

## Porokeratotic eccrine ostial and dermal duct nevus: A report of three cases

Sir,

Porokeratotic eccrine ostial and dermal duct nevus is a rare non-hereditary eccrine hamartoma. This disorder of keratinization usually occurs at birth or in early childhood but may sometimes present in adults. It generally involves acral areas, especially palms and soles and is characterized by multiple punctate pits or keratotic papules and plaques with comedo-like plugs in a linear distribution.<sup>[1]</sup> The histopathological hallmark of porokeratotic eccrine ostial and dermal duct nevus is the presence of a cornoid lamella with subjacent acrosyringium.

We report three men aged 40, 20 and 16 years with porokeratotic eccrine ostial and dermal duct nevus. Details of the cases are summarized in Table 1. All three patients had involvement of the lateral border of left hand. Lesions on the palmar aspect appeared to be pits or plugged comedos whereas lesions on the lateral aspect were linear and keratotic and also violaceous in one patient [Figure 1]. In all three patients, the rest of the mucocutaneous and systemic examination did not reveal any abnormality.

Histopathological examination in all patients revealed epidermal acanthosis and a deep epidermal invagination of a parakeratotic column with underlying absence of granular layer (cornoid lamella) and dilated acrosyringium beneath the cornoid lamella [Figure 2]. Thus, a final diagnosis of porokeratotic eccrine ostial and dermal duct nevus was made in all three patients based on clinical and histopathological examination. Patients were treated with topical keratolytics and

emollients and were counseled regarding the nature of the disease.

This rare entity was first reported as comedo nevus of palms by Marsden *et al.* and was later rechristened porokeratotic eccrine ostial and dermal duct nevus by Abell and Read.<sup>[1]</sup> Recently, the term porokeratotic adnexal ostial nevus has been proposed as a superset encompassing both porokeratotic eccrine ostial and dermal duct nevus and a related condition “porokeratotic eccrine and hair follicle nevus” where the cornoid lamella overlies both eccrine acrosyringia and hair follicular infundibula.<sup>[2]</sup> Clinically, porokeratotic eccrine ostial and dermal duct nevus has punctate pits with comedo-like plugs typically localized to palms and soles. However, lesions at other sites as well as Blaschkoid and systemized patterns have been reported.<sup>[1,3]</sup> Lesions may become verrucous, especially on hair-bearing skin beyond the palms and soles.<sup>[3]</sup> The histopathological hallmark of the condition is the cornoid lamella centered on dilated acrosyringium.<sup>[2]</sup>

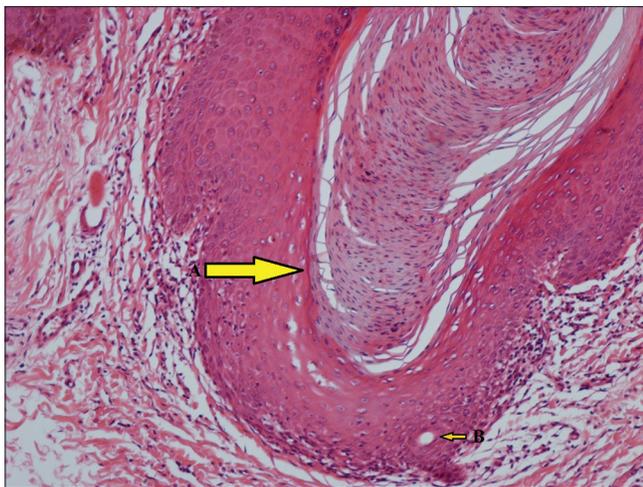
Various pathogenetic mechanisms have been proposed for porokeratotic eccrine ostial and dermal duct nevus such as, (a) abnormally dilated parakeratotically plugged acrosyringium, (b) lack of carcinoembryonic antigen expression, (c) keratinization defect, (d) increased proliferation of basal keratinocytes, (e) genetic mosaicism and (f) somatic GJB2 gene mutation (which provides instructions for making a protein called gap junction beta 2, more commonly known as connexin 26).<sup>[1,3-5]</sup> A person with porokeratotic eccrine ostial and dermal duct nevus having a somatic mutation in GJB2 may have a child with syndromes such as keratitis-ichthyosis-deafness syndrome, Vohwinkel syndrome, Bart-Pumphrey syndrome and non-epidermolytic palmoplantar keratoderma with deafness that are associated with GJB2 mutations and they should be counseled about this risk.<sup>[5]</sup>

**Table 1: Details of the three cases of porokeratotic eccrine ostial and dermal duct nevus being reported**

| Gender | Age of onset (years) | Age at presentation (years) | Distribution of lesions                                                                                  | Morphology of lesions                                                                                                                                                                                              | Figure          |
|--------|----------------------|-----------------------------|----------------------------------------------------------------------------------------------------------|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|-----------------|
| Male   | 30                   | 40                          | Thenar eminence of left palm, ulnar aspect of left thumb extending up to wrist joint in a linear pattern | Multiple, well-defined, discrete, punctate pits and keratotic papules with comedo-like plugs on the thenar eminence. A few keratotic papules and plaques having verrucous appearance on left thumb and wrist joint | Figure 1a and b |
| Male   | 12                   | 20                          | Ulnar aspect of left thumb extending to wrist joint in a linear pattern                                  | Multiple, discrete, comedo-like plugged punctate pits coalescing to form violaceous plaque. A few lesions had verrucous appearance                                                                                 | Figure 1c       |
| Male   | 11                   | 16                          | Thenar eminence of left palm                                                                             | Multiple, discrete, comedo-like plugged pits                                                                                                                                                                       | Figure 1d       |



**Figure 1:** (a) Multiple, punctate pits and keratotic papules with comedo-like plugs on thenar aspect of left palm. (b) A few keratotic papules on ulnar border of thumb extending proximally to the wrist joint. (c) Multiple, comedo-like plugged punctate pits overlying on violaceous papules and plaques on ulnar aspect of left thumb extending proximally to the wrist joint. (d) Multiple, comedo-like plugged pits present on thenar aspect of left palm



**Figure 2:** Epidermal acanthosis and a deep epidermal invagination of a parakeratotic column with underlying absent granular layer (cornoid lamella, arrow A) centered on dilated acrosyringium (arrow B) (H and E,  $\times 400$ )

Most commonly, porokeratotic eccrine ostial and dermal duct nevus is a localised lesion. However, systematized or bilateral porokeratotic eccrine

ostial and dermal duct nevus may be associated with breast hypoplasia, palmoplantar keratoderma, psoriasis, hemiparesis, seizure disorder, scoliosis, polyneuropathy, hyperthyroidism, developmental delay, onychodystrophy or squamous cell carcinoma.<sup>[1,2]</sup> Differential diagnoses of porokeratotic eccrine ostial and dermal duct nevus include linear porokeratosis, nevus comedonicus, inflammatory linear verrucous epidermal nevus, verrucous epidermal nevus, linear psoriasis and punctate keratoderma.<sup>[1,2]</sup> Porokeratotic eccrine ostial and dermal duct nevus and linear porokeratosis are difficult to distinguish both clinically and histopathologically, especially on non-acral sites. Clinically, non-acral porokeratotic eccrine ostial and dermal duct nevus lack typical keratotic comedo-like lesions, are less scaly and have an appearance similar to conventional porokeratosis. Histopathological features of linear porokeratosis and porokeratotic eccrine ostial and dermal duct nevus may overlap and a diagnosis of linear porokeratosis may be made if adnexa are not captured in the particular section

showing a cornoid lamella. Conversely, a misdiagnosis of porokeratotic eccrine ostial and dermal duct nevus may be made if the cornoid lamella in a conventional case of porokeratosis is associated with underlying adnexa. Thus, multiple biopsies and deeper sections should be examined and correlated with with clinical features to distinguish porokeratotic eccrine ostial and dermal duct nevus and linear porokeratosis.<sup>[2]</sup> The other differential diagnoses lack cornoid lamella on histopathology and are easier to exclude.

Various therapies such as topical keratolytics, corticosteroids, calcipotriol, anthralin, tar, urea phototherapy, cryotherapy, electrocautery, surgical excision and ultrapulsed CO<sub>2</sub> laser have been proposed for porokeratotic eccrine ostial and dermal duct nevus but with limited benefit.<sup>[1]</sup>

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#### Conflicts of interest

There are no conflicts of interest.

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