

# A Case of Kaposi Varicelliform Eruption in Darier-White Disease

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## GOAL

To understand Darier-White disease (DWD) and its potential complications

## OBJECTIVES

Upon completion of this activity, dermatologists and general practitioners should be able to:

1. Recognize the clinical presentation of DWD in patients.
2. Manage the treatment of DWD.
3. Discuss the complications associated with DWD.

**CME** Test on page 32.

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*Darier-White disease (DWD), otherwise known as keratosis follicularis, is a rare disorder of keratinization of the epidermis, mucous membranes,*

*and nails. It is autosomal dominant in transmission. Patients with DWD are prone to frequent superinfection including the rare complication of Kaposi varicelliform eruption. It is postulated that a defect in cell-mediated immunity may contribute to this predisposition.*

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**D**arier-White disease (DWD) is an autosomal-dominant disorder characterized by abnormal keratinization of the epidermis, mucous membranes, and nails. It normally presents in the first or second decade of life and is characterized by numerous pruritic papules that range from flesh colored to yellow or brown. These papules often occur in the seborrheic distribution and may



**Figure 1.** Umbilicated vesicles coalescing into 4×8-cm plaques in the axilla.

coalesce into larger plaques. Men and women are equally affected, and patients with the disease are prone to frequent superinfection.<sup>1</sup>

### Case Report

A 19-year-old man with a history of DWD presented to his family physician reporting a 2-day history of a burning sensation and painful rash around his lips and eyes. Results of a physical examination showed eczematous and impetigenous lesions on his upper lip and red papules under both eyes measuring 3 mm in diameter. The patient was diagnosed with impetigo, started on oral amoxicillin/clavulanate and topical bacitracin, and discharged.

Two days later, the patient presented to the emergency department with a temperature of 100.5°F. He complained of increasing numbers of painful lesions that had spread to his axillae and across his face. The emergency department physician reaffirmed the diagnosis of impetigo, changed the patient's antibiotics to levofloxacin, added a lotion containing camphor and menthol to his therapeutic regimen, and discharged the patient.

The next day, the patient was transported to the emergency department via ambulance complaining of worsening rash and an elevated temperature of 102.9°F. He complained of severe oral pain, and the rash had spread over his periorbital and perioral areas, axillae, and groin. The physicians noted crusting and drainage over the lesions. The patient was discharged with naproxen and tramadol added

to his medication regimen. A dermatology consult was ordered.

On presentation to the dermatologist 2 days later, the patient had plaques of erosions noted on his groin that measured 4×6 cm. Additionally, his axillae showed umbilicated vesicles coalescing into 4×8-cm plaques bilaterally (Figure 1). Numerous umbilicated vesicles were present on his face in a seborrheic distribution (Figures 2 and 3). A more detailed history revealed that several days prior to the initiation of the rash the patient had kissed his girlfriend, who had a cold sore. Results of a Tzanck smear showed multinucleated giant cells. The patient was diagnosed with Kaposi varicelliform eruption, and valacyclovir was added to his therapeutic regimen, after which he noted rapid improvement of his symptoms.

### Comment

DWD is a rare autosomal-dominant genodermatosis. It is characterized by a pruritic, warty, flesh-colored to yellow or brown papular rash that is most often found in a seborrheic distribution. The nails often will have a typical notching of the free edge and can show alternating red and white longitudinal bands under the nails. The disease typically presents between the ages of 6 and 20 years but may develop later in life.<sup>2</sup>

Results of a histologic examination show acantholytic dyskeratosis characterized by suprabasal clefting, acantholytic keratinocytes in the cleft



**Figure 2.** Kaposi varicelliform eruption on the face with umbilicated vesicles and perioral erosions.

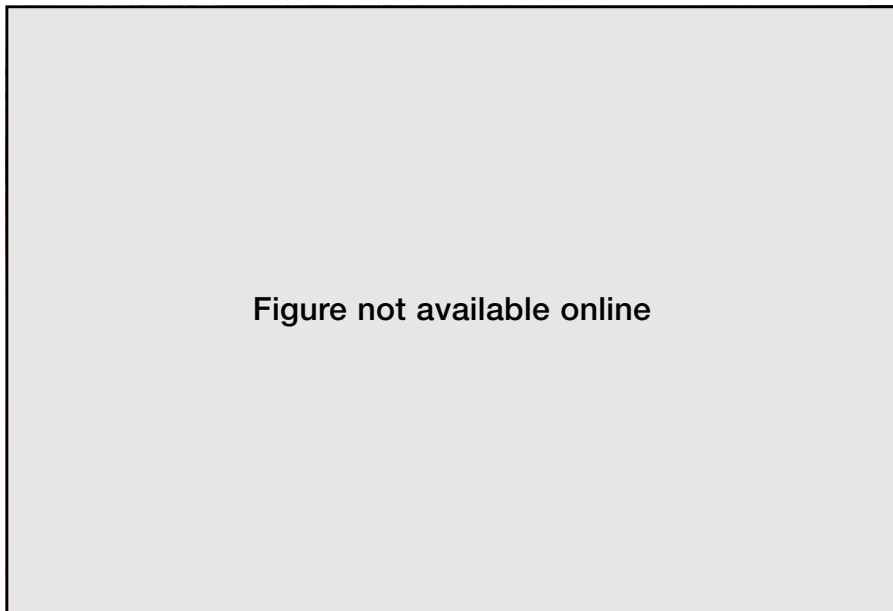


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**Figure 3.** Periorbital involvement of Kaposi varicelliform eruption.

spaces called *corps ronds*, and dyskeratotic cells in the spinous layer and stratum corneum called *corps grains*. The genodermatosis has been linked to a mutation of the *ATP2A2* gene (chromosome 12q23-24). This gene encodes a sarcoplasmic or endoplasmic reticulum calcium ATPase. This mutation may interfere with normal cell growth and differentiation.<sup>3</sup>

Management of DWD includes avoiding exacerbating factors such as UVA and UVB exposure, heat, perspiration, and mechanical trauma. This

may be accomplished by wearing cool clothing and sunscreen. Topical steroids, oral and/or topical retinoids, and a moisturizing regimen including emollients containing urea or lactic acid are important mainstays of treatment.<sup>3</sup> Oral antibiotics are often used when indicated because patients frequently become infected with *Staphylococcus aureus*.

Patients with DWD are more susceptible to widespread viral infections such as eczema vaccinatum and, as evidenced by this patient, Kaposi varicelliform eruption.<sup>4</sup> This complication is most

commonly caused by herpes simplex virus.<sup>5</sup> DWD joins atopic dermatitis as a contraindication to smallpox vaccination of US military personnel because of the increased risk of eczema vaccinatum in patients with DWD. Although not definitive, it is postulated that patients with DWD and atopic dermatitis share epidermal immune defects that predispose them to such outbreaks. In one study of 10 patients with DWD, lymphocyte transformation test results showed there was statistically enhanced responsiveness to phytohemagglutinin and concanavalin A mitogens, which may suggest lymphocytes in DWD have alterations in their immunoregulation.<sup>6</sup> Another study of patients with DWD showed anergy to common skin test antigens; in addition, their peripheral lymphocytes failed to produce lymphokine in vitro.<sup>7</sup> The potential pathogenesis of such apparent defects in cell-mediated immunity in DWD with respect to Kaposi varicelliform eruption is still unknown and warrants further investigation.

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