RESEARCH PAPER

Dowling–Degos Disease with Facial and Flexural Involvement: A Clinicodermoscopic Case

Dr. Yogalakshmi M[1], Dr. Nourin Basheer[2], Dr. Sowmya P[3]

[1,2,3]Junior Resident, Department of DVL, Sree Balaji Medical College and Hospital, Chennai, Tamilnadu, India.

Corresponding author addresses:

Dr. Yogalakshmi M, Junior Resident, Department of DVL, Sree Balaji Medical College and Hospital, Chennai, Tamilnadu, India.

Email: myog1998@gmail.com

Received Date: 22/03/2025 | Accepted: 28/03/2026 | Published Date: 25/04/2026

ABSTRACT

Background: Dowling–Degos disease (DDD) is a rare genodermatosis characterized by progressive reticulate hyperpigmentation predominantly affecting flexural areas. Although classical involvement includes axillae and other flexures, facial lesions are uncommon and may mimic freckles or lentigines, posing diagnostic challenges. Dermoscopy has emerged as a valuable non-invasive tool for identifying characteristic features of this condition.

Case Presentation: We report a case of a 24-year-old male presenting with asymptomatic, gradually progressive hyperpigmentation over flexural areas and face. Clinical examination revealed reticulate hyperpigmented macules over the axillae and antecubital fossae, along with multiple freckle-like macules over the face. Dermoscopic evaluation demonstrated irregular reticulate pigmentation, follicular plugging, and brown antler-like projections, consistent with Dowling–Degos disease.

Management and Outcome: The patient was managed with topical tretinoin, hydroquinone, and strict photoprotection. At follow-up, mild improvement in facial pigmentation was observed, while flexural lesions remained largely unchanged.

Conclusion: This case highlights an atypical presentation of Dowling–Degos disease with facial involvement and emphasizes the importance of dermoscopy in diagnosis. Early recognition helps avoid misdiagnosis and unnecessary interventions.

Keywords: Dowling–Degos disease, Reticulate pigmentation, Dermoscopy, Facial hyperpigmentation, Flexural pigmentation.

How to cite this article: Yogalakshmi M, Basheer N, Sowmya P. Dowling–Degos Disease with Facial and Flexural Involvement: A Clinicodermoscopic Case. Int J Drug Deliv Technol. 2026;16(54s): 1-4. DOI: 10.25258/ijddt.16.54s.1

Source of support: Nil.

Conflict of interest: None

INTRODUCTION

Dowling–Degos disease (DDD) is a rare, autosomal dominant genodermatosis characterized by progressive reticulate hyperpigmentation predominantly affecting flexural areas such as the axillae, groin, inframammary regions, and antecubital fossae. The condition, which was first independently reported by Dowling and Freudenthal (1938) and Degos and Ossipowski (1954), is thought to be a follicular keratinization disorder with subsequent pigmentary change [1,2]. Clinically, comedo-like lesions and pitted perioral scars are frequently seen together with hyperpigmented macules organized in a lace-like or reticular pattern[3]. Mutations in genes controlling epidermal development, including KRT5, which codes for keratin 5, a structural protein necessary for basal keratinocyte integrity, have been connected to the molecular etiology of DDD [4]. Notch signaling pathways that control melanocyte–keratinocyte interactions may be disrupted by further mutations involving POFUT1 and POGLUT1 [5]. The distinctive reticulate pigmentation is caused by these anomalies, which lengthen rete ridges and promote melanin deposition in the basal layer [6].

Atypical presentations are becoming more widely acknowledged, even though flexural involvement is thought to be the hallmark of DDD. In particular, facial involvement is rare and can be difficult to diagnose since it might resemble other pigmentary conditions including freckles, lentigines, or post-inflammatory hyperpigmentation [7]. Facial lesions might indicate a different version of the illness or an expanded phenotype. A useful non-invasive method for assessing pigmentary abnormalities, such as DDD, is dermoscopy. Follicular pits or plugs, irregular brown projections, and a reticulate pigment network that histologically correlates to branching rete ridges are characteristic dermoscopic characteristics [8]. These results help differentiate DDD from its clinical mimickers, including dyschromatosis universalis hereditaria and acanthosis nigricans.

Histopathologically, DDD is characterized by follicular involvement without considerable inflammation, increased basal pigmentation, and extension and branching of rete ridges that resemble "antler-like" patterns [6]. However, when dermoscopic and clinical results are usual, biopsy is not always required. DDD is frequently misdiagnosed or underdiagnosed due to its rarity and

inconsistent presentation. In order to better identify aberrant presentations, particularly those involving non-flexural parts like the face, it is crucial to report such instances with clinicodermoscopic correlation. We describe a case of Dowling-Degos disease with considerable face pigmentation and characteristic flexural involvement, emphasizing the need of dermoscopy in making the diagnosis.

CASE PRESENTATION

A 24-year-old male presented with gradually progressive hyperpigmentation over flexural areas and face for the past 6–8 years. There was no pain, drainage, or itching connected with the lesions. It began subtly in late adolescence and progressed slowly in both severity and intensity. There was no prior history of photosensitivity, systemic disease, medication use, or inflammation. There was no contribution from family history. Upon cutaneous inspection, the axillae and antecubital fossae showed many hyperpigmented macules grouped in a reticulate pattern. Skin marks were somewhat accentuated, and the pigmentation was more noticeable inside skin folds. There was no evidence of induration, scaling, or erythema.

A facial inspection revealed a large number of distinct, symmetrically distributed dark brown macules that resembled freckle-like coloration over the nose, cheeks, and periorbital area. Dermoscopy of the lesions revealed brown star-shaped projections with perifollicular accentuation, follicular plugging, and uneven reticulate pigmentation, all of which are indicative of Dowling-Degos disease. A diagnosis of Dowling-Degos disease with facial and flexural involvement was obtained based on dermoscopic findings and clinical morphology.

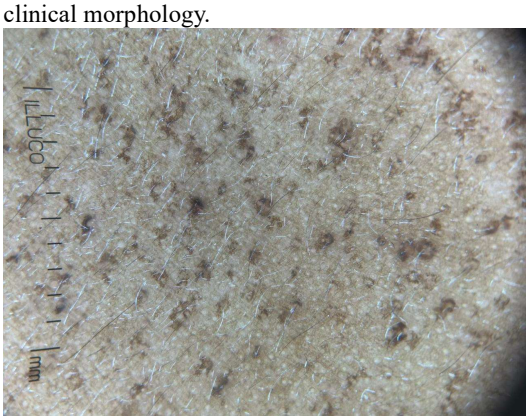

Figure 1: Dermoscopic Features of Dowling–Degos Disease

This dermoscopic image demonstrates a characteristic reticulate (net-like) pattern of hyperpigmentation composed of irregular brown interconnected lines and projections. Dark keratin plugs, which look as brown to black specks and highlight several follicular apertures, indicate follicular involvement. In addition, there are branching projections that resemble stars and antlers, which histopathologically match long, branching rete ridges. The backdrop appears reticular generally because to the diffuse light brown coloration. These results help distinguish Dowling-Degos disease from other pigmentary conditions like freckles or acanthosis nigricans and are very indicative of the condition.

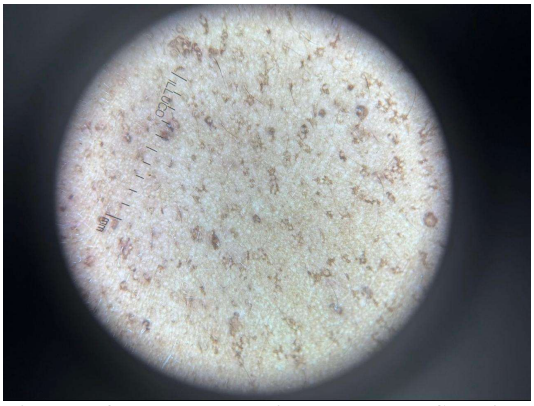

Figure 2: Dermoscopic Pattern Showing Reticulate Pigmentation in Dowling–Degos Disease

A diffuse reticulate network of light to dark brown pigmentation forms an uneven mesh-like structure in this dermoscopic picture (circular field view). Keratin plugs, which show up as dark brown to black specks and accentuate numerous follicular apertures, indicate significant follicular involvement. Long, branching, antler-like projections that resemble elongated rete ridges on histopathology are scattered throughout the network. There are sporadic fine vellus hairs visible in the background, which has a uniform light brown color.

These dermoscopic characteristics help distinguish Dowling-Degos disease from other pigmentary illnesses that lack this unique follicular and reticulate pattern, such as freckles, lentigines, or acanthosis nigricans.

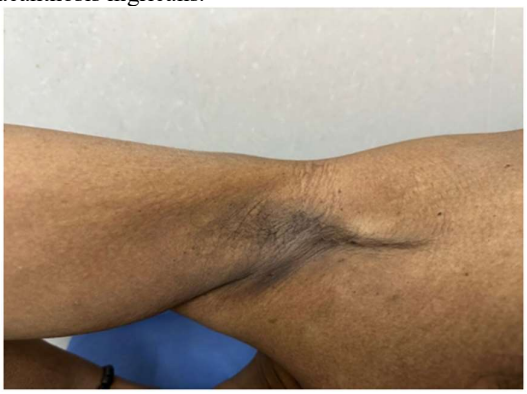

Figure 3: Flexural Reticulate Hyperpigmentation in Dowling–Degos Disease

Clinical images showing reticulate hyperpigmentation over axilla and antecubital fossa. Along skin creases, pigmentation is accentuated, and hair-bearing follicles are somewhat thicker and more

prominent. It can be distinguished from acanthosis nigricans by the absence of erythema, scaling, or velvety texture.

Figure 5: Clinical images showing bilateral facial hyperpigmented macules in Dowling–Degos disease.

This composite figure includes three views (left lateral, right lateral, and frontal) demonstrating facial involvement:

Left lateral view: displays a number of distinct, dark brown macules on the side of the nose and cheek. The lesions have a speckled appearance and are folliculocentric and uniformly distributed.

Right lateral view: confirms bilateral involvement, which is characteristic of pigmentary diseases like DDD, by revealing a comparable symmetrical distribution of hyperpigmented macules.

Frontal view: features many freckle-like macules and diffuse involvement of the middle face, particularly the nose and malar areas. There is no erythema or scaling, and the pigmentation is consistent.

Although the lesions generally resemble ephelides (freckles), the diagnosis of Dowling-Degos disease is supported by the concomitant flexural involvement and dermoscopic findings. Even though it is less frequent, facial involvement is an extended manifestation of the illness.

Treatment

Management of Dowling–Degos disease remains largely symptomatic and focused on cosmetic improvement, as no definitive curative therapy exists. In order to improve epidermal turnover and lessen pigmentation, the patient in this instance was started on topical tretinoin (0.025%) used every night. Topical hydroquinone (2%) was recommended as a depigmenting treatment for facial lesions in order to prevent the formation of melanin. Strict photoprotection with a broad-spectrum sunscreen (SPF ≥30) was also recommended for the patient in order to stop further pigmentary accentuation. General precautions were also advised, such as reducing friction in flexural regions and practicing good cleanliness. Although they were postponed at first, sophisticated treatments including chemical peels (glycolic acid, salicylic acid) and laser modalities (Q-switched Nd:YAG laser, fractional lasers) were suggested as possible choices for refractory or cosmetically unpleasant lesions.

Outcome and Follow-up

The patient showed a slight but discernible improvement in face hyperpigmentation at the three-month follow-up, with a decrease in the severity of macules across the nose and cheeks. Flexural lesions, on the other hand, responded very little, which is in line with the fact that DDD is known to be resistant to therapy in these regions. There were no new lesions found, and the pigmentation that was already there did not worsen. The patient expressed satisfaction with the minor cosmetic improvement and good adherence to treatment. There were no side effects associated with topical drugs. The condition usually has a chronic, slowly progressing course with varied response to medication, thus the patient was recommended to continue treatment and maintain long-term follow-up.

DISCUSSION

Dowling–Degos disease is a rare pigmentary disorder with characteristic flexural reticulate hyperpigmentation. The current example shows both considerable face pigmentation, a rare but increasingly recognized characteristic, and classical involvement of flexures [7]. In DDD, facial lesions might resemble freckles or lentigines, which frequently results in an incorrect diagnosis. Melanosome transfer and keratinocyte differentiation are impacted by mutations in KRT5, POFUT1, and POGLUT1[4,5]. The classic reticulate pattern seen clinically and dermoscopically is caused by these molecular anomalies, which also cause basal layer hyperpigmentation and rete ridge elongation [6]. Dermoscopy is an essential clinical tool, particularly for unusual appearances. The dark projections, uneven reticulate pigmentation, and follicular pits observed in our patient are diagnostic findings that coincide with histological characteristics [8]. This non-invasive technique aids in preventing needless biopsies.

Acanthosis nigricans, freckles, lentigines, and Galli-Galli illness are examples of differential diagnosis. Acanthosis nigricans has velvety thickening instead of reticulate pigmentation, in contrast to DDD. Flexural predominance and dermoscopic follicular alterations are absent in freckles and lentigines [3]. There is no known cure for DDD, and treatment is still difficult. Hydroquinone, laser treatments, and topical retinoids all offer varying degrees of cosmetic improvement [9]. Patient counseling and long-term follow-up are necessary due to the disease's chronic and progressive character. This example demonstrates the significance of identifying unusual DDD manifestations and the function of dermoscopy in verifying diagnosis. Misdiagnosis and needless procedures can be avoided with greater understanding.

CONCLUSION

Dowling–Degos disease is a rare pigmentary disorder with characteristic flexural involvement but

may present with atypical features such as facial pigmentation. The significance of clinicodermoscopic correlation in making a diagnosis, especially in atypical presentations, is shown by this case. A useful non-invasive diagnostic method that improves accuracy and lessens the necessity for biopsy is dermoscopy. Since the condition is benign but has major cosmetic effects, early detection and adequate counseling are crucial. Reporting such occurrences helps doctors distinguish it from other pigmentary illnesses and advances our understanding of the disease spectrum.

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