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Single Case

Unilateral Linear Punctate Palmoplantar Keratoderma: A Case Report

Chanisa Kiatsurayanon[a] Jinda Rojanamatin[a] Poonawis Sudtikoonaseth[a] Kowit Kampirapap[a] Mingkwan Wichaidit[a] François Niyonsaba[b, c]

[a]Institute of Dermatology, Department of Medical Services, Ministry of Public Health, Bangkok, Thailand; [b]Atopy (Allergy) Research Center, Juntendo University Graduate School of Medicine, Tokyo, Japan; [c]Faculty of International Liberal Arts, Juntendo University, Tokyo, Japan

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Abstract
Punctate palmoplantar keratoderma (PPPK) is a rare entity with an estimated prevalence rate of 1.17/100,000. PPPK usually presents with bilateral asymptomatic, tiny, hyperkeratotic punctate papules and plaques on the palmoplantar surface. Among the PPPK varieties, the linear presentation is much rarer, and so far there have been only 3 case reports. Here, we report the case of a 27-year-old female Thai patient who presented to our outpatient clinic with unilateral asymptomatic linear thickening lesions on her right sole since childhood. There were no similar lesions on other parts of the body. A histopathologic examination revealed epidermal hyperplasia and hyperkeratosis without columns of parakeratosis or cornoid lamella. The other examinations were normal. The clinical and histological contexts were consistent with a diagnosis of unilateral linear PPPK. The patient was treated with topical 10% urea cream and 10% salicylic acid cream twice daily. To the best of our knowledge, this is the first reported case of unilateral linear PPPK in Thailand, and the fourth reported case worldwide.

KARGER

Chanisa Kiatsurayanon
Institute of Dermatology, Department of Medical Services, Ministry of Public Health
420/7 Rajvithi Road
Phayathai District, Rajthevee, Bangkok 10400 (Thailand)
E-Mail chanisa.kiatsurayanon@gmail.com

Introduction

Palmoplantar keratodermas (PPKs) are a heterogeneous group of disorders characterized by abnormal hyperkeratosis affecting the palms and soles [1]. PPKs can be classified based on whether they are inherited or acquired. Inherited PPKs can be further divided clinically into 3 distinct subgroups: diffuse PPK, focal PPK, and punctate PPK [1].

Case Presentation

A 27-year-old female Thai patient presented to our outpatient department with unilateral asymptomatic linear thickening lesions on her right sole since childhood. The lesions first appeared as punctate small plaques that gradually increased in number and extended in linear fashion along the plantar surface. The patient had no history of a traumatic event prior to the appearance of the lesions. There were no similar lesions on any other parts of the body, and none of her family members had similar lesions. A review of systems did not reveal any symptoms of systemic involvement such as fever, chronic weight loss, or anorexia. The patient is a Chinese-Thai interpreter in China, and she denied any history or current use of drugs, alcohol, tobacco, or any other medication.

A dermatologic examination revealed unilateral localized skin-colored-to-yellowish hyperkeratotic papules and plaques distributed in linear configuration on the right plantar surface (Fig. 1). Based on the clinical presentation, we made the differential diagnoses of linear punctate PPK (PPPK), linear punctate porokeratosis, linear epidermal nevus, callus, and verruca vulgaris. We performed a 4-mm punch biopsy, and the specimen showed epidermal hyperplasia with hypermelanization and hyperkeratosis without columns of parakeratosis or cornoid lamella (Fig. 2). Furthermore, the dermis showed subtle inflammation. Based on the histopathologic findings combined with the clinical features, the final diagnosis of unilateral linear PPPK was made.

Discussion

PPPK is a rare entity of PPK with an estimated prevalence rate of 1.17/100,000 [1]. PPPK typically presents as bilateral asymptomatic tiny hyperkeratotic punctate papules/plaques on the palmoplantar surface [1]. Among the PPPK varieties, the linear presentation is much rarer, and its incidence remains unknown to date [1]. Although the exact etiology of PPPK is not fully understood, both genetic and environmental factors are believed to play some roles in triggering the disease [1, 2]. Autosomal dominant, recessive, mitochondrial, X-linked, and acquired forms of PPKs have all been described [2, 3]. Autosomal dominant PPPK, which is known to be the main mode of inheritance, has recently been found to be linked with 2 loci on chromosomes 15q22–15q24 and 8q24.13–8q24.21 [3].

Unilateral presentation of PPPK is rare and may be explained by genetic mosaicism [4]. Until recently, there have been only 3 reports of unilateral linear PPPK [4], and some cases have been associated with nail abnormalities such as longitudinal ridging, onychoschizia, onychorrhexis, trachyonychia, and notching [5]. Although associated systemic involvement is uncommon, patients with PPPK may have an increased risk of developing some malignant conditions such as gastrointestinal, lung, and breast cancer as well as Hodgkin's lymphoma [1]. Thus, longitudinal follow-up is mandatory in cases of hereditary PPPK.

Unfortunately, PPPK lesions do not resolve spontaneously, and treatment is generally symptomatic. Topical keratolytics, systemic retinoids, liquid nitrogen, PUVA (psoralen plus ultraviolet A), re-PUVA, and pairing have been used with variable success [1, 2]. Our patient was treated with topical 10% urea cream and 10% salicylic acid cream twice daily. After treatment, the lesions showed slight improvement; however, the patient was lost to follow-up, as she had already gone back to China.

In conclusion, we report a typical case of unilateral linear PPPK. Our patient presented with clinical findings and histopathologic features compatible with linear PPPK. So far, this is the fourth case of unilateral linear PPPK reported in the world, and the first one in Thailand. After diagnosis, patients with PPPK should be scheduled for long-term follow-up, because they may have an increased risk of developing some cancers. Generally, no treatment is needed for PPPK, but doctors may offer symptomatic treatment to alleviate the symptoms and improve disfigurement from PPPK lesions.

Statement of Ethics

The authors confirm that the patient provided written informed consent to use her photos for publication.

Disclosure Statement

The authors have no conflicts of interests to disclose.

References

1 Oztas P, Alli N, Polat M, Dagdelen S, Ustün H, Artüz F, Erdemli E: Punctate palmoplantar keratoderma (Brauer-Buschke-Fischer syndrome). Am J Clin Dermatol 2007;8:113–116.

2 Lieberman MR, Kober M, Lowenstein EJ, Heilman E: Punctate palmoplantar keratodermas: case reports and a review of the literature and terminology. J Dermatol Plast Surg 2016;1:1003.

3 Gao M, Yang S, Li M, Yan KL, Jiang YX, Cui Y, Xiao FL, Shen YJ, Chen JJ, Liu JB, Xu SJ, Huang W, Zhang XJ: Refined localization of a punctate palmoplantar keratoderma gene to a 5.06-cM region at 15q22.2–15q22.31. Br J Dermatol 2005;152:874–878.

4 O'Toole A, O'Malley M: Unilateral keratoderma in a mother and her son. J Cutan Med Surg 2012;16:288–290.

5 Gupta R, Mehta S, Pandhi D, Singal A: Hereditary punctate palmoplantar keratoderma (PPK) (Brauer-Buschke-Fischer syndrome). J Dermatol 2004;31:398–402.

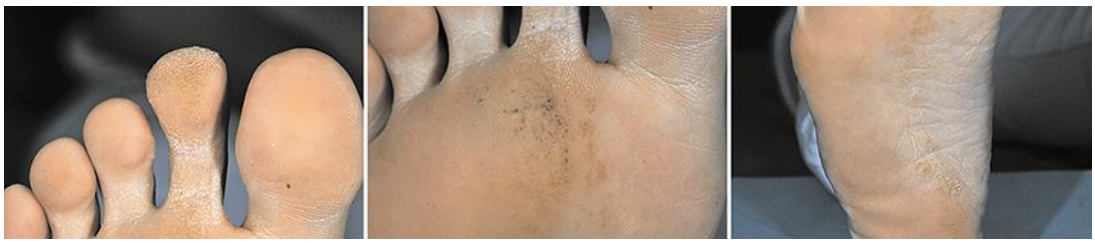

Fig. 1. Unilateral localized skin-colored-to-yellowish hyperkeratotic papules and plaques were distributed in a linear pattern on the right plantar surface of the patient.

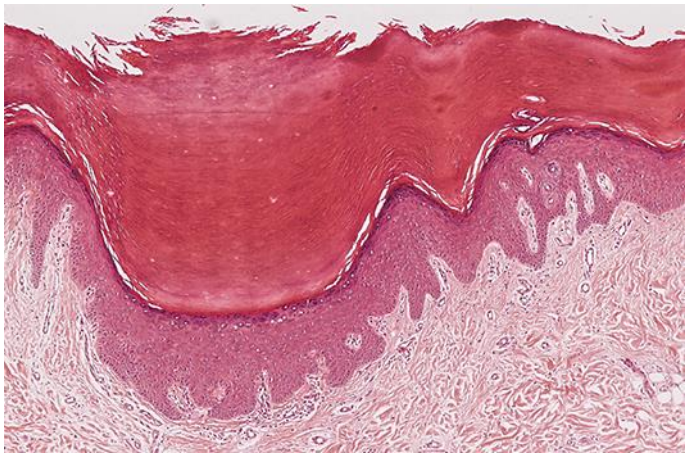

Fig. 2. Histopathology showed epidermal hyperplasia with hypermelanization and hyperkeratosis without columns of parakeratosis or cornoid lamella. H&E. ×10.