Linear Darier's disease: A case with bilateral presentation

Anal Jyoti Bordoloi, Khagendra Narayan Barua

Department of Dermatology, Gauhati Medical College and Hospital and Dr. K.N.Barua Institute of Dermatological Sciences (BIDS), Guwahati, Assam, India

ABSTRACT

Darier's disease is an autosomal dominant genodermatosis characterized by a persistent eruption of hyperkeratotic greasy papules mainly over the seborrheic sites of the body, usually associated with nail abnormalities and sometimes with mucous membrane lesions. The lesions typically occur in the younger age group and are associated with pruritus. We report here an atypical case of Linear Darier's disease with bilateral presentation in a middle-aged adult that could be confused with conditions such as lichen planus, inflammatory linear verrucous epidermal nevus, and wart.

Key words: Darier's disease, genetic mosaicism, linear

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Address for correspondence:
Dr. Anal Jyoti Bordoloi, Department of Dermatology, Gauhati Medical College and Hospital, Guwahati - 781 032, Assam, India.
E-mail: ajbordoloi@gmail.com

INTRODUCTION

Darier's disease, also known as Darier–White disease or keratosis follicularis is an autosomal dominant dermatoses characterized by warty, hyperkeratotic papules that are particularly distributed over the seborrheic sites of the body. It may be associated with nail dystrophy, palmoplantar pits, "cobblestoning" of oral mucosa[2,11] and sometimes with neuropsychiatric abnormalities. The condition was first described by Darier and White in 1889. The average age of onset is usually during childhood and adolescence, with no gender predilection. Darier's disease in a localized pattern was first described in 1906 by Kreibich and since then various clinical variants have been described in the form of unilateral, linear, segmental, or zosteriform Darier's disease. The term "Acantholytic Dyskeratotic Epidermal Nevus" is used synonymously with linear Darier's disease.[4]

CASE REPORT

A 55-year-old male patient presented to our clinic with the chief complaint of recurrent, pruritic eruption over his scalp, forehead, face, and neck region since the last seven years. The eruption first appeared on the right side of his forehead as some linearly distributed papules and had remained stable for about five years without any symptoms due to which the patient did not seek any medical advice. But since then they spread in a linear pattern to involve the right side of his face, neck, and scalp as well as to the left side of his forehead and neck. There was associated pruritus and the lesions aggravated on sun exposure. He also complained of loss of hair from the scalp over the areas where these lesions appeared. There was no history of similar lesions in the family.

On examination, hyperkeratotic, skin colored to hyperpigmented, grouped as well as scattered papules arranged in a linear pattern were seen involving the right side of his forehead, right cheek, and left side of forehead [Figure 1a, b and c]. Some of the lesions had a violaceous hue but without any whitish streaks on the surface. A similar distribution was also seen on right side of his neck [Figure 1d]. These were not associated with erythema or scaling. On further examination, a few hyperkeratotic papulonodular eruptions were seen over the vertex of his scalp [Figure 1e]. The surrounding skin showed some pigmented macules and was slightly atrophic with loss of hair.

Nail examination did not show any abnormality and the oral mucosa was also normal.

With the differential diagnosis of inflammatory linear verrucus epidermal nevus (ILVEN), linear lichen planus, warts, seborrheic keratosis, and linear Darier's disease we did a punch biopsy from the lesional skin over the forehead and the scalp. The histopathological examination revealed a circumscribed area of hyperkeratosis with acantholytic dyskeratosis and suprabasal

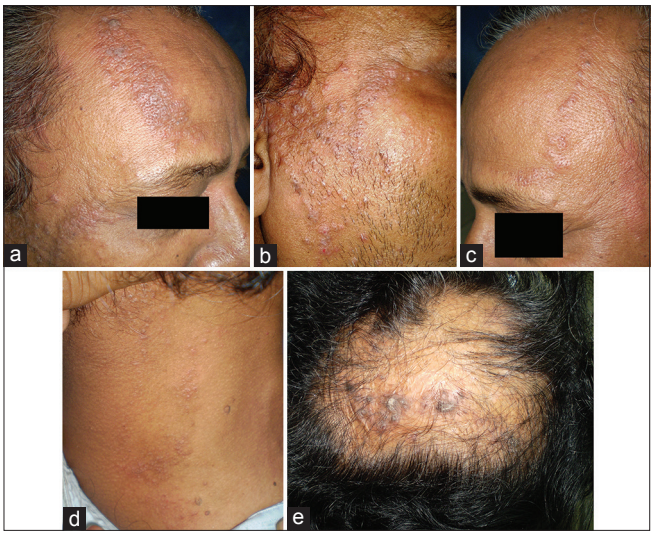

Figure 1: (a) Linear grouped hyperkeratotic and violaceous papules over forehead. (b) Similar lesions on the face. (c) Lesions on left side of forehead. (d) Lesions over neck. (e) Lesions over scalp

cleft formation. Within the focus, the epidermis showed scattered acantholytic dyskeratotic cells (corps ronds) [Figure 2a and b]. The stratum corneum showed an array of parakeratotic dyskeratotic cells (grains). The underlying dermis showed sparse superficial perivascular lymphohistiocytic infiltrate. With the above findings, a diagnosis of "linear Darier's disease" was made. The patient was at first started on topical retinoic acid (0.05%) to be applied once in the evening for one month. However, as he did not show any significant improvement, he was started on a course of oral isotretinoin 20 mg daily to which his skin lesions responded.

DISCUSSION

Darier's disease, a dominantly inherited skin condition, is characterized by hyperkeratotic papules coalescing to form warty plaques on symmetrical areas of the face, trunk, and extremities. Mutations of the ATP2A2 gene located in chromosome 12 encoding SERCA2, which has an important role in calcium signal transduction, have been identified as the molecular basis of this condition.[1] It has been found that selective inhibition of SERCA pumps interfere with the formation of intercellular junctions and cellular adhesion.

The localized form of Darier's disease is rare and was first described by Kreibich in 1906.[2] This variant often lacked other features that were associated with typical Darier disease and the skin lesions were usually confined to a limited area. It has been postulated to reflect genetic mosaicism occurring during embryogenesis. It has been estimated that in 10% of cases of Darier's disease, the lesions are distributed in a linear pattern following the Lines of Blaschko. The most commonly affected sites are the trunk and the limbs. A family history is usually

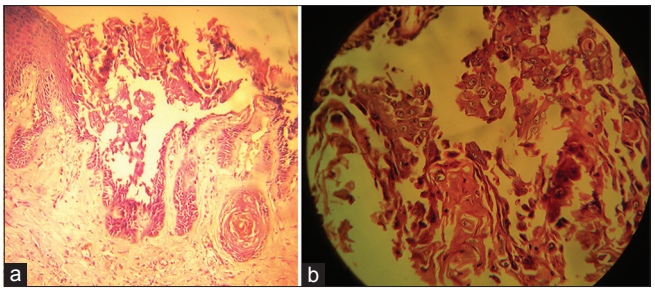

Figure 2: (a) Histopathology showing suprabasal cleft formation, acantholysis, and parakeratotic dyskeratotic cells, H and E, ×10. (b) High power magnification showing corps-ronds and grains, H and E, ×40

absent and rarely other associated features of Darier's disease have been reported.[3]

Aggravating factors are the same as seen in diffuse Darier's disease like light, heat, sweating, and friction. Pregnancy has also been shown to exacerbate this condition. The onset of the disease is usually in the third or fourth decade of life and there is no sexual predilection. Starink and Woerdeman reported seven cases showing unilateral, linear, or zosteriform patterns, without other findings and suggested the name "Acantholytic Dyskeratotic Epidermal Nevus."[4] Munro and Cox described a patient who had unilateral acantholytic dyskeratotic skin lesions with nail and palm changes of Darier's disease.[5] According to Happle's classification of cutaneous mosaicism for genetic disorders, segmental Darier's disease is of two types. Type 1 form retains the heterozygosity of mutation and the skin outside the segmental lesion is normal, whereas Type 2 is rarer, more severe, and is characterized by linear streaks over a background of generalized Darier's disease.[6,7,8] Our case probably belonged to the Type 1 form because it was localized and the surrounding skin was also normal.

Involvement of the scalp in linear Darier's disease has been rarely reported in the literature.[9,10] Moreover, the bilateral involvement of the linear variant as was seen in our case is also a striking feature, which has not been reported, to the best of our knowledge. A recurrent form of Linear Darier's Disease aggravated by sunlight was reported by Plantin *et al*.[11]

Treatment modalities include topical agents such as salicylic acid, lactic acid, and retinoids such as retinoic acid and tazarotene.[13] Systemic retinoids mainly isotretinoin have also been shown to be effective. If medical therapy is ineffective, carbon dioxide laser and erbium: YAG laser have been used for chronic, recalcitrant cases.[14,15]

Our patient, a middle-aged individual, had lesions distributed over the face and neck, which is quite uncommon for the linear variant of Darier's disease. Other features such as the presence of bilateral lesions, involvement of the scalp, and the lack of the typical "dirty warty" look of the papules made this an unusual presentation.

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