



Case report

Report of three rare cases of Dowling Dego's disease

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Abstract

Dowling Dego's disease is a rare autosomal dominant condition. It is caused by loss of function mutations in keratin 5 gene (kRT5) situated in keratin gene cluster on 12q13. We are reporting three cases of Dowling Dego's disease out of which two belong to same family. The clinical manifestations and histopathology were analyzed. All the three patients of which two were mother and son had symmetric, reticulate hyperpigmentation, pitted scars and acneiform eruption over face (predominantly perioral area, nasolabial folds), pinnae, upper trunk, thighs were seen. Palms, soles and scalp were spared. Buccal mucosa also showed reticulate hyperpigmentation. In addition the female patient had vulval reticulate hyperpigmentation and hidradenitis suppurativa affecting axillae, groins and inframammary folds.

Key words: Dowling Degos disease, Hidradenitis suppurativa, Reticulate pigmentary disorders

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Dowling Dego's disease is a rare autosomal dominant condition, caused by loss of function mutations in keratin 5 gene (kRT5) situated in keratin gene cluster on 12q13. It usually presents post pubertally with multiple, small, round, pigmented macules distributed over sites like neck, face, arms, back, axillae and groins¹. Here we present three cases, two of which belong to the same family and one from a different family.

Case 1

A 19 year old male presented with multiple asymptomatic tiny black colored spots over face, neck, trunk, groin folds and thighs since 10 years, slowly increasing in numbers.

Cutaneous examination revealed symmetric, reticulate hyperpigmentation which were round in shape and about 1-1.5 mm in size, pitted scars on bilateral palms and acneiform eruption over face

(predominantly in perioral area, nasolabial folds), pinnae, upper trunk, thighs were seen. In addition they were also hypopigmented macules distributed along the body which were asymmetrical. Palms, soles and scalp were spared of hyperpigmentation. Buccal mucosa also showed reticulate hyperpigmentation (Fig 1, Fig 2 and Fig 3).

Case 2

A 49 year old female, who was mother of case 1, had clinical features of small round hyperpigmented macules of about 1 mm in size on face, pinnae, upper trunk, thighs, axillae, inframammary area, groins and cubital fossae (Fig 4 and Fig 5).

Case 3

A 35 year old female, presented with features of reticulate hyperpigmented macules distributed over

face, upper trunk, thighs, inframammary folds, buttocks and vulva. In addition she had Hydradenitis suppurativa (Fig 6).



Fig 1. Hyperpigmentation over upper back



Fig 2. Pitted scars over palms



Fig 3. Hypopigmented macule over chest



Fig 4. Hyperpigmented macules in inframammary region



Fig 5. Hyperpigmentation in cubital fossa



Fig 6. Hyperpigmented macules over face

Histopathology of all 3 cases showed hyperkeratosis of epidermis, elongation, lateral fusion and hyperpigmentation of tips of rete ridges with perivascular lymphocytic infiltrate characteristic of Dowling Deigo's disease.

All three patients were counseled about the disease and advised regular use of sunscreen and moisturizers, as there is no specific treatment.

Discussion

Dowling Deigo's disease also known as reticular pigmented anomaly of the flexures is a rare autosomal dominant condition. It was first described in 1938 as reticulate pigmented anomaly of the flexures by Dowling and Freudenthaland. It was subsequently named "dermatose pigmentaire reticuledes plis" by Degos and Ossipowski in 1954². It presents post pubertally with multiple small, round pigmented macules that resemble freckles. The most common site are axillae groins, the intergluteal and inframammary folds, neck, scalp, trunk and arms and vulva^{1,3,4}. Pigmentation is asymptomatic, symmetrical and progressive and the degree of pigmentation varies, but in some patients the lesions are almost confluent, giving a brown or black reticulate lace-like pattern. Dark brown papules may also develop at affected sites. Other features that may be present include scattered comedo-like lesions referred to as dark dot

follicles and pitted acneiform scars near the angles of the mouth. It may be associated with Hidradenitis suppurativa^{5,6}. Nails and hair are normal. Histopathology shows distinctive form of acanthosis, characterized by an irregular elongation of thin branching rete ridges, with a concentration of melanin at the tips. The condition involves the follicular infundibulum, and in some cases there is follicular plugging. The melanocyte count is normal^{7,8}.

The second patient describes here has overlap of reticulate acropigmentation of Kitamura in view of presence of pitted scars on bilateral palms⁹⁻¹².

Similarly Singh et al reported two cases of same family with follicular lesions mimicking chloracne and perioral pits and other keratinizing disorder as ichthyosis vulgaris which is not associated with classical case of Dowling Degos disease¹³.

There is no specific treatment available for this disease. In a case series of three patients done by Bhagwat et al, patients were treated with oral isotretinoin 20 mg / day and topical retinoids for two months without appreciable results¹⁴.

Hence the patients in our case series were counseled about the disease and advised regular use of sunscreens and moisturisers .

Conclusion

We are reporting three cases of Dowling Degos disease in view of rarity of this condition, which requires a high index of suspicion and careful clinical and histopathological evaluation for its diagnosis.

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Conflict of interest: None

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