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Case Report

Circumscribed palmar hypokeratosis: case report and literature review

局限性掌部角化減少症：病例報告及文獻綜述

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Circumscribed palmo-plantar hypokeratosis is a rare entity. The morphology consists of a long-standing, solitary well-circumscribed macule with a sharp and mildly scaly border. It is usually found on the palms or soles. We report a case of 73-year-old Chinese woman who presented with such a lesion, with emphasis on the clinical and histopathological features. This is followed by a brief literature review.

局限性掌跖角化減少症是一種罕見病。形態上表現為長期單一的皮膚斑，邊界清淅伴有細鱗屑，常見於手掌或腳掌。我們報告了一例 73 歲的華裔婦女出現這種病變，重點探討其臨床和組織病理學特徵，並接續簡短的文獻綜述。

Keywords: Circumscribed, hypokeratosis, palmar, plantar

關鍵詞：局限性、角化減少症、掌部、跖部

Introduction

Circumscribed hypokeratosis is a rarity in clinical dermatology. Since the first description by Pérez et al,[1] the entity circumscribed palmar or plantar hypokeratosis has been increasingly recognised as a distinct clinicopathological entity.

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The typical clinical scenario consists of a long-standing, asymptomatic solitary depressed macule with a sharply defined border over the palms and soles.

The differential diagnoses include porokeratosis of Mibelli, palmoplantar psoriasis, ruptured pompholyx if the lesions concerned are tiny, and Bowen's disease. Diagnosis requires histopathology. Histopathologically, there is hypogranulosis and marked thinning of stratum corneum. By definition, cornoid lamellation and epidermal dysplasia have to be absent before a diagnosis of hypokeratosis can be made.

Despite the accumulating experience, the pathogenesis remains unknown, and treatment is

not satisfactory. We describe a 73-year-old Chinese woman who presented with a solitary lesion over her left palm for a decade. To the best of our knowledge, this is the first local case reported in Hong Kong.

Case report

A 73-year-old lady presented with an asymptomatic macular eruption over her left palm for one decade. There was no preceding trauma history, exposure to chemicals or insect bite. The eruption was slowly enlarging. She was healthy all along. Clinically it was a circumscribed depressed light-pinkish macule, located at the thenar eminence of her left palm. It measured 1x0.9 cm across (Figure 1a). The surface of the macule was smooth and skin markings were well preserved. There was no increase in temperature, tenderness, pulsatility or palpable mass underneath. The border of the macule was sharp and irregular, step-like and slightly scaly (Figure 1b).

Dermoscopically, there were linear rows of whitish dots and thin streaks arising from a background of erythema (Figure 1c). There was no abnormal pigment network or vascular pattern. The border was stair-like, with sloping towards the inner macular region (Figure 2a). Scaling was intermittently observed along the entire course of the border. Incisional skin biopsy was extended to include the depressed macule, the border and the adjacent normal skin (Figure 2b).

Histopathologically, there was orthohyperkeratosis with a stair-like, drop-off of stratum corneum at the border of the macule (Figure 2c). The macular region showed a marked reduction in stratum corneal thickness (Figure 3). Fraying and hyper-eosinophilia was observed along the inner free edge of the ortho-keratin (Figure 4a). The thickness of the stratum corneum returned to normal outside the orthohyperkeratotic border (Figure 2c). Hypogranulosis and hypokeratosis were present within the region of depressed macule (Figure 4b), while both the dermis and subcutis were unremarkable. There was no cornoid lamellae, cytoplasmic or intra-nuclear viral inclusion, spongiotic dermatitis or epidermal keratinocyte dysplasia.

The patient opted for a trial of topical calcipotriol ointment. Apart from mild irritation, the treatment was well-tolerated. The patient is currently under regular follow-up.

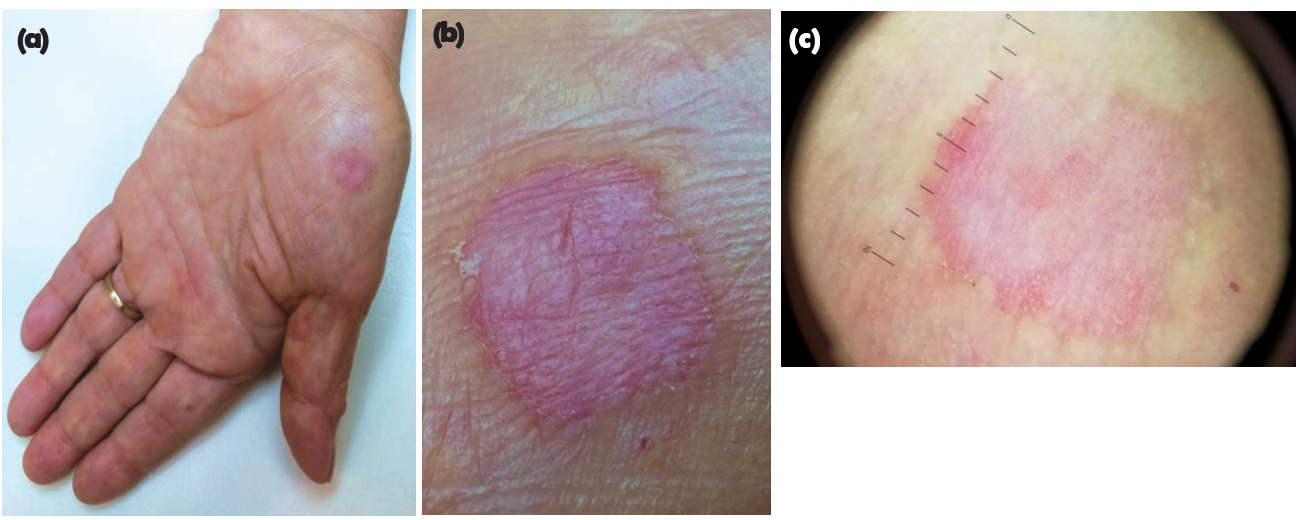

Figure 1. (a) A solitary asymptomatic macule over the thenar eminence of the left palm of the patient. (b) Close-up view of the asymptomatic macular eruption, featuring a depressed erythematous macule with a sharply defined border with focal scaling. (c) Dermoscopic view showing white dots and thin white streaks arising in an erythematous background.

Discussion

Since Pérez et al published their first case series in 2002, there have been less than a hundred case reports of palmoplantar hypokeratosis.[1] Perez et al proposed faulty epidermal maturation as the responsible mechanism, given a slow disease progression and an absence of trauma history in their case series.[1] Obermoser et al, on the contrary, opined that late onset of clinical disease plus progressive lesional enlargement in their case series spoke against a developmental anomaly or congenital malformation.[2] Chronic, minor repetitive trauma,[3,4] human papilloma virus infection,[5] primary keratinisation defect on acral sites,[6] have all been implicated as potential triggers. Other studies have shown a differential expression of various keratins in lesional and non-lesional keratinocytes, as demonstrated by immunohistochemical methods.[7-9] Further attempts at characterisation of the various keratins at the site of hypokeratosis have revealed

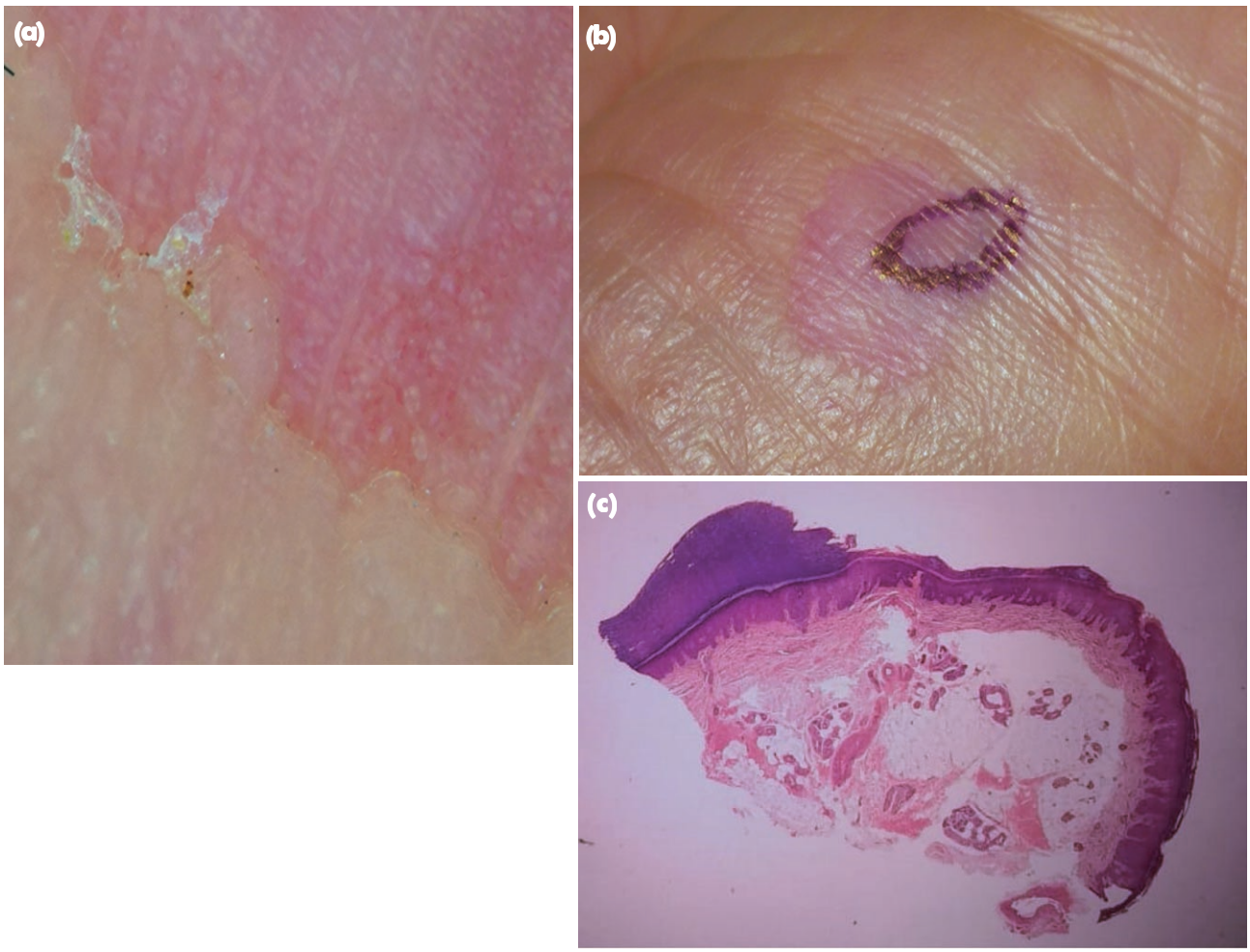

Figure 2. (a) Magnified dermoscopic view. A relative prominence of white dots and thin streaks within the macular erythema corresponding to the acrosyringial apparatus. It is surrounded by an irregular scaly border which is stair-like, sloping downward and inward towards the erythematous area. (b) Pre-operative clinical photo of the site of incisional skin biopsy. (c) Scanning magnification on one half of the bisected skin specimen. The depressed macule was located on the right-hand side of this specimen. An abrupt reduction of stratum corneal thickness is striking even at this scanning power view. Towards the other end of the specimen, the corneal thickness returned to normal after passing through the orthohyperkeratotic border of the lesion. (H&E stain, 10x)

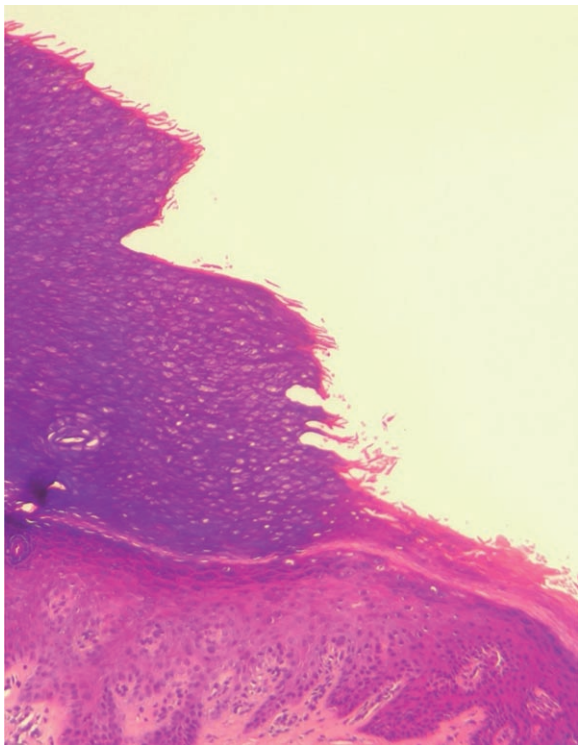

Figure 3. Medium power of the photomicrograph, taken at the border of the depressed macule. The stair-like drop-off configuration of the stratum corneum can be clearly seen. There is a gradual decrease in thickness of the stratum granulosum in the region of hypokeratosis. (H&E stain, 100x).

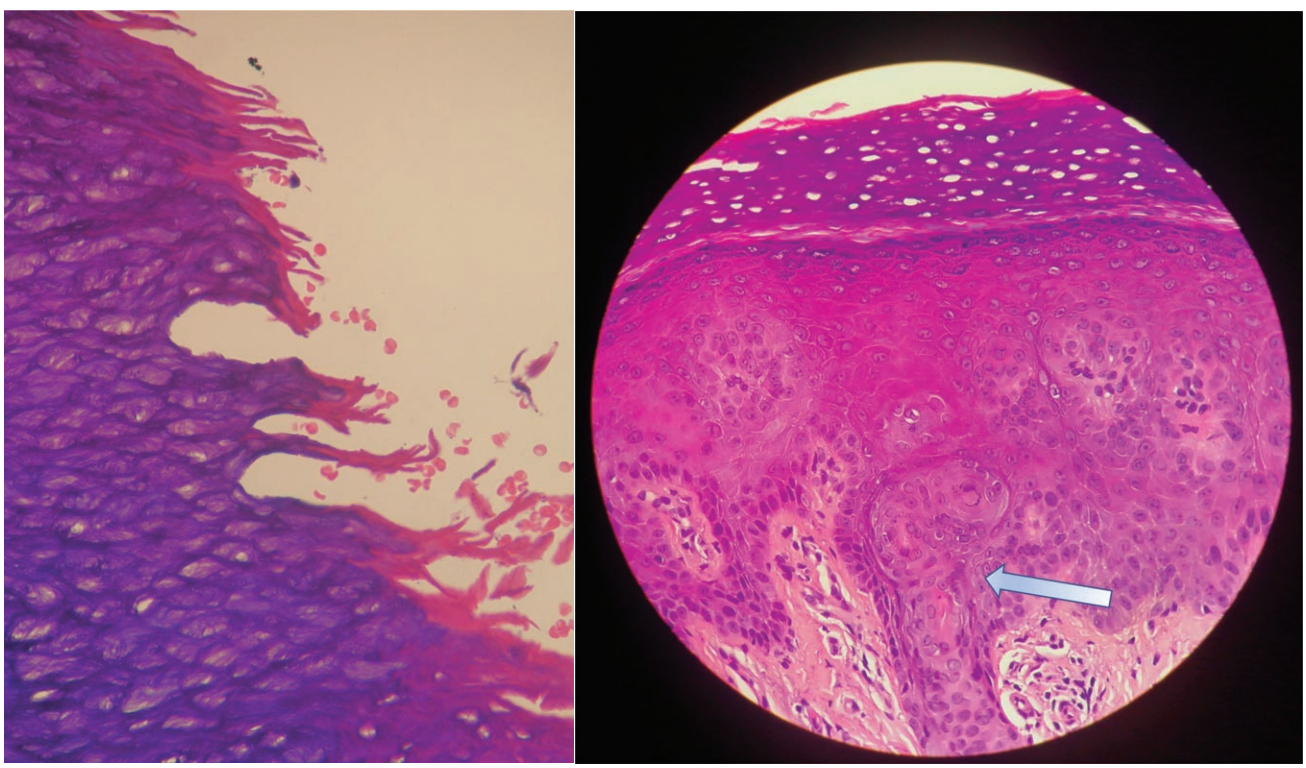

Figure 4. (a) High power view showing the fraying and hypereosinophilia of keratinous material along its inner free edge. (H&E stain, 400x). (b) Medium power view taken within the site of depressed macule. Hypokeratosis and hypogranulosis are both evident. The acrosyringium is seen at the lower centre of the photomicrograph (depicted by a blue-white arrow). White dots or thin streaks under dermoscopy are accentuated as a result of the reduced thickness of the overlying stratum corneum. (H&E stain, 200x)

a failure of palmo-plantar type keratinisation, and subsequent regression towards the truncal type inter-follicular differentiation. Corneocyte fragility and enhanced desquamation process are believed to be other contributory factors in the pathogenesis of hypokeratosis.[10] Despite all these novel findings, as the definitive link between the clinical phenotype and inherent genetic predisposition is unknown, the exact pathogenic mechanisms leading to the abnormal keratinisation patterns remain speculative.

The lesion commonly occurs on the volar aspects of palms and soles. Stepped desquamation along the rimming border, white spots, red dots and diffuse erythema are considered to be reliable dermoscopic signs.[11] The white dots as observed dermoscopically like in our case represents acrosyringia. The perceptible erythema is attributed to dilated superficial dermal capillaries, as accentuated by the thinned horny layer from above.[11] Nishimura et al proposed that chronic dermal inflammation may also contribute to the visually perceptible erythema.[11]

Diagnosis requires a skin biopsy. The incision should extend to include the lesion together with the surrounding normal skin. Histopathologically, the border of the depressed macule will typically reveal stair-like compact orthokeratotic keratin. Fraying and gradual thinning of the orthokeratin towards the centre of the lesion is a consistent feature. Within the depressed macule, hypokeratosis associated with hypogranulosis is pathognomonic. We consider that hypokeratosis-hypogranulosis and stair-like compact ortho-hyperkeratosis be viewed as a diagnostic *sine qua non* in its own right.

Circumscribed hypokeratosis usually exists alone. According to the literature, the clinical course of circumscribed hypokeratosis seems to be chronic and indolent. For tiny pruritic lesions where spongiotic reaction pattern is evident histopathologically, ruptured pompholyx should be considered as a differential diagnosis. Other important differential diagnoses include Bowen's disease, porokeratosis of Mibelli, and palmoplantar psoriasis. Histopathology is required to distinguish between them. Chronicity, absence of symptoms, and slow clinical progression are common features shared by porokeratosis, hypokeratosis and Bowen's disease. Therefore, it is possible that there may be a link between them. Further studies are required.

Treatment of this condition is generally unsatisfactory irrespective of treatment modality. Success has been reported with topical calcipotriol,[6] cryotherapy,[4] and laser therapy.[12] Curative surgical excision is feasible as long as the lesion is less than 1 cm in diameter. In view of the indolent biological behaviour of circumscribed hypokeratosis, aggressive treatment is not recommended. If treatment outcome remains unsatisfactory, expectant management is always a feasible option to consider.

Conclusion

We herein describe the first report of circumscribed palmar hypokeratosis in our locality. It is not a difficult diagnosis to make, provided that one is cognisant of such clinicopathological entity and the diagnostic *sine qua non*. Circumscribed hypokeratosis is a benign, rare and chronic disease which can be overlooked in a busy clinic. Although dermoscopy examination can provide clues, histopathology remains the gold standard for diagnosis. So far, treatment is not clearly defined and expectant treatment can be the best choice.

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