Reticulate Acropigmentation of Kitamura involving Face & Tongue- a Rare Entity

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Abstract: Reticulate acropigmentation of Kitamura (RAPK) is a rare, autosomal dominant disorder first described in Japan characterised by a reticulate pattern of slightly atrophic, angulated, hyperpigmented macules affecting the acral areas of the body. We, herein, describe a unique case of an 11 year old Indian child with reticulate acropigmentation of Kitamura affecting his whole body including face & tongue which is a rare entity.

Keywords: autosomal dominant, reticulate acropigmentation of Kitamura, genodermatoses

INTRODUCTION

Reticulate acropigmentation of Kitamura (RAPK) was first described by Kitamura and Akamatsu in Japanese patients in 1943.[1] It is characterized by angulated, slightly atrophic, hyperpigmented macules that are arranged in a reticulate pattern typically on the dorsal aspect of hands and feet. The condition is inherited in an autosomal dominant fashion and skin changes begin to develop during childhood. [2] There is also involvement of palms and soles in the form of irregular interruptions of the dermatoglyphics and fine pits. RAPK is predominantly characterized by acral reticulate pigmentary lesions and fine reticulate brown pigmentation on dermoscopy.3 Facial & tongue involvement, as seen in our case, is unusual in RAPK.

CASE REPORT

An 11-year-old male presented with a 7 year history of asymptomatic and progressive brownish-black discoloration over his tongue followed by the dorsa of hands and feet. Pigmentation progressed slowly to involve the dorsal and ventral surfaces of the extremities, neck, chest and face. His past medical history was unremarkable. There was no history of photosensitivity. There was no history of handling any chemical directly or any significant history of drug intake. There was a positive family history for the same, his younger brother being the affected one alongwith positive history of consanguinity among parents.

Cutaneous examination revealed punctate, irregular, slightly atrophic, brown macules arranged in a reticulate pattern on tongue, dorsa of hands and feet, the extensor and flexor surfaces of forearms, flexor aspects of neck and chest [Fig. 1-5]. Brownish pigmentation was also present on the palms with pits and the fingernails were dystrophic [Fig.6]. Black-brownish reticulate macules were also seen over nose, bilateral cheeks and chin [Fig.1]. Systemic examination did not reveal any abnormality. Routine investigations including complete blood counts, urinalysis, liver function tests, renal function tests and serum electrolytes were within normal limits.

Skin biopsy was taken from the back which showed structure of skin with epidermis and dermis. Epidermis was thinned out with hyperpigmentation of basal keratinocytes. Superficial dermis showed few clusters of melanocytes. Also seen with perivascular and periadnexal lymphocytic infiltrate.There was melanin incontinence in the papillary dermis [Fig.7]. This confirmed the diagnosis of RAPK.

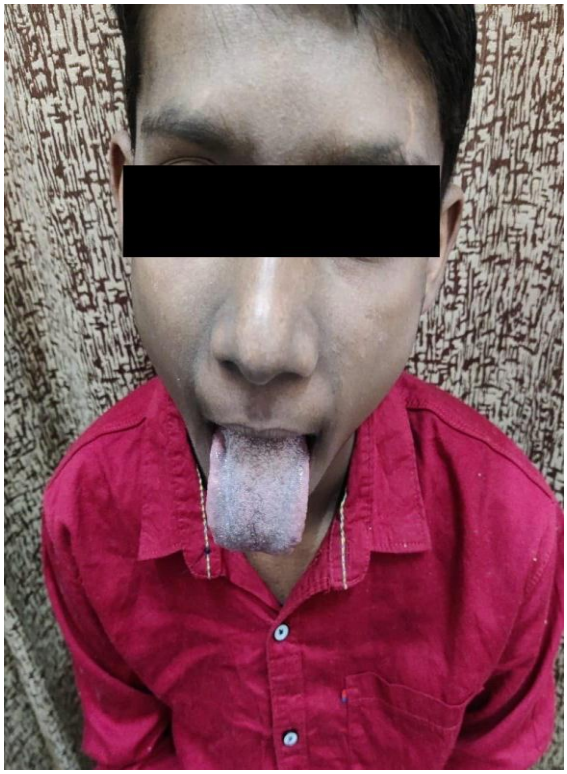

Fig.1

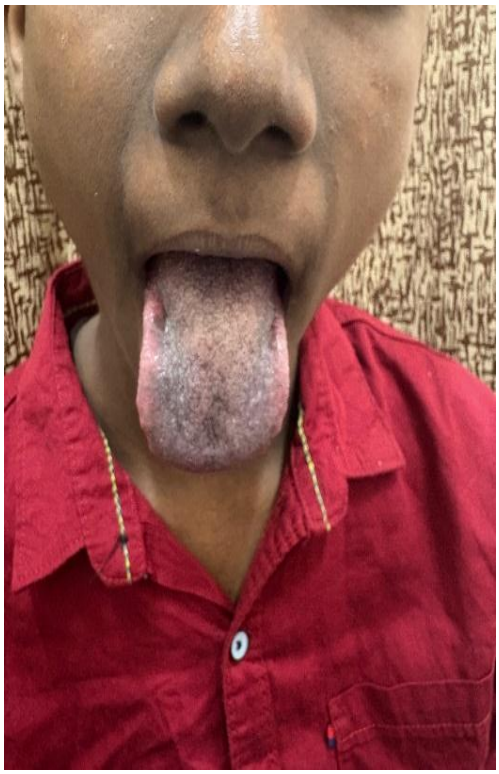

Fig.2

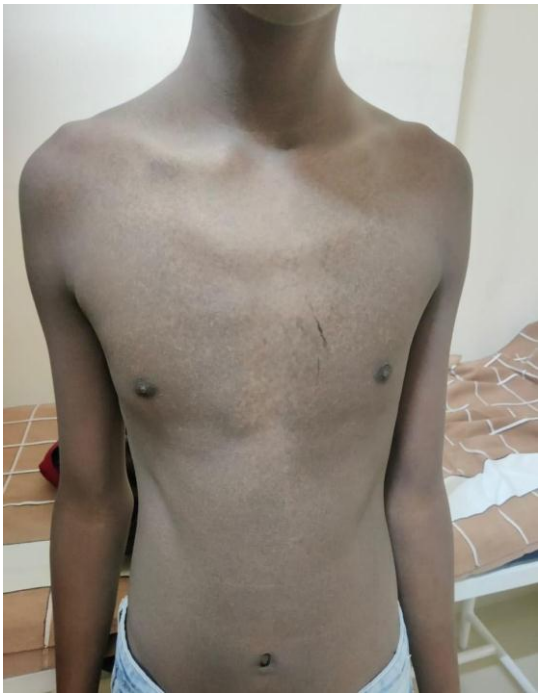

Fig.3

Fig.4

Fig.5

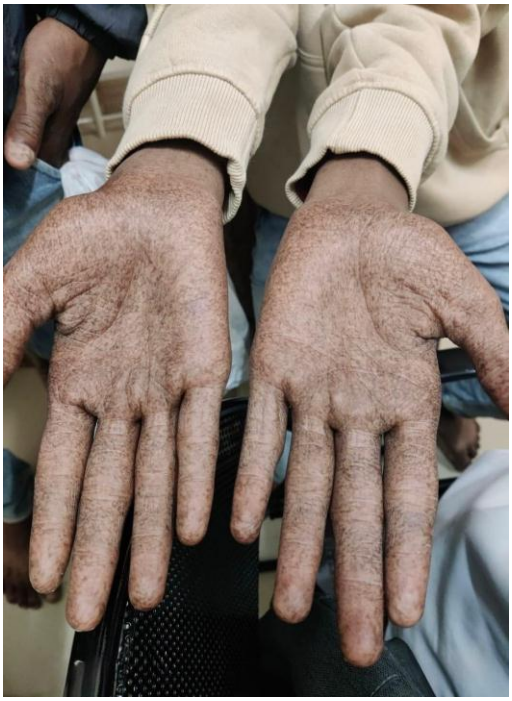

Fig.6

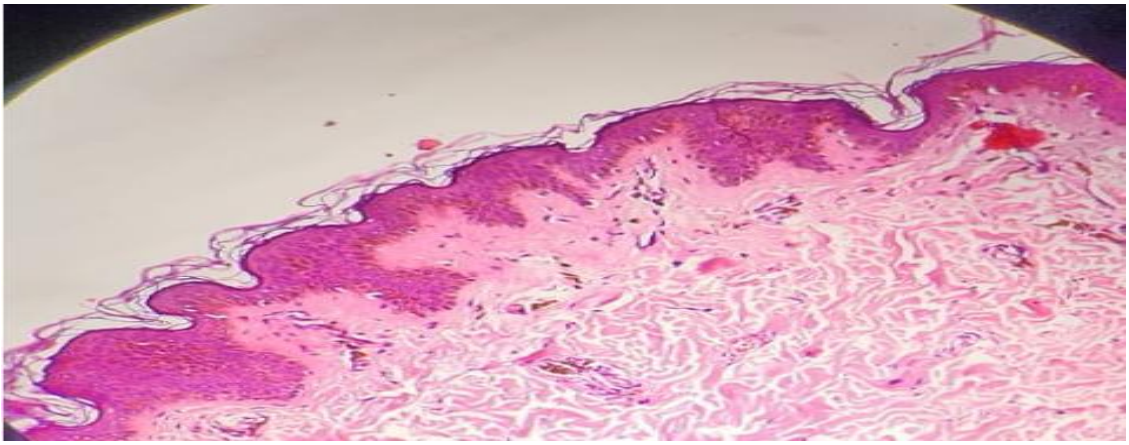

Fig.7

DISCUSSION

Reticulate acropigmentation of Kitamura is a rare genodermatoses that was first described in Japan. However, rare familial cases have been described from India, Turkey, Iran, Saudi Arabia, Italy, and Latin America. [4] It follows an autosomal dominant mode of inheritance with high penetrance. The characteristic presentation of RAPK is pigmented, angulated, irregular freckle-like lesions with atrophy on the surface arranged in a reticulate pattern on the dorsa of hands and feet.[2] The lesions usually start in the first and second decades of life and gradually extend onto the extremities and rarely on the face and eyelids. The lesions gradually darken over time. Sunlight may aggravate the condition. Pits and breaks in the dermatoglyphics are found over the palms, soles and dorsal phalangeal surfaces. [5] Our patient presented with reticulate hyperpigmentation over tongue which is a rare entity since 4 years of his age followed by the rest of body.

Histopathological examination showed characteristic epidermal atrophy associated with club-like elongation of the rete ridges and an excess of melanin in the basal layer. [6]

The differential diagnosis of RAPK include diseases presenting with reticulate or punctuate hyperpigmentation such as Dyskeratosis congenita, Dyschromatosis universalis hereditaria, Franceschetti-Jadassohn's syndrome, Dermatopathia pigmentosa reticularis, Reticular acropigmentation of Dohi (RAPD) and Dowling Degos disease (DDD). [7]

Reticulate acropigmentation of Dohi is also an acral type of dyschromatosis (dyschromatosis symmetrica herediteria) that usually starts in infancy or early childhood as hypopigmented and hyperpigmented macules over the dorsa of the hands and feet. [8] The differentiation between RAPD and RAPK is based on clinical and histological findings. In RAPK, there are usually no hypopigmented macules and histologically hyperpigmented lesions show epidermal atrophy, elongation of rete ridges and increased number of DOPA positive melanocytes, while in RAPD only increased or decreased basilar pigmentation is present in hyperpigmented or hypopigmented lesions respectively with occasional melanin incontinence in hypopigmented lesions.

In Dyschromatosis universalis hereditaria mottled pigmentation is present involving the trunk, extremities, face, palms, soles and occasionally oral mucosa. Dyskeratosis congenita follows autosomal dominant mode of inheritance and is caused by dyskerin gene mutation. Reticulate telangiectatic hyperpigmentation interspersed with areas of hypopigmentation involves face, neck, trunk and upper thighs with sparing of distal extremities as in RAPK. The pharyngeal, oral, anorectal mucosae show leukokeratosis while nails are dystrophic in dyskeratosis congenita.

Dermatopathia pigmentosa reticularis in an autosomal dominant disorder and reveals a characteristic triad of generalized reticulate hyperpigmentation, non-cicactricial alopecia and onychodystophy.

Recently, there have been increasing reports on the coexistence of RAPK and DDD in the same patient. [9] A mutation in ADAM 10 gene encoding a zinc metalloprotease, a disintegrin and metalloprotease domain-containing protein 10 (ADAM 10) has been identified in RAPK family. [10] ADAM 10 is known to be involved in the ectodomain shedding of various substrates in the skin. KRT 5 mutation has been thought to be causative in DDD. [11] Thus, genetic studies reveal RAPK and DDD as distinct entities.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent/assent.

Footnotes

Source of Support: Nil

Conflict of Interest: None declared

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