CASE REPORT

Late-onset solitary porokeratotic eccrine ostial and dermal duct nevus

Mitchell Arbogast, MD, a Daniel Bax, BS, b Gyorgy Paragh, MD, b and Paul N. Bogner, MD b,c
Durham, North Carolina, and Buffalo, New York

Key words: acrosyringial ducts; cornoid lamellae; cutaneous horn; porokeratotic adnexal ostial nevus; porokeratotic eccrine ostial and dermal duct nevus; porokeratotic eccrine and hair follicle nevus; porokeratotic lesion.

INTRODUCTION

Porokeratotic eccrine ostial and dermal duct nevus (PEODDN) is a rare porokeratotic dermatosis, predominantly found along the lines of Blaschko on the distal extremities and thorax. 1 PEODDN is morphologically diverse, most commonly consisting of multiple papules to expansive plaques, and classically presenting at birth or in early life. 2-4 Here we present a case of PEODDN that is atypical in clinical morphology, presentation, and anatomic location.

REPORT OF A CASE

An African-American man in his mid-30s presented with the complaint of a “sore” on his right buttock. This slowly enlarging, nonpruritic lesion had been present for more than 1 year and had been painful upon sitting. He admitted to frequently picking at the area. He denied previous history of similar lesions and prior skin changes in the area. The patient denied significant sun exposure and any family history of dermatologic malignancy. Medical history was significant for poorly controlled type II diabetes mellitus and heavy tobacco smoking.

Physical examination of the right buttock found a hyperpigmented relatively well-demarcated 2-cm plaque with multiple 3- to 4-mm-thick yellow-brown hornlike projections (Fig 1). No other significant lesions were noted in the area. At this time, a specific clinical diagnosis was unclear, and the leading differential diagnoses included a large verruca and, less likely, a morphologically unusual squamous cell carcinoma. Because of the diagnostic uncertainty and the symptomatic nature of the lesion, the patient was offered biopsy.

Partial shave biopsy sections of the right buttock lesion found irregular epidermal acanthosis with papillomatosis and overlying hyperkeratotic stratum corneum. Streaky fibrosis of the superficial dermal collagen, vascular ectasia, and perivascular mononuclear cell inflammation were seen. The sections lacked evidence of viral cytopathic effect, and findings at the time were suggestive of lichen simplex chronicus with cutaneous horn. Given the marked hyperkeratosis, the absence of considerable pruritus, and the lack of improvement even after reportedly stopping manual irritation of the area, urea 40% cream as spot treatment with occlusion was recommended.

At the patient’s return visit 2 months later, he reported very limited improvement. The lesion continued to cause the patient considerable discomfort, and shave removal was offered and subsequently performed. Histologic evaluation of the complete lesion found an unusual verrucous process with partially endophytic growth. There was, again, no histologic evidence of a viral etiology, but the dermis contained patchy fibrosis and inflammation. Most strikingly, numerous cornoid lamellae were present (Fig 2, see also supplemental eSlide: VM04582), many associated with underlying

Abbreviation used:
PEODDN: porokeratotic eccrine ostial and dermal duct nevus

JAAD Case Reports 2018;4:434-6.
2352-5126
© 2018 by the American Academy of Dermatology, Inc. Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).
https://doi.org/10.1016/j.jdcr.2017.12.008
acrosyringial ducts (Fig 3). This characteristic pathologic combination supported the diagnosis of PEODDN, despite the odd clinical location and gross morphology. Upon follow-up, 3 and 12 months after the shave removal, the site was well healed and asymptomatic, with no evidence of recurrence and no other concerning skin lesion in the area.

**DISCUSSION**

PEODDN falls within the category of porokeratotic dermatoses and is further characterized as a rare benign hamartoma of eccrine ducts, with few reported cases worldwide. Lesions begin as papules with raised annular edges and central columns of keratosis, with the possibility of lateral expansion to form plaques.1 There are several potential clinical morphologies, ranging from typical constellations of small papules to unusual large solitary lesions.2,3 Disseminated variations of expansive plaques following the lines of Blaschko have also been described.5 Lesions range from pink to hyperpigmented with varying levels of keratosis.2,3 Anatomically, lesions typically cluster unilaterally or bilaterally on distal extremities or on the thorax.5 It presents predominantly at birth or early in life.1 On presentation, patients can be asymptomatic, but pruritus, pain, and hyperhidrosis or anhidrosis have been reported.1,3,6,7

**Fig 1.** Two-centimeter right buttock lesion on initial presentation with 3- to 4-mm thick yellow-brown hornlike projections.

**Fig 2.** Classic cornoid lamella show a stacked tier of parakeratosis overlying a gap in the stratum granulosum with underlying keratinocyte dyskeratosis. (Hematoxylin-eosin stain; original magnification: ×100.)

**Fig 3.** Cornoid lamella with stacked parakeratosis arising at the opening of an eccrine duct. Duct lumen is seen in the dermis directly below. (Hematoxylin-eosin stain; original magnification: ×100.)
Given its features and the usual early presentation, many investigators consider PEODDN as a special type of epidermal nevus. However, some cases of adult-onset PEODDN have also been reported.\textsuperscript{3,8} Autosomal dominant mutations in \textit{GJB2}, the gene encoding the gap junction protein connexin 26, have been associated with PEODDN. Levinsohn et al\textsuperscript{9} found that a single somatic mutation in that allele can be responsible for PEODDN. There are significant familial implications to the diagnosis, as children of those with PEODDN have increased risk of development of associated keratitis-ichthyosis-deafness syndrome.\textsuperscript{9}

As a group, porokeratotic lesions are defined histologically by the presence of coronoid lamellae: columns of parakeratosis arising from thinned epidermis associated with regional dyskeratosis and loss of the stratum granulosum. The coronoid lamellae of PEODDN are associated generally with underlying eccrine ducts, which is the classic histologic property of this lesion.\textsuperscript{1}

In this patient’s case, and in others reported in the literature, cornoid lamellae can also be seen arising in the ostia of hair follicles. Given this variability in location, alternative and more inclusive nomenclature for this group of lesions has been proposed.\textsuperscript{10} The diagnosis of both “porokeratotic adnexal ostial nevus” and “porokeratotic eccrine and hair follicle nevus” would be appropriate.

Conservative treatment options include topical agents such as keratolytics or topical retinoids, but results have been variable. For symptomatic and isolated lesions, excision and CO\textsubscript{2} laser ablation have shown better results.\textsuperscript{3}

This case features a distinct, large, solitary lesion appearing on the buttock of an adult male. The odd coalescing keratotic spires of the lesion obscured the initial clinical diagnosis, and histopathology was central to identifying this atypical presentation of PEODDN. The temporal, anatomic, and morphologic findings in the case are in stark contrast to the typical presentation of PEODDN. Singular lesions of this entity are rarely described, but the unusual site of the lesion is perhaps its most unique feature, with only one previous description of this type in the literature.\textsuperscript{3} The case presented previously by Beer and Medenic\textsuperscript{2} was also a lesion on the right buttock of similar size and had verrucous components. However, their patient was asymptomatic, and the lesion was described additionally as pink and crateriform, as opposed to our lesion.

Our case expands the reported variety of gross presentations for PEODDN, and reinforces the importance of including PEODDN in the differential diagnosis of verrucous lesions even when solitary and localized to unusual sites like the buttock.

REFERENCES

1. Abell E, Read S. Porokeratotic eccrine ostial and dermal duct nevus. \textit{Br J Dermatol}. 1980;103:435.
2. Beer K, Medenica M. Solitary truncal porokeratotic eccrine ostial and dermal duct nevus in sixty-year-old man. \textit{Int J Dermatol}. 1996;35(2):124-125.
3. Wang N, Meola T, Orlow S, et al. Porokeratotic eccrine ostial and dermal duct nevus: a report of 2 cases and review of literature. \textit{Am J Dermatopathol}. 2009;31:582-586.
4. Warren B, Verbov J, Kokai G. Porokeratotic eccrine ostial and dermal duct nevus. \textit{Pediatr Dermatol}. 2006;23(5):465-466.
5. Cambiaghi S, Gianotti R, Caputo R. Widespread porokeratotic eccrine ostial and dermal duct nevus along Blaschko lines. \textit{Pediatr Dermatol}. 2007;24(2):162-167.
6. Naraghi M, Nikoo A, Goodarzi A. Porokeratotic eccrine ostial and dermal duct nevus. \textit{Case Rep Dermatol Med}. 2013;2013:3.
7. Birol A, Erkek E, Bodzothan O, et al. A case of porokeratotic eccrine ostial and dermal duct naevus of late onset. \textit{JEADV}. 2004;18:619-621.
8. Stooft T, Starink T, Nieboer C. Porokeratotic eccrine ostial and dermal duct nevus. Report of a case of adult onset. \textit{J Am Acad Dermatol}. 1989;20(5):924-927.
9. Levinsohn JL, McIliff JM, Antaya RJ, et al. A somatic p.G45E GJB2 mutation causing porokeratotic eccrine ostial and dermal duct nevus. \textit{JAMA Dermatol}. 2015;151(6):638-641.
10. Kazakov D, McKee P, Michal M, et al. Porokeratotic Adnexal Ostial Nevus. \textit{Cutaneous Adnexal Tumors}. 1st ed. Philadelphia, PA: Wolters Kluwer Health; 2012:164-166.