UNILATERAL DARIER’S DISEASE

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ABSTRACT

Darier’s disease (DD) is an uncommon genodermatosis characterized by the symmetrical eruption of keratotic reddish-brown papules occurring in the seborrheic areas of the body. The unilateral form of this disease is rare and could result from genetic mosaicism in this autosomal dominant disorder. We describe an unusual case of unilateral DD affecting the upper right side of his trunk.

Key words: Darier’s disease, unilateral Darier’s disease, keratosis follicularis

INTRODUCTION

Darier’s disease (DD) is an autosomal dominant skin disorder that is characterized by multiple symmetric keratotic papules, loss of epithelial adhesion, and abnormal keratinization\(^1\). Unilateral, focal form is the one of the rare type of the DD\(^2\). We present a case of DD that involved the upper right side of his trunk.

CASE REPORT

A 17 year old man had a one year history of yellow-brown keratotic papules on the right side of his trunk (Figure 1). There were no exacerbating factors such as sunlight, heat, or sweating. His family history was negative for any similar skin problem, and he had no children. Previous treatments with topical corticosteroids had been unsuccessful. No other lesions were detected on examination of the rest of the skin and the mucosa. The nails were normal in appearance. All routine hematological investigations were normal. Histopathology from the hyperkeratosis, papillomatosis, acanthosis, suprabasal cleft formation, dyskeratotic keratinocytes (Figure 2). These findings were consistent with the diagnosis of DD.
DISCUSSION

Darier’s disease (DD), also known as Darier-White disease or keratosis follicularis, was initially described in 1889. DD is an autosomal dominant disorder with altered keratinization of the epidermis, nails and mucous membranes. Mutations in a

Figure 1. Skin lesions at trunk.

Figure 2. Histopathology.
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sarcoendoplasmic reticulum Ca\(^+2\) ATPase isoform 2b (SERCA 2b) is the main association of DD

DD usually runs a chronic course with exacerbations and remissions. Exacerbations can be caused by hot weather, excessive sun exposure, medications, steroids or mechanical trauma.

DD is characterized by a symmetric eruption of keratotic, reddish brown papules that occur in seborrheic areas. Punctate keratoses or pits are frequently noted on the palms and soles. The nails show subungual hyperkeratosis, fragility, and splintering, with white and red alternating longitudinal bands and triangular nicking of the distal nail plate. The trunk is the most frequent site. The disease usually begins in adolescence or early adult life.

A localized form of DD was first described by Kreibich in 1906. Since that time, numerous individual cases have been reported, and there has been debate in the literature about how to classify this entity. Because of the age at onset, the absence of other signs of DD, and the negative family history in these patients, it has been suggested that the term acantholytic dyskeratotic epidermal nevus be used instead of localized DD.

The clinical differential diagnosis includes seborrheic dermatitis, Hailey-Hailey disease, and Grover disease.

Oral retinoids have made life easier for patients with severe DD, but many do not require such aggressive therapy. Acitretin appears more effective than isotretinoin. Systemic antibiotics and anti-herpes simplex therapy are also very useful. Topical corticosteroids may reduce inflammation, topical antimicrobials may help control infection and topical retinoids may slightly decrease scaling, but none is extremely successful. We treated his with topical mometasone furoate 0.1% ointment alternating with tretinoin 0.05% cream. He obtained partial regression of the disease after two months of treatment.

Unilateral DD should be remembered in the differential diagnosis of localized keratotic papules.

COMPETING INTERESTS

The author certifies that the author does not have any actual or potential conflict or competing interest.

REFERENCES