Dermoscopy of Erythrokeratoderma Variabilis

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A 24-year-old male, born of third degree consanguineous marriage, presented with aymptomatic dark colored scaly lesions over neck, trunk, and extremities since 6 months of age, with aggravation in winters and relief with emollients. Dermatologic examination revealed multiple, confluent, irregularly-shaped, sharply demarcated, hyperpigmented scaly plaques, symmetrically distributed over neck, trunk, and extremities. [Figure 1] There was hyperlinearity of soles and palms. Scalp, face, mucosae, genitalia, hair, and nails were spared. Dermoscopy of plaque over neck showed white scaly lines over focal areas of dark brown to light brown hyperpigmentation and white scales. It also showed linear crista cutis and sulcus cutis like pattern similar to acanthosis nigricans [Figure 2a]. Dermoscopy from lower back revealed white scales arranged around focal areas of hyperpigmentation consisting of brown dots and globules [Figure 2b]. Skin biopsy from neck showed hyperkeratosis with papillomatosis and focal hypergranulosis. Pigmentation and mild vacuolar degeneration of basal layer with melanophages, and perivascular lymphohistiocytic infiltrate in the superficial dermis was present [Figure 3]. Based on these findings, diagnosis of Erythrokeratoderma variabilis (EKV) Mendes da Costa hyperkeratotic type was considered.

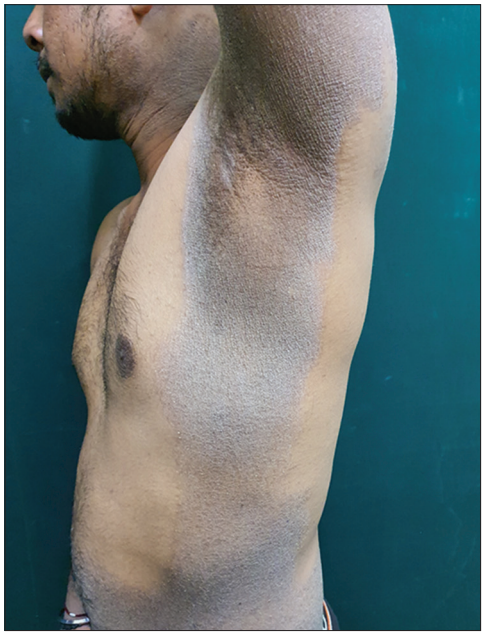

Figure 1: Multiple, confluent, irregularly-shaped, sharply demarcated, hyperpigmented scaly plaques with an erythematous border at places, distributed over neck, axilla and lateral aspect of trunk

Erythrokeratodermas (EK) are a rare group of keratinizing disorders broadly classified into erythrokeratoderma variabilis (EKV) and progressive symmetric erythrokeratoderma (PSEK) with overlapping features in few patients.[1] EKV is an autosomal dominant disorder, presenting at birth or during infancy with co-existence of transient, migratory and fixed erythematous, annular, or hyperkeratotic plaques predominantly over extensor surfaces of the distal limbs, trunk and palmoplantar keratoderma (PPK) in approximately 50% individuals.[2] Lesions of PSEK are similar to EKV but are nonmigratory typically sparing the trunk and

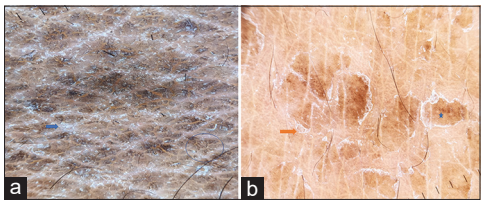

Figure 2: (a) Dermoscopy of plaque over neck shows reticular pattern of white scaly lines (blue arrow) surrounding focal areas of dark brown to light brown hyperpigmentation. It shows linear crista cutis and sulcus cutis like pattern with cristae showing hyperpigmentation. (blue circle) (Dermlite DL4, polarized 10×). (b) Dermoscopy from lower back revealed white scales (orange arrow) arranged around focal areas of hyperpigmentation consisting of brown dots and globules. (Dermlite DL4, polarized 10×)

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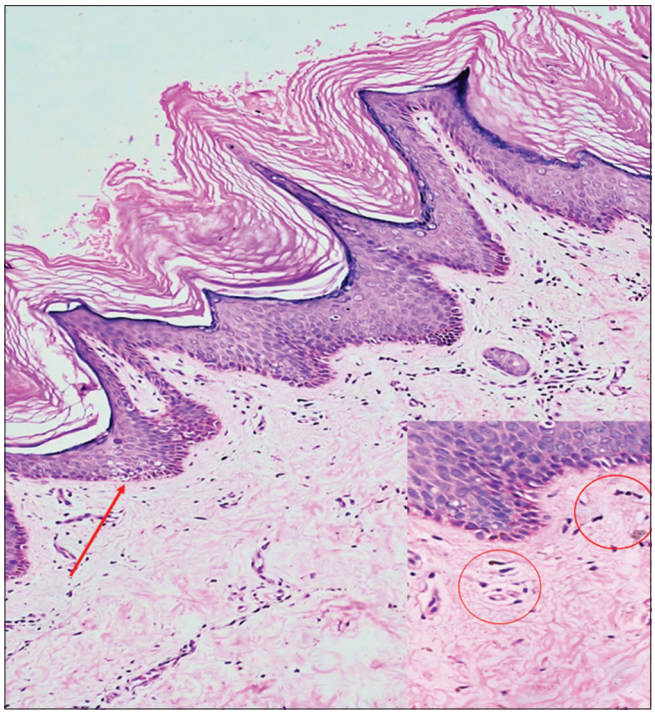

Figure 3: Skin biopsy from neck showed hyperkeratosis with papillomatosis and focal hypergranulosis. Pigmentation and mild vacuolar degeneration of basal layer (red arrow) with melanophages (red circle) and perivascular lymphohistiocytic infiltrate in the superficial dermis was present. (H & E, 10×)

has greater incidence of PPK. Dermoscopy of PSEK has been described in a single case report and depicts white scaly lines over focal area of hyperpigmentation.[3] The possible histopathologic correlates of various dermoscopic findings in our case are; white scales corresponds to hyperkeratosis, white scaly lines to focal hypergranulosis (similar to Wickham's striae), brown dots, and globules to melanophages and linear crista cutis and sulcus cutis like pattern with dark brown hyperpigmentation corresponding to acanthosis and papillomatosis similar to acanthosis nigricans. EKV seems to show a distinctive dermoscopic pattern and may be helpful to differentiate other dermatoses such as pityriasis rubra pilaris which shows whitish or yellowish keratotic plugs and linear vessels over a yellow background, and psoriasis with white scales and regular dotted vessels over a pinkish background.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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