

Porokeratotic eccrine ostial and dermal duct nevus (PEODDN): A case report

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Porokeratotic Eccrine Ostial and Dermal Duct Nevus (PEODDN) is a rare disorder of keratinization that clinically resembles comedo nevus but occurs on the palms and soles and is characterized by cornoid lamella in the histopathology. We hereunder report a young male with mildly itchy papules on the lateral surface of the left foot who was diagnosed with PEODDN upon biopsy of the lesion. We briefly reviewed the several aspects of the condition in our paper.

Keywords: cornoid lamella, parakeratosis, porokeratotic eccrine ostial and dermal duct nevus, porokeratosis

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INTRODUCTION

The term Porokeratotic Eccrine Ostial and Dermal Duct Nevus (PEODDN) was first coined in 1980 by Abell and Read¹. However, it was first described by Marsden in 1979². PEODDN is a very rare skin condition which is classified as porokeratotic dermatoses. It is characterized by cornoid lamella which is a column of parakeratotic cells and is associated with dyskeratosis in the spinous layer as well as reduction in the number of granular zone cells. It is in close association with subjacent acrosyringia³. Hereunder, we report a case with this rare condition.

CASE REPORT

A 25-year-old male came to our outpatient clinic with a ten-year history of a mildly- itchy lesion on

his left foot. No other symptoms were detected. Past history as well as family and drug history was unremarkable. The lesion was unilateral and limited to the lateral surface of left foot. It consisted of multiple erythematous papules which were distributed in a linear fashion and plugs were seen in the center of most lesions (Figure 1, 2). No other cutaneous lesions were noted. General physical examination showed no abnormality. A skin biopsy showed cornoid lamella which is exclusively associated with eccrine acrosyringia (Figure 3).

DISCUSSION

To date, only 42 cases (Medline search) of PEODDN have been reported. Many of them present at birth or at young ages although some may occur in adults or even in the elderly³. Male



Figure 1. Multiple erythematous papules which were distributed in a linear fashion on the lateral aspect of foot



Figure 2. Multiple erythematous plantar papules and plugs were seen in the center of most lesions

predominance is seen in most of the studies, although the number of cases is too few to make a conclusion. For example, in a retrospective study on ten cases, the male/female ratio was 7:3⁴ while in a review of 24 cases, this ratio was 12:10 (two unknown)⁵. Our case was diagnosed after 10 years, similar to a case series of ten patients in which the duration between onset and diagnosis is from several months to seventeen years⁴.

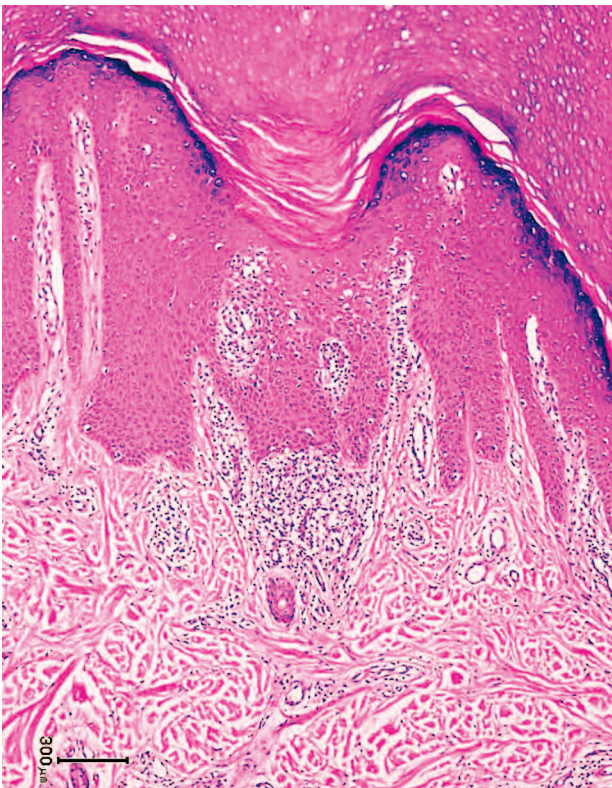


Figure 3. Cornoid lamella which is exclusively associated with eccrine acrosyringia (H&E*40)

In reported cases, the lesions are mainly located on extremities, as in our case. Trunk, forehead and neck involvement have also been reported. The lesions may also be bilateral, generalized, or occur along Blaschko lines⁶⁻¹⁴. It is usually asymptomatic, although it may be accompanied by a mild pruritus, hyperhidrosis or anhydrosis. Association with other conditions is rare and includes neurological problems (sensory polyneuropathy, seizure, developmental delay, deafness, and hemiparesis), scoliosis, palmoplantar keratoderma, onychodysplasia, alopecia, and hyperthyroidism^{9,15,16}. There is also a report of association with Bowen disease¹⁷.

Etiologically, it has been proposed that the invagination of the epidermis may result from an abnormal clone of epidermal cells which leads to the formation of cornoid lamella¹⁸. Another hypothesis suggests that the invagination is a dilated acrosyringial and dermal duct which is keratin-plugged¹⁹.

Histopathology is the mainstay of diagnosis. As we mentioned earlier, cornoid lamella with the involvement of acrosyringia is pathognomonic for PEOODN. It is usually associated with the dilation of eccrine duct. Differential diagnoses include porokeratosis plantaris discreta, inflammatory linear verrucous epidermal nevus, nevus comedonicus, linear epidermal nevus, linear psoriasis, spiny keratoderma, linear porokeratosis, congenital unilateral punctate porokeratosis and porokeratosis of Mibelli^{3,15}.

Although the condition is benign, most treatment modalities fail to show beneficial results. Steroids,

5-FU, retinoids (both topical and oral), cryosurgery, photo and laser therapy and keratolytics all have been used but have shown limited efficacy^{3,14,20}.

In conclusion, we presented a case of PEODDN and briefly reviewed the literature on the topic. Because of the rarity of this condition, each diagnosed case of PEODDN should be reported to enhance our knowledge regarding this condition.

REFERENCES

1. Abell E, Read SI. Porokeratotic eccrine ostial and dermal duct naevus. *Br J Dermatol* 1980; 103: 435-41.
2. Marsden RA, Fleming K, Dawber RP. Comedonaevus of the palm--a sweat duct naevus? *Br J Dermatol* 1979;101:717-22.
3. Wang NS, Meola T, Orlow SJ, Kamino H. Porokeratotic eccrine ostial and dermal duct nevus: a report of 2 cases and review of the literature. *Am J Dermatopathol* 2009;31:582-6.
4. Masferrer E, Vicente MA, Bassas-Vila J, Rovira C, González-Enseñat MA. Porokeratotic eccrine ostial and dermal duct naevus: report of 10 cases. *J Eur Acad Dermatol Venereol* 2010;24:847-51.
5. Sassmannshausen J, Bogomilsky J, Chaffins M. Porokeratotic eccrine ostial and dermal duct nevus: a case report and review of the literature. *J Am Acad Dermatol* 2000;43 (2 Pt 2):364-7.
6. Warren RB, Verbov JL, Kokai GK. Porokeratotic eccrine ostial and dermal duct nevus. *Pediatr Dermatol* 2006;23:465-6.
7. Birol A, Erkek E, Bozdoethan O, Koçak M, Atasoy P. A case of porokeratotic eccrine ostial and dermal duct naevus of late onset. *J Eur Acad Dermatol Venereol* 2004;18: 619-21.
8. Aloï FG, Pippione M. Porokeratotic eccrine ostial and dermal duct nevus. *Arch Dermatol* 1986;122:892-5.
9. Jamora MJ, Celis MA. Generalized porokeratotic eccrine ostial and dermal duct nevus associated with deafness. *J Am Acad Dermatol* 2008;59 (2 Suppl 1):S43-5.
10. Murata Y, Nogita T, Kawashima M, Hidano A. Unilateral, systematized, porokeratotic eccrine ostial and dermal duct nevi. *J Am Acad Dermatol* 1991;24 (2 Pt 1):300-1.
11. Bhattacharya SN, Singal A, Bhattacharya G, Singh N. Porokeratotic eccrine ostial and dermal duct nevus as nevus unius lateris in an Indian girl. *J Dermatol* 1998;25:682-4.
12. Beer K, Medenica M. Solitary truncal porokeratotic eccrine ostial and dermal duct nevus in a sixty-year-old man. *Int J Dermatol* 1996;35:124-5.
13. Cambiaghi S, Gianotti R, Caputo R. Widespread porokeratotic eccrine ostial and dermal duct nevus along Blaschko lines. *Pediatr Dermatol* 2007;24:162-7.
14. Del Pozo J, Martínez W, Vereá MM, Yebra-Pimentel MT, García-Silva J, Fonseca E. Porokeratotic eccrine ostial and dermal duct naevus: treatment with carbon dioxide laser. *Br J Dermatol* 1999;141:1144-5.
15. Rasi A, Tajziechi L. Late-onset porokeratotic eccrine ostial and dermal duct nevus associated with sensory polyneuropathy and hyperthyroidism. *Arch Iran Med* 2008;11:218-20.
16. Kroumpouzou G, Stefanato CM, Wilkel CS, Bogaars H, Bhawan J. Systematized porokeratotic eccrine and hair follicle naevus: report of a case and review of the literature. *Br J Dermatol* 1999;141:1092-6.
17. Coras B, Vogt T, Roesch A, Landthaler M, Hohenleutner U. Bowen's disease on porokeratotic eccrine ostial and dermal duct nevus. *Dermatol Surg* 2007;33:496-9.
18. Bergman R, Lichtig C, Cohen A, Friedman-Birnbaum R. Porokeratotic eccrine ostial and dermal duct nevus. An abnormally keratinizing epidermal invagination or a dilated, porokeratotically plugged acrosyringium and dermal duct? *Am J Dermatopathol* 1992;14:319-22.
19. Stoof TJ, Starink TM, Nieboer C. Porokeratotic eccrine ostial and dermal duct nevus. Report of a case of adult onset. *J Am Acad Dermatol* 1989;20 (5 Pt 2):924-7.
20. Leung CS, Tang WY, Lam WY, Fung WK, Lo KK. Porokeratotic eccrine ostial and dermal duct naevus with dermatomal trunk involvement: literature review and report on the efficacy of laser treatment. *Br J Dermatol* 1998;138:684-8.