Follicular Dowling Degos Disease with Lichen Planus in Two Siblings: A Rare Association

Pooja Agarwal, Ashish Jagati, Sabha Neazee, Shefali V. Patel

Department of Dermatology, SCL Hospital, N.H.L. Medical College, Ahmedabad, Gujarat, India

Sir,

Dowling-Degos disease (DDD) is a rare autosomal dominant genodermatosis, characterized by multiple, small, asymptomatic, bilaterally symmetrical, progressive, pigmented macules, pitted scar or comedo-like lesions; over the face, neck, arms, trunk, and intertriginous areas. It has also been referred to as "Pigmented reticulate anomaly of flexures".[1] Its association with perioral pitted scars, epidermoid/trichilemmal cysts, chloracne-like lesions, hidradenitis suppurativa, seborrheic keratoses, palmar pigmentation, and rosacea-like lesions (Haber's syndrome) have been reported in the literature.[2] We hereby present a case of Familial follicular Dowling Degos with lichen planus in two brothers.

A 28-year-old man visited our outpatient clinic with chief complaints of intensely pruritic skin lesions over the lower trunk for 15 days. On cutaneous examination, multiple discrete, polygonal, 1–3 mm diameter papules with violaceous hue over back and lower abdomen were noted. Clinical diagnosis of lichen planus was made [Figure 1]. On further examination multiple asymptomatic, comedo-like lesions and atrophic pitted scars over face, upper chest, and back were noted. History suggested that these asymptomatic lesions were present since childhood. He was accompanied by his 23-year old younger brother who had lesions suggestive of LP over trunk for 10 days and comedo-like and pitted scar like lesions over face, trunk, and flexures since childhood [Figure 2a-d]. Dermoscopy of the comedo-like lesion from face showed thick hyperkeratotic, hyperpigmented follicular plug [Figure 3a and b]. Biopsy from two representing lesions, papular lesion, from trunk and pitted scar-like lesion from face of both brothers. Biopsy from face showed dilated hair follicles with keratinous plug in the infundibulum, elongated rete ridges, focal increase in melanocyte, and horn cysts like changes in the epidermis with normal interfollicular epithelium. Histological changes were suggestive of follicular Dowling degos [Figure 4a-d]. Lesion from lower back showed irregular epidermal hyperplasia with lymphocytic infiltrate at dermo-epidermal junction suggestive of lichen planus [Figure 5a-c]. Based on clinical histological and dermatoscopic finding final diagnosis of follicular Dowling Degos associated with lichen planus was made.

DDD is a rare genodermatosis transmitted by autosomal dominant pattern but can also occur sporadically. It was first described initially by Dowling and Freudenthal in 1938 and Degos and Ossipowski in 1954.[3] It is due to dysfunctional mutations in the keratin 5 gene (KRT5) located on chromosome 12q leading to haploinsufficiency in the keratin gene resulting in defective melanosome uptake and defect in hair follicle structure and sebaceous glands.[4]

The affected patients are usually females in their thirties or forties having acquired progressive hyperpigmentation predominantly in flexures (axillae, groins, intergluteal and inframammary folds, anti-cubital fossa, neck), whereas trunk, arms, face, scalp, and genitalia are also involved in some cases. Isolated involvement of the genitalia has also been reported.[3] Other notable features include pitted follicular, perioral, and facial scars in patients with no previous history of

Address for correspondence:
Dr. Ashish Jagati,
OPD Room No. 16, Department of Dermatology,
Smt NHL Medical College and SCL General Hospital Saraspur, Ahmedabad - 380 018, Gujarat, India.
E-mail: jagatiashish@gmail.com

Access this article online

Website: www.idoj.in

DOI: 10.4103/idoj.IDOJ_309_20

Quick Response Code:

How to cite this article: Agarwal P, Jagati A, Neazee S, Patel SV. Follicular dowling degos disease with lichen planus in two siblings: A rare association. Indian Dermatol Online J 2021;12:459-61.

Received: 30-Apr-2020. **Revised:** 11-Jun-2020. **Accepted:** 22-Aug-2020. **Published:** 12-May-2021.

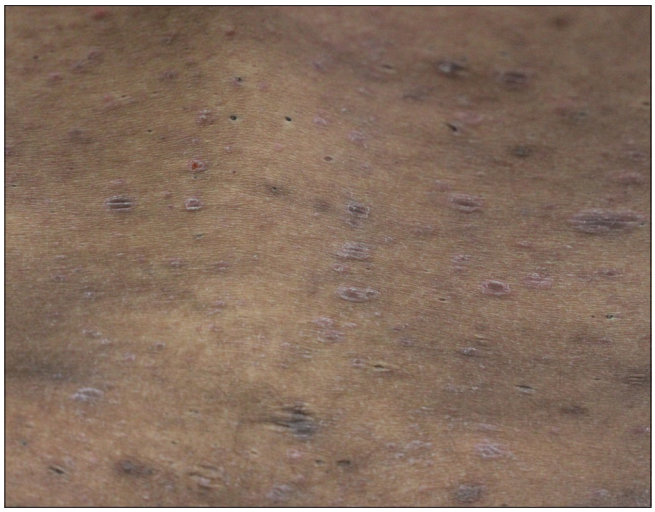

Figure 1: Multiple discrete, polygonal, violaceous papules over back

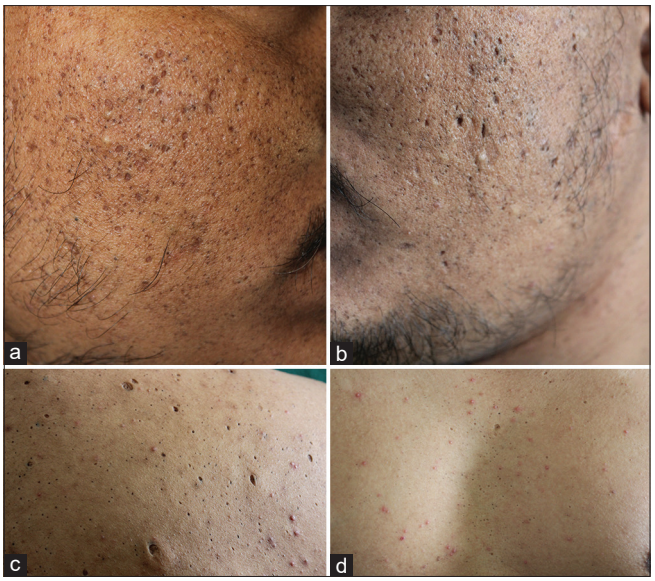

Figure 2: Multiple asymptomatic, comedo-like lesions and atrophic pitted scars over. (a and b) Face (c) Back (d) Upper chest

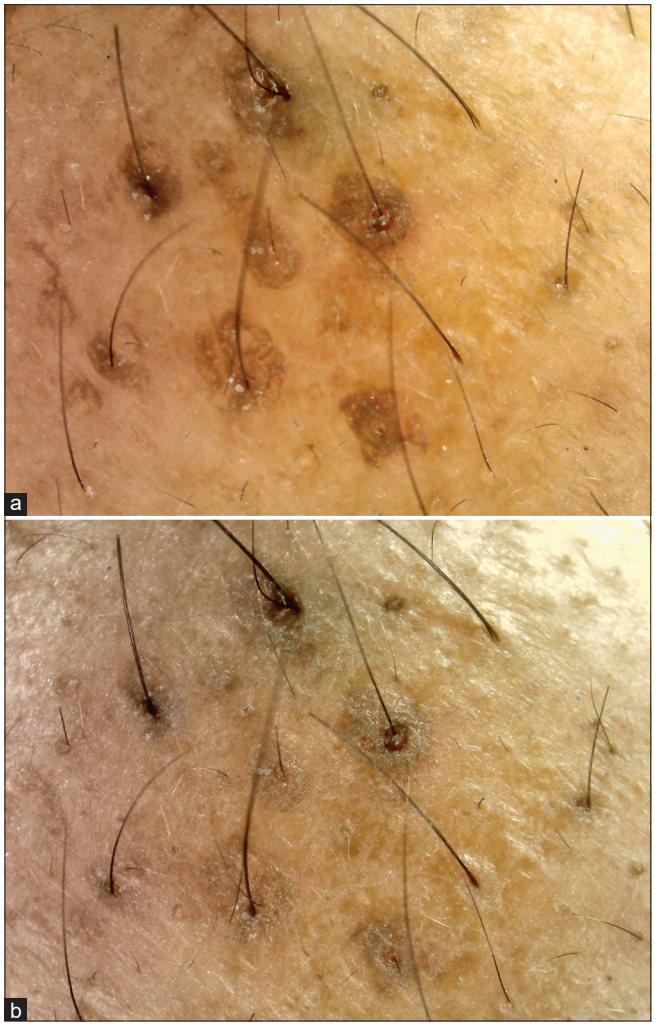

Figure 3: Thick hyperkeratotic, hyperpigmented follicular plug on dermoscopy (a) polarized (b) non-polarized

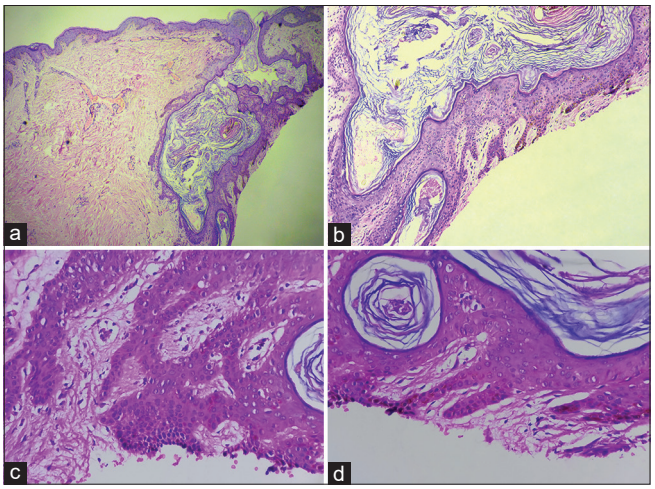

Figure 4: Elongated rete ridges, focal increase in melanocyte and horn cysts like changes in the epidermis dilated hair follicles with keratinous plug in the infundibulum on histology (H and E; a 40x, b 100x, c and d 400x)

acne and hyperkeratotic comedo-like follicular papules in the neck and/or back (dark dot follicles). It is usually asymptomatic but pruritus of the affected areas can be present. Histopathology of the lesion demonstrates dusky, dappled disfigurements, and dark dot depressions and disclosing digitate down growth delving dermally giving an antler like appearance.[5]

Variants of DDD are generalized as galli-galli disease-(acantholytic cell Variant), follicular DDD, haber`s syndrome (pigmented keratotic papules on axilla, neck, and torso with pitted scars on face and persistent facial erythema), pigmentation reticularis Faciei et -Colli.[2] There are several reports of an increased prevalence of epidermal cysts, hidradenitis suppurativa, keratoacanthomas, and perianal squamous cell carcinoma among those diagnosed with DDD.[6] DDD have been reported in several rare association in past like Hidradenitis suppuritiva, Dariers disease, Reticulate acropigmentation of kitamura, Malignant

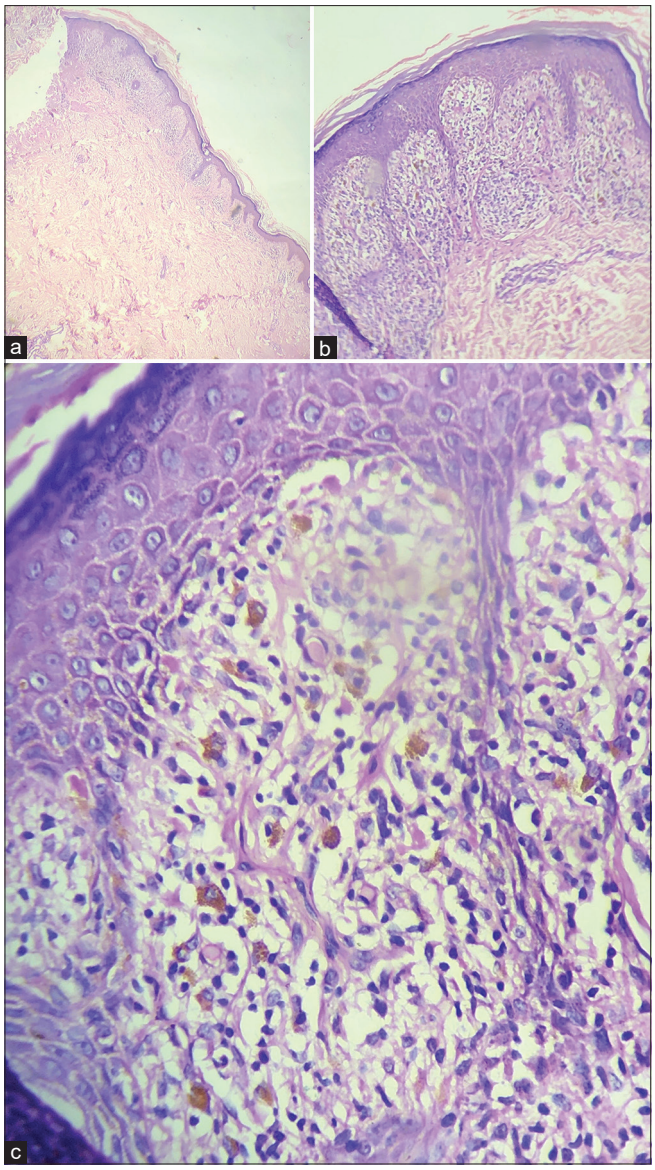

Figure 5: Irregular epidermal hyperplasia with lymphocytic infiltrate at dermoepidermal junction (H and E; a 40x, b 100x, c 400x)

melanoma, and Psoriasis.[7-11] To best of our knowledge simultaneous overlap of follicular Dowling Degos and lichen planus has not been reported.

To conclude, an uncommon entity like Dowling Degos, though limited to skin and relatively harmless, should be diagnosed promptly as it has multiple variants and have increased risk of other concerning cutaneous diseases.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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