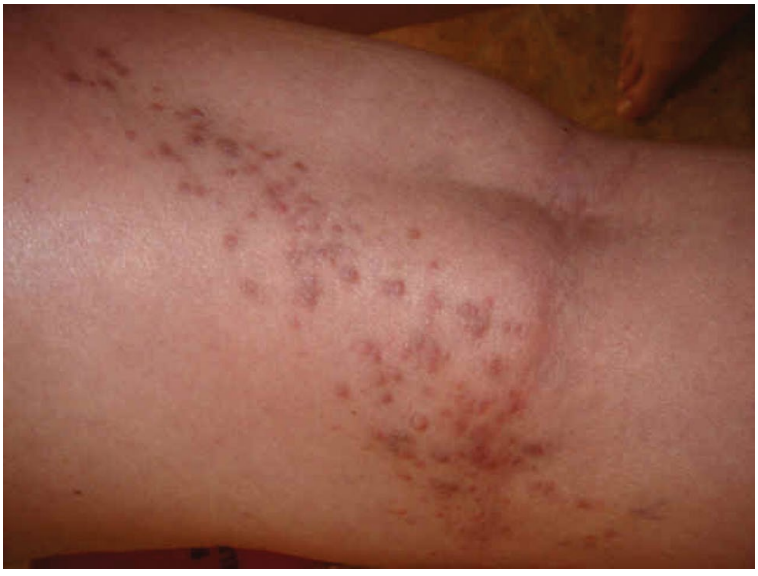
What Is Your Diagnosis?



A 60-year-old woman presented for evaluation of brown papules in the left popliteal area. The papules were mildly pruritic and had developed over the last 3 months. Her history was remarkable for hypertension, fibromyalgia, and breast cancer treated with a right radical mastectomy. Her medications included gabapentin and furosemide.

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The authors report no conflict of interest.

The Diagnosis: Linear Darier Disease

Our patient presented with brown papules in the left popliteal area (Figure 1). Biopsy results revealed hyperkeratosis, parakeratosis, and acantholytic dyskeratosis (Figure 2). Corps ronds were identified. The presence of acantholytic dyskeratosis raised a differential diagnosis that included Darier disease (keratosis follicularis) and Grover disease (transient acantholytic dermatosis) as well as an isolated and solitary acantholytic dyskeratoma. The biopsy findings and clinical history supported a diagnosis of linear Darier disease. Although some previously reported cases were classified as an unusual zosteriform epidermal nevus with acantholysis or linear Grover disease, current genetic studies suggest that genetic alterations and mosaicism account for the striking and unusual findings in these cases.[1-3]

Darier disease has been associated with abnormalities in the gene encoding Ca^{2+} adenosine triphosphatase, *ATP2A2,* on chromosome 12q23-24.1.[4] The gene is involved in protein formation for the sarcoplasmic/endoplasmic reticulum.[5-7] The resultant abnormality in calcium ion concentration is thought to have an impact on desmosomal components, leading to acantholysis.[8] Some individuals in the same kindred with the same genetic abnormality may have different degrees of clinical involvement, indicating that environmental factors and other genetic factors are important in disease expression. A single wild-type functioning gene seems to be insufficient to protect against skin lesions that are caused by the presumed postzygotic mutation in these cases.

Lesions typically present during the teenaged years and are precipitated by friction, heat, sunlight, and humidity. Yellow-brown papules develop and some lesions become lichenified because of repeated rubbing and trauma. Some lesions may become malodorous. Treatment with cool compresses, emollient creams, and topical steroids are helpful. Avoiding friction and sweating as well as keeping involved areas cool also is helpful. Oral retinoids may be of value in severe cases.[9] Laser and photodynamic therapy also have been reported to be of benefit.[10,11] Tazarotene

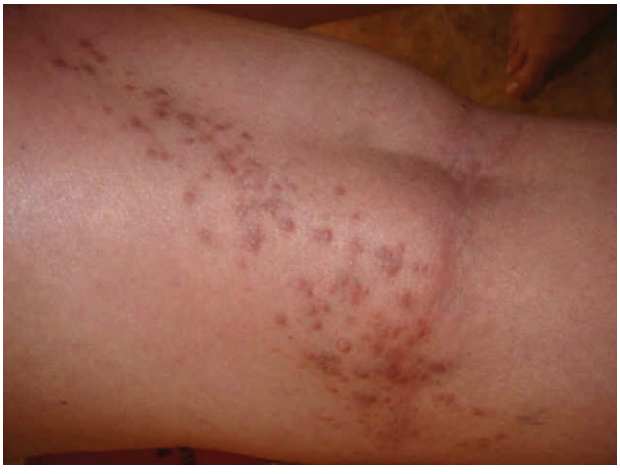

Figure 1. Brown papules in a linear distribution in the popliteal area.

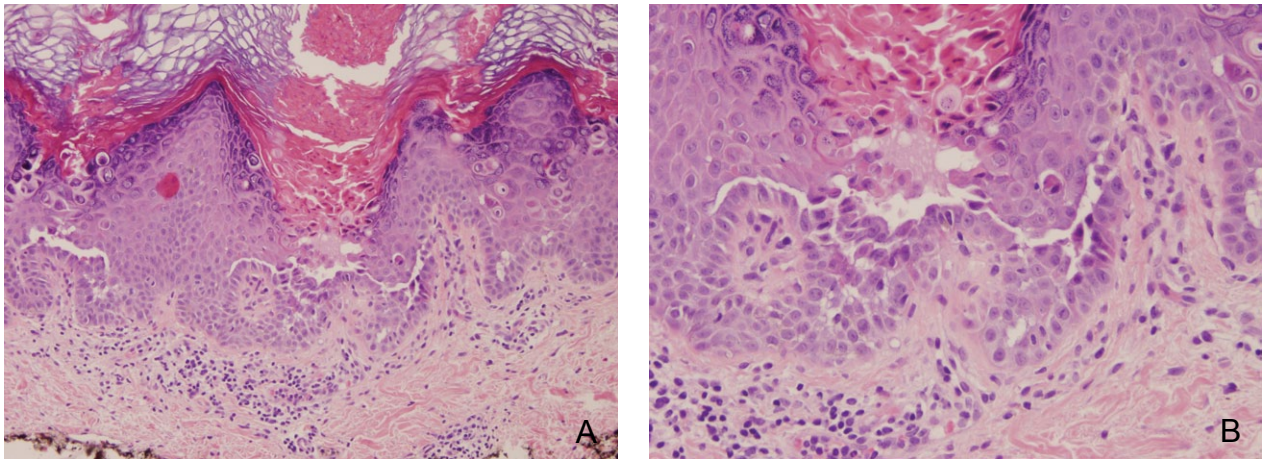

Figure 2. Hyperkeratosis, parakeratosis, and acantholysis are evident (A) as well as acantholytic dyskeratosis (B)(H&E; original magnifications ×20 and ×40, respectively).

gel 0.1% may be of benefit for localized disease,[12] as in this case. Patients also should be closely watched for eczema herpeticum or other complications.

The differential diagnosis includes an inflammatory linear verrucous epidermal nevus, lichen striatus, and granulomatous dermatitis occurring after herpes zoster infection. The biopsy findings as well as the clinical history exclude all of these entities.

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