Dermoscopy of Zosteriform and Swirling Pattern Type 1 Segmental Darier Disease

Dear Editor,

Segmental Darier disease (DD) is a rare disease with around 40 described English literature cases. It is hypothesized that one of the causes of the disease is a post-zygotic somatic mutation for the calcium ATPase pump, only present in lesional skin. There are two types of segmental DD: type 1, where lesions follow Blaschko's lines unilaterally, and type 2, characterized by focal areas of increased severity in patients with generalized DD (1). Type 1 segmental DD is not easily diagnosed due to the lack of positive family history, the late onset of the disease in the third or fourth decade of life, and lack of DD-associated features. The differential diagnosis of type 1 segmental DD includes acquired papular dermatoses distributed in linear or zosteriform fashion, such as lichen planus, psoriasis, lichen striatus, or linear porokeratosis (2).

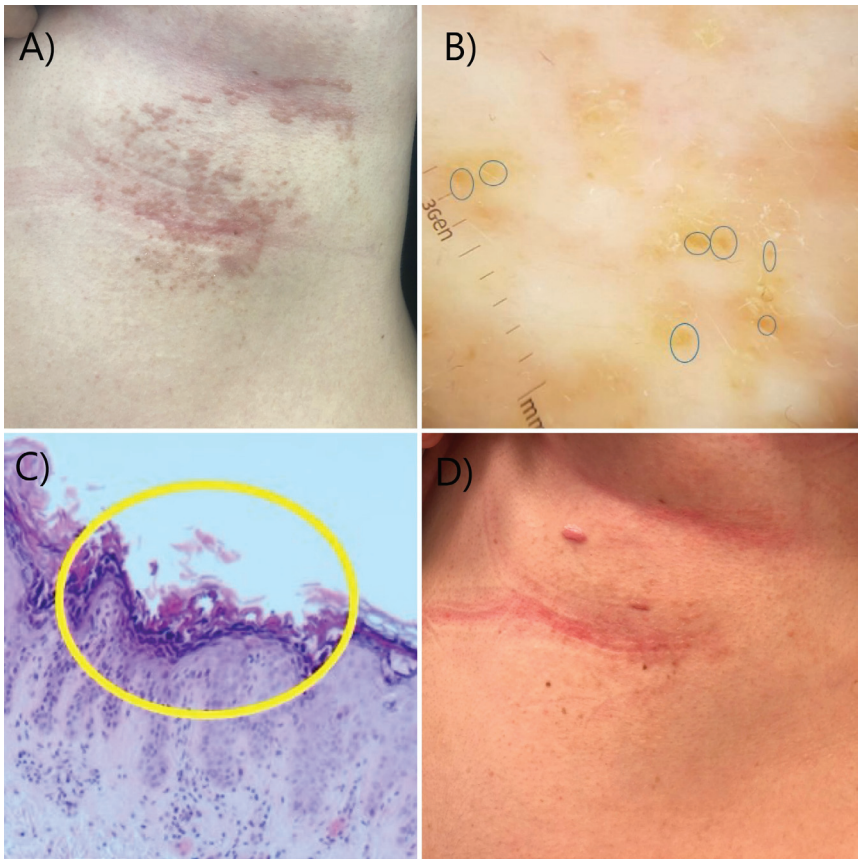

Figure 1. Physical examination showed light brownish to reddish keratotic small papules arranged in a swirling pattern on the left abdomen and inframammary area (a). Polarized light dermoscopic examination (×10 magnification) showed polygonal or roundish yellowish/brown areas surrounded with whitish structureless areas (b). The histopathological correlations for dermoscopic brownish polygonal or round areas are hyperkeratosis, parakeratosis, and dyskeratotic keratinocytes, which were present in the biopsy specimen (hematoxylin and eosin ×20) (c). Marked improvement was observed after three months of topical retinoid therapy, with only a scar visible on the skin biopsy site (d).

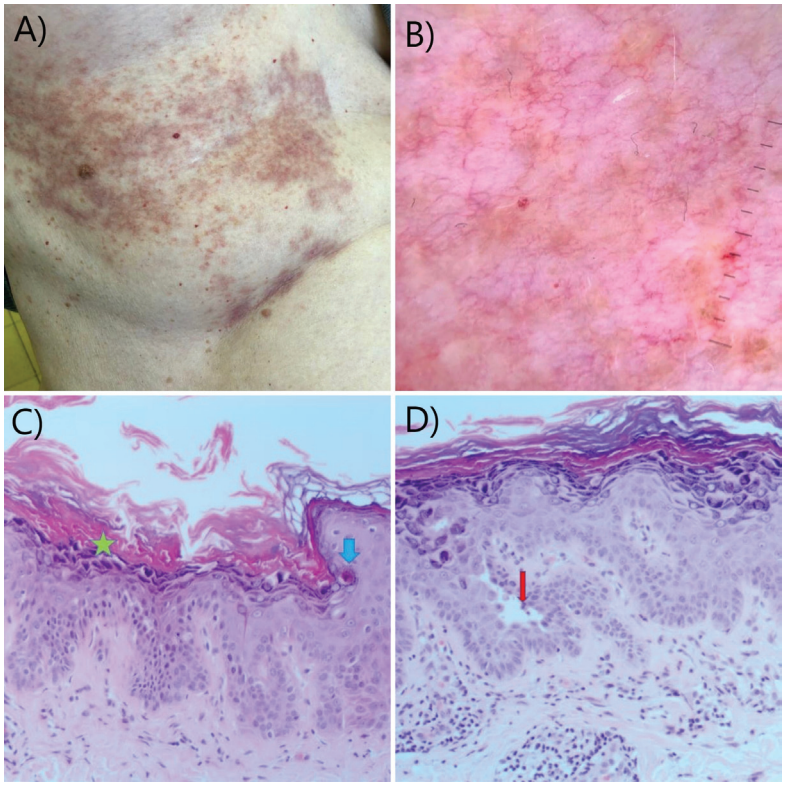

Figure 2. Physical examination sjowed small red-brown papules, eroded papules, and some yellowish crusts arranged in a zosteriform pattern on the right side of the upper abdomen (a). Polygonal, roundish, yellowish areas surrounded with whitish and reddish structureless areas were observed on dermoscopy (×10 magnification) (b). Histology mainly showed compact orthokeratosis and small foci of parakeratosis, a marked granular layer with dyskeratotic keratinocytes in the form of "corp ronds" (blue arrow) and "grains" (green star), and foci of suprabasal acantholysis (red arrow) (hematoxylin and eosin ×40) (c, d).

We report two cases of segmental DD, of which the first case was a 43-year-old woman who presented with pruritic skin changes five years in duration and a history of seasonal aggravation. On examination, light brownish to reddish keratotic small papules were observed on the left abdomen and inframammary area, arranged in a swirling pattern (Figure 1, a). Dermoscopy showed polygonal or roundish yellowish/brown areas surrounded with whitish structureless areas (Figure 1, b). The histopathological correlations for dermoscopic brownish polygonal or round areas are hyperkeratosis, parakeratosis, and dyskeratotic keratinocytes, which were present in the biopsy specimen (Figure 1, c). The patient was prescribed 0.1% tretinoin gel, which led to marked improvement (Figure 1, d).

The second case was a 62-year-old woman who presented with a flare of small red-brown papules, eroded papules, and some yellowish crusts arranged in a zosteriform pattern on the right side of the upper abdomen (Figure 2, a). Dermoscopy showed polygonal, roundish, yellowish areas surrounded with whitish and reddish structureless areas (Figure 2, b). Histopathology mainly revealed compact orthokeratosis and small foci of parakeratosis, marked granular layer with dyskeratotic keratinocytes, and foci of suprabasal acantholysis consistent with the diagnosis of DD (Figure 2, d, d). The patient was prescribed topical steroid cream and 0.1% adapalene cream, which also led to improvement.

In both of our cases, a final diagnosis of type 1 segmental DD was established based on clinico-histopathologic correlation, since acantholytic dyskeratotic epidermal nevus could not have been ruled out only based on the histopathology report as it is clinically and histologically indistinguishable from segmental DD. However, the late age of onset and aggravation resulting from external factors such as heat, sunlight, and sweat supported the diagnosis of segmental DD. Although the final diagnosis of type 1 segmental DD is typically established based on clinico-histopathological correlation, we find dermoscopy particularly useful in aiding the diagnosis by eliminating differential diagnoses and being aware of their well-known dermoscopic patterns.

References:

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Anamaria Balić, Joan Garces Rene, Jaka Radoš

Zagreb University Hospital Centre, Department of Dermatology and Venereology, University of Zagreb School of Medicine, Zagreb, Croatia

Corresponding author:
Anamaria Balić, MD
University Hospital Centre Zagreb
Department of Dermatology and Venereology
Kišpatićeva 12
Zagreb, Croatia.
jovicanamaria@gmail.com

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