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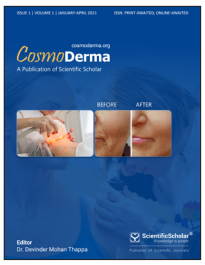

Spot the Diagnosis

A curious case of unilateral hypo-pigmented macules

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Quick Response Code:

A 67-year-old male presented with complaints of light colored lesions over the left side of trunk since childhood. He also gave history of development of raised itchy and fluid filled lesions over the light colored lesions for 5–6 years. There was a history of summer exacerbation of these itchy lesions. There was no family history of similar complaints.

On examination, hypo-pigmented macules arranged in blaschkoid distribution were present on the left side of trunk, not crossing the midline [Figure 1]. Few vesicles and lichenoid papules coalescing to form plaques were also present over the hypopigmented macules [Figure 2a and b].

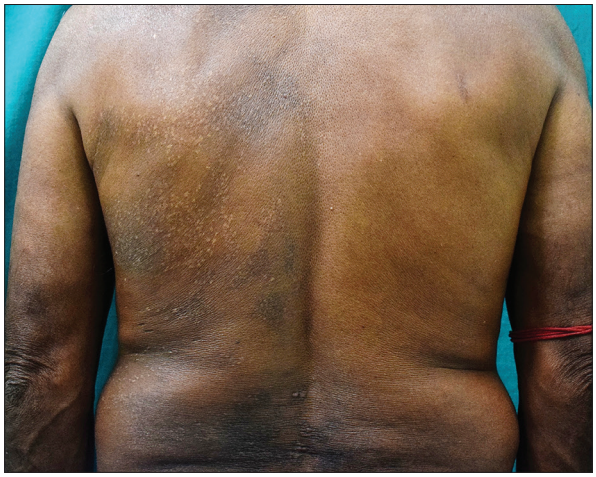

Figure 1: Hypo-pigmented macules arranged in blaschkoid distribution present on the left side of trunk, not crossing the midline.

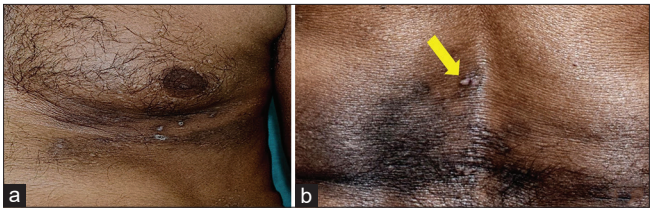

Figure 2: Lichenoid papules present in inframammary area the (a) and (b) Lichenoid papules coalescing to form plaques on lower back as depicted by the yellow arrow.

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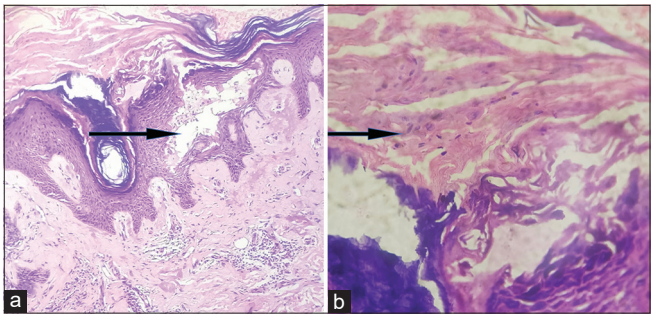

Figure 3: (a) Split in the malphigian layer of stratum corneum showing acantholytic cells on H&E staining as depicted by the black arrow under ×40 magnification. (b) Dyskeratotic cells seen within the stratum corneum as depicted by the black arrow on H&E staining under ×100 magnification.

Histopathological examination of the H&E stained biopsy specimen taken from the lichenoid papule showed a split in malphigian layer with acantholytic cells [Figure 3a] under ×40 magnification. On high power (×100), dyskeratotic cells were seen within the stratum corneum [Figure 3b].

What is your diagnosis?

Answer:

Diagnosis: Segmental Darier Disease

DISCUSSION

Darier disease is a rare autosomal dominant disorder caused by mutation in ATP2A2 gene, with a prevalence of about 1:30,000–1:100,000.[1] It is clinically characterized by the eruption of dirty warty papules and plaques in a seborrheic distribution. Involvement of the flexural areas may be seen in some. Nails can show V-shaped nicking at the free margin as well as white and red longitudinal bands. Oral mucosa, particularly the palate, may show papules, giving a cobblestone appearance. Furthermore, palmoplantar pits can be present.

The pathogenesis of Darier disease is related to the mutation in the ATP2A2 gene located on chromosome 12, which causes dysfunction of the sarcoplasmic/endoplasmic reticulum Ca^{2+} ATPase pump (SERCA2). Intracellular calcium is essential for signal transduction, disturbance of which leads to abnormalities in keratinization (dyskeratosis) and loss of intercellular adhesions leading to acantholysis.[2]

About 10% of patients of Darier disease can present with the localized disease variants such as unilateral, linear, segmental, and zosteriform.[1] Segmental Darier disease is classified into two phenotypes, Type 1 which is caused by a postzygotic mutation in embryo leading to a mosaic pattern of skin involvement and Type 2 which occurs in patients with generalized Darier disease with additional postzygotic mutation in the other alleles of ATP2A2.[1] Our patient belonged to Type 1.

Various triggering factors have been described such as heat, sweating, friction, infection, and drugs such as lithium and calcium channel blockers.[3,4]

Histopathology shows supra basal split with acantholytic cells and presence of dyskeratotic cells like grains in stratum corneum (corps grains) and ronds in stratum spinosum (corps ronds).

Treatment options include avoiding triggering factors, intermittent topical steroids, topical retinoids, and topical antibiotics. For extensive disease, oral retinoids, cyclosporine, or oral doxycycline can be used.

Our patient showed improvement with oral retinoids and topical steroids.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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