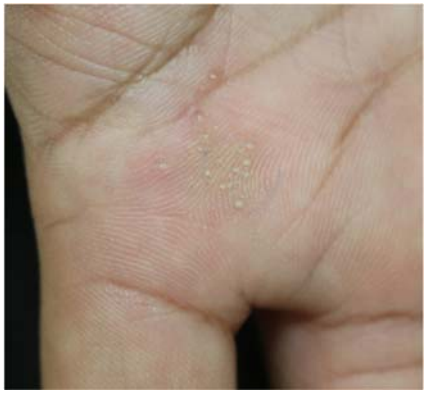
Case 11
A 28 year-old Thai woman from Chachoengsao

Chief complaint: Localized pitted papules on right palm since childhood


(Fig. 11.1)

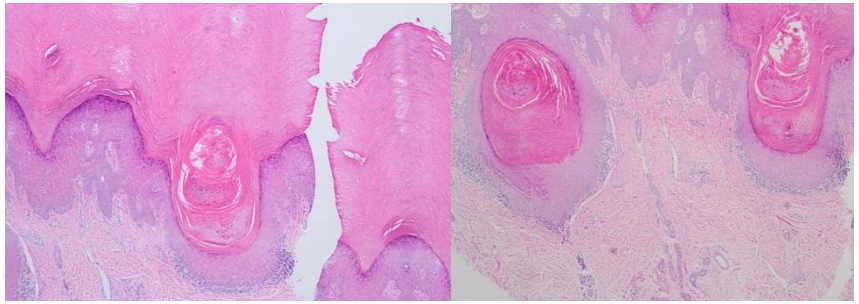
Present illness: The patient presented with non-progressive asymptomatic plaque on right palm since childhood. The lesion was spiky and rough which needed to be trimmed often. There were no mucosal or nail abnormalities. Hair and teeth were normal. She denied any previous treatment or trauma. There was no history of similar rash in any member of her family.

Past history:
- Tuberculosis lymphadenopathy, currently on anti-tuberculosis drugs
- Non-alcoholic fatty liver disease

Physical examination:
HEENT: Anicteric sclera
Lymph node: Not palpable
Lungs: Normal breath sound, no adventitious sound
Abdomen: No hepatosplenomegaly

Dermatological examination: (Fig. 11.1)
Well-circumscribed spiky keratotic papules coalescing to form plaques with pits on right palm

Histopathology (S17-011192, Right palm) (Fig. 11.2)


(Fig. 11.2)

- Marked compact hyperkeratosis, hypergranulosis, and epidermal papillomatosis
- Deep widely dilated eccrine duct containing column of parakeratosis and dyskeratotic vacuolated keratinocytes

Diagnosis: Porokeratotic eccrine ostial and dermal duct nevus

Treatment:
- Topical keratolytic agent: 20% urea cream apply on lesion

- Reassurance and education

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Discussion:

Porokeratotic eccrine ostial and dermal ductal nevus (PEODDN) is a rare, benign congenital eccrine hamartoma. It was first described by Marsden et al. in 1979 as "comedo nevus of the palm"[1] and was later renamed porokeratotic eccrine ostial and dermal duct nevus by Abell and Read in 1980.[2] Since that time approximately 50 cases have been reported.

The pathogenesis is still unknown. But various pathogenic mechanisms have been proposed, such as, (a) abnormally dilated parakeratotic 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 encode connexin 26, a gap junction protein).[2-5] Mutation in GJB2 gene suggests that PEODDN may be a mosaic form of keratitis ichthyosis deafness (KID) syndrome and are at risk of conceiving children with KID syndrome.

The condition is congenital and usually appears at birth or in early childhood, though late-onset presentations occur. [6,7] A review of the literature by Valks et al. showed that the frequency of late onset disease may be as high as 26%.[8] Gender distribution is nearly equal. Clinically, the lesions are asymptomatic or mildly pruritic. PEODDN can present as multiple linear pits or keratotic papules and plaques, typically localized on the palms and soles as linear or band-liked distribution. However, lesions at other sites as well as blaschkoid and systemized patterns have been reported.[2,3] Rarely, systematized or bilateral PEODDN may be associated with breast hypoplasia, palmoplantar keratoderma, psoriasis, hemiparesis, seizure disorder, scoliosis, polyneuropathy, hyperthyroidism, developmental delay, onychodystrophy or squamous cell carcinoma.[2]

Histopathology is diagnostic of PEODDN which shows orthokeratosis, and a column of parakeratosis occupying an invagination of the epidermis, which, at the base of the column shows loss of granular cells.[8] Dilated acrosyringia at the base of the invaginations point to an eccrine origin.

Differential diagnoses of porokeratotic eccrine ostial and dermal duct nevus include linear porokeratosis, nevus comedonicus, inflammatory linear verrucous epidermal nevus (ILVEN), verrucous epidermal nevus, linear psoriasis and punctate keratoderma.[1,2] The condition can be differentiated from ILVEN by the absence of extremely pruritic bands of eczematous or psoriasiform lesions. PEODDN and linear porokeratosis are quite similar both clinically and histopathologically, especially on non-acral sites.

The course of PEODDN is usually stationary. Long term involution of the lesions has been observed rarely.[10] The lesions are benign and no malignant transformation had been reported. The ideal therapeutic regimen for this condition remains elusive and generally unsatisfactory. Small and localized lesions may be surgically excised. There was little success with topical corticosteroids, tar, psoralen ultraviolet A, ultraviolet B, anthralin, keratolytics, and retinoids. Good results with laser therapy, in the form of ultrapulse CO_2 laser, combined erbium/CO_2 laser and combined curettage/CO_2 laser have been reported. [9,11,12]

References:

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