Case Report

Dowling Degos Disease : Classic Clinicohistopathologic Presentation

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ABSTRACT

Dowling Degos Disease (DDD) is a rare genetic disease of the skin, characterised by flexural brown pigmented macules, comedo-like papules on back, neck and pitted perioral or facial scars. We hereby present the case of 38 year old male with hyperpigmented macules on neck, axillae and cubital fossa showing characteristic histopathological features of reticulate pigmentation, acanthosis, hyperkeratosis, increased pigmentation of basal layer and slight digitation of rete ridges.

Introduction -

Dowling Degos disease is an uncommon genodermatosis of autosomal dominant transmission with variable penetrance which usually occurs in 3rd to 4th decade of life with no sex or race predilection.[1] Most cases have been reported from Asian and Mediterranean countries.[2]

In one chinese family a gene locus was mapped to 17p13.3, while another study identified a heterozygous frame shift mutation in V1 domain of keratin 5.[2]

The disease is characterised by acquired reticulated skin hyperpigmentation, which begins in the armpits, groin and later spreads to other skin folds. It can also affect wrist, antecubital fossa, popliteal fossa, face, scalp, scrotum and vulva.[3] Lesion do not change with sun exposure. Comedolytic black lesion on face, back & in the same areas described above can also be observed[4] as well as cribriform scars and perioral acne in patients with no history of acne. Other features like mental retardation and pilar cyst may be present.[5] We present this case due to rarity of its presentation.

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Case Report -

A male patient, aged 38, residing at Kamptee, Nagpur, presented to us with hyperpigmented lesions on neck **(fig. 1)**, axillae **(fig. 2)** and cubital fossa **(fig. 3)** for last 4 years. Pitted scars were present on upper part of back and face **(fig.4 & 5)**. Patient reported similar clinical condition of his father and brother. For variety of reasons these relatives had not been submitted to clinical evaluation.

Biopsy was performed from hyperpigmented macule on axillae which showed acanthosis, hyperkeratosis, increased pigmentation of basal layer and slight digitation of rete ridges *(fig.6)*. The outcome of the clinical and histopathological test pointed conclusively to Dowling Degos disease.

Discussion -

Dowling Degos disease (DDD) is a late onset genodermatosis. The reticulate hyperpigmentation is admixed with and sometimes composed of lentigo-like brown macules; small brown papules with variable hyperkeratosis may also develop. These findings progressively increase over time, initially appear in axillae and groin with later involvement of intergluteal and inframammary folds, neck, trunk and inner aspects of arm and thighs. Some patients report pruritus of affected flexural areas. Comedo-like lesions on back and neck, pitted perioral scars, epidermoid cysts[1], hidradenitis suppurativa represent additional features in some patients.[1] Histopathological finding of the disease include moderate orthokeratosis or hyperkeratosis, thinning of suprapapillary

epithelium and elongation of papillae with basal layer hyperpigmentation. These thread like growth of epidermis have the appearance of "antlers" and generally involve the follicle with follicular plug. A perivascular lymphohistiocyte infitrate in papillary dermis and pseudo horny cysts can also be observed.[3]

Differential Diagnosis -

Acanthosis nigricans is distinguished clinically by velvety plaques and histopathologically by less pronounced elongation of rete ridges, in addition there is no follicular involvement.[1]

According to few literatures Acropigmentation of kitamura, Galli Galli disease and Habers disease are considered as differential diagnosis of DDD.[1,5]

Reticulate acropigmentation of kitamura (RAPK) is sporadic autosomal dominant disease of unknown origin, clinical features consists of hyperpigmented atrophic macules on dorsum of hands and feet. It onset in childhood. The lesion darken with time and worsen with sun exposure. Pitting on the palm and sole and dorsa of fingers can also be found.[1,5]

Galli-Galli disease is acantholytic varient of DDD which presents in people age between 15 and 56. Clinical symptoms includes presence of hyperpigmentation of flexures together with itching and sometimes with erythematous, scaly papules on these sites as well as on the trunk and proximal extremities. Histopathology resembles that of DDD but with foci of acantholysis.[6]

Harber's disease is characterised by photosensitive facial rosacea-like rash which develops in adolescence, followed by the appearance of to keratotic papules, comedones-like lesion, cribriform scars, reticulate hyperpigmentation of trunk, proximal extremities and armpits.[1]

A typical histopathological examination with compatible clinical features are enough for diagnosing DDD, as we did in this case.

Topical hydroquinone, tretinoin, adapalene and corticosteroids have been used with varying success. Improvement following treatment with the erbium : YAG laser also been reported.[1]

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Conflict of interest : Nil reported

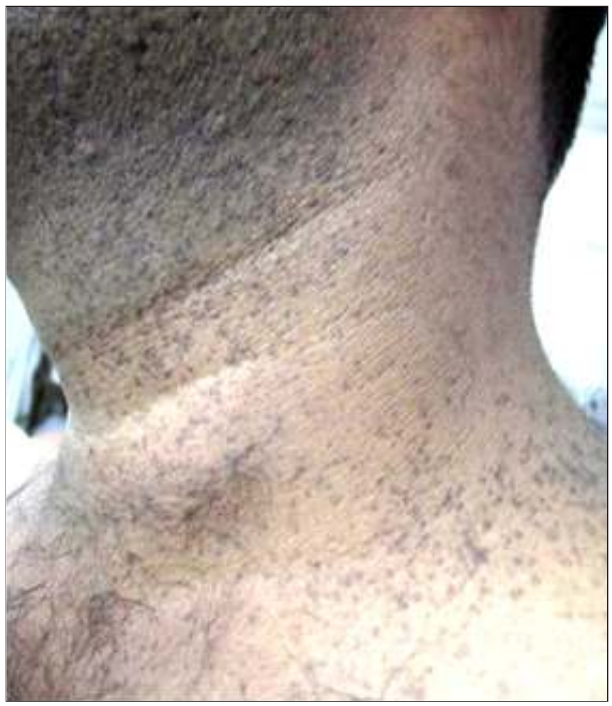

Fig 1 : **Hyperpigmented lesions on neck**

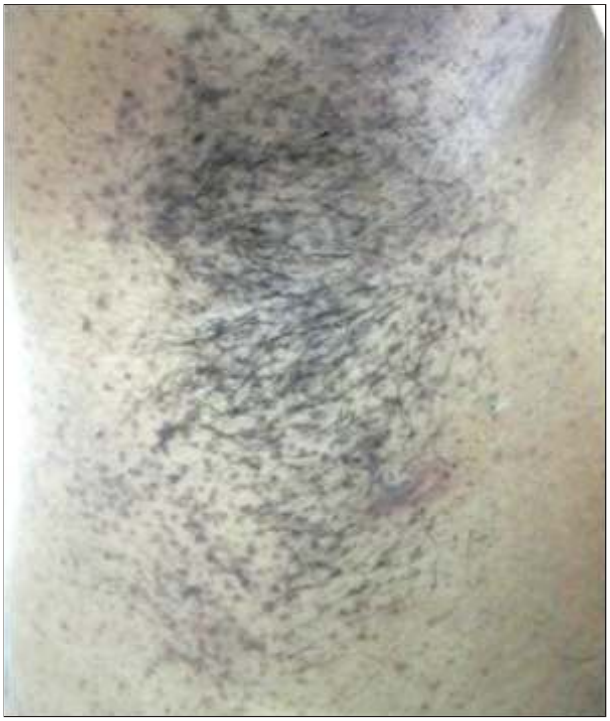

Fig 2 : **Hyperpigmented lesions in Axilla**

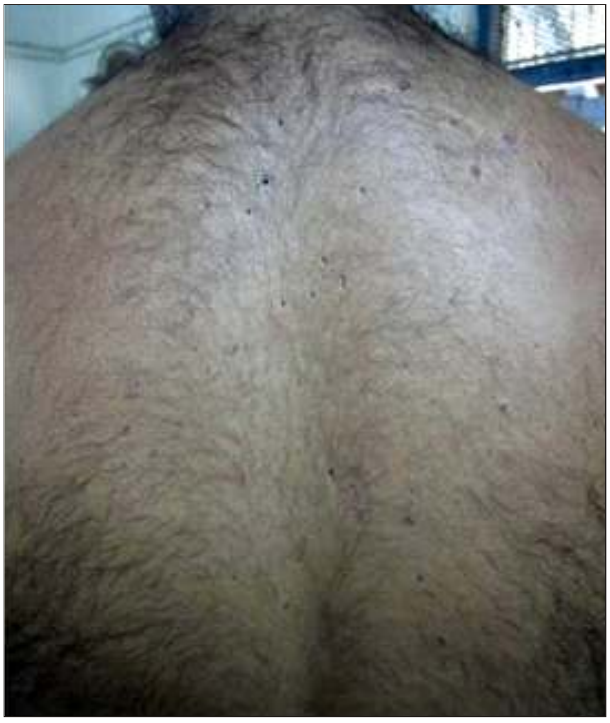

Fig 4 : **Pitted scars on back**

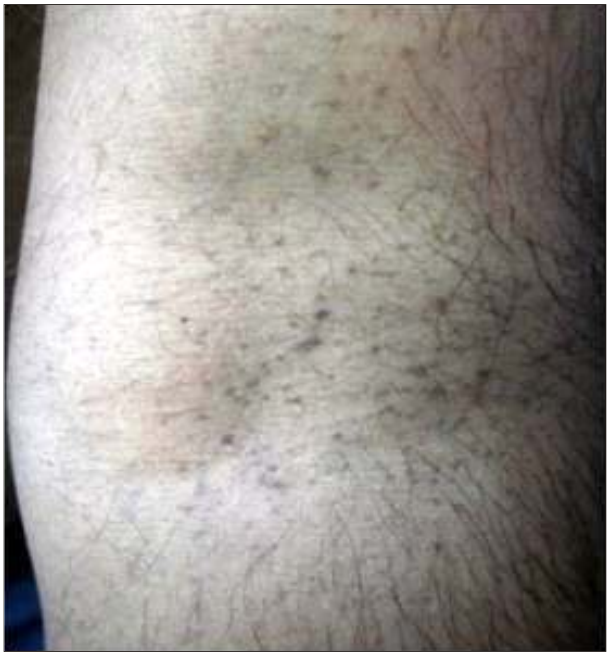

Fig 3 : **Hyperpigmented lesions in Anticubital fossa**

Fig 5 : **Pitted scars on face**

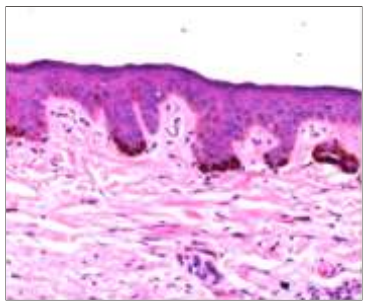

Fig 6 : **Histopathology of Macule showing : Acanthosis, hyperkeratosis, increased pigmentation of basal layer and slight digitation of rete ridges**