GENETICS AND GENODERMATOSES

RETICULATE ACROPIGMENTATION OF KITAMURA : A RARE CASE REPORT IN A FAMILY WITH 19 CASES OF DOWLING DEGOS DISEASE

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BACKGROUND: Reticulate hyperpigmentary disorders are a group of rare genetic pigmentary abnormalities which includes Reticulate Acropigmentation of Kitamura (RAPK), Dowling-Degos Disease, Reticulate Acropigmentation of Dohi and Galli-Galli disease. RAPK was first described in Japan in 1943 by Kitamura and Akamatsu.

OBSERVATIONS: A 20-year-old female born of non consanguinous marriage, presented with asymptomatic but progressive dark admixed white colored lesions on her hands, feet and neck since 7 years of age with history of photoexaggeration.There was history of similar complaints in many members of the family. Dermatological examination revealed mildly atrophic,hyperpigmented with hypopigmented macules in a reticulate pattern involving the dorsa of the hands and feet, front and sides of neck, and axillae. There were breaks in palmar dermatoglyphics.
1. Biopsy from dorsum of right hand of the patient, showed subtle epidermal proliferation and hyperpigmentation predominantly of the lower dermis. Upper epidermis was less pigmented. The pigment is also seen within the melanophages in the papillary dermis. The number of melanocyte in epidermis was within normal limits. Consistent with RAPK.
2. Biopsy from the patients father showed circumscribed foci of epidermal proliferation and hyperpigmentation. Epidermis in these areas showed subtle proliferation in the form of delicate elongated and confluent rete ridges that showed “antler like” branches at places. The pigment is concentrated at the bottoms of rete ridges and is also seen within melanophages in the papillary dermis. The proliferative rete ridges show accentuation around plugged and dilated follicular infundibula. These features were consistent with Dowling Degos disease.

KEY MESSAGE: To evaluate and trace the family tree in a patient having overlapping features of Acropigmentation of Kitamura and Dowling Degos disease in a family with 17 cases of Dowling Degos Disease, which has not been previously reported in the literature