the publication of all patient photographs and medical information was provided by the authors at the time of article submission to the journal stating that all patients gave consent for their photographs and medical information to be published in print and online and

with the understanding that this information may be publicly available.

Correspondence to: Tyler Long, DO, Department of Dermatology, LewisGale Hospital Montgomery, 3700 S Main St, Blacksburg, VA 24060. E-mail: [Longjtyler@yahoo.com](mailto:Longjtyler@yahoo.com).

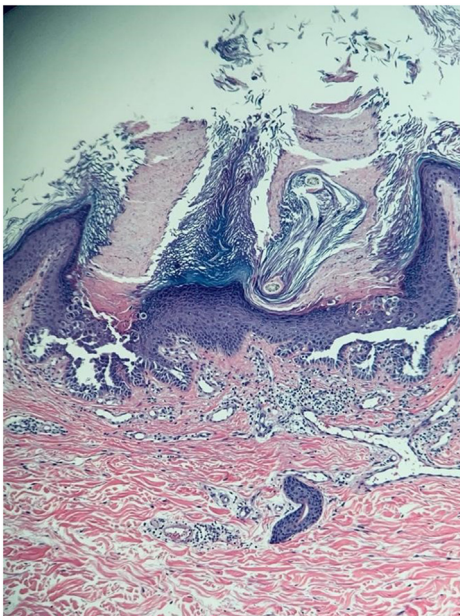
JAAD Case Reports 2022;28:136-7.  
2352-5126

© 2022 by the American Academy of Dermatology, Inc. Published by Elsevier, Inc. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

<https://doi.org/10.1016/j.jdc.2022.08.017>



**Fig 1.** Photo of the patient demonstrating brown keratotic papules in linear distribution along the left lateral abdomen.



**Fig 2.** Biopsy showed acantholytic dyskeratosis. No herpes or varicella viral cytopathic changes were identified (hematoxylin and eosin:  $\times 40$ ).

disease exacerbation following the stressors of infection.

To our knowledge, an association has not been observed among COVID-19 infections and linear Darier disease. However, COVID-19 has been associated with many cutaneous manifestations such as rash, pernio-like acral lesions, urticaria, macular erythema, vesicular eruption, papulosquamous eruption, and retiform purpura.<sup>6</sup> The underlying pathophysiology that best explains this spectrum of dermatologic presentations is believed to be from an overactive immune response to the viral nucleotides as well as the presence of the viral particles in the cutaneous vasculature that interact with keratinocytes.<sup>6,7</sup>

Recently, a case has been reported of a patient who presented with a flare of Darier disease 2 days

after receiving her COVID-19 vaccination.<sup>3</sup> Therefore, an association between COVID-19 vaccination and Darier disease has been observed, but the authors were unable to find a natural COVID-19 infection associated with linear Darier disease presentation. PubMed was queried for “Darier” AND “COVID.” Our patient had no prior history of Darier disease, so we believe his infection precipitated development of disease despite his advanced age and the fact that a majority of cases develop in childhood.<sup>1</sup> This presentation adds to our understanding of variable presentation of linear Darier disease.

The primary management goal in Darier disease is to control symptoms while avoiding triggers, wearing loose and cotton clothing, and using emollients and topical corticosteroids to control itching.<sup>4</sup> In extensive or severe disease, topical and oral retinoids have been found to be effective. Our patient’s rash resolved partially after initiation of topical tazarotene 0.05%.

Dermatologists and other providers should be aware of this association due to the current nature/condition of the global pandemic. This case adds to our understanding that COVID-19 can not only incite flares of Darier disease and its variants but can precede the initial onset of disease. When differentiating between linear lesions, it’s important to consider linear Darier disease as a viable diagnosis to not delay in starting supportive management to improve the patient’s quality of life.

#### Conflicts of interest

None disclosed.

#### REFERENCES

1. Takagi A, Kamijo M, Ikeda S. Darier disease. *J Dermatol*. 2016; 43(3):275-279. <https://doi.org/10.1111/1346-8138.13230>
2. Medeiros PM, Alves NR, Trujillo JM, Silva CC, Faria PC, Silva RS. Segmental Darier’s disease: a presentation of difficult diagnosis. *Anais brasileiros de dermatologia*. 2015;90(3 Suppl 1):62-65. <https://doi.org/10.1590/abd1806-4841.20153581>
3. Elbæk MV, Vinding GR, Jemec G. Darier’s disease flare following COVID-19 vaccine. *Case Rep Dermatol*. 2021;13(2):432-436. <https://doi.org/10.1159/000517256>
4. Cooper SM, Burge SM. Darier’s disease: epidemiology, pathophysiology, and management. *Am J Clin Dermatol*. 2003;4(2):97-105.
5. Burge SM, Wilkinson JD. Darier-White disease: a review of the clinical features in 163 patients. *J Am Acad Dermatol*. 1992;27(1): 40-50. [https://doi.org/10.1016/0190-9622\(92\)70154-8](https://doi.org/10.1016/0190-9622(92)70154-8)
6. Freeman EE, McMahon DE, Lipoff JB, et al. The spectrum of COVID-19-associated dermatologic manifestations: an international registry of 716 patients from 31 countries. *J Am Acad Dermatol*. 2020; 83(4):1118-1129. <https://doi.org/10.1016/j.jaad.2020.06.1016>
7. Gianotti R, Veraldi S, Recalcati S, et al. Cutaneous clinico-pathological findings in three COVID-19- positive patients observed in the metropolitan area of Milan, Italy. *Acta Derm Venereol*. 2020;100:adv00124. <https://doi.org/10.2340/0015555-3490>