Dermoscopy: A Valuable Tool in the Diagnosis of Flexural Acantholytic Dyskeratosis Disorders

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A 44-year-old man with skin phototype V, presented with multiple pigmented lesions over the neck and bilateral axilla from childhood with history of occasional blistering. Cutaneous examination revealed multiple pigmented discrete and confluent keratotic scaly papules and plaques over the axilla and lateral aspect of the neck [Figure 1]. Examination of palms, soles, scalp, nails, and oral mucosa did not reveal any abnormality.

Dermoscopic examination (HEINE DELTA 20®,10 × magnification) under nonpolarized contact dermoscopy over both axilla and neck revealed a targetoid pattern; a central variable shaped crater with angulated margin, surrounded by a white homogenous area/border and peripheral blue-gray pigmentation and radial streaks. The central crater at places had a gray-white homogenous area, brown dots, and dotted and linear vessels [Figure 2]. White dots corresponding to normal eccrine duct openings and brown keratotic plugs of crusting were also seen.

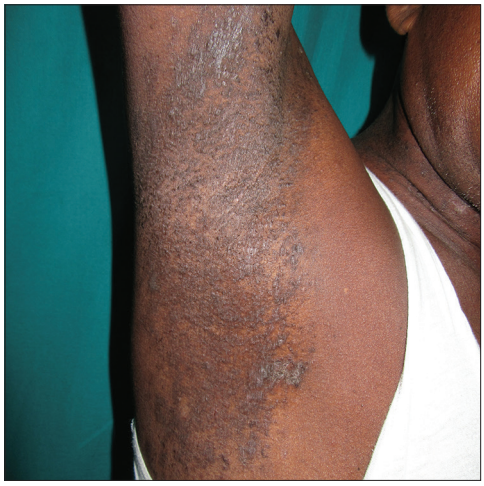

Figure 1: Discrete and confluent pigmented keratotic scaly papules and plaques over the axilla

Histopathological examination showed epidermal hyperplasia, supra-basal cleft, villi, acantholysis with multiple acantholytic cells in the lacunae, and occasional dyskeratotic cells consistent with Hailey-Hailey disease [Figure 3a and b].

The early lesion of Hailey-Hailey disease (HHD) is typified by the presence of multiple erythematous to pigmented keratotic scaly papules over the flexures. As the disease progress, vesiculo-pustules and crusted erosions appear due to the friction, heat, and secondary infection.[1] The early lesions, as in our case, can be mistaken for other disorders like atopic dermatitis, inverse psoriasis, pemphigus vegetans, candidal intertrigo, Fox-Fordyce disease, follicular Dowling-Degos Disease, and Galli-Galli disease. Other acantholytic dyskeratotic disorders that can mimic HHD are flexural Darier disease and Grover's disease. Dermoscopy of vesiculo-pustules in HHD has shown pinkish-white areas with pink furrows and whitish areas in

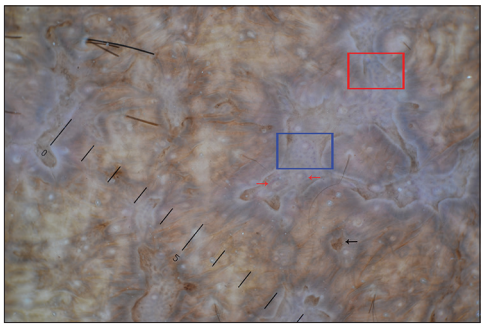

Figure 2: Dermoscopy (HEINE DELTA20® non-polarized contact dermoscope, 10 × magnification) showing a central irregular crater with angulated margin, surrounded by a white homogenous area/border and peripheral blue-gray pigmentation and radial streaks. The floor of the crater has gray-white homogenous area interspersed with gray dots (red square) along with brown dots (black arrow) and dotted (blue square) and linear vessels (red arrows)

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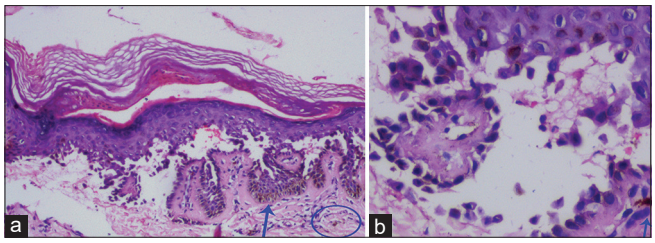

Figure 3: (a) Epidermal hyperplasia and supra-basal cleft with increased basal pigementaion (arrow) and pigment incontinence (circle) (H and E,10 ×). (b) Villi, acantholysis with multiple acantholytic cells in the lacunae and few dyskeratotic cells. (Arrow pointing pigment incontinence) (H and E,40 ×)

crumpled fabric appearance.[2] Dermoscopic features described for Darier's disease are central roundish or polygonal yellow/brown areas with white halo surrounded by pinkish homogenous area, and giant pseudocomedones (dilated oval openings with raised or flat borders and central brown/yellow hyperkeratotic plugs).[3]

Darier-like type of Grover's disease showed polygonal, star-shaped yellowish/brownish areas with a peripheral whitish halo on dermoscopy.[4] Dowling Degos disease is another flexural disorder that can have an irregular star-shaped brown outline on a red-brown background on dermoscopy, and Galli Galli disease, which is its acantholytic variant has been reported to have central brown, mottled area surrounded by whitish halo, similar to that of afore mentioned conditions.[5,6] Thus, all these flexural disorders with acantholytic dyskeratosis have overlapping reproducible features on dermoscopy. This helps in differentiating these disorders from other flexural conditions like inverse psoriasis (regularly distributed dotted vessels in the absence of scales), eczema including atopic dermatitis (yellowish crusts and dotted vessels in patchy distribution in acute stage, white scales and clusters of red dots in chronic stage), Fox-Fordyce disease (hair follicle-centered papules, traumatized terminal hairs, and blackheads) and Langerhans cell histiocytosis (reddish-purple areas in a geographic pattern and brown dots).[7-9]

Burge, who did an extensive study on HHD, has pointed out that the morphology of lesions can vary immensely, and misdiagnosis is common even in patients with positive family history.[1] Dermoscopy offers a quick and reliable way to bring to the fore a diagnosis of HHD while ruling out some close differential diagnoses. The central variable-shaped crater corresponds to the erosion produced by acantholytic dyskeratois process, white homogenous area/border to acathotic epidermis, and the blue-gray colour corresponds to the increased epidermal pigmentation/pigment incontinence. It should be kept in mind that although dermoscopy may give a clue in the diagnosis of HHD, biopsy is mandatory to rule out other flexural disorders with acantholytic dyskeratosis as dermoscopic findings may overlap in these conditions. In conclusion, a dermoscopic pattern of targetoid appearance can be a useful tool in differentiating HHD from other flexural mimics.

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Conflicts of interest

There are no conflicts of interest.

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