Acral freckling with palmar pits

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A 32-year-old female patient presented with asymptomatic hyperpigmentation of both the forearms and hands since 15 years. The pigmentation initially began over the hands and gradually extended to involve the forearms. Patient's mother and sister also had similar complaints. Clinical examination revealed multiple hyperpigmented macules configured in a reticulate pattern involving the dorsal aspect of hands [Figure 1] and extensor aspects of both forearms [Figure 2]. Palmar aspects of both the hands had multiple fine pits [Figure 3]. Rest of the cutaneous examination was unremarkable. A 4 mm punch biopsy from the hyperpigmented region revealed slightly atrophic epidermis with an intensely melanized basal layer [Figure 4]. Based on the clinical and historical findings, a diagnosis of reticulate acropigmentation of Kitamura (RAK) was considered and confirmed by histopathological examination.

Reticulate acropigmentation of Kitamura is an autosomal dominantly inherited disorder characterized by reticulate macular hyperpigmentation involving the dorsa of hands and feet that begin in first or second decade and gradually progress to involve the extremities proximally and the trunk. It was first described by Kitamura and Akamatsu in 1943.[1] The condition is essentially asymptomatic except for the cosmetic concerns.[2] The hyperpigmentation is attributed to the increased number of active melanocytes in the basal layer and to the increased transfer of melanosomes to adjacent keratinocytes.[3] A characteristic feature of this condition is the presence of palmar pits and linear irregular interruptions in the dermatoglyphics. The reticulate hyperpigmentation may be seen involving the axillae as well. However, presence of palmar pits differentiates RAK from the reticular pigmented anomaly of the flexures (Dowling-Degos' disease [DDD]). Interestingly, there are several reports of RAK being associated with DDD[1,4,5] and it was proposed that DDD and RAK were different ends in the same spectrum of reticulate pigmentary anomalies.[6] However, exome sequencing of four family members in a pedigree with RAK has shown that mutations affecting the *ADAM10* gene (known to be involved in the ectodomain shedding of various substrates in the skin) as the cause of this disease. No mutations in the *KRT5* gene (the cause of DDD) were identified in any of these subjects and hence, it was concluded that RAK is entirely distinct from DDD.[7] Histopathologically, the hyperpigmented macules exhibit slight epidermal atrophy and heavily melanized basal layer with the increased number of dihydroxyphenylalanine (DOPA)-positive melanocytes, reminiscent of solar lentigo. Electron microscopy has shown melanosome complexes within the keratinocytes, dendrites filled with numerous melanosomes and melanosome complexes within the melanocytes.[3] No systemic

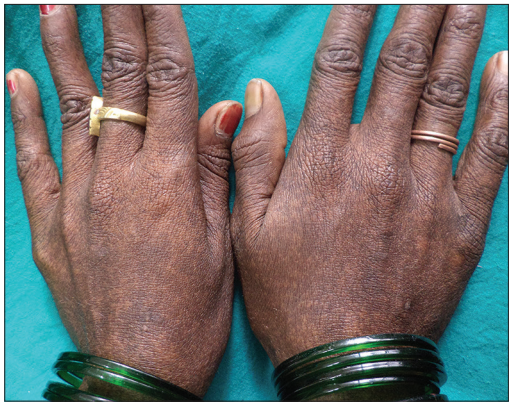

Figure 1: Reticulate freckle-like hyperpigmentation of the dorsa of hands

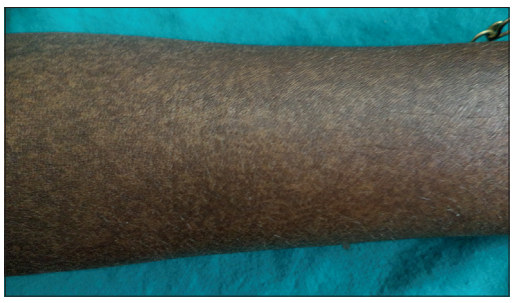

Figure 2: Hyperpigmented macules in a reticular fashion over the extensor surface of forearms

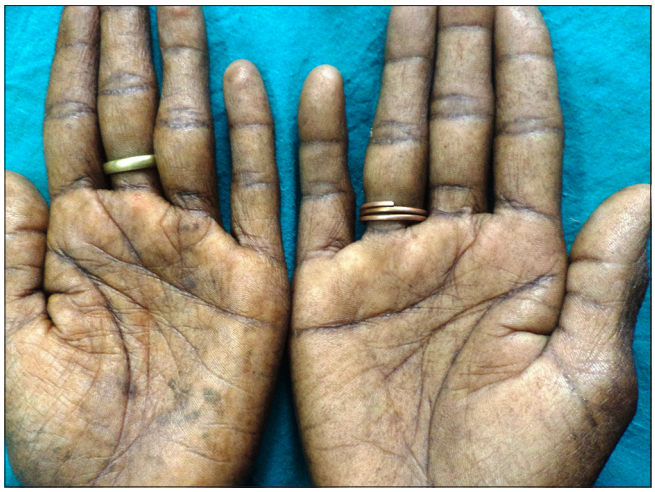

Figure 3: Fine pitting of the palms, especially prominent over the volar aspects of digits

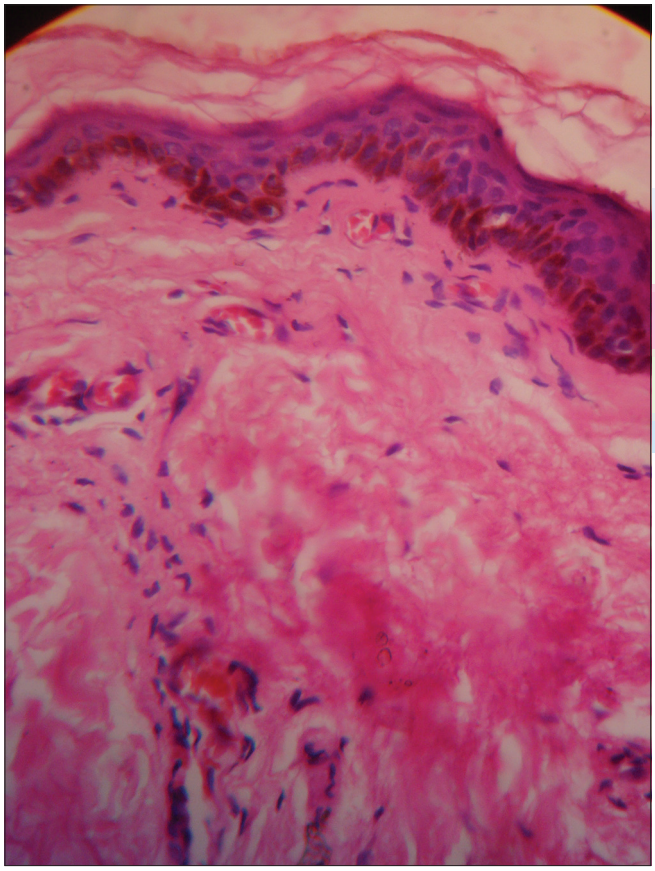

Figure 4: Increased melanization of the basal layer with slightly atrophic epidermis (H and E, ×40)

abnormalities are associated with RAK, although the absence of terminal phalanges in a 23-year-old female with RAK has been reported.[8]

The most important differential diagnosis is reticulate acropigmentation of Dohi which shares the same inheritance pattern and histopathological features. It is characterized by hyper- and hypo-pigmented macules in a reticulate fashion located on the extensor aspects of the extremities that may be associated with freckle-like pigmentation of the malar area. The lesion begin to appear earlier in life and become static by puberty. No palmar pitting is seen. Other differential diagnoses include dyskeratosis congenita, dermatopathia pigmentosa reticularis, dyschromatosis universalis hereditaria and Galli–Galli disease all of which exhibit reticulate pigmentary anomaly.[2] Palmar and/or plantar pitting is seen in several other conditions like Gorlin syndrome (nevoid basal cell carcinoma syndrome), pitted keratolysis, punctate keratoderma, and chronic arsenic poisoning. Treatment of the hyperpigmentation in RAK is difficult.

REFERENCES

1. Tang JC, Escandon J, Shiman M, Berman B. Presentation of reticulate acropigmentation of kitamura and dowling-degos disease overlap. J Clin Aesthet Dermatol 2012;5:41-3.
2. Adya KA, Inamadar AC, Palit A. Reticulate dermatoses. Indian J Dermatol 2014;59:3-14.
3. Mizoguchi M, Kukita A. Behavior of melanocytes in reticulate acropigmentation of Kitamura. Arch Dermatol 1985;121:659-61.
4. Shen Z, Chen L, Ye Q, Hao F, Vitorino Mdos S, Yang X, *et al.* Coexistent Dowling-Degos disease and reticulate acropigmentation of kitamura with progressive seborrheic keratosis. Cutis 2011;87:73-5.
5. Al Hawsawi K, Al Aboud K, Alfadley A, Al Aboud D. Reticulate acropigmentation of Kitamura-Dowling Degos disease overlap: A case report. Int J Dermatol 2002;41:518-20.
6. Kocatürk E, Kavala M, Zindanci I, Zemheri E, Koç MK, Sarigül S. Reticulate acropigmentation of Kitamura: Report of a familial case. Dermatol Online J 2008;14:7.
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. el-Hoshy K, Hashimoto K. Bony anomalies in a patient with reticulate acropigmentation of Kitamura. J Dermatol 1996;23:713-5.

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