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Darier's Disease: The Benefits of Dermoscopy

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Abstract **Case Report**

Darier disease is a rare genodermatosis transmitted in autosomal dominance, with more or less complete penetrance and variable expressivity within each generation. It combines mainly follicular papulokeratotic lesions, predominantly in seborrheic areas, with acral involvement and highly suggestive nail lesions. The disease has a chronic course, marked by bacterial or viral superinfections. Knowledge of this disease has advanced considerably in recent years, with the gene responsible for it now located on the long arm of chromosome 12. Dermoscopic aspects of this dermatosis are poorly reported in the literature. We describe the clinical and dermoscopic analysis of lesions in a patient treated in our clinic for Darier's disease. Dermoscopic signs can help guide the diagnosis of Darier's disease.

Keywords: Darier Disease, Dermoscopy, Genodermatosis.

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INTRODUCTION

Darier disease is a rare genodermatosis. It generally occurs in young people and affects both sexes. It manifests as yellowish, brown or pinkish keratotic micropapules, covered with an adherent crust, confluent in sheets, giving a rough appearance. The contribution of dermoscopy in this dermatosis is little reported in the literature. We describe the clinical and dermoscopic analysis of lesions in a patient treated in our clinic for Darier's disease.

CLINICAL OBSERVATION

40-year-old man, with no family history of similar cases, presenting with erythematobrunate, pruritic lesions scattered over the body, aggravated in summer and when sweating profusely. Dermatological examination revealed brownish, hyperkeratotic, profuse papules forming lichenified plaques predominating in seborrheic areas (Fig. 1), clear papular lesions on the forearms, backs of hands, legs and backs of feet, with red and white longitudinal striae on the nails (Fig. 2). Dermoscopy revealed several polygonal, star-like or oval, rounded structures in the form of yellowish and sometimes brownish areas of varying size, surrounded by a thin whitish halo (Fig. 3). Histological examination of the patient's skin biopsy revealed hyperparakeratotic areas of varying size in the epidermis, dyskeratosis with round bodies and grains, and focal acantholysis with suprabasal cleft formation in favor of Darier's disease (Fig.4). Therapeutically, the patient was started on acitretin (0.5 mg/kg/dr) with adjuvant symptomatic treatment.

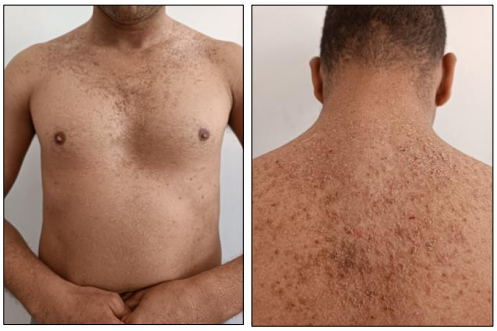

Figure 1: brownish, hyperkeratotic, profuse papules forming lichenified plaques, predominantly in seborrheic areas

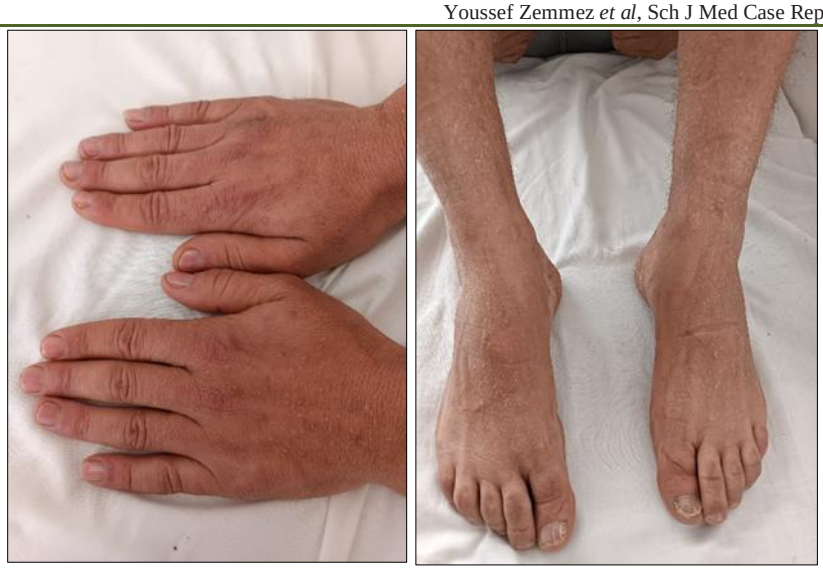

Figure 2: clear papular lesions on forearms, backs of hands, legs and backs of feet

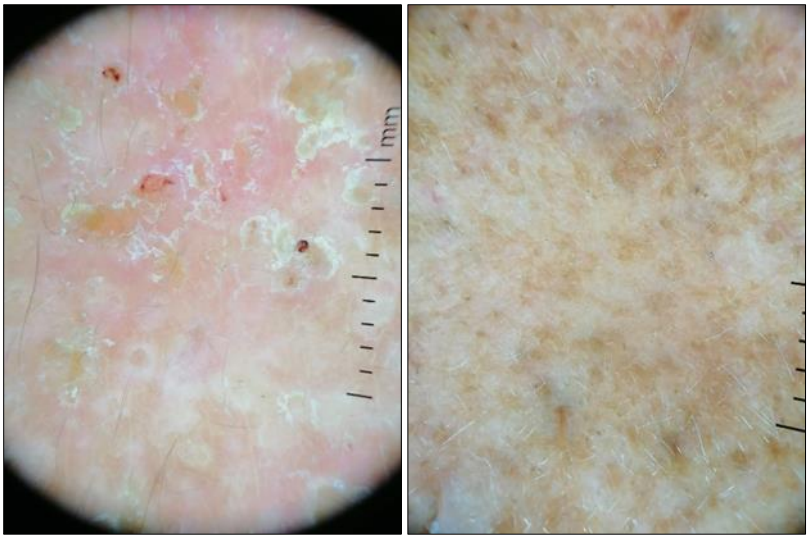

Figure 3: polygonal, star-like or oval, rounded structures in the form of yellowish and sometimes brownish areas, of various sizes, surrounded by a thin whitish halo

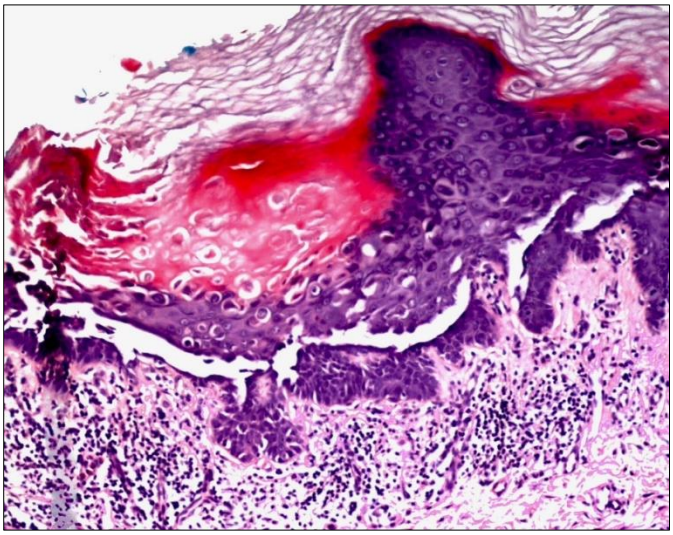

Figure 4: hyperparakeratotic areas of varying size in the epidermis, dyskeratosis with round bodies and grains, and focal acantholysis with suprabasal cleft formation

DISCUSSION

Darier disease is a rare autosomal inherited disorder [1]. It is secondary to mutations in the 12q-localized ATP2 A2 gene, encoding the ça ++ ATPase secretory pathway of the endoplasmic reticulum causing changes in calcium-dependent intracellular signals, resulting in loss of cell adhesion in the epidermis, leading to acantholysis and apoptosis [2]. Its sex ratio is 1, and no differences between ethnic groups have been identified [1-3]. The disease generally begins between the ages of 6 and 20, with a peak at the onset of puberty (11-15 years) [2]. Primary lesions are keratotic papules, sometimes crusted, ranging from red to brown, which develop preferentially in a "seborrheic" distribution involving the trunk, scalp, face and neck. These lesions tend to become confluent and may form papillomatous masses [2].

Few publications have examined the dermoscopy of this disease. The dermoscopic aspects described are the presence of several polygonal, star-shaped or rounded-oval yellowish and brownish areas of different sizes, surrounded by a fine whitish halo [4]. In our patient, dermoscopic examination of the The main histological features observed in Darier disease are acantholysis and dyskeratosis. Acantholysis is due to a loss of cellular adhesion and is responsible for suprabasillar clefts. Dyskeratosis corresponds to premature differentiation of keratinocytes. These dyskeratotic cells are described as "round bodies" in the squamous layer and "grains" in the stratum corneum [1]. An anatomo-clinical correlation has been established to better understand these dermoscopic aspects. Indeed, polygonal structures correspond histologically to hyperkeratosis, while round bodies correspond to large, clear epidermal cells [4]. Our publication highlights the value of dermoscopy as an examination that can help clinicians better identify this dermatosis and differentiate it from other acantholytic disorders affecting the folds. Management consists in avoiding triggering factors (sun, maceration) and applying photoprotection measures. Oral retinoids are the reference treatment for extensive forms [3]. Multiple lesions revealed the same dermoscopic features described in the literature.

CONCLUSION

Dermoscopy can be considered as a useful complementary non-invasive support to confirm the diagnosis of darier's disease and eliminate the other main clinical differential diagnoses.

Declaration of Links of Interest: The authors declare that they have no ties of interest.

REFERENCES

1. Engin B, Kutlubay Z, Erkan E, Tüzün Y. Darier disease: A fold (intertriginous) dermatosis. Clin Dermatol. 2015, 33:448–451.
2. Buteică E, Burada F, Stoicescu I, Stănoiu B, Georgescu CV. Darier disease and Hailey-Hailey disease. Rom J Morphol Embryol. 2007, 48:423-6.
3. Valois P-Y, Girault, J-J, Morand. Maladie de Darier. EMC –Dermatologie. 2016, 11:1-9.
4. Lacarrubba F, Verz AE, Errichetti E, Stinco G, Micali G. Darier disease: Dermoscopy, confocal microscopy, and histologic correlations J Am Acad Dermatol. 2015, 73:e97-9.