Porokeratotic Eccrine Ostial and Dermal Duct Nevus and Porokeratotic Eccrine and Hair Follicle Nevus: Is Nomenclature "Porokeratotic Adnexal Ostial Nevus" More Appropriate?

Sir,

Porokeratotic eccrine ostial and dermal duct nevus (PEODDN) is a rare adnexal hamartoma of which only fewer than 70 cases have been reported.[1] Additionally, cases other than PEODDN showing cornoid lamella overlying both eccrine acrosyringia and hair follicles have been also described as porokeratotic eccrine and hair follicle nevus (PEHFN).

A 10-year-old boy presented with asymptomatic papular lesions over medial right palm since birth that have been extending progressively over forearm in a linear fashion [Figure 1]. He was the first child born to non consanguineous parents after an uneventful gestation and birth. He was immunized to his age and had normal developmental milestones. His other medical history and clinical examination were unremarkable. His 8-year-old brother and other family members were reportedly healthy. Cutaneous examination showed multiple discrete and coalescing at places, pitted comedo-like papules with a central pigmented keratotic plug arranged in a linear pattern over medial half of the palm, wrist, and forearm. Biopsy from a palm lesion showed focal hyperkeratosis and acanthosis, a column of parakeratosis overlying a thin or near absent granular layer, and a cornoid lamella overlying eccrine acrosyringia, suggestive of porokeratotic eccrine ostial and dermal duct nevus [Figure 2]. He did not consult further.

This 20-year-old girl had multiple asymptomatic papular lesions over right foot arranged in a linear pattern. These had been there since early childhood as far as she could remember. They have been progressively extending from toes to ankle [Figure 3]. She was the first child of non consanguineous parentage born after an uneventful pregnancy. Her other medical history and systemic examination were unremarkable. Other family members were reportedly healthy. Cutaneous examination showed multiple discrete skin-colored papules arranged in a linear pattern and coalescing at places to form plaque over dorsolateral aspect of right foot extending from little toe to lateral malleolus. Biopsy from a lesion showed features of focal hyperkeratosis and acanthosis; granular layer and insignificant parakeratosis overlying pilosebaceous ostia; and dilated, prominent, and tortuous eccrine acrosyringia with normal secretory portion suggestive of PEHFN [Figure 4]. She was prescribed topical salicylic 6% ointment for twice daily application. She did not follow up further.

Clinically, PEODDN presents either as comedo-like papules with central pits and keratotic plugs within or keratotic papules and plaques resembling linear verrucous epidermal nevus involving palmoplantar skin as in our case-1, and/or extremities (mostly unilateral) as in case-2. The lesions over trunk are uncommon. Concurrent seizures, left hemiparesis and scoliosis, deafness and delayed development, hyperthyroidism and polyneuropathy, or breast hypoplasia have been reported rarely.[2-4] Squamous cell carcinoma and Bowen disease arising within PEODDN too have been documented.[2,5]

It is usually congenital with no defined inheritance pattern but may have onset at early childhood or occasionally late in life without any gender predilection.[3] The pathogenesis of PEODDN has been recently attributed to mutations in gene GJB2.21 Cx26 encoding connexin 26 (Cx26), a gap junction protein in cochlea, cornea, and skin that is also associated with palmoplantar keratoderma, keratitis-ichthyosis-deafness (KID) syndrome, hystrix-like

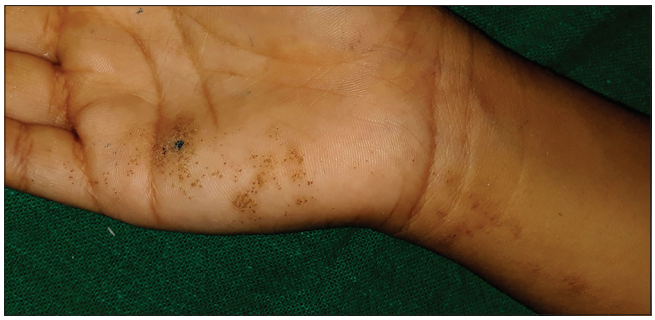

Figure 1: Multiple pitted comedo-like papules with a central pigmented keratotic plug arranged in a linear pattern over palm, wrist, and forearm

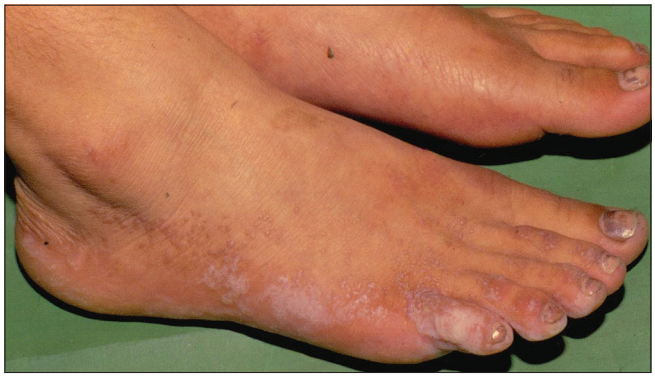

Figure 3: Multiple discrete skin-colored papules arranged in a linear pattern, coalescing at places to form plaque extending from toes to lateral malleolus of right foot

ichthyosis with deafness syndrome, Vohwinkel syndrome, and Bart-Pumphrey syndrome.[6-8] Since KID syndrome is related to such an extent that PEODDN has been even considered its mosaic form by some workers.[7,8]

Linear and punctate porokeratosis of the palms and soles remains the closest differential but cornoid lamella overlying eccrine acrosyringia or hair follicle ostia, the characteristic histological features of PEODDN, will be differentiating.[1,9] Nevus comedonicus, linear epidermal nevus or inflammatory linear epidermal nevus, warts, and porokeratoma are other infrequent differentials. On the other hand, accurate differentiation between PEODDN and PEHFN may sometimes be difficult. Some workers have even questioned existence of both entities being distinct from each other and proposed alternative nomenclature "porokeratotic adnexal ostial nevus (PAON)".[2] In view of the observations made in our two cases, both PEODDN and PEHFN appear to represent a single entity with varied presentations of a disorder involving different levels of adnexal structures, eccrine acrosyringia in palmoplantar lesions or hair follicle ostia, and eccrine acrosyringia in lesions of hair bearing skin. The nomenclature PAON, thus, seems appropriate especially when the two entities are indistinguishable clinicopathologically.

Progressive improvement over a period up to 26 years may occur but it tends to persist.[10,11] Although we could not ascertain therapeutic outcome in our patients,

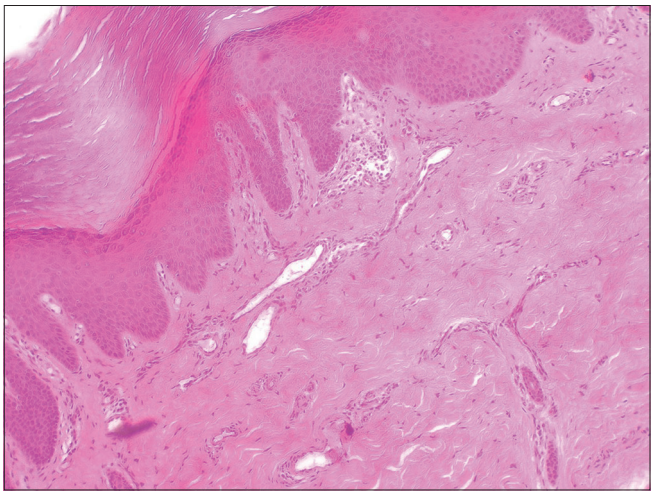

Figure 2: Characteristic histology of palmar porokeratotic eccrine ostial and dermal duct nevus (PEODDN) featuring focal hyperkeratosis and acanthosis, a column of parakeratosis overlying a thin or near absent granular layer, and a cornoid lamella over eccrine acrosyringia (H and E, ×40)

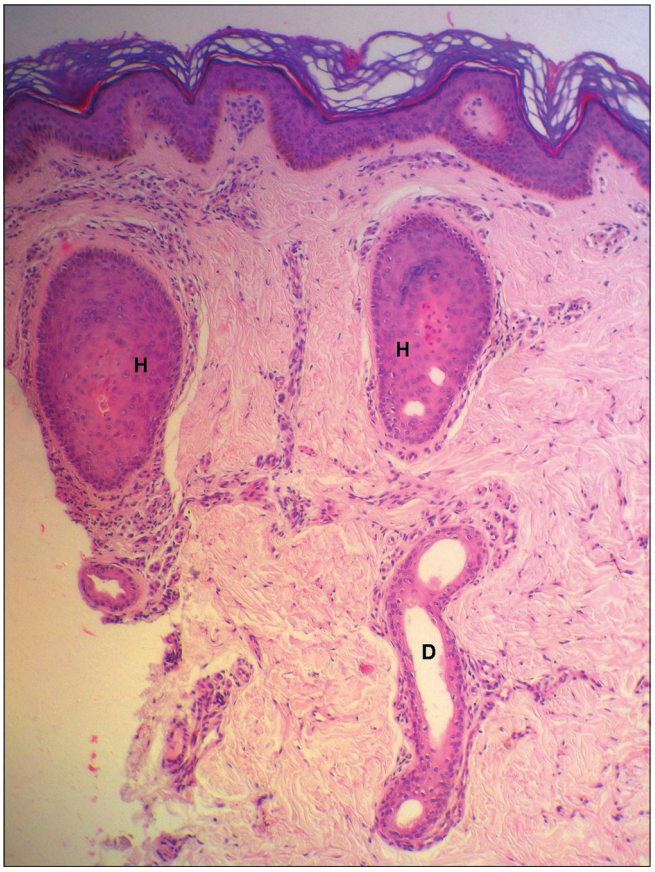

Figure 4: Characteristic histology of porokeratotic eccrine and hair follicle nevus (PEHFN) of hair bearing skin featuring focal hyperkeratosis and acanthosis, and a parakeatotic column overlying a thin granular layer over eccrine duct (D) and hair follicle (H) ostia (H and E, ×40)

most cases have been treated for cosmetic reasons with topical calcipotriol, corticosteroids, dithranol, coal tar, 5-fluorouracil, or keratolytic ointments, tazarotene gel, or systemically with acitretin, psoralen plus ultraviolet-A, or tetracyclines with variable success. Small and localized

lesions are amenable to surgical excision, ablation with cryotherapy, electrodessication, or electrocautery, while carbon dioxide laser alone or in combination with erbium laser and photodynamic therapy may have better outcome.[1,2,9] The plethora of therapies used for treating PEODDN itself reflects their unsatisfactory efficacy and recurrences are not uncommon.

Contributors' statement

BC obtained, compiled, analyzed all dataand prepared initial draft. AS helped in data obtaining, compiling, and literature search.VKM analyzed and interpreted data, drafted, and critically evaluated the manuscript for important intellectual content. All authors were involved in the revision of the draft manuscript and have agreed to the final content.

Statement of ethics

Informed consent was obtained from parents/patient for publication of material. All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975 as revised in 2013.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

Bhumika Chowdhary, Anuj Sharma, Vikram K. Mahajan

Department of Dermatology, Venereology and Leprosy, Dr. R. P. Govt. Medical College, Kangra (Tanda), Himachal Pradesh, India

Address for correspondence:
Dr. Vikram K. Mahajan, Department of Dermatology, Venereology and Leprosy, Dr. R. P. Govt. Medical College, Kangra (Tanda) - 176 001, Himachal Pradesh, India.
E-mail: vkm1@rediffmail.com

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