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Dermoscopy: a useful auxiliary tool in the diagnosis of type 1 segmental Darier's disease

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ABSTRACT Type 1 segmental Darier's disease is a blaschkolinear variant of Darier's disease resulting from a postzygotic mosaicism. Since it usually lacks diagnostic clues typical of the generalized form, including positive family history of the disease, nail and mucosal abnormalities, and/or acral involvement, its distinction from other acquired inflammatory blaschkolinear dermatoses may often be quite challenging, thus requiring histopathological examination to reach a definitive diagnosis. We report a case of type 1 segmental Darier's disease with its dermoscopic findings in order to show the usefulness of dermoscopy in assisting the noninvasive identification of this condition.

Introduction

In approximately 10% of cases, Darier's disease (DD) may present in a localized pattern as the result of a postzygotic mosaicism in the ATP2A2 gene (type 1 segmental DD) [1]. Such a variant is characterized by hyperkeratotic reddish-brownish papules having a linear distribution along Blaschko's lines, which frequently get worse during the summer [1]. Since type 1 segmental DD usually lacks diagnostic clues typical of the generalized form, i.e., positive family history of the disease, nail and mucosal abnormalities, and/or acral involvement, its distinction from other acquired inflammatory blaschkolinear dermatoses may often be quite challenging, thus requiring histopathological examination to reach a definitive diagnosis [1]. We here describe for the first time the use of dermoscopy in a patient with type 1 segmental DD.

Case presentation

A 25-year-old Caucasian man presented with a two-year history of slightly itchy skin lesions on the right side of his abdomen, which were reportedly worse in the summer months. There was no family history of similar problems. Physical examination showed numerous hyperkeratotic reddish-brownish papules in a blaschkoid distribution (Figure 1a). No mucosal or nail lesions were present. On polarized light

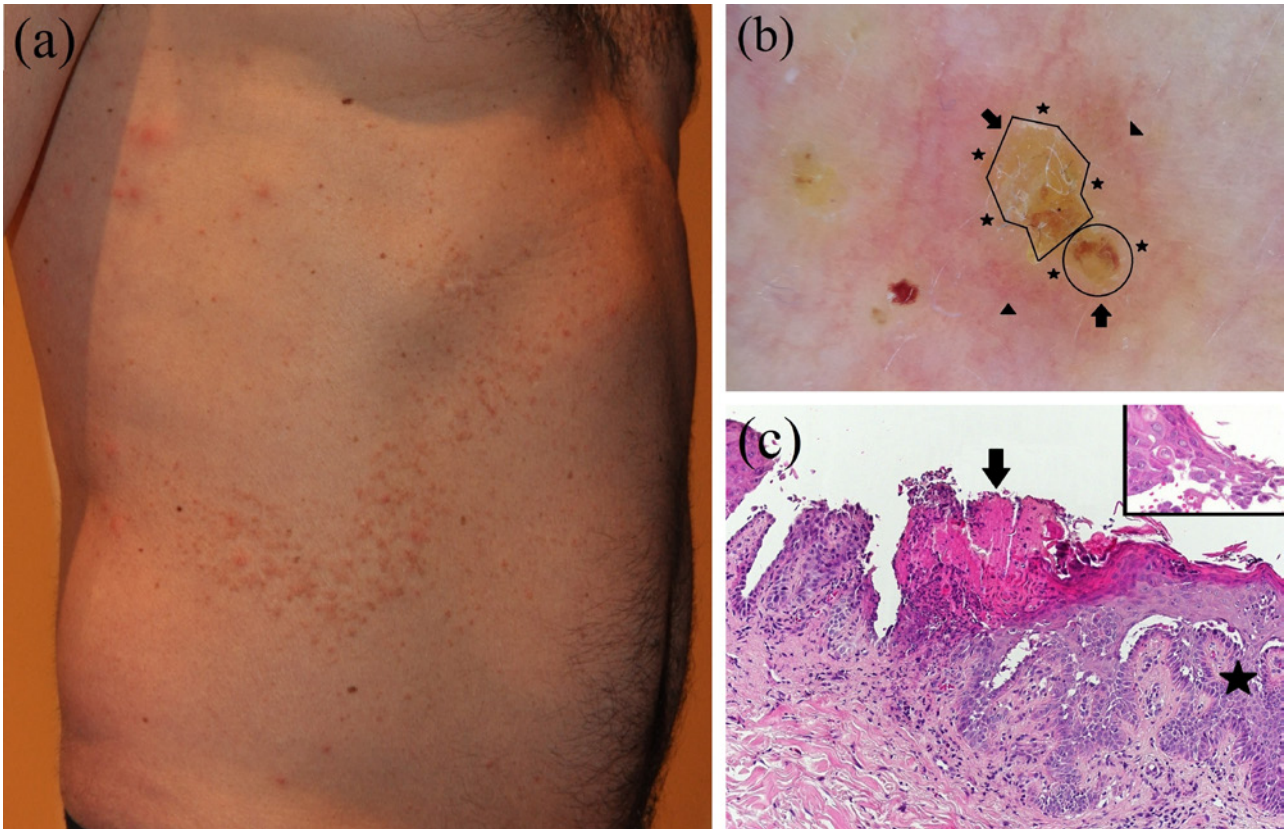

Figure 1. Physical examination shows numerous hyperkeratotic reddish-brownish papules in a blaschkoid distribution on the right side of the abdomen; unrelated sparse lesions of folliculitis and the scar at the biopsy site are also evident on the right subaxillary region and costal arch, respectively (a). Polarized light dermoscopic examination (x10 magnification) of two contiguous papules displays centrally located, polygonal (outlined in black) or roundish (outlined in black), yellowish/brownish areas (black arrows) presenting a whitish halo (black stars), which are surrounded by a pinkish homogeneous structureless area with some dotted vessels (black arrowheads) (b). Histology shows compact hyperkeratosis (black arrow), parakeratosis, acanthosis (black stars), foci of suprabasal acantholysis, dyskeratotic cells (better visible in the box) in the form of "corp ronds" and "grains", and superficial dermal chronic inflammation (hematoxylin and eosin stain x100) (c). [Copyright: ©2016 Errichetti et al.]

dermoscopic examination (carried out with DermLite DL3 x10; 3Gen, San Juan Capistrano, CA, USA), most papules displayed the same aspect, namely, centrally located, roundish or polygonal, yellowish/brownish areas of various sizes (often with a whitish halo) that were surrounded by a pinkish homogeneous structureless areas (with or without linear and/or dotted vessels) (Figure 1b). Histology revealed compact hyperkeratosis, parakeratosis, acanthosis, foci of suprabasal acantholysis, dyskeratotic cells in the form of "corp ronds" and "grains," and superficial dermal chronic inflammation (Figure 1c), thus leading to the diagnosis of type 1 segmental DD. The patient was treated with tretinoin 0.05% cream (once a day) with moderate improvement of the clinical picture after three weeks.

Discussion

During the last years, the use of dermoscopy in "general" dermatology has considerably increased [2-6]. The first dermoscopic description of DD dates back to 2004, when Vázquez-López et al reported a study on five patients. The authors found a variable vascular pattern, including red dots, red lines, or erythema, and constant nonvascular structures, which they named "giant pseudocomedones," consisting in dilated oval openings with raised or flat borders and central brown or yellowish hyperkeratotic plugs [7]. However, according to our recent study on 11 patients, DD may display a more complex dermoscopic pattern characterized by a centrally located polygonal, star-like or roundish-oval shaped yellowish/brownish area, surrounded by a more or less thin whitish halo, overlying a pinkish homogeneous structureless area, with or without whitish scales and dotted and/or linear vessels presenting a whitish halo [8]. Indeed, we observed a similar appearance in the present "localized" instance. In terms of dermoscopic-pathological correlation, the central yellowish-brownish area and its whitish halo would correspond to the compact hyperkeratosis and acanthosis (Figure 1b, c), respectively, while the pinkish background and vessels would be due to the dermal inflammation [8-9].

By reporting the dermoscopy of this case of type 1 segmental DD, we would like to point out the usefulness of such

a low-cost and noninvasive technique in assisting the recognition of this variant of DD. In fact, even though the definitive diagnosis of such a condition relies on histopathological analysis, dermoscopy may be used as an auxiliary tool to rule out the other major acquired inflammatory papular dermatoses which may present in a blaschkolinear fashion and enter into the differential diagnosis [10], including lichen planus, which is typically characterized by Wickham striae (seen as round, linear, reticular or annular pearly-whitish structures) [2]; psoriasis, showing white scales and symmetrically/regularly distributed dotted vessels on a light or dull red background [2,3] and lichen striatus, displaying vessels of mixed morphology (dotted and/or linear) over a pinkish background, with or without whitish scales (personal observations).

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