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CASE REPORT

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A case of porokeratotic adnexal ostial nevus misdiagnosed as wart

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Abstract

Porokeratotic adnexal ostial nevus may be misdiagnosed as wart based on clinical appearance and morphology. An accurate diagnosis through skin biopsy is crucial, as is the inclusion of rare disorders such as porokeratotic adnexal ostial nevus in the differential diagnosis.

KEYWORDS

eccrine hamartoma, porokeratotic adnexal ostial nevus, porokeratotic eccrine and hair follicle nevus, porokeratotic eccrine ostial and dermal duct nevus

1 | INTRODUCTION

Because of its low prevalence and the relatively high prevalence of wart, porokeratotic adnexal ostial nevus (PAON) may be misdiagnosed as wart. The clinical courses and respective treatment are significantly different; skin biopsy is crucial, as is the inclusion of rare disorders such as PAON in the differential diagnosis.

PAON is a rare benign nevoid disorder that presents as palmoplantar comedo-like lesions with small pits filled by keratotic plugs or keratotic papules and plaques with linear arrangement.[1] A definitive diagnosis is made from the histopathology examination, which shows a cornoid lamella with a dilated intraepidermal eccrine duct.[2] Based on the similar histopathological characteristics of porokeratotic eccrine ostial and dermal duct nevus (PEODDN) and porokeratotic eccrine and hair follicle nevus (PEHFN), the term porokeratotic adnexal ostial nevus (PAON) has been proposed. PAON presents at birth or early in childhood, usually involving the distal extremities. Occurrence in the proximal extremities, trunk and face or widespread involvement have been reported.[3] We report a case of PAON with late onset that clinically misdiagnosed as wart due to the rarity of the condition.

2 | CASE PRESENTATION

A 15-year-old otherwise-healthy Korean boy presented with a 6-year history of multiple papules on his fingers. As the skin lesions had been recalcitrant to laser treatments and cryotherapies, he was referred to our clinic. The physical examination revealed punctate pits and hyperkeratotic papules in a linear arrangement. The keratotic papules had comedo-like plugs that had coalesced to form linear verrucous plaques on the first and second fingers of his right hand (Figure 1A, B). Based on a clinical diagnosis of palmoplantar wart, he was prescribed periodic bleomycin intralesional injections and cryotherapy. Despite these repeated procedures, the skin lesion did not respond and a punch biopsy was therefore taken from one of the papules.

The histologic examination revealed epidermal invagination with a parakeratotic column, or a cornoid lamella, centered on an acrosyringial duct (Figure 1C). The granular layer

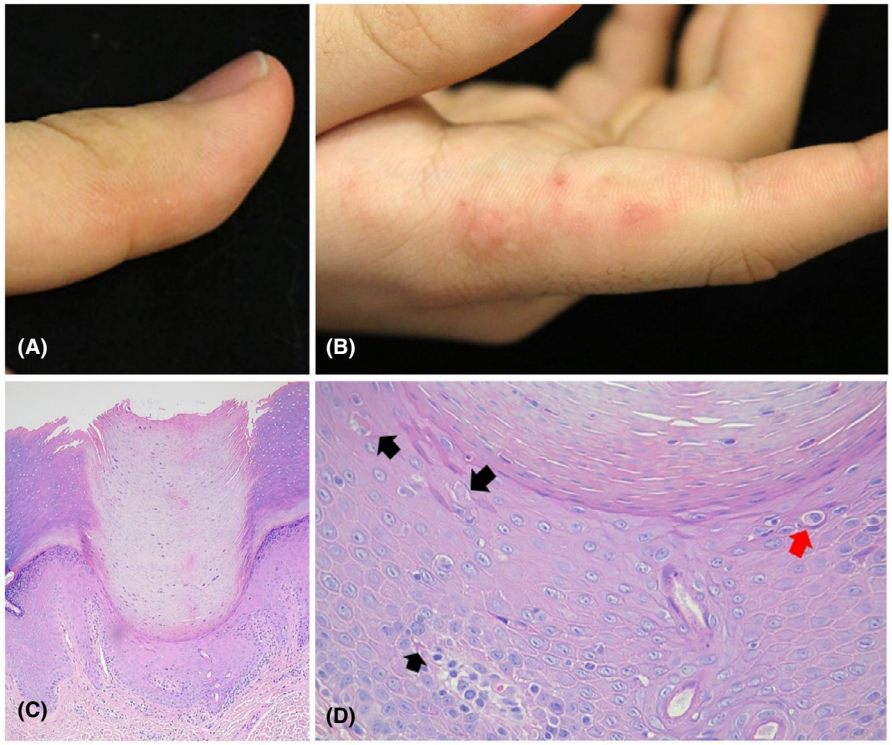

FIGURE 1 A,B) Hyperkeratotic papules with comedo-like plugs and punctate pits that coalesced into linear verrucous plaques on the first and second fingers of the patient's right hand. C) Histology revealed a cornoid lamella overlying the dilated acrosyringia (H&E, ×100). D) At the base of the plug, the underlying granular layer is absent and a few dyskeratotic cells (black arrow) and vacuolization (red arrow) are observed (H&E, ×400)

was absent at the base of the epidermal invagination, and dyskeratotic keratinocytes and vacuolization were observed. Intraepidermal eccrine ducts below the cornoid lamella were slightly tortuous and dilated but had no morphologic abnormalities (Figure 1D). As these findings were consistent with PAON, the patient was prescribed topical retinoid and emollients. In addition, the patient and his family were educated about the refractory nature of the disease.

3 | DISCUSSION

PAON typically presents as multiple linear hyperkeratotic papules or punctate pits on extremities, most often at birth or in early childhood.[4,5] Upon review of the literature, at least 81 cases of PEODDN have been reported but many of these cases had their onset at birth, unlike our case.[3] An association with deafness, hemiparesis, hyperthyroidism, polyneuropathy, and seizures has been reported, but the relationship remains uncertain and most patients with PAON are otherwise healthy.[6] The term PAON was recently proposed to encompass both PEHFN and PEODDN. The latter presents as a cornoid lamella with a dilated eccrine acrosyringium underneath, while both the eccrine acrosyringia and the hair follicles are involved in PEHFN. Both PEHFN and PEODDN may be variants of the same process in which the affected adnexal structure and level of involvement differ.[3]

The exact pathogenesis of PAON is unclear; given the clinical and histopathological resemblance of PAON and keratitis-ichthyosis-deafness (KID) syndrome, a somatic mosaic mutation in GJB2, coding for connexin 26, a gap junction protein, has been proposed as the cause of PAON.[7] The increased intracellular calcium concentration induced by this mutation is thought to lead to epidermal hyperproliferation and the eventual formation of the cornoid lamella.[8]

A definite diagnosis is made according to the histopathological examination, but due to the rarity of PAON a biopsy is often delayed. The differential diagnosis includes palmoplantar wart, nevus comedonicus, punctate porokeratotic keratoderma, porokeratosis plantaris discreta, and punctate porokeratosis.[4] Since PAON tends to form a linear arrangement, it should also be differentiated from dermatoses with this same pattern, such as linear epidermal nevus, linear psoriasis, and linear porokeratosis.[4]

Palmoplantar wart, nevus comedonicus, linear epidermal nevus, and linear psoriasis can be ruled out by histopathological examination since they lack a cornoid lamella. Punctate porokeratotic keratoderma, porokeratoses plantaris discrete, punctate porokeratosis and linear porokeratosis are dermatoses that may also include cornoid lamellation. Cornoid lamella may also occur incidentally, such as in inflammatory, hyperplastic, and neoplastic conditions of the skin.[9] However, only in PAON is the cornoid lamella centered over dilated acrosyringia, with a scant lymphocytic infiltration of the superficial dermis.

There are various treatment modalities, including ablative laser therapy, topical steroid, retinoid, emollient, calcipotriol, 5-fluorouracil, and cryotherapy, but their success is often only partial.[10] CO2 laser and topical retinoids with emollients have consistently yielded clinical improvement

to some extent,[4,5] but conservative treatment is the preferred approach. The course of PAON is usually indolent; progressive improvement through a period up to 16 years has been reported.[11] A case of Bowen disease arising in a patient with PEODDN suggests the need for long-term follow-up. [12]

4 | CONCLUSIONS/FUTURE DIRECTIONS

PAON is a very rare disorder of keratinization with eccrine involvement. Because of its low prevalence and the relatively high prevalence of palmoplantar wart, palmoplantar wart is typically diagnosed in a patient with hyperkeratotic papules that form verrucous plaques on the extremities. As the clinical course and treatment of PAON and wart can differ significantly, an accurate diagnosis is crucial. This case report demonstrates the importance of skin biopsy and the need to consider rare disorders such as PAON in the differential diagnosis of refractory warty lesions.

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CONFLICT OF INTEREST

The authors have no conflict of interest to declare.

AUTHOR CONTRIBUTIONS

BRK: first author, responsible for the introduction, case presentation, discussion, and conclusion. RK: responsible for the introduction, discussion, conclusion, and paper submission. JYB: responsible for review and revision. YWC: responsible for review and revision. HYC: responsible for the clinical message, differential diagnosis, discussion, and conclusion. MYL: Principal investigator, responsible for discussion and review.

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REFERENCES

1. Requena L, Sangueza O. *Cutaneous Adnexal Neoplasms*. Cham: Springer; 2017:41-45.
2. 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;31:582-586.
3. Goddard DS, Rogers M, Frieden IJ, et al. Widespread porokeratotic adnexal ostial nevus: Clinical features and proposal of a new name unifying porokeratotic eccrine ostial and dermal duct nevus and porokeratotic eccrine and hair follicle nevus. *J Am Acad Dermatol*. 2009;61:1060.e1-14.
4. Petkiewicz S, Baltz J, Cornejo K, Deng A, Wiss K. Widespread presentation and spontaneous regression of porokeratotic eccrine ostial and dermal duct nevus. *JAAD Case Rep*. 2018;4:972-975.
5. Arbogast M, Bax D, Paragh G, Bogner PN. Late-onset solitary porokeratotic eccrine ostial and dermal duct nevus. *JAAD Case Rep*. 2018;4:434-436.
6. Llamas-Velasco M, Hilty N, Kempf W. Porokeratotic adnexal ostial anevus: review on the entity and therapeutic approach. *J. Eur. Acad. Dermatol. Venereol*. 2015;29:2032-2037.
7. Easton JA, Donnelly S, Kamps MA, et al. Porokeratotic eccrine nevus may be caused by somatic connexin26 mutations. *J. Invest. Dermatol*. 2012;132:2184-2191.
8. Levinsohn JL, McNiff JM, Antaya RJ, Choate KA. A somatic p. G45E GJB2 mutation causing porokeratotic eccrine ostial and dermal duct nevus. *JAMA Dermatol*. 2015;151:638-641.
9. Patterson J. *Weedon's skin pathology*. 4 th ednLondon: Churchill Livingstone-Elsevier. 2016;299.
10. Kim WJ, Choi SR, Lee HJ, Kim DH, Yoon MS. Porokeratotic eccrine ostial and dermal duct nevus showing partial remission by topical photodynamic therapy. *Ann Dermatol*. 2011;23:s322-325.
11. Llamas-Velasco M, Hilty N, Kempf W. Porokeratotic adnexal ostial naevus: review on the entity and therapeutic approach. *J Eur Acad Dermatol Venereol*. 2015;29:2032-2037.
12. Coras B, Vogt T, Roesch A, Landthaler M, Hohenleutner U. Bowen's disease on porokeratotic eccrine ostial and dermal duct nevus. *Dermatol Surg*. 2007;33:496-499.

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