Short Communication

Progressive symmetric erythrokeratoderma in a child treated successfully with tazarotene

Sir, erythrokeratoderma is a heterogeneous group of inherited cornification disorders which presents with erythematous hyperkeratotic plaques which are either migratory or stationary. Progressive symmetric erythrokeratoderma (PSEK) is a rare subtype characterized by nonmigratory, hyperpigmented, symmetric plaques that are usually distributed on the extremities, buttocks and sometimes the face. Very few cases of PSEK are reported from India.

A 10-year-old girl presented with dry, scaly skin lesions involving her neck, axillae and popliteal fossae since the age of 9 months. There was neither a family history of similar lesions nor any seasonal variation. The skin lesions had a recurrent and relapsing course. Cutaneous examination revealed symmetric well-defined, scaly plaques involving the cubital fossae (**Figure 1**), wrists and popliteal fossae. Rippled hyperkeratosis was present over the axillae and neck (**Figure 2**). There was no palmoplantar keratoderma and systemic examination was normal. Histopathological examination of the skin biopsy showed hyperkeratosis, acanthosis and mild papillomatosis with superficial perivascular infiltrate in the dermis suggestive of progressive symmetric erythrokeratoderma (PSEK) [**Figures 3**].

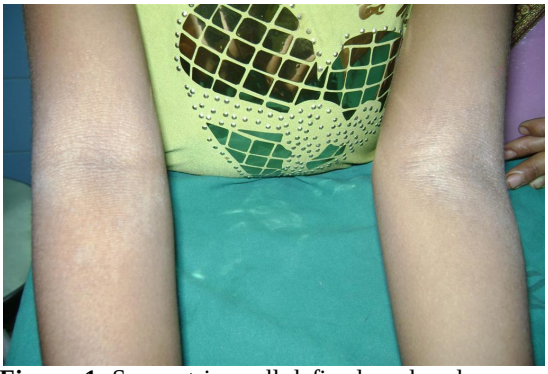

Figure 1 Symmetric well-defined scaly plaques involving the cubital fossa

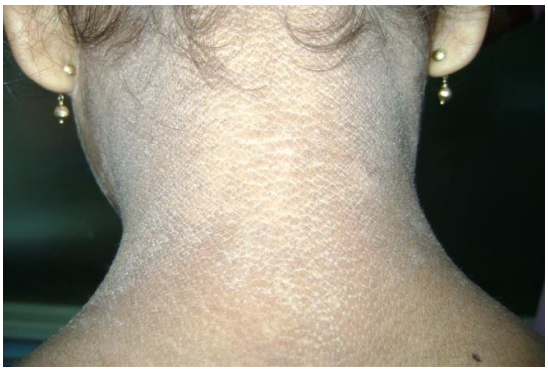

Figure 2 Rippled hyperkeratosis on neck.

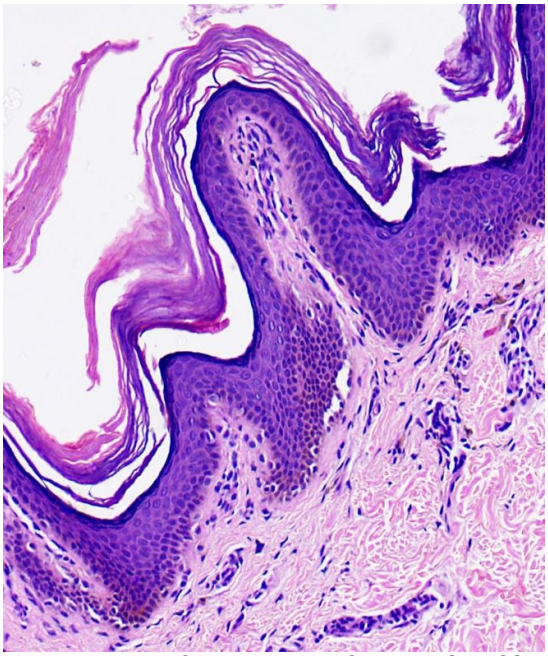

Figure 3 Hyperkeratosis, acanthosis and mild papillomatosis with superficial perivascular infiltrate in the dermis

Discussion

Progressive symmetric erythrokeratoderma (PSEK) also known as Gottron's syndrome is a rare autosomal dominant genodermatoses

characterized by large, fixed, geographic and symmetrical fine scaly erythematous plaques over the knees, elbows, shoulder girdle, hands and feet. Onset of PSEK is usually in early childhood. The condition progresses over the next few years and then becomes stable with the morphology, colour and sites remaining constant over time. A positive family history may be elicited in only about 50 percent patients, rest of cases are due to spontaneous mutation of the loricrin gene. Loricrin is a major structural component of the cornified cell envelope, formed beneath the plasma membrane of stratified squamous epithelial cells during terminal differentiation. Association of PSEK with palmoplantar keratoderma, ataxia and syndactyly has been reported.[1]

PSEK needs to be differentiated from erythrokeratoderma variabilis. Unlike in PSEK, the lesions in erythrokeratoderma variabilis fluctuate in their extent and configuration, involve the abdomen and thorax in addition to the extremities and show seasonal variation. Biopsy findings essentially are a psoriasiform hyperplasia with focal parakeratosis and well preserved granular layer. There is no suprapapillary thinning of the epidermis or Munro's microabscesses as seen in psoriasis. The other differentials that need to be considered are psoriasis and pityriasis rubra pilaris which can aptly be ruled out on the basis of histopathology.[2,3]

Therapeutic options for PSEK include emollients, topical and oral retinoids. Our patient was treated with tazarotene and emollients with which her condition improved remarkably.

References

1. Verma R, Jaiswal AK, Vaishampayan SS, Baveja S. Progressive symmetric erythrokeratoderma. *Indian J Dermatol* 1999; **42**: 84-5.
2. Brahma D, Jain VK. Erythrokeratoderma variabilis. *Indian J Dermatol Venereol Leprol* 2003; **69** (Suppl): S5-S6
3. Chu DH, Arroyo MP. Progressive and symmetric erythrokeratoderma. *Dermatol Online J* 2003; **9** (4): 21**.**

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An interesting case of idiopathic patterned hypomelanosis – a new entity?

Sir, the practice of dermatology sometimes presents us with riddles, implication of which goes beyond the boundaries of hitherto published wisdom. Here, we present a case of idiopathic patterned hypomelanotic condition which showed unique histopathological features.

A 20-year-old male student presented to our hospital with asymptomatic, non-scaly, hypomelanotic macules over chest, abdomen and back for six months. The lesions were insidious in onset, with no history of pre-existing dermatosis. Morphologically three different types of lesions were present: oval, arcuate and annular (**Figure1**). All lesions had well-defined borders with no sensory change, anhidrosis or alopecia. Cutaneous and cranial nerve examination depicted no abnormality. Sensory and motor functions were absolutely normal. Patient did not have any history of kala-azar or previous nodular or ulcerated lesions. Careful family history could not elicit