

Egyptian Dermatology Online Journal

Volume 6 Number 1

Dowling Degos Disease

H. Gammaz, H. Amer, F. Ibrahim & M. Bagdady

Egyptian Dermatology Online Journal 6 (1): 11

AL- Haud Al- Marsoud Hospital, Cairo, Egypt

Submitted: April 15, 2010

Accepted: May 1, 2010

e-mail: hananderma@hotmail.com

Summary:

A 30 years old female presented with small, rounded, pigmented macules that resemble freckles which were affecting her face, infra-mammary regions and axillae. Histopathological examination revealed hyperkeratosis, acanthosis, papillomatosis and hypermelanosis.

Introduction:

Dowling Degos disease is a rare condition inherited via an autosomal dominant gene. It presents during adult life most frequently in the 4th decade as numerous, small, rounded pigmented macules that resemble freckles. The axillae and groins are the usual sites, although other areas may be involved including inter-gluteal folds, infra-mammary folds, neck, scalp, trunk and arms [1]. Involvement of genitalia has been described [2]. Pigmentation is symmetrical and progressive but otherwise asymptomatic [1].

Case presentation:

A 30 years old female presented to our outpatient clinic at Al-Haud Al-Marsoud Hospital complaining of progressive small, rounded, pigmented macules that resemble freckles. The lesions were found affecting her face (**fig. 1**), both infra-mammary regions (**fig. 2**) and axillae (**fig. 3**). On examination, we found comedo like lesions, pitted acneform scars near the angles of the mouth (**fig. 4**).



Fig 1: Small, round, pigmented macules that resemble freckles.



Fig 2: Pigmented macules affecting the infra-mammary areas.



Fig 3: Pigmented macules affecting axillae.

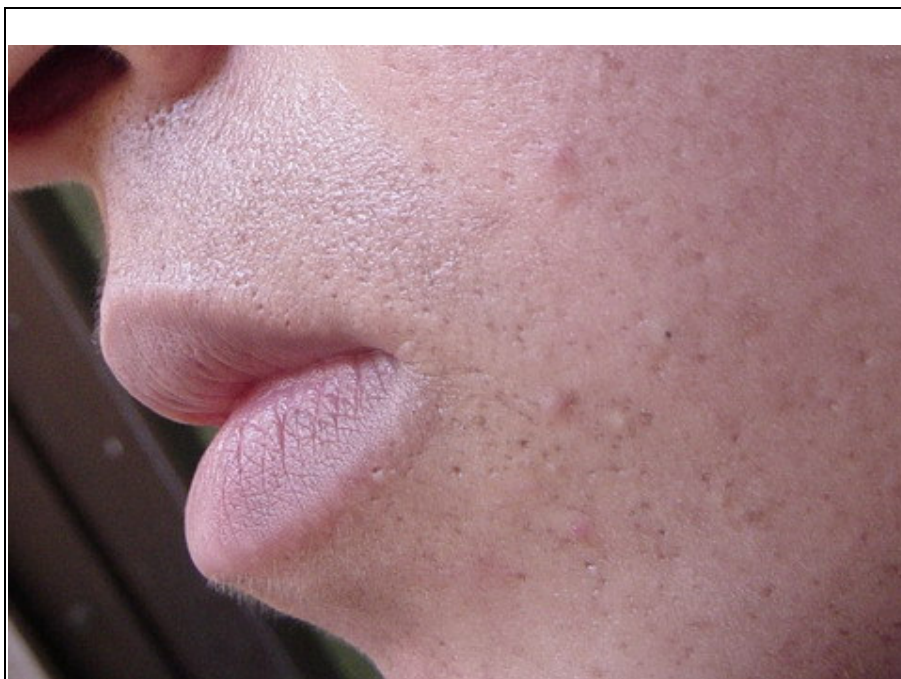


Fig 4: Comedo- like lesions and pitted acne-form scars near the angles of the mouth.

Our differential diagnoses included Dowling Degos disease, acanthosis nigricans, multiple basal cell papillomas, epidermal nevi and axillary freckles of neurofibromatosis.

Histopathological examination of the specimen which was stained by Haematoxylin & Eosin (**fig. 5-7**) revealed hyperkeratosis, acanthosis, papillomatosis and hypermelanosis. Also there was branching heavily melanized epidermal downward proliferation arising from the lower border of the epidermis and small horn cysts were found within the epidermal proliferation. The histopathological findings confirmed that we were dealing with a case of Dowling Degos disease.

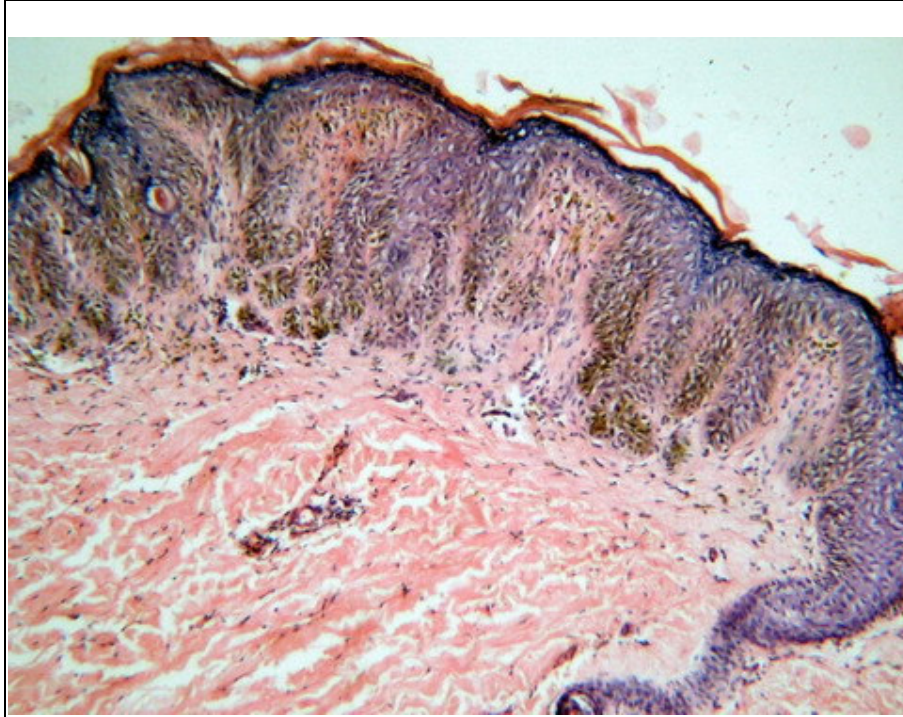


Fig 5: H&E stained section showing hyperkeratosis, acanthosis, papillomatosis and hypermelanosis.

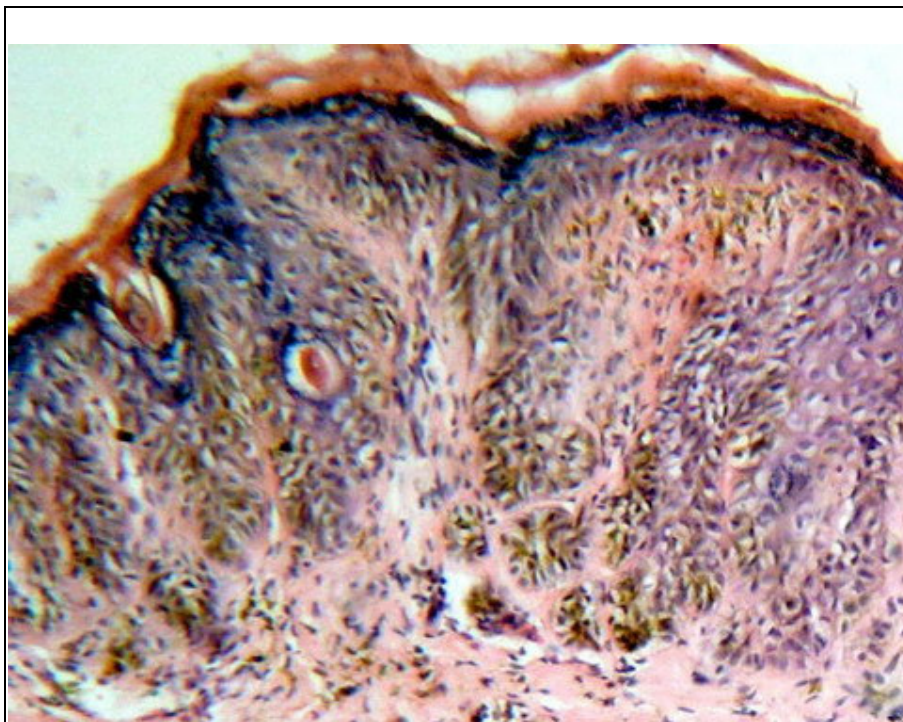


Fig 6: Branching heavily melanized epidermal downward proliferations arising from the lower border of epidermis.

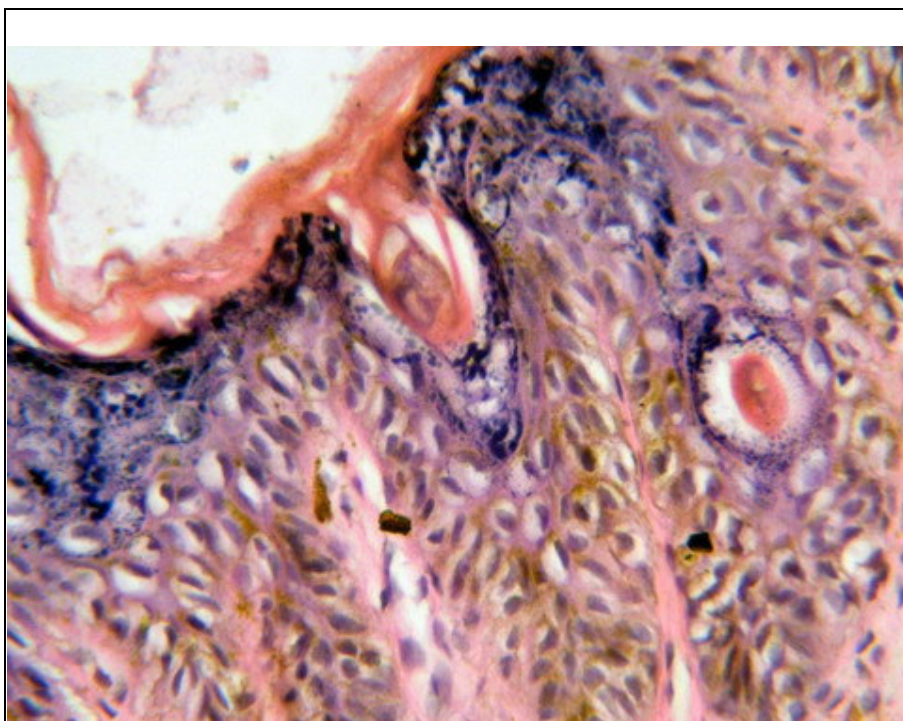


Fig 7: Small horn cysts within the epidermal proliferations.

Discussion:

In Dowling Degos disease, the degree of pigmentation varies, but in some patients the lesions are almost confluent giving a brown or black lace like pattern [3]. Other reported features are comedo- like lesions (dark dot follicles) and pitted acneform scars occurring near the angle of the mouth [4].

Reported associations of the disease are hidradinitis suppurativa, mental retardation or trichilemmal cysts [5].

It has been suggested that Dowling Degos disease is one part of a spectrum of conditions that are characterized by reticulate pigmentation, this would include some autosomal dominant conditions as Kitamura's acropigmentation reticularis [6].

Successful treatment of Dowling Degos disease with Er: YAG laser pulse energy between 1000 and 1200 mJ three consecutive passes leads to good results [7].

References

1. Wu YH, Lin YC. Generalized Dowling-Degos disease. J Am Acad Dermatol 2007; 57(2): 327- 334.
2. Milde P, Goerz G, Plewing G. Dowling Degos disease with exclusively genital manifestation. Hautarzt. 1992 Jun; 43(6) 369- 372.
3. Oiso N, Tsuruta D, Ota T, Kobayashi H, Kawada A. Spotted and rippled reticulate hypermelanosis: a possible variant of Dowling-Degos disease. Br J Dermatol 2007; 156(1): 196- 198.
4. Georgescu EF, St?nescu L, Popescu CF, Com?nescu M, Georgescu I. Dowling-Degos disease. Rom J Morphol Embryol 2010; 51(1): 181- 185.
5. Dixit R, George R, Jacob M, Sudarsanam TD, Danda D. Dowling-Degos disease: Hidradinitis suppurativa and arthritis in mother and daughter. Clin Exp Dermatol 2006; 31(3): 454- 456.
6. Thami GP, Jaswal R, Kanwar AJ, Radotra BD, Singh IP. Overlap of reticulate acropigmentation of Kitamura and Dowling-Degos disease in four generations. Dermatol 1998; 196(3): 350- 351.
7. Wenzel J, Tappe K, Gerdson R, Uerlich M, Philipp-Dormston W, Bieber T, Petrow W. Successful treatment of Dowling-Degos disease with Er:YAG laser. Dermatol Surg 2002; 28 (8): 748- 750.