# CIRCUMSCRIBED ACRAL HYPOKERATOSIS

ALEXANDRU OANȚĂ*, MARIUS IRIMIE**, VERONICA ILIESCU*, TIBERIU TEBEICĂ***, SMARANDA ȚĂREAN*

**Summary**

*Circumscribed acral hypokeratosis is a condition characterized by an acquired, solitary, well-defined and depressed, asymptomatic lesion occurring in a middle-aged or advanced-age woman. In the pathogenesis of the condition, the existence of an aberrant epidermal clone is incriminated as a cause of epidermal hyperproliferation and increased corneocyte fragility.*

*The case presented is that of a 75-year-old patient consulted for a clinical and histopathological lesion of circumscribed acral hypokeratosis located at the level of the fifth finger of the left hand. The treatment consisted of cryotherapy without an improvement in the lesion.*

***Key words:*** *circumscribed acral hypokeratosis.*

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## Introduction

Circumscribed acral hypokeratosis (CAH) is a condition described by Perez et al. [1] in 2002. Subsequently further cases were published.[2-6] At present, the condition is recognized as a distinct entity. Patients are usually middle-aged or advanced-age women who have an acquired, solitary, asymptomatic, well-defined and depressed lesion, especially palmar (thenar and hypothenar eminence) but also plantar, with a long evolution. In the pathogenesis of the condition, the existence of an aberrant epidermal clone responsible for a localized disorder of keratinization is incriminated as a cause of the condition.

## Clinical case

A 75-year-old female patient is consulted for a lesion located at the level of the fifth finger of the left hand for about 18 months. The dermatological examination revealed a well-defined, oval lesion sized 3/2 cm, with an asymptomatic, red, depressed central area located at the external side of the fifth finger of the left hand (Figure 1).

The histopathological examination revealed a well-defined area of hypokeratosis contrasting with the compact keratosis layer of the adjacent epidermis, with sudden transition. At the level of the hypokeratosis lesion there is hypogranulosis, slight irregular acanthosis of the malpighian layer and vascular hyperplasia in the underlying dermal papillae (Figures 2 and 3).

The treatment administered consisted of cryotherapy of the lesion, without a clear improvement of the lesion.

* SC Dermamed SRL Brașov.
** Faculty of Medicine Transilvania University Brașov.
*** Dr. Leventer Centre - Bucharest.

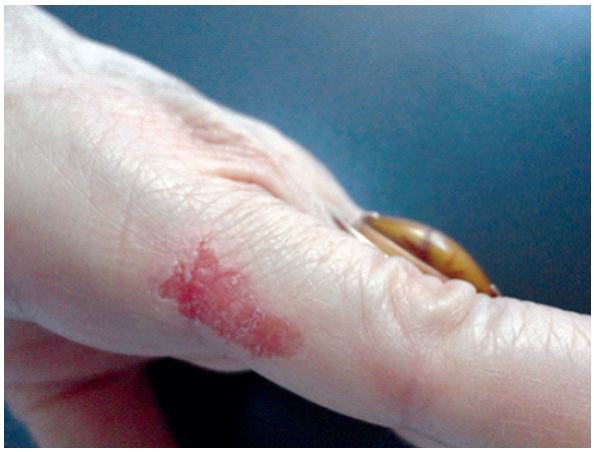

*Fig. 1 – Circumscribed acral hypokeratosis - clinical aspect*

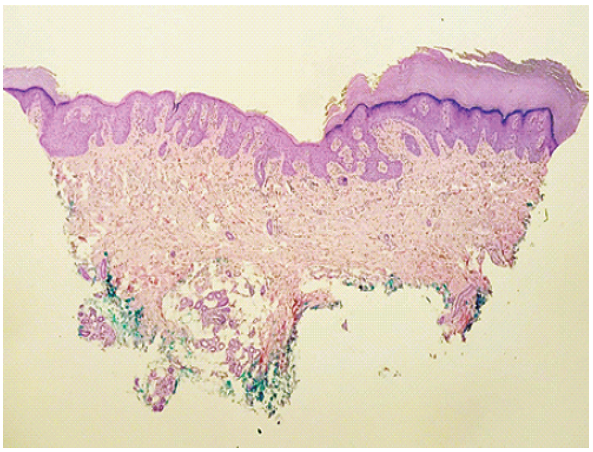

*Fig. 2 – Circumscribed acral hypokeratosis – histopathological aspect (HE stain, 10X)*

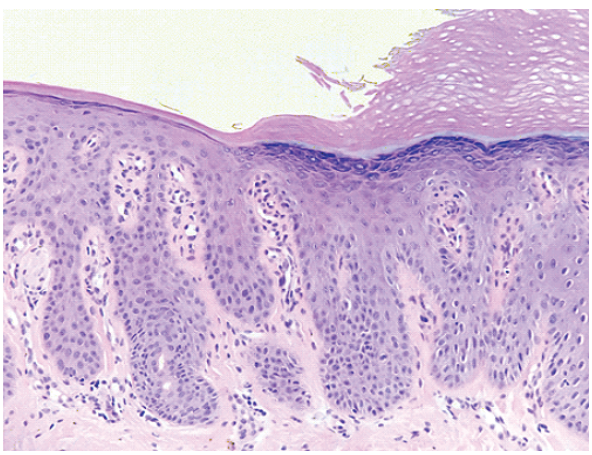

*Fig. 3 – Circumscribed acral hypokeratosis – histopathological aspect (HE stain, 20X)*

## Discussions

CAH, also known as circumscribed palmar or plantar hypokeratosis, was defined by Perez in 2002.[1] The clinical and histopathological aspects of CAH are distinctive. The condition is usually found in middle-aged and advanced-age women, the average age being 60 years. Clinically, they occur as round or oval asymptomatic lesions with a diameter of 1 to 4 cm, very well defined, shallow, with erythematous base, no expansion after appearance and with an evolution duration of up to 40 years.[2] Usually there is only one lesion, but the coexistence of two lesions has also been described. The localization of the lesions is particularly palmar, especially on the thenar and hypothenar eminences and rarely plantar on the medial area.

Dermatoscopy may be useful in the diagnosis of CAH, with two characteristic signs reported. The first sign, the edges of the lesion are "*en marches d'escadier*" or geological layers, observing

a diminution of the stratum corneum from the outside to the interior of the lesion.[7] The second sign, the centre of the hypokeratosis lesion has a homogenous, erythematous appearance, the superficial dermal vessels are visible and have a "dotted line" appearance, and some vascular loops may be present.[8]

The histopathological appearance of CAH is characterized by a sudden decrease in the thickness of the stratum corneum at the site of the lesion. The lesion centre has an orthokeratosis appearance, but sometimes a parakeratosis area can be highlighted. The granular layer is usually slightly diminished although sometimes it may be normal or thicker. Dilated and slightly sinuous capillaries are present in the papillary dermis.

The clinical differential diagnosis of CAH is set by comparison with porokeratosis of Mibelli, Bowen's disease and less with post-traumatic bullae, pitted keratolysis, eccrine poroma, and spinocellular carcinoma. Bowen's disease may present as an erythematous plaque with more or less hyperkeratosis characteristics. Unlike CAH, in Bowen's disease the diameter of the lesion increases slowly. In the porokeratosis of Mibelli, at palpation, a peripheral relief *"en chemin de rond"* corresponding to the lamellar corneal is present, while in the CAH the squamous margin indicates the concave depression of the lesion.

In the CAH pathogenesis the main hypothesis is based on the existence of an aberrant epidermal clone that would be responsible for the localized disorder of keratinization. This clone would present a lack of expression of certain keratins.[9] Repeated local trauma was also incriminated for the occurrence of CAH especially in the dominant hand.[10, 11] The presence in the epidermis of the alternation of areas of hypo- and hypergranulosis with irregular acanthosis and sometimes with the presence of keratohyalin granules, changes met in verruca vulgaris as well as detection by a number of authors of HPV type 4 and 6 have suggested HPV infection as a possible causative factor of CAH (3, 4), an infirmed hypothesis. If in the vast majority of cases published, CAH is an acquired lesion, very rare publications recall the existence of a congenital CAH.[12]

Jarret[2] and Boffa[6] have successfully used cryotherapy in the treatment of CAH, having as an explanation the destruction of the abnormal clone followed by repopulation with normal epidermal cells. Urbina et al.[13] showed improvements in CAH following the application of the topical calcipotriol, which models the growth of the epidermis.[14,15] In contrast, Mensing et al.[16] have found the topical calcipotriol ineffective in the treatment of CAH. Photodynamic therapy was also attempted, and topical corticoids and retinoids proved to be ineffective.[5,17]

Our case presents a lesion in an elderly woman located at the external side of the fifth finger of the left hand with clinical and histopathological aspects characteristic of CAH. In conclusion, CAH is a rare condition, whose knowledge allows the clinical diagnosis to be established. In the pathogenesis of HCA, an epidermal hyperproliferation is incriminated, accompanied by increased corneocyte fragility due to the existence of an aberrant epidermal clone.

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Conflict of interest
NONE DECLARED

*Correspondance address:* Marius Irimie
Faculty of Medicine Transilvania University Brașov
E-mail: marius_irimie2002@yahoo.com