

## Parakeratosis Pustulosa: A Rare Disease

Ayşe Serap Karadağ,<sup>1</sup> MD, Serap Güneş Bilgili,<sup>2\*</sup> MD, İbrahim Halil Yavuz,<sup>2</sup> MD, Aysegül Erdem,<sup>3</sup> MD, Göknur Özaydın Yavuz,<sup>2</sup> MD

Address: <sup>1</sup>İstanbul Medeniyet University, Department of Dermatology, Goztepe Trainig and Research Hospital, Istanbul, Turkey <sup>2</sup>Yuzuncu Yil University Faculty of Medicine, Van, Turkey, <sup>3</sup>İstanbul Medeniyet University, Department of Pathology, Goztepe Trainig and Research Hospital, Istanbul, Turkey

E-mail: dretuba\_oz@hotmail.com

\* Corresponding Author: Dr. Serap Güneş Bilgili, Yuzuncu Yil University, Faculty of Medicine, Department of Dermatology, Van, Turkey

Published:

J Turk Acad Dermatol 2015; 9 (4): 1594c4

This article is available from: <http://www.jtad.org/2015/4/jtad1594c4.pdf>

**Keywords:** Eczematoid eruption, Parakeratosis pustulosa

### Abstract

**Observation:** Parakeratosis pustulosa is predominantly seen in children as an eczematoid eruption adjacent to the free margin of nail, extending to the dorsal nail fold. This entity a distinct but less commonly known skin disease. Differential diagnosis should be considered Hallopeau's acropustulosis, palmoplantar pustulosis, palmoplantar psoriasis, Andrew's pustular bacteridid, dermatophyte infection, and contact dermatitis. Histopathological assessment are essential in the diagnosis.

### Introduction

Parakeratosis pustulosa (PP), is also named as Hjorth-Sabouraud syndrome, is a rare chronic dermatosis. Most of the cases are diagnosed before age 5. Fingers and nails are the most commonly affected locations. The etiology is unclear and it usually presents with erythematous and squamous plaques [1]. Herein, we present a nine year-old boy presenting with treatment-resistant erythematous, pustular lesions in the big toe for six months.

### Case Report

A 9 year-old boy was brought to the Dermatology polyclinic with complaints of thickened scaly patches on the left great toe for six months. Disease started initially with scaling and gradual thickening of skin with pustular eruption. It isn't recovered after treatment with topical medication such as corticosteroids, antibiotics, antimycotics etc. No

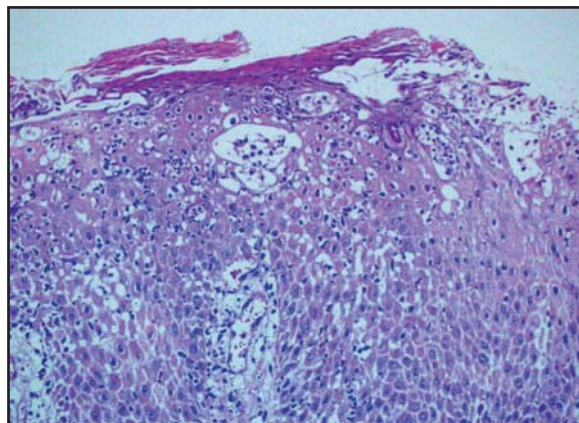
history of new shoes, synthetic socks. Family history was normal. Dermatological examination of the left great toe revealed eczematous scaly patches and pustular eruption (Figures 1 and 2). Potassium hydroxide examination of the skin scrapings showed no evidence of fungal infection. Bacterial culture was sterile. Routine haematolo-



**Figure 1.** Eczematous scaly patches and pustular eruption on the left great toe



**Figure 2 .** Eczematous scaly patches and pustular eruption on the left great toe



**Figure 3 .** Hyperkeratosis, parakeratosis, mild acanthosis, and papillomatosis with infiltrate around the dilated blood vessels (H-E x20)

gical and biochemical investigations were within normal limits. Histopathology of the lesion revealed hyperkeratosis, parakeratosis, mild acanthosis, and papillomatosis with infiltrate around the dilated blood vessels (**Figure 3**). Based on the clinical, and histopathological findings, he was diagnosed with parakeratosis pustulosa. The patient was treated with topical calcipotriol, which resulted in marked improvement of within two weeks period.

### Discussion

The incidence of PP is not completely known due to under-diagnosis of PP. The first serious study done by Hjorth and Thomsen investigated 91 cases with PP. The etiology is unclear and there is no genetic penetrance of PP. However, PP may have relationship with psoriasis, contact dermatitis and onychomycosis. PP is more frequently seen in girls before the age of five. In contrast, our case was boy and at age nine. PP is usually affected thumb, and middle toes [2]. In our case, a foot involvement was observed. Lesions usually start with pink or skin-color, and mildly squamous plaques. Vesicular or pustular changes can be seen in the lesions. Onycholysis is commonly present in distal nails. Subungual hyperkeratosis and nail bed changes can be accompanied by PP. However, nail pitting is rarely seen. Nail involvement can cause deformities [3, 4]. Erythema and squamous changes are frequently seen in distal nails, particularly in the pulp. However, pain and itching is not typically present.

The diagnosis of PP can be made based on clinical and histopathological findings. However, minor and major diagnostic criteria have been described in recent years. The major criteria are frequently seen in children, benign process and spontaneous remission, exclusion of fungal infection with KOH examination. The minor criteria are negative family history, rare and transient pustules, non-responsiveness to topical emollients, non-specific histopathological findings [1]. Hyperkeratosis, parakeratosis, pustule, acanthosis, mild ectocytosis, papillomatosis, and lymphocytic infiltration can be seen in the histopathological examination. The histopathological findings are nonspecific because these findings can be seen in psoriasis and contact dermatitis as well [5].

The differential diagnoses of PP include Hallopeau acrodermatitis, pustular psoriasis, contact dermatitis, atopic dermatitis, tinea pedis, and paronychia. A determination of the major and minor criteria can help to make accurate diagnosis of PP [6].

There is no specific treatment for PP [7]. Topical emollient, corticosteroids are commonly used. Topical tretinoin, and calcipotriol can be effective. The disease has good prognosis and spontaneous remission but it does not have to be seen in all PP cases [1, 2, 3, 4, 5, 6]. In our case, topical calcipotriol was administered with remarkable remission.

In the case of pustular lesions on the plantar region in the childhood, parakeratosis pustulosa should be considered in the differential diagnosis.

## References

1. Mahajan VK, Ranjan N. Parakeratosis pustulosa: A diagnostic conundrum. Indian J Paediatr Dermatol 2014; 15: 12-15.
2. Hjorth N, Thomsen K. Parakeratosis pustulosa. Br J Dermatol 1967; 79: 527-532. PMID: 6061563
3. Cronin E, Samman PD. Parakeratosis pustulosa. Clin Exp Dermatol 1986; 11: 643-645. PMID: 3665153
4. Tosti A, Peluso AM, Zucchelli V. Clinical features and long-term follow-up of 20 cases of parakeratosis pustulosa. Pediatr Dermatol 1998; 15: 259-263. PMID: 9720686
5. Avci O, Gunes AT. Parakeratosis pustulosa with dyskeratotic cells. Dermatology 1994; 189: 413-414. PMID: 7873832
6. Pandhi D, Chowdhry S, Grover C, Reddy BS. Parakeratosis pustulosa--a distinct but less familiar disease. Indian J Dermatol Venereol Leprol 2003; 69: 48-50. PMID: 17642830
7. Richert B, André J. Nail disorders in children: Diagnosis and management. Am J Clin Dermatol 2011; 12: 101-112. PMID: 21348541