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DOWLING – DEGOS DISEASE – A CASE REPORT

Jayakar Thomas[1*], Hemadharshini B[2], Tamilarasi S[2], Asha D[2], Manoharan D[3]

[1]Professor and Head, [2]Residents, [3]Professor, Department of Dermatology,
Sree Balaji Medical College and Bharath University, Chennai, Tamilnadu, India.

Corresponding Author: - **Jayakar Thomas**
E-mail: jayakarthomas@gmail.com

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ABSTRACT
Dowling-Degos disease is a rare pigmentary disorder characterized by reticulate pigmented macules of the flexures. Many variants of this disease and its overlap with other reticulate pigmentary disorders have been reported in the literature. We report a case of 50-year-old man who presented with reticulate pigmentation of almost all the flexural creases and perioral pits.

INTRODUCTION

Reticulate pigmentary disorders as a group, characterized by various intensities of brown or black hyper pigmented macules [1]. In many disorders they are angulated, freckle-like and tend to join at their ends to form a reticulate pattern. In some conditions there are hypo pigmented macules in-between areas of hyperpigmentation. Dowling Degos disease is a reticulate pigmentary disorder in which pigmentation are confined to the flexures. Wilson Jones and Grice first discovered it in 1938 [2].

CASE REPORT

A 50- year old male came with complaints of blackish discoloration of skin over the face, neck and trunk for the past 2 months. The lesions were asymptomatic, initially appeared over the neck, which gradually progressed to axilla and groin and to other parts of the body. No history of similar complaints in the family.

Dermatological examination revealed symmetrical reticulate pigmentation of flexures involving face, neck, bilateral axilla, groin and cubital fossa and inframammary creases. Pigmentation was also noted in the lower back and scrotum. Perioral pits were present. Palms and soles were spared. Systemic examinations were normal. Routine investigations done and were within normal limits. A skin biopsy was performed from the pigmentation in the neck. Histopathology revealed hyperkeratosis, acanthosis and one elongated rete ridge related to hair follicle (antler horn like appearance), supra papillary thinning with sparse melanin in the papillae and concentrated melanin at the tip of the rete ridge, which is characteristic of Dowling Degos disease.

DISCUSSION

Synonyms: Dark dot disease, reticulate pigment anomaly of flexures. [3]

Reticulate pigmentary disorders are a group of heterogeneous disorders, comprising of several inherited disorder that are characterized by reticulate pigmentation, other cutaneous and extra- cutaneous malformations. The true reticulate disorders classically include Dyskeratosis congenital, Dowling Degos disease, Naegeli Franceschetti Jadassohn syndrome, Dyschromatosis symmetrica hereditaria, Acropigmentation of Kitamura, X linked reticulate pigmentary disorder and Dyschromatosis universalis hereditaria.

Dowling Degos disease (DDD) is rare disorder of autosomal dominant inheritance [4], with variable

phenotypic expression. A genetic defect in keratin 5(loss of functional mutation) [5] has been reported. It is usually Sporadic. DDD is a progressive pigment disorder, characterized by reticulate pigmentation of the flexures, comedone – like papules on the back and neck. The onset is typically during the third to fourth decade of life. Lesions initially start as a macule; gradually coalesce to form "lace-like" or "reticulate" pigmentation of flexures [6]. This pigmentation is progressive and symmetrical. Lesions usually occur at the sites of groin and axilla initially, which later progress to neck, infra mammary creases, trunk, proximal arms and the antecubital fossa. Perioral pits and comedones are the hallmark of the disease. Occurrences of hyperkeratotic follicular papules of neck and axilla, epidermoid cysts have been reported. Hyperkeratotic papules occur at the sites of friction. The association of hidradenitis suppurativa (acne inversa), keratoacanthoma and squamous cell carcinoma with DDD has been reported [7, 8]. Hypo pigmented macules of the trunk have been also reported as one of the association.

Histology of a skin biopsy taken from a pigmented lesion shows an atrophied epidermis, increased pigmentation of the basal cell layer, finger-like rete ridges with thinning of the suprapapillary epithelium. This results in an "antler-like" pattern [9] arising from the under surface of the epidermis and hair follicles, very characteristic. Dermal melanophages and a perivascular infiltrate composed of lymphocytes and histiocytes are also observed.

Differential diagnosis:

Acanthosis nigricans can be clinically distinguished by velvety plaques and histologically by less pronounced elongation of rete ridges [5]. In addition, there is no follicular involvement. Patients with neurofibromatosis type 1 develop lentigines (oftentimes referred to as 'freckles') in the axillae and groin, but this disorder can be easily differentiated from DDD.

Variants of DDD:

Haber's syndrome is characterized photosensitive rosacea like facial eruptions that develop during adolescence, followed by the appearance of keratotic papules, prominent comedones, pitted scars and by reticulate pigmentation on the trunk and proximal extremities as well as axilla [10]. Galli- Galli disease has the same clinical and histologic features as of DDD, with exception of presence of suprabasal non-dyskeratotic acantholysis in biopsy specimens of lesional skin[11]. Reticulate acropigmentation of kitamura is characterized by slightly depressed, well-demarcated brown/black macules of the extensor surface of the skin, rarely involves flexures.

CONCLUSION

Treatment: Topical hydroquinone, corticosteroids, adapalene and tretinoin have been used with varying success. Effective treatment with the Erbium: YAG laser was reported in one patient.

Figure 1. Clinical picture showing reticulate pigmentation of the neck and face. Perioral pits noted.

Figure 2. Clinical picture showing reticulate pigmentation of the axilla and groin (flexures

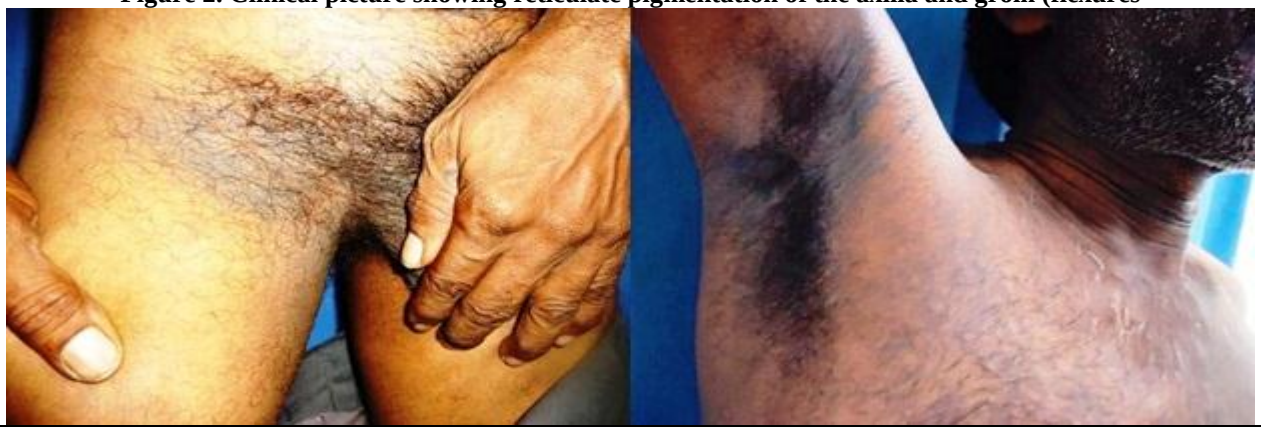

Figure 3. Clinical picture showing mottled pigmentation over the back and inframammamary creases

Figure 4. Histopathology of skin (H&E section, magnification 25x10), showing hyperkeratosis, acanthosis and one elongated rete ridge related to hair follicle (antler horn like appearance)

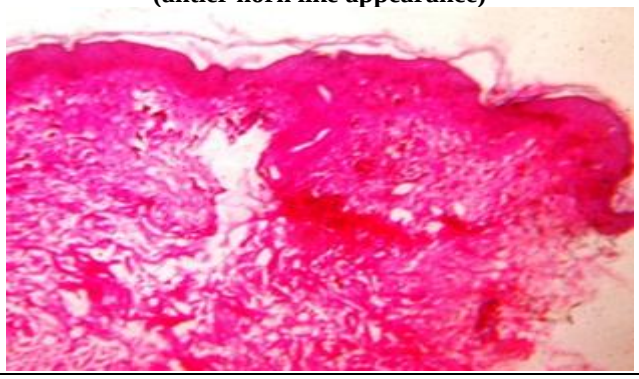

Figure 5. Microscopic view (magnification 40x10) revealing supra papillary thinning with sparse melanin in the papillae and concentrated melanin at the tip of the rete ridge

REFERENCES

1. Griffiths WAD. (1984) Reticulate pigmentary disorder-a review. *Clin Exp Dermatol*, 9, 439-50
2. Wilson-Jones E, Grice K. (1978) Reticulate pigmented anomaly of the flexures. *Arch Dermatol*, 114, 1150-7.
3. Sardana K, Goel K, Chug S. (2013) Reticulate pigmentary disorder. *Indian J Dermatol Venereol Leprol*, 79, 17-29.
4. James WD, Berger TG, Elston DM. (2006) Disturbances of Pigmentation, Pigmented Anomalies of the Extremities. Andrews' Diseases of the Skin Clinical Dermatology. 11th ed. China, Elsevier's, 855-6.
5. Bolognia JL. (2000) Disorders of hypopigmentation and hyperpigmentation. In, Harper J, Orange H, Prose N, editors. Textbook of Pediatric Dermatology. 2nded. United States, Blackwell Science, 868-70.
6. Esterly NB, Basalge E. (2006) Genetic Epidermal Syndromes, Disorders characterized by Reticulated Hyperpigmentation. In, Nordlund JJ, editor. The pigmentary system, physiology and pathophysiology. 2nd ed. United States, Blackwell Science, 780-808.
7. Li M, Hunt MJ, Commens CA (1997). Hidradenitis suppurativa, Dowling Degos disease and perianal squamous cell carcinoma. *Australas J Dermatol*, 38, 209-11.
8. Fenske NA, Groover CE, Lober CW, Espinoza CG. (1991) Dowling-Degos disease, hidradenitis suppurativa, and multiple keratoacanthomas. A disorder that may be caused by a single underlying defect in pilosebaceous epithelial proliferation. *J Am Acad Dermatol*, 24, 888-92.
9. Kim YC, Davis MD, Schanbacher CF, Su WP. (1999) Dowling-Degos disease (reticulate pigmented anomaly of the flexures), A clinical and histopathologic study of 6 cases. *J Am Acad Dermatol*, 40, 462-7.
10. Wenzel G, Petrow W, Tappe K, Gerdsen R, Uerlich WP, Bieber T. (2009) changing a concept-controversy on the confusing spectrum of the reticulate pigmented disorders of the skin. *J Cutan Pathol*, 36, 44-8.
11. Hanneken S, Rütten A, Pasternack SM, Eigelshoven S, El Shabrawi-Caelen L, Wenzel J, *et al*. (2010) Systematic mutation screening of KRT5 supports the hypothesis that Galli-Galli disease is a variant of Dowling-Degos disease. *Br J Dermatol*, 163, 197-200.