A Rare Case Report: Unilateral Punctate Palmoplantar Keratoderma

Nadir Bir Olgu Sunumu: Unilateral Punktat Palmoplantar Keratoderma

Nadir Bir Keratoderma Olgusu / A Rare Case of Keratoderma

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Özet

Brauer-Fischer-Buschke Sendromu (Punktat Palmoplantar Keratoderma) nadir görülen, genetik geçişli bir deri hastalığıdır. Unilateral lineer tutulum ise çok nadirdir ve halen sıklığı tam olarak bilinmemektedir. Biz burada el ve ayaklarında sarımtırak renkte lineer, punktat, hiperkeratotik ağrılı lezyonları bulunan 21 yaşındaki bir erkek hasta sunuyoruz. Bildiğimiz kadarıyla literatürde bildirilen üçüncü olgudur.

Anahtar Kelimeler

Keratoderma; Palmoplantar; Unilateral

Abstract

Brauer-Fischer-Buschke Syndrome (Punctate Palmoplantar Keratoderma) is a rare inherited genetic skin disorder. Unilateral linear presentation is a very rare entity of punctate keratodermas and the exact incidence is currently unknown. We report herein a 21-year-old male patient presenting with painful yellowish punctate hyperkeratotic linear lesions on his right palm and sole. To the best of our knowledge our case is the third one reported in the literature.

Keywords

Keratoderma; Palmoplantar; Unilateral

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Introduction

Punctate palmoplantar keratoderma (PPK) is a rare genodermatosis characterized by hyperkeratinization. Although, the exact etiology is unknown, some environmental and genetic factors have been implicated as possible etiologic factors [1]. The Prevalence is estimated to be around 1,17/100.000 [2,3]. Patients usually present with asymptomatic, multiple punctate, irregularly distributed, hyperkeratotic and verrucous lesions with sizes varying between 2-8mm in diameter occurring on both the palms and soles.

Case Report

A 21-year-old male patient presented to our outpatient clinic with complaints of multiple, painful, firm, yellowish to brown lesions on his right palm and sole. The patient claimed that his complaints started nearly 16 years ago over several areas of his palm and sole, and spread throughout the whole palm and sole within the succeeding years. Family history for similar complaints was negative and the patient neither had any history of food allergy, weight loss, and systemic disease nor did he have a regular drug usage history. He also did not have any similar lesions on the rest of his body. Routine blood samples, right hand, right foot and chest radiographies, electrocardiography and abdominal ultrasonography investigation were insignificant.

On dermatologic examination multiple, punctate and plaque-like lesions, bigger in size at pressure sites, were observed (Figure 1, zoomed-in view). While linear and punctate yellowish-brown hyperkeratotic lesions sized between 2 to 9 mm in diameter, plaques were 1 to 4 cm in diameter (Figure 1). The left palmoplantar region was lesion-free (Figure 2). The patient claimed that he sustained pain on performing activities such as walking



Figure 1. The zoomed-in view shows linear and punctate yellowish-brown hyperkeratotic lesions



Figure 2. Note the lesion-free left palmoplantar



Figure 3. Significant orthokeratotic hyperkeratosis and acanthosis with an intact dermis (HEx40)

or holding something. A 4-mm punch biopsy specimen revealed significant orthokeratotic hyperkeratosis and acanthosis with an intact dermis on histopathology (Figure 3). Given the clinical symptoms and histopathological findings the patient was diagnosed as “unilateral punctate palmoplantar keratoderma”.

Discussion

Palmoplantar keratodermas (PPKs), as the name implies, constitutes a group of diseases characterised by hyperkeratosis of the palms and soles. PPK is classified into acquired and hereditary forms. The hereditary form is further sub-classified into diffuse, focal and punctate types, according to the epidermal involvement and clinical pattern. While the diffuse type affects the palmoplantar surface uniformly, the focal type involves hyperkeratosis mainly on areas exposed to pressure and recurrent friction. Punctate PPK differs from these entities by multiple

small, hyperkeratotic papules, spicules or nodules partially or completely involving the palm and the sole [1]. Linear presentation is rare and unilateral linear presentation is even more uncommon. We were able to find only two other similar cases reported in the literature [2,4].

The exact incidence of punctate PPK is not known. It is more common among males (64,6%) and while almost half of the patients develop punctate PPK in their first decade of age, the incidence is higher between 11-20 ages (32,9 %)[5]. Although the exact etiology is currently unknown, genetic and environmental factors are believed to influence the course of the disease. The chromosome 15q22-q24 locus has previously been linked to punctate PPK [6].

The differential diagnosis of punctate PPK should include calluses, corns, linear punctate porokeratosis, porokeratotic eccrine ostial and dermal duct nevus (PEODDN), arsenical keratosis, secondary syphilis, AIDS associated keratoderma and striate PPK [2]. Treatment approaches focus on relieving the hyperkeratosis related discomfort. Although several topical alternatives such as keratolytics, salicylic acid and retinoids are being widely used they haven't proved to be efficient [1,7]. Studies on the use of systemic etretinate at 0.5–1.0 mg/kg dosage, on the contrary, reported better outcome results [8]. Because our patient refused to receive systemic acitretine due to its side effects; we used topical keratolytics for almost six weeks. Although hyperkeratosis partially resolved and a subjective pain relief was achieved with topical treatment, recurrence was observed shortly after the cessation of treatment.

To the best of our knowledge this case is the third one reported in the literature [2]. Our case is unique in that, it was subject of a disturbing pain (rated with visual analog scale). Moreover, the Dermatology Life Quality Index revealed a considerable impact of the disease on the life quality of the patient, which we believe is another important factor that should be taken into consideration when caring for these patients.

Competing interests

The authors declare that they have no competing interests.

References

1. Oztas P, Alli N, Polat M et al. Punctate Palmoplantar Keratoderma (Brauer-Buschke-Fischer Syndrome). American Journal of Clinical Dermatology 2007;8(2):113-116.
2. Sharma S, Barman KD, Garg VK et al. Unilateral linear punctate palmoplantar keratoderma. Indian J Dermatol Venereol Leprol 2012;78(1):85-8.
3. Stanimirovic A, Kansky A, Basta-Juzbasic A, Skerlev M, Beck T. Hereditary palmoplantar keratoderma, type papulosa, in Croatia. J Am Acad Dermatol 1993;29:435–7.
4. O'Toole A, O'Malley M. Unilateral keratoderma in a mother and her son. J Cutan Med Surg. 2012 Jul-Aug;16(4):288-90.
5. Mahajan PM, Gharpuray MB, Kulkarni V. Clinical and histopathological study of palmoplantar keratoderma. Indian J Dermatol Venereol Leprol 1994;60:258-61.
6. Martinez-Mir A, Zlotogorski A, Londono D, Gordon D, Grunn A, Uribe E, et al. Identification of a locus for type I punctate palmoplantar keratoderma on chromosome 15q22–q24. J Med Genet 2003;40:872–8.
7. Özdemir M, Gümüşel M, Toy H. Tip 1 Herediter Punktat keratoderma. Turkiye Klinikleri J Dermatol 2006;16:201-204.
8. Judge MR, Mclean WHI, Munro CS. Disorders of keratinisation. In: Burns T, Breathnach S, Cox N, Griffiths C, editors. Rook's textbook of dermatology. 8th ed. Oxford: Blackwell Science; 2010. p. 19.112–3.

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