INDIAN J DERMATOL VENEREOL LEPROL 1986 ; 52 : 48-51

DOWLING DEGOS DISEASE
(Report of 5 cases in a family)

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Fifteen members of a large family were known to be affected by a reticulate pigmentary anomaly, chiefly of the flexures, of whom five members were examined and found to be affected by Dowling Degos disease. The family tree supports autosomal dominant transmission, with female preponderance. The unusual features noted include distribution of the lesions to the acral parts, palmar pigmentation and seborrhoeic keratoses like lesions.

Key words : Reticulate pigmentation, Dowling Degos disease.

Dowling Degos disease (DDD) is a rare genodermatosis, characterized by spotted and reticulate pigmentation of the flexures.[1-4] An earlier family report in which eleven members were affected by the disease suggested an autosomal dominant transmission.[5] Other reports were also suggestive of the genetic nature and familial distribution of the disease, though the exact pattern was not made out. To our knowledge, there is no earlier report on this condition from India. We examined 5 cases of DDD belonging to 3 generations of a large family. In addition, ten other members of the family were also known to be affected.

Case Reports

Case 1

A 58-year-old north Indian woman had noticed pigmented macules developing and persisting in the peri-orbital regions, axillae and groins, starting at the age of 12-13 years. Later, in adult life, the cubital fossae, the infra-mammary regions, the neck, the forehead, the trunk and the limbs were involved progressively, the macules coalescing to give rise to a reticulate pigmentation. Subsequently, at the age of about 50 years, she developed multiple pigmented papular and warty lesions on the neck, the trunk and extremities. These were 2-5 mm in diameter and resembled seborrhoeic keratosis (Fig. 1). Both her mother and grand-mother

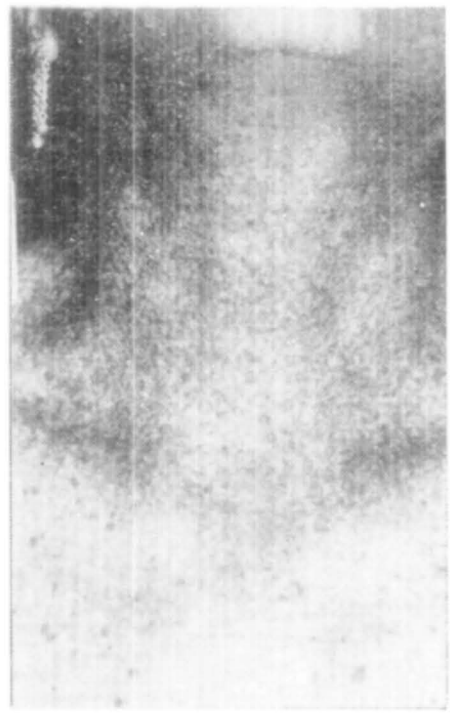

Fig. 1. Hyperpigmented macules and soft pigmented papules.

had been similarly affected with pigmentation, though her mother, who died at a relatively early age, did not develop seborrhoeic keratoses-like lesions. There was no history of consanguinity. Only 2 males were known to be affected in the family and both of them had mild disease, limited to a few pigmented macules on the face

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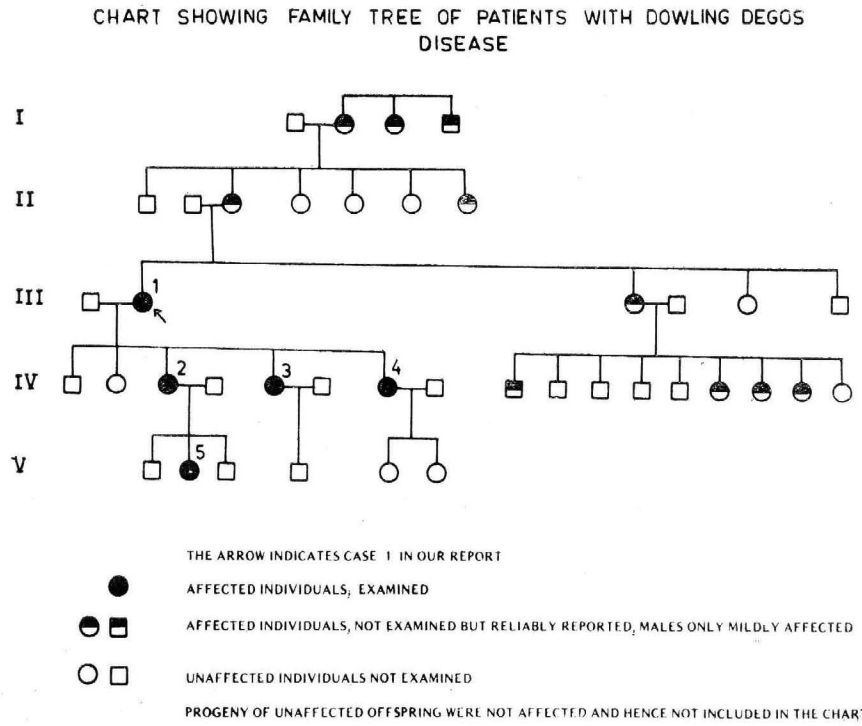

Fig. 2. Family tree suggesting autosomal dominant inheritance.

and flexures (Fig. 2). Examination revealed that the patient had, in addition to the pigmentation and seborrhoeic keratoses-like lesions, diffuse palmo-plantar keratoderma with discrete pigmented macules, 2-4 mm in diameter, on the palms. There were no significant systemic findings.

Cases 2, 3 and 4

Patients 2, 3 and 4 were three daughters of case 1, aged 31 years, 29 years and 26 years respectively. All of these patients had noted pigmented macules in the peri-orbital region between the ages of 10-12 years. Subsequently, they developed pigmentation of the flexures; lesions beginning as hypopigmented macules which gradually became hyperpigmented(Fig. 3). Numerous hypopigmented macules 1-3 mm in diameter were noted on the forearms and legs in cases 3 and 4. All the three patients had prominence of the palmar creases and case 2, in addition, had pigmented macules on the palms. None of them had the wide-spread pigmentation or seborrhoeic keratoses-like lesions, manifest in their mother.

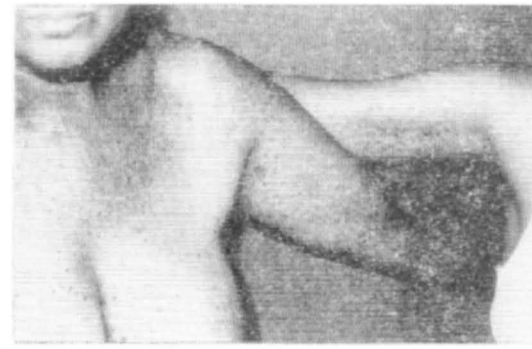

Fig. 3. Sisters showing pigmentation and seborrhoeic keratosis-like lesions.

Case 5

This patient was the 11-year-old daughter of case 2 who had begun to develop hyperpigmented macules on the face a few months ago. No other abnormality was seen in her.

Routine haemogram, urine and stools examination and skiagram of the chest in all the 5 patients were within normal limits. Skin biopsies taken from the pigmented macules in cases 1, 2 and 4 showed characteristic features of Dowling-Degos disease, namely : typical delicate pigmented epithelial down-growths, resembling adenoid seborrhoeic keratoses (Fig. 4). Skin

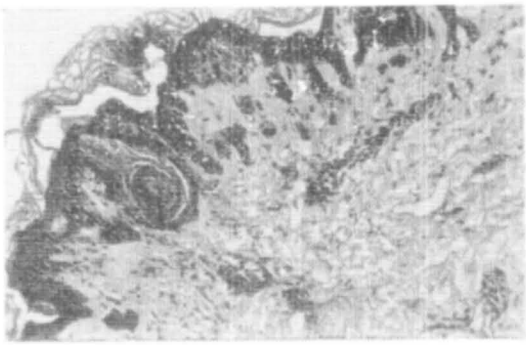

Fig. 4. Epidermis showing slight acanthosis, its projections into the dermis and intense pigmentation (H and E ×100).

biopsy of a seborrhoeic keratoses-like lesion from the chest in case 1 revealed a similar picture (Fig. 5). Biopsy from a macule in case 3, who was relatively mildly affected, was unremarkable.

Comments

Dowling Degos disease is a distinct clinical entity characterised by reticulate and spotted pigmentation of the flexures, more commonly seen in females.[3,5,6] Wilson Jones and Grice[2] in their review of the DDD noted familial occurrence of the disease. Later reports by Crovato et al[5] and Brown[7] pointed to an autosomal dominant transmission. The report by Brown of a family with six members being

Fig. 5. Biopsy of a papule showing similar picture of acanthosis and digitate budding into the dermis (H and E ×100).

affected suggested an autosomal dominant trait with possibly variable penetrance, variable expressivity and delayed onset. Our report also supports the same, with a relatively mild disease in the affected males.

The other features of interest are distribution of the lesions to areas other than flexures, their occurrence on the palms also and hyperlinear palms in all the patients. This brings into focus, the view regarding the overlap of certain clinical features of the various pigmentary genodermatoses.[8,9] Rebora and Crovato[9] in a recent review, studied a number of patients and families with an overlap between Dowling Degos disease, Haber's syndrome and Kitamura's acropigmentation and suggested that the term DDD should be extended to a group of autosomal dominant genodermatoses that are characterised clinically by reticulate pigmented macules, hyperkeratotic follicular lesions or pitted scars, and facial erythema in varying degrees of clinical severity, and histopathologically by a constant picture of digitate budding and extensive proliferation of the epidermis and follicular walls.

One of our patients (case 1) had numerous seborrhoeic keratoses-like lesions, which from the family history seemed to be inherited along with the pigmentary anomaly, manifesting only

later in life. Multiple seborrhoeic keratoses have been described in the case reported by Howell and Freeman[4] and in one of the ten cases reported by Wilson Jones and Grice,[3] though they make no comment on the inheritance of these lesions along with the pigmentary anomaly. None of our patients had the pin-head sized pigmented comedones and pitted follicular pores described by Wilson Jones and Grice[3] and other authors.[5,7]

Because of the histopathological resemblance of DDD to adenoid seborrhoeic keratoses, it would be worthwhile to follow-up these cases to see if widespread seborrhoeic keratoses develop in these patients in later life.

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