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Clinicopathologic challenge

Small annular lesions on the nose of a young Caucasian woman

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What is your diagnosis?

Case report

An otherwise healthy 20-year-old Caucasian woman presented with small flat papules (1–3 mm in diameter) showing a hyperkeratotic and desquamative rim and a slightly depressed center, located exclusively on the nose (Fig. 1a). They were noted 2 months before and were asymptomatic. On dermoscopy, they appeared faintly erythematous and displayed a white-yellowish keratotic annular border (Fig. 1b). At the total body examination, no other skin lesion was found, and the patient did not report a family history of similar conditions.

A biopsy was performed on the ridge of a papule, and histological examination revealed a narrow column of tightly fitted parakeratotic cells with underlying hypogranulosis (Fig. 2).

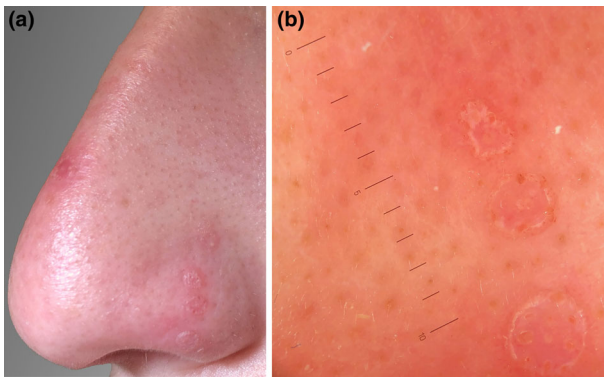

Figure 1 (a) Clinical appearance of the lesions: small flat papules located on the nose, 1–3 mm in diameter, with a hyperkeratotic-desquamative rim and a slightly depressed center. (b) Dermoscopic appearance of the lesions: faintly erythematous round areas with a white-yellowish keratotic collar

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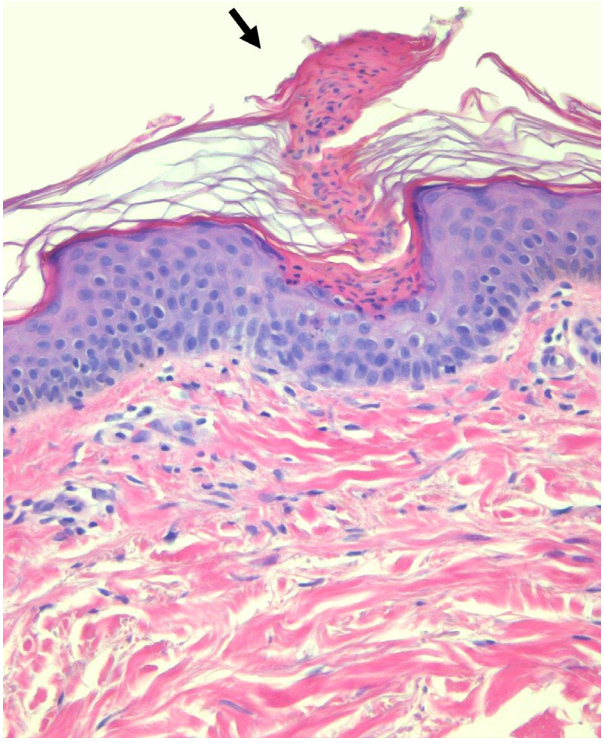

Figure 2 Skin histopathology of the biopsy, performed on the ridge of a lesion: presence of a narrow column of tightly fitted parakeratotic cells with underlying hypogranulosis (hematoxylin and eosin, ×20)

Diagnosis

Localized porokeratosis of the nose.

Discussion

Porokeratosis (PK) was first described by Mibelli in 1893 and is thought to be a disorder of keratinization, but the definitive pathogenesis remains unclear. It has been proposed that PK is caused by a proliferation of abnormal clones of epidermal cells, and multiple genetic loci have been associated with PK. It can be inherited as an autosomal dominant disorder, but many cases appear to be sporadic.[1,2]

There are many clinical variants of PK, including classic porokeratosis of Mibelli, disseminated superficial actinic porokeratosis (DSAP), linear porokeratosis, punctate porokeratosis (and its variant porokeratosis palmaris et plantaris disseminata), porokeratosis ptychotropica, and eruptive disseminated porokeratosis.[1,2]

On histopathology, the hallmark of the disorder is a well-distinct column of parakeratotic cells extending within the stratum corneum, called "cornoid lamella." Beneath the cornoid lamella, the epithelium is often thinned because of the presence of hypogranulosis or agranulosis, and vacuolar changes at the dermal–epidermal junction can also be observed. In the dermis, perivascular inflammatory infiltrate can be an additional finding.[2]

On dermoscopy, all variants of PK are characterized by a specific clue, that is the keratotic white track having two free edges (double-track sign) at the periphery of the lesion.[3,4] Clearly, this dermoscopic sign corresponds to the histological cornoid lamella.[3]

Trunk and extremities are the most frequently affected sites by PK; the occurrence on the face, including the nasal and perinasal area, represents an unusual clinical finding. PK of Mibelli has been seldom reported with a facial presentation (single or multiple lesions), predominantly in adolescent or adult subjects.[5,6] Congenital linear PK, which is thought to be a genetic mosaicism following the Blaschko lines, has also been described on the face as an uncommon location.[7] Furthermore, DSAP, which is a more common condition than Mibelli and linear PK, is rarely described with exclusive facial localization, although 15% of patients with the typical disseminated variant have facial involvement.[8]

However, beyond the more defined variants, PK turns out to be a heterogeneous disorder. For example, a number of cases have been reported in which PK on the face was the sole manifestation of the disorder (mostly in Middle-East and Indian population) and appeared to be induced and exacerbated by sun exposure.[9–11] Sharquie et al. named it Solar Facial Porokeratosis (SFP), considering this disease a new variant of PK.[9] This one was characterized by a predominance of the female sex among the affected individuals, and the age of onset was typically the second and third decades of life, with no elements of inheritance. In addition, in SFP, the risk of malignant degeneration (especially squamous cell carcinoma) seemed to be absent, unlike the known risk of the variants mentioned above, such as PK of Mibelli, linear PK, and DSAP.[9]

To date, multiple variants have been described, but the pathogenesis of PK remains not completely understood. It remains to be determined whether the presentation on the face may constitute, at least in some cases, a disease variant in its own right or whether it represents an unusual presentation of one of the several subtypes.

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