

DOWLING DEGOS¹*Dr. Nandita Patel (MBBS), ²Dr. Chirag Desai (MBBS) (DDVL) and ³Dr. Sharmila Patil (MD DDV)

Department of Dermatology, Venerology and Leprosy, Dr. D Y Patil Hospital, School of Medicine, Nerul, Navi Mumbai, Maharashtra, India.

***Corresponding Author: Dr. Nandita Patel (MBBS)**

Department of Dermatology, Venerology and Leprosy, Dr. D Y Patil Hospital, School of Medicine, Nerul, Navi Mumbai, Maharashtra, India.

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ABSTRACT

A 45 years old female presented with multiple, round, light and dark coloured lesions on her body since 15 years. There were no other systemic complaints. On cutaneous examination multiple, asymptomatic, small, round, symmetrically distributed, hypopigmented to dark brown and black coloured macules were present over the neck, axilla, flexures, trunk and groins. There were also multiple, hyperpigmented papules present over the face and trunk. Additionally, multiple comedo like lesions were present over the face and nape of neck. The lesions were increasing in number with the age of the patient. The patient was a known case of hyperthyroidism for 3 years and was on treatment for the same. On detailed questioning, it was revealed that the patient had a family history for same, affecting her mother, daughter and son. Histopathological examination of the papule revealed elongated rete ridges shaped like antler horns and reticulated pattern at one focus. So a final diagnosis of Dowling Degos Disease was made on the basis of the above findings. This case is being reported for its rarity.

KEYWORDS: Dowling Degos Disease, Reticulate pigmentary disorders of flexures, comedo like lesion.**INTRODUCTION**

Dowling Degos Disease is a rare benign autosomal dominant genodermatosis affecting the flexural areas. It is a type of reticulated pigmentary anomaly. The latter group includes many inherited conditions showing reticular type of hyperpigmentation with several other cutaneous manifestations. Dowling Degos Disease is characterized by multiple, symmetrical, asymptomatic, small, round, black to dark brown or white coloured, flat lesions in axillae, groins, flexures and few on face with multiple, symmetrical, dark brown coloured, small, raised lesions on trunk especially on chest and scattered dark dot type follicles/comedo like lesions present on face appearing at any age.

In this case, histopathology revealed increased melanin deposition in epidermis with elongation of rete ridges forming reticulated pattern at a single focus. Antler horn cyst formation at a few places was also seen.

Clinical features with characteristic histopathological findings make the diagnosis easy and straightforward.

Herein, we report a rare case with familial association of Dowling Degos Disease.

CASE REPORT

A 45 years old female, presented to our out patient department with multiple, round, black and white coloured lesions on her body since the last 15 years.

Lesions were multiple, symmetrically distributed, asymptomatic, small(0.5-1cm), round, hyperpigmented macules(freckles like) in axillae (Figure 1), groins, flexures, few on the face and hypopigmented macules on upper extremities and lower extremities with a progressive course. Multiple, symmetrical, hyperpigmented, small papules(seborrheic keratosis like) were seen on the trunk(especially on chest) (Figure 2) with comedo like lesions on face and nape of the neck (Figure 3).

Additionally, the patient had generalized, fine, polygonal, skin coloured scales on trunk and lower extremities, more prominent below the knees. Hand and feet were spared.

On detailed history, it was revealed that the patient's mother, daughter and son were also suffering from the same lesions of which mother and daughter were available for examination.

Co-incidentally, the patient was a known case of hypothyroidism for the last 3 years and was on treatment for the same. There were no other systemic comorbidities.

Routine investigations (complete blood count, liver function test, renal function test, urine routine and microscopy, serum lipids) were within normal limits. Thyroid profile was deranged.

Dowling Degos Disease, chloracne, comedonal Darier's, Kitamura disease were considered as differential diagnosis on the basis of history and clinical examination.

Skin biopsy of the hyperpigmented macule and papule revealed increased melanin deposition in the basal layer with reticulated elongation of rete ridges at a single focus. Antler horn cyst like formation was also seen at few places (Figure 4).

On the basis of clinical features and characteristic biopsy findings, the patient was labelled as a case of Dowling Degos Disease.



Figure-1.

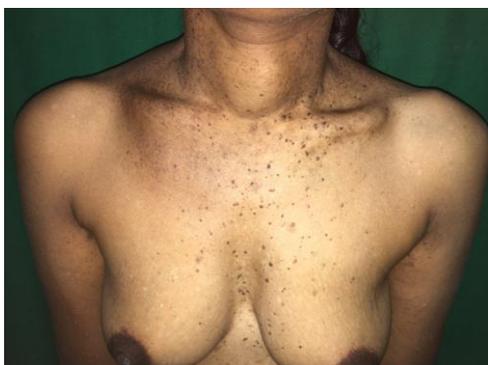


Figure-2.



Figure-3.

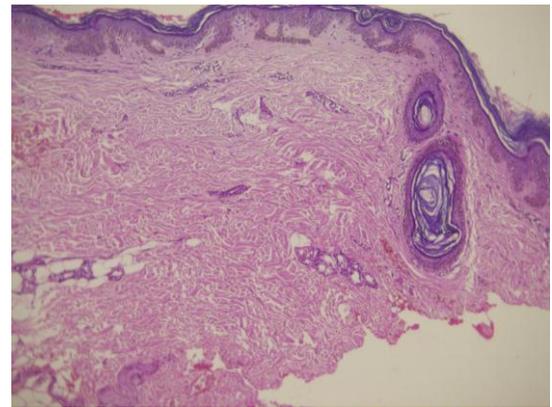


Figure-4.

DISCUSSION

Reticulated pigmentary disorders comprise several types of inherited reticulated disorders characterized by hyperpigmented macules (freckles like) and papules with other cutaneous and extra-cutaneous manifestations.^[1] Dowling Degos Disease is a rare type of genodermatosis caused by functional mutation of the keratin 5 gene.^[2] It is an autosomal dominant disease with a female preponderance (noted in few case studies) which is true in our case also (3 females out of 4 cases).^[3,4,5,6]

But recent studies indicate a possible role of follicular pathogenesis as seen in our case also. The specific indicators include comedo like lesions, clustered and punctate papules (as seen in Dowling Degos Disease), association with hidradenitis suppurativa.^[6,7,8,9] Interestingly, this patient also had ichthyosis vulgaris which is an inherited keratinizing disorder caused by mutation of the filaggrin gene (unrelated to Dowling Degos Disease).^[2] In several case reports, some authors noted unusual features like palmar pigmentation, acral distribution of lesions, seborrheic keratosis and pitted follicular pore.^[10] Seborrheic keratosis and pitted follicular pores were noted in our patient with sparing of palms and soles.^[9]

Skin biopsy findings are ascertained but have been mentioned in few studies. A case study of Kim *et al.*^[6] on 6 cases of Dowling Degos Disease showed basal layer

hyperpigmentation with elongated rete ridges and suprapapillary thinning. A single biopsy showed antler horn like cyst out of 6 cases which is evident in our case.

Hereby we report a rare case of Dowling Degos Disease with familial involvement.

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