Case Report

Reticulate Acropigmentation of Kitamura: A rare hyperpigmented genetic skin disorder

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ABSTRACT

Reticulate acropigmentation of Kitamura is an exceptionally rare autosomal dominant genodermatosis characterized by reticulate hyperpigmented macules that interrupt the dermatoglyphics, primarily affecting the dorsa of the hands and feet. We report a case of a 65-year-old man who presented with multiple bilateral symmetrical, hyperpigmented, well-defined, discrete macules and plaques over the neck, both shoulders, both axillae, both cubital fossae, both knees, both legs, and the bilateral dorsa of the hands, present for approximately 1½ months with recent progression over the preceding 15 days. A positive family history in the patient's brother supported the autosomal dominant inheritance pattern. Skin biopsy confirmed the diagnosis, demonstrating hyperkeratosis, irregular acanthosis, filiform elongation of rete ridges, and increased melanocytes within the elongated epidermal projections. The importance of clinicopathological correlation in achieving an accurate diagnosis of this rare pigmentary disorder is emphasized.

Key words: Genodermatosis, Hyperpigmentation, Reticulate acropigmentation of Kitamura, Skin biopsy

Reticulate acropigmentation of Kitamura (RAPK) is a rare genodermatosis with an autosomal dominant pattern of inheritance, high penetrance, and a predilection for individuals of Asian, particularly Japanese heritage [1]. First reported in Japan, the condition predominantly affects females, though cases in males have been described [2]. The global prevalence is estimated at <1/million, and the condition is seldom reported outside Asia [3]. RAPK is differentiated from other reticulate pigmentary disorders by its distinctive combination of sharply demarcated reticulate hyperpigmented macules affecting the dorsa of the forearms and hands, palmoplantar pitting, and characteristic interruptions of the dermatoglyphics [1,3]. Although primarily a cutaneous disorder, rare extra-dermatological manifestations, including bony anomalies involving the terminal phalanges, have been documented [4,5]. The condition may be misdiagnosed as other heritable reticulate pigmentary disorders, making awareness of its clinical, dermoscopic, and histopathological features crucial for accurate diagnosis [4].

This case is reported due to its rare late-age presentation, classical clinico-histopathological correlation, and strong familial occurrence, suggesting autosomal dominant inheritance. It also highlights the diagnostic overlap of RAPK with other reticulate pigmentary disorders, emphasizing the importance of clinicopathological correlation in avoiding misdiagnosis and unnecessary investigations.

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CASE REPORT

A 65-year-old man presented with a chief complaint of multiple bilateral symmetrical, hyperpigmented, well-defined, discrete macules and plaques over the neck, both shoulders, both axillae, both cubital fossae, both knees, both legs, and the bilateral dorsa of the hands of approximately 1½ months duration, with accelerated progression over the preceding 15 days. The patient reported that the lesions first appeared on both legs before extending over the remainder of the body. A positive family history was elicited: the patient's brother had similar cutaneous findings on his hands, consistent with the autosomal dominant inheritance pattern of RAPK. There was no history of prior treatment or similar episodes in the patient himself.

Cutaneous examination confirmed multiple bilateral symmetrical, hyperpigmented, well-defined, discrete macules and plaques at the sites described above (Fig. 1a-g). The hyperpigmented lesions on the palmar surface demonstrated palmar pitting and characteristic interruptions of the dermatoglyphics (Fig. 2).

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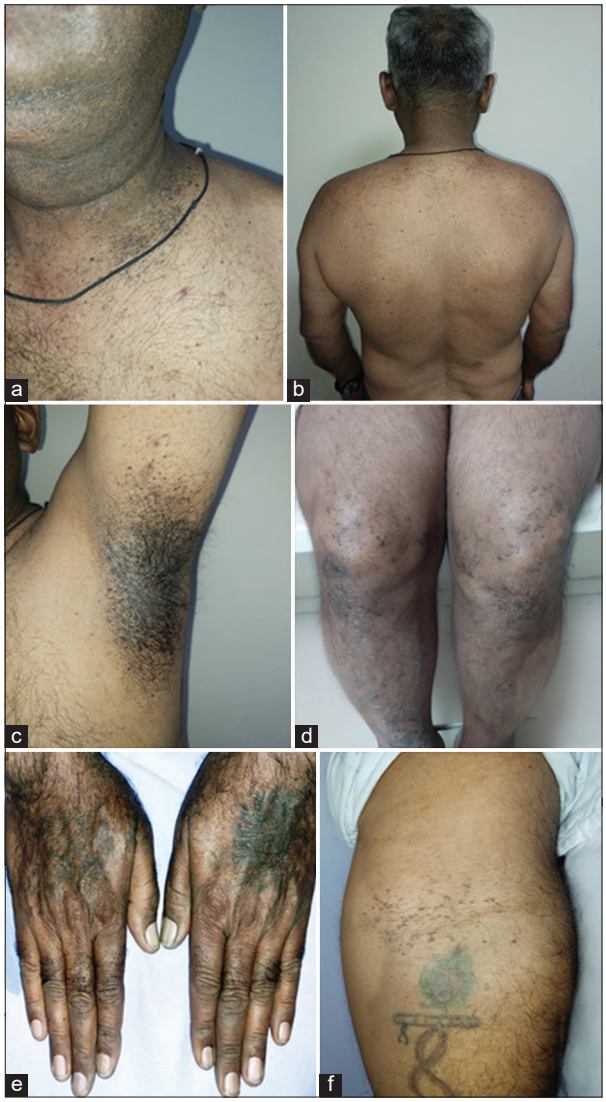

Figure 1: Clinical photographs of a typical reticulate acropigmentation of kitamura patient. Patient had reticular pigmented macules on the neck and both shoulders (a and b), axilla, both knees with both legs (c and d), both cubital fossa (e and f), and bilateral dorsum of hands

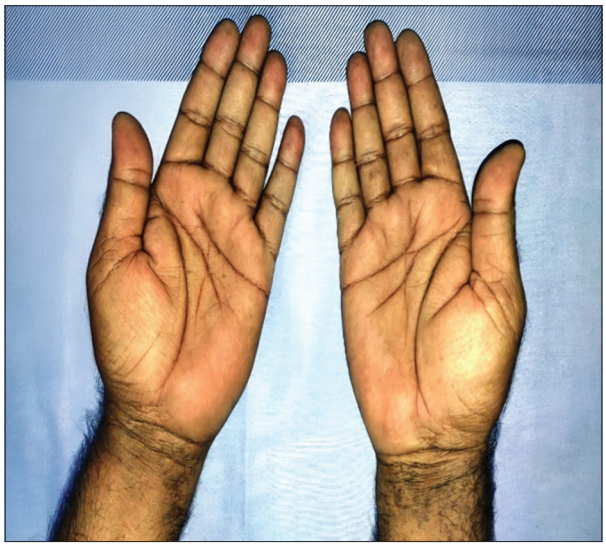

Figure 2: Clinical photograph showing palmar pitting and breaks in the dermatoglyphics

A skin biopsy was performed, and the specimen was submitted for histopathological examination. On gross examination, grey-white, skin-covered tissue was received, measuring 0.2×0.2×0.2 cm in aggregate. All tissue was processed in a single block and subjected to formalin-fixed paraffin-embedded sectioning.

Microscopic examination of hematoxylin and eosin -stained sections revealed hyperkeratosis, irregular acanthosis, focal thinning of the lining epithelium, and filiform elongation of the rete ridges (Fig. 3a). Increased melanocytes were noted within the filiform epidermal elongations, with scanty mononuclear infiltrate in the superficial dermis. No acantholysis was identified. The remainder of the dermis showed a fibro-collagenous tissue with scanty adnexal elements. The subcutis was unremarkable (Fig. 3b). The above findings are classical for RAPK, and the absence of acantholysis is an important feature helping to exclude reticulate acropigmentation of Dohi (RAD), and related acantholytic pigmentary disorders.

Based on the clinical presentation, family history and histopathological features, a diagnosis of RAPK was established. Dermoscopy of the pigmented lesions was not done due to cost-restraints. The patient was managed conservatively with topical corticosteroids, white soft paraffin, and oral antihistamines, resulting in symptomatic improvement.

DISCUSSION

RAPK belongs to a heterogeneous group of heritable dyschromatoses. The condition typically manifests in the second or third decade of life, although onset in the first decade has been described, with the development of atrophic hyperpigmented papules or macules on the dorsal acral surfaces that coalesce into a characteristic reticular, net-like pattern. Sun exposure may aggravate the lesions. Additional cutaneous features include palmoplantar pitting and interruptions of the dermatoglyphics [1-3].

Rarely, extra-dermatological manifestations occur; the only published case of RAPK with bony involvement described the absence of terminal phalanges of the second, third, and fourth toes [4,5]. The positive family history in our patient, with the patient's brother having similar acral lesions, is consistent with the autosomal dominant inheritance pattern.

The histopathological hallmarks of RAPK include hyperkeratosis, epidermal atrophy, and irregular elongation of rete ridges with increased melanin in the basal layer and increased melanocytes within the elongated epidermal projections. Superficial perivascular infiltrates and dermal melanin incontinence may also be present [5]. Importantly, the absence of acantholysis helps differentiate RAPK from RAD and related disorders [6].

The molecular pathogenesis of RAPK involves loss-of-function mutations in ADAM10, the gene encoding a disintegrin and metalloproteinase 10, which is an activator

of the Notch signaling pathway. ADAM10 mediates ectodomain shedding of membrane-bound substrates, including L1-CAM, CD44, E-cadherin, N-cadherin, the interleukin-6 receptor, and CD30. Dysregulation of this pathway is believed to underlie the aberrant melanocyte distribution and epidermal architectural changes observed in RAPK [1,7].

Dermoscopy constitutes a valuable, non-invasive adjunct to clinical diagnosis. In RAPK, dermoscopy of pigmented lesions demonstrates multiple discrete hyperpigmented globules in an irregular pattern with a dark brown to tan background, overlying black dots, and superficial fine white scaling. Dermoscopy of the palmar lesions characteristically reveals canaliform structures oriented perpendicular to the dermatoglyphic ridges, corresponding to the dermatoglyphic interruptions seen clinically [1,8]. These dermoscopic findings closely correlate with the histopathological substrate and can support or even preclude the need for invasive biopsy in classical cases.

The differential diagnosis of RAPK includes a spectrum of inherited reticulate pigmentary disorders such as dyskeratosis congenita, dyschromatosis universalis hereditaria, Naegeli–Franceschetti–Jadassohn syndrome, dermatopathia pigmentosa reticularis, RAD, and Dowling–Degos disease (DDD) [9]. Although these entities share overlapping clinical features, they can be distinguished based on respective distribution patterns, associated systemic involvement, and characteristic histopathological findings (Table 1).

RAD typically demonstrates suprabasal acantholysis on histology, a feature not seen in RAPK, which instead shows epidermal atrophy with increased basal melanocytes and filiform elongation of rete ridges [6]. In contrast, DDD, an autosomal dominant condition, predominantly involves flexural areas such as the axillae, neck, and groin, and may present with comedo-like lesions and perioral pitted scars. Histologically, it is characterized by branching pigmented rete ridges associated with follicular infundibular involvement [10]. Syndromic reticulate pigmentary disorders, on the other hand, are usually distinguished by additional systemic manifestations, which were absent in the present case.

Published literature has described occasional atypical presentations of RAPK, including rare skeletal abnormalities and clinical overlap with DDD. These reports emphasize the phenotypic variability of reticulate pigmentary disorders and underscore the importance of histopathological confirmation, particularly in atypical or late-onset cases where clinical overlap may lead to diagnostic uncertainty [3-5].

Regarding management, effective treatment options for RAPK remain limited. Both topical and systemic retinoids have demonstrated minimal benefit. Topical azelaic acid has been associated with greater improvement in some reported cases. The erbium: YAG laser, used traditionally for other pigmentary disorders, represents a potential therapeutic modality. In the present case, topical corticosteroids and white soft paraffin combined with oral antihistamines produced symptomatic improvement, consistent with prior observations of conservative management [2,9].

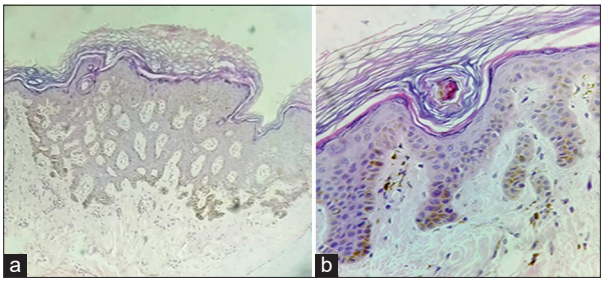

Figure 3: (a) Microphotograph showing hyperkeratosis, irregular acanthosis, focal thinning of the lining epithelium, and filiform elongation of rete ridges (hematoxylin and eosin [H&E], ×100). (b) Increased melanocytes within the filiform epidermal elongations (H&E, ×400).

Table 1: Key differentiating features of common reticulate pigmentary disorders [4,6,8,10]

Feature	RAPK	RAD (RAPD)	DDD	Dyskeratosis congenita	Naegeli syndrome
Distribution	Dorsal acral, proximal spread	Dorsal acral, symmetric	Flexural areas	Neck, chest (reticulate)	Trunk, neck
Hypo-pigmented lesions	Absent	Present (mixed)	Rare	Absent	Absent
Dermatoglyphic breaks	Present	Absent	Absent	Absent	Absent (reduced)
Palmar pitting	Present	Absent	Absent	Absent	Absent
Systemic features	Rare bony anomalies	None	Comedo-like lesions	Nail dystrophy, Bone marrow failure	Dental anomalies, hypohidrosis
Histology key feature	Filiform rete ridges+↑ melanocytes	Basal melanosis	Follicular infundibular projections	Variable	Variable
Gene	*ADAM10*	*ADAR1*	*KRT5*	*DKC1/TERC*	*KRT14*

RAD: Reticulate acropigmentation of Dohi, RAPK: Reticulate acropigmentation of Kitamura, DDD: Dowling-Degos disease

CONCLUSION

RAPK is a rare genodermatosis characterized by acral reticulate pigmentation, palmoplantar pitting, and dermatoglyphic disruption. Late presentation, as in this case, emphasizes the need for awareness of its full clinical spectrum. Clinicopathological correlation is essential for accurate diagnosis and differentiation from other reticulate pigmentary disorders.

DECLARATION OF PATIENT CONSENT

All appropriate patient consent was obtained in writing. The patient provided consent for the publication of his clinical images and other case-related information. The patient understands that his name and initials will not be published and that every effort will be made to preserve anonymity, though complete anonymity cannot be guaranteed.

REFERENCES

1. Adya KA, Inamadar AC, Palit A. Reticulate acropigmentation of kitamura: A dermoscopic perspective. Indian Dermatol Online J 2020;11:128-30.
2. Vasudevan B, Verma R, Badwal S, Pragasam V, Moorchung N, Badad A. A case of reticulate acropigmentation of kitamura: Dowling degos disease overlap with unusual clinical manifestations. Indian J Dermatol 2014;59:290-2.
3. MalaCards: The Human Disease Database. Reticulate Acropigmentation of Kitamura. 2021. Available from: https://www.malacards.org/card/reticulate_acropigmentation_of_kitamura [Last accessed on 2026 Feb 22].
4. El-Hoshy K, Hashimoto K. Bony anomalies in a patient with reticulate acropigmentation of Kitamura. J Dermatol 1996;23:713-5.
5. Weedon D. Disorders of pigmentation. In: Weedon's Skin Pathology. 3rd ed. Edinburgh: Churchill Livingstone Elsevier; 2010. p. 281-99.
6. Alfadley A, Al Ajlan A, Hainau B, Pedersen KT, Al Hoqail I. Reticulate acropigmentation of Dohi: A case report of autosomal recessive inheritance. J Am Acad Dermatol 2000;43 1 Pt 1:113-7.
7. Kono M, Sugiura K, Suganuma M, Hayashi M, Takama H, Suzuki T, *et al*. Whole-exome sequencing identifies ADAM10 mutations as a cause of reticulate acropigmentation of kitamura, a clinical entity distinct from dowling-degos disease. Hum Mol Genet 2013;22:3524-33.
8. Bolognia JL, Schaffer JV, Cerroni L, editors. Dermatology. 4th ed. Philadelphia, PA: Elsevier; 2018.
9. Köktürk A, İkizoğlu G, Baz K, Kaya TI. A case with features of reticulate acropigmentation of kitamura and dowling-degos disease. Turk Arch Dermatol Venereol 2002;36:286-9.
10. Wu YH, Lin YC. Generalized dowling-degos disease. J Am Acad Dermatol 2007;57:327-34.

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