Progressive Symmetric Erythrokeratodermia with Overlying Psoriasis as Wolf’s Isotopic Phenomenon: A Case Report

Dear Editor,

Progressive symmetric erythrokeratodermia (PSEK) is a rare genodermatosis characterized by well-defined erythematous hyperkeratotic plaques in a symmetrical distribution. Darrierin 1911 first described this condition; Gottron renamed it.[1] The usual onset is in infancy, with progression during childhood followed by a stable course. We report a rare and probably the first case of sporadic onset PSEK with psoriasis developing over it as Wolf’s isotopic phenomenon associated with psoriatic arthritis.

A 32-year-old male, born out of a nonconsanguineous marriage, having normal growth, presented with multiple asymptomatic skin lesions since 1 year of age. The lesions started as red scaly plaques involving the foot’s right ankle and dorsum, gradually involving the hands and feet, bilateral knees, elbows, and the sacral area. Over the next 7 years, it stabilized except for mild exacerbations during the winters.

On examination, multiple well-defined, erythematous, hyperkeratotic plaques with mild scaling were noted over the feet [Figure 1a], hand [Figure 1b], knees [Figure 1c] including palmoplantar keratoderma. All other mucocutaneous and systemic examinations and routine investigations were normal. Dermoscopy [Figure 1d] supported the clinical diagnosis of PSEK. Histopathology [Figure 2] showed a sparse superficial perivascular lymphocytic infiltrate with mild focal spongiosis, lamellated parakeratosis and orthokeratosis, and slight epidermal hyperplasia. He was counseled about the prognosis, and keratolytic use was advised.

One year later, he developed a few hyperkeratotic papules over and above the PSEK plaque of the left dorsum of the hand with concomitant pain on the left plantar surface. Oral isotretinoin was started suspecting the winter exacerbation of PSEK, and orthopedics consultation for the out-of-proportion disabling pain was obtained. The PSEK plaque improved with isotretinoin, but the new papules persisted along with the eruption of similar erythematous, hyperkeratotic well-defined papules all over the pre-existing PSEK plaque of the hand [Figure 3a], knee [Figure 3b], and elbow [Figure 3c]. He gradually developed pain in the bilateral knee, back, left wrist, and left little finger dactyilitis. It aroused the suspicion of psoriasis over the pre-existing PSEK.

Dermoscopy [Figure 3d] showed regularly placed dotted blood vessels along with fine scaling over an erythematous background, and histopathology [Figure 4] confirmed clinical diagnosis. The ESR and CRP were raised; rheumatoid factor and anticyclic citrullinated peptide antibodies (anti-CCP) were negative. X-rays showed mild reactive changes, and a diagnosis of psoriatic arthritis according to the Classification of Psoriatic Arthritis (CASPAR)’s criteria was made.

Wyburn–Mason reported an unrelated disease at the previously healed skin lesion site, and Wolf termed it an “isotopic phenomenon.” Psoriasis has been reported to develop over previous lesions of herpes zoster, pemphigus vulgaris, and striae distensae.[2-4] To the best of our knowledge, no other disease over PSEK has been reported to date. There are few case reports of association of PSEK with ataxia, peripheral neuropathy, nephrotic syndrome, syndactylism, keratosis pilaris, bilateral cortical cataract, mental retardation, narcolepsy, convulsions, and isotopic phenomenon.

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deafness, delayed intellectual milestones, dermatophytosis, high arched palate, fissured tongue, and sternal depression (pectus-excavatum).\textsuperscript{1,5-7} However, our patient did not had any such association.

Dermoscopy is playing a crucial role in making a better clinical diagnosis. Erythrokeratoderma variabilis, psoriasis, and pityriasis rubra pilaris (PRP) are considered differentials of PSEK. Psoriasis and PRP can be easily differentiated from PSEK with dermoscopy and clinicopathological correlation. In EKV, the lesions are more transient and precipitated by extremes of temperature, emotional
stress, friction, and sun exposure. Face and palmoplantar involvement are less as compared to PSEK.

Our patient developed new lesions of psoriasis within the pre-existing PSEK plaques. It may occur due to Wolf’s isotopic or because of some unknown association between psoriasis and PSEK at the genetic level or might be merely a coincidence. Mutation in patients with erythrokeratoderma is well-known. Although sporadic cases occur due to new genetic mutations, we could not do the genetic analysis of the patient for logistic reasons. However, both psoriasis and PSEK lesions were well controlled on oral methotrexate in our patient.

**IRB statement**

Consent for publication has been obtained from the patient. In our institute, the case report does not require ethical approvals.

**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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Nil.

**Conflicts of interest**

There are no conflicts of interest.

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