

GENETICS AND GENODERMATOSES

DOWLING-DEGOS DISEASE- A RARE RETICULATE PIGMENTARY DISORDER

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Background: Dowling-Degos disease is a rare autosomal dominant form of reticulate genodermatosis with variable penetrance, characterized by reticular pigment pattern that is most pronounced in flexural areas. Reticulate pigmentary disorders include a spectrum of acquired and congenital conditions which differ in age of onset and distribution of lesions.

Observation: A 21-year-old male patient presented with multiple asymptomatic dark coloured lesions over the face since one year. The lesions were first noticed on the face and then gradually increased in number to involve the flexor aspects of elbows, wrists and axillae. The patient was born of a non-consanguineous marriage and there was no history of similar complaints in the family. There was no history of other skin diseases. His general physical examination was within normal limits. Skin examination revealed multiple hyperpigmented macules in a reticulate pattern over the face, cubital fossa ,flexure aspect of wrists and axillae. Multiple pitted acneform scars were seen on the face, concentrated mainly over the perioral area. The mucosa, nails, teeth and hair were normal. On investigation, complete blood count, blood chemistry, liver and renal function tests were all within normal limits. Histopathological examination was suggestive of Dowling-Degos Disease.

Conclusion: Although Dowling-Degos disease is limited to the skin and is relatively harmless, it is important to differentiate it from other reticulate pigmentary disorders as some of them are associated with malignancies, cosmetic concerns, psychological disturbances and difficulty in getting identity within a community due to loss of fingerprints.





