History and clinical findings

A 91-year-old woman was referred for a second opinion on an acquired and asymptomatic but suspect lesion on the palm of her right hand. Skin examination revealed a 1.0 × 0.6 cm, sharply circumscribed, symmetrical, erythematous, depressed area on the right thenar eminence (Figure 1). The patient reported that the lesion had developed suddenly almost three years ago, but could not remember any causative event or trauma to the region. Her medical history included previous excision of two basal cell carcinomas on unrelated skin sites. She had no systemic comorbidities and reported no other complaints at the time of consultation.

Histological findings

Histopathological evaluation of a 4 mm punch biopsy specimen showed a hyperplastic epithelium without atypia and abrupt thinning of the stratum corneum in the lesional area (Figure 2). The epidermis in this area showed hypogranulosis. The transition between the normal and hypokeratotic area was abrupt. There was no inflammation, atypia or corneal lamella.

Figure 1 Clinical finding on the right thenar eminence in overview (a) and close-up (b) showing a sharply circumscribed, erythematous, depressed area with a step-like configuration of the rim.

Figure 2 Histological examination in overview (a) and detail (b) showing a hyperplastic epithelium without atypia or inflammation, an abrupt thinning of the corneal layer as well as hypogranulosis in the lesional area (hematoxylin-eosin stain, original magnification x 30 (a) and x 100 (b)).
Diagnosis:
Circumscribed palmar hypokeratosis

Discussion

Circumscribed palmar or plantar hypokeratosis (CPH) is a rare epidermal differentiation defect of the skin, characterized by localized reduction of the stratum corneum and first described in 2002 by Perez et al. [1]. Its typical clinical presentation is an atrophic, well-circumscribed, erythematous, depressed macule or patch. It shows predilection for the thenar and hypothenar regions of the palm or the medial aspect of the sole [2, 3]. Multiple lesions have occasionally been described [4]. Clinical presentation and localization in our patient were both typical of CPH.

Circumscribed palmar or plantar hypokeratosis presents mostly in older adult patients and in the elderly [5]. Less than 100 cases of CPH have been published so far; only one of these was congenital (a 10-year-old African American boy) [6]. Diagnosis of CPH is based on its distinctive clinical characteristics, including the macroscopic and dermoscopic appearance as well as the palmoplantar distribution and typical histological findings.

However, the etiology of CPH is still unclear. Presumptions range from traumatic damage through infection with human papilloma virus to genetic alterations with consecutive loss of adhesion in the corneal layer or keratinization defects. Our patient could not remember any trauma as a trigger for the lesion, although it has been shown that CPH manifests at sites of initial traumatic damage, more frequently of the dominant hand [7]. It has been hypothesized that long-term cumulative or repetitive blunt trauma can alter the keratinization process [8]. Other findings support the theory of a virus-induced epidermal disorder, as increased proliferation markers and p53 protein in lesional keratinocytes could be interpreted as indirect viral involvement [7, 9]. We cannot comment on the possibility of viral involvement in our patient, as an HPV-PCR was not part of our diagnostic workup. Ultrastructural biopsy investigation supports the concept of CPH as an abnormal, localized keratinization defect, expressed morphologically in the corneal and granular layers [2, 6, 10, 11]. In one recent case, keratin 10 and keratin 16 were expressed in a mutually exclusive manner at the boundary between the affected and unaffected skin (K16+ in affected skin, K10+ in unaffected skin) [12]. However, the exact etiology and the underlying pathomechanism is still unclear, and in our patient we can only assume that some sort of cumulative trauma and possibly a genetic predisposition may have been causative.

Circumscribed palmar or plantar hypokeratosis is a rare and commonly misdiagnosed condition. If patients (especially elderly patients) present with a solitary, well-circumscribed, erythematous, depressed macule or patch on the palm or sole with abrupt thinning of the corneal layer at the margins, one should keep CPH in mind as a potential differential diagnosis.

Dermoscopy can facilitate differentiation between the two main differential diagnoses, Bowen’s disease and porokeratosis. Dermoscopic features of CPH include two main findings, which we have also observed in our patient. The first is a “geological stratification” type or step-like configuration of the rim, with varying thickness of the epidermal layers and the margin of the lesion appearing slightly thickened. The second is homogeneous erythema with dotted vessels in the central area. In contrast, Bowen’s disease does not show raised margins, and coiled or glomerular vessels are found instead of dotted vessels. However, in porokeratosis the vascular pattern is missing and the margin has a characteristic whitish rim without a step-like configuration [13]. Isolated basal cell carcinoma in atypical anatomical regions such as the palms is another differential diagnosis, but it is rare and few cases have been published so far [14].

Typical histopathological findings of CPH are a well-demarcated central area with decreased thickness of the stratum corneum and a diminished granular layer without inflammation [3]. A non-invasive alternative to skin biopsy of suspected lesions – although rarely available – is the use of optical coherence tomography, which can facilitate diagnosis in patients reluctant to undergo skin biopsy, or be used to identify preclinical skin lesions amenable to early treatment in patients with known CPH [15].

As CPH is a benign skin defect, with only one report of associated actinic keratosis [7], treatment is not mandatory if malignancy or malignant degeneration can be ruled out clinically. Available treatment options are based on individual case reports: cryotherapy, photodynamic therapy, topical calcipotriol and fluorouracil have been shown to be effective [9, 10, 16, 17].

Treatment and follow-up

Considering her benign and symptom-free condition, we complied with our patient’s wish to have no treatment. A follow-up after eight months did not show any enlargement, evolution or signs of malignant transformation.

Conflict of Interest
None.

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