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Case Report

Reticulate acropigmentation of Kitamura: A familial case with eyelid involvement

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Introduction

Reticulate pigmentary disorders of the skin, a recently described group includes Dowling–Degos disease (DDD), Reticulate acropigmentation of Kitamura (RAPK), Haber's syndrome (HS), Galli–Galli disease (GGD) and reticulate acropigmentation of Dohi (RAD).¹ Although these reticulate pigmentary disorders do not affect the general health of the involved patients, they are a stigma for the affected patients.

Amongst the above pigmentary disorders, RAPK is a rare autosomal dominant genetic disorder of unknown origin that

characteristically presents as freckle-like, irregular, depressed atrophic macules arranged in a reticular pattern mainly on the dorsum of hands and feet, lesions predominantly acral. Eyelid involvement has been reported in very few cases till date.

A twenty-three years old female patient presented with dark colored lesions over distal aspect of all four extremities since the age of fourteen years. There was history of progressive increase in pigmentation and the area of involvement over the last nine years. Lesions initially appeared over the dorsum of both hands and later increased to involve both forearms, lower one-third of both legs and dorsum of both feet. Familial history was significant as similar skin lesions were present in the grandfather, father, maternal aunt and brother. There was no history of oral ulcers, photosensitivity, joint pains or application of any irritants or chemicals to the affected areas prior to the onset of lesions.

Dermatological examination revealed punctate, irregular, slightly atrophic, brown macules arranged in a reticular pattern on the dorsa of the hands and feet, distal half of both forearms and distal one-third of both legs [Fig. 1]. Multiple, pinpoint, freckle-like, hyperpigmented macules were also present over both upper eyelids and cheeks [Fig. 2]. Both palms showed presence of palmar pits.

Skin biopsy from the forearm showed hyperpigmentation of basilar keratinocytes along with an increase in the number of melanocytes [Fig. 3]. Digitated elongation of hyperpigmented rete ridges was present while in papillary dermis

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Fig. 1 – Reticulate hyperpigmented macules present over both hands and feet.

Fig. 2 – Multiple pinpoint, freckle-like, hyperpigmented macules present over upper eyelids and cheeks.

melanin-laden macrophages were seen. On the basis of clinical and histopathological findings, a diagnosis of Reticulate acropigmentation of Kitamura (RAPK) was made.

Discussion

Reticulate pigmentary disorders of the skin include Dowling–Degos disease, RAPK, Haber's syndrome, Galli–Galli disease and reticulate acropigmentation of Dohi. Galli–Galli disease is a rare genodermatosis with features of reticulate pigmentation along with hyperpigmented brown macules and papules involving flexural surfaces with characteristic histopathologic feature of suprabasal acantholysis. Reticulate

Fig. 3 – Histopathological examination showing hyperpigmentation of basilar keratinocytes along with an increase in the number of melanocytes (H&E ×40).

acropigmentation of Dohi is a dyschromic disorder characterized by hyperpigmented and hypopigmented macules with a reticulate pattern, localized on the dorsum of hands and feet, and appears predominantly in Japanese patients while Haber's syndrome is a reticulate pigmentation disorder with characteristic early onset, persistent rosacea like eruption on the face along with verruciform, pigmented, keratotic papules found in axillae, neck and trunk.¹

RAPK is an autosomal dominant inherited condition with a slight preponderance in females.² The typical hyperpigmented, atrophic, angulated macules generally start in the first and second decades of life over the dorsum of the hands and gradually extend onto the extremities. Pits and breaks in the dermatoglyphics are found on the palms, soles and dorsal phalangeal surfaces, which are considered characteristic for this condition.³ Lesions over face and eyelids are rare. Sharma et al reported a familial case of RAPK with periorbital involvement along with lesions over eyelids and face.⁴ Reticulated pigmentation on the dorsum of hands and feet has been reported in a few patients with Dowling–Degos disease (DDD) and several authors have proposed that RAPK and DDD are related disorders.^{5,6} In DDD characteristic lesions include flexural hyperpigmented macules, pitted perioral scars and comedo-like hyperkeratotic dark follicular papules over neck and axillae. The histopathology of RAPK is very similar to that of DDD, but clinically, in RAPK palmar and plantar pits are present and there is slight depression of the pigmented lesions. Histopathological examination in RAPK classically shows epidermal atrophy associated with club-like elongation of the rete ridges and an excess of melanin in the basal layer.

Treatment of reticulate pigmentary disorders especially RAPK and DDD is generally not very effective, however, Kameyama et al reported successful treatment of a case of RAPK with 20% Azelaic acid.⁷ Various studies mention use of topical hydroquinone, topical and systemic retinoids, topical corticosteroids and phototherapy with minimal improvement. Few case reports mention good response with Er:YAG-laser in management of Dowling–Degos disease.⁸

This involvement of the face and eyelids in RAPK, which has seldom been reported in the literature, prompted us to report this case of familial RAPK with eyelid involvement.

Conflicts of interest

All authors have none to declare.

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