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CONTROVERSIES IN DERMATOLOGY

Circumscribed Palmar or Plantar Hypokeratosis 10 Years After the First Description: What Is Known and the Issues Under Discussion☆

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Abstract This review of the literature on palmoplantar hypokeratosis, a process that was first indentified only 10 years ago, discusses the current state of our understanding, the therapeutic options available, and the debate about etiology. Forty-four publications reporting 69 cases were found. Palmar or plantar hypokeratosis occurs mainly in women (76.8%) and age at the time of a first visit to a physician ranges from 42 to 84 years. Most cases present between the ages of 51 and 70 years. The majority of patients have had solitary lesions usually located on the right palm, particularly in the regions of the thenar (in 44/79 lesions [55.7%]) or hypothenar eminences (in 11/79 lesions [13.9%]). In only 8 cases was there a history of prior trauma at the site. Studies using polymerase chain reaction techniques to identify human papillomavirus involvement were negative in most cases. These hypokeratotic lesions are localized epidermal depressions formed by an abrupt thinning of the stratum corneum, providing a singular histopathologic feature. This condition can currently be considered a localized keratinization disorder affecting zones where there is a thick stratum corneum. The precipitating cause is unknown and a definitive treatment remains to be found. The mechanism would be the localized failure of a clone of keratinocytes during differentiation toward normal palmoplantar hyperkeratinization.

PALABRAS CLAVE
Capa córnea;
Hipoqueratosis;
Palmas y plantas;

Hipoqueratosis circunscrita palmar o plantar. Conocimientos y controversias tras 10 años de su descripción

Resumen Se revisa el estado actual de un nuevo proceso y los avances que han ido apareciendo en la literatura respecto a su estudio, posibilidades terapéuticas y controvertida etiología, tras

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justo una década de su descripción inicial. Hemos encontrado 44 publicaciones al respecto, con 69 casos. Predomina en mujeres (76,8%), con edades entre los 42 y 84 años para toda la casuística en el momento de la consulta, siendo el rango más frecuente de aparición de las lesiones entre los 51 y 70 años. La gran mayoría de las veces fueron lesiones únicas y se localizaron predominantemente en la palma derecha, preferentemente en la región tenar, con 44/79 lesiones (55,7%) y luego en la hipotenar, con 11/79 lesiones (13,9%). Solo en 8 casos se obtuvo el antecedente de un traumatismo previo. Los estudios mediante PCR en búsqueda de papiloma virus humano han sido negativos la gran mayoría de las veces. Las lesiones consistieron en una depresión focalizada de la epidermis producida por una brusca disminución de espesor de la capa córnea, delimitando un singular concepto histolopatológico. En la actualidad puede ser considerada como un trastorno focalizado de la queratinización, que afecta zonas con un estrato córneo grueso, cuya causa precipitante es desconocida y su tratamiento definitivo aún por dilucidar. Se produciría por un fallo clonal localizado de los queratinocitos hacia una diferenciación hiperqueratósica normal, de tipo palmo-plantar.

Introduction

Increased thickness of the corneal layer of the skin, or hyperkeratosis, is described in possibly all the analytical indices of dermatopathology and dermatology texts. In contrast, the term *circumscribed palmar or plantar hypokeratosis*, brought into usage a decade ago to describe an abrupt reduction in the thickness of the corneal layer in a circumscribed area of the palm or sole, is much less common.[1] For the origin of this term we must go back to the year 2002, when it was used to describe a series of 10 patients from Germany, Chile, and Spain, reported by Pérez et al.[2]

Since that time, further cases with identical clinical and histopathologic characteristics have been reported in other countries, including Austria, Korea, United States, France, Italy, Japan, Malta, New Zealand, and Peru, leading gradually to the characterization of a new entity.

The purpose of this article has been to review the advances that have been made in this disease a decade after its first description, with impartial contributions from different countries across the globe, by professionally independent authors. Our aim has been to describe the current state of knowledge of the disease and to review the literature on advances in research into the disease, on the therapeutic options available, and on the debate about its etiology.

Clinical Presentation

A search performed in a number of databases at the time this article was being written revealed 44 papers on circumscribed palmar or plantar hypokeratosis. Sixty-nine cases have now been reported.[2–44] Their clinical characteristics are listed in Table 1 and are summarized in Table 2. This is an acquired disorder that is more common in women, with a female to male ratio of 3.3 to 1, and it has been reported in patients aged between 42 and 84 years (mean age, 64.6 years) at the time of first consultation (one outlying case with lesions present from birth[44] was excluded from this calculation). In a large majority of cases (60 patients) the lesions were single; there were only 8 cases with 2 lesions,[3,11,13,20,21,27,29] one with 3 lesions,[31] and an exceptional case with 13 lesions in a linear distribution[21] (also excluded from the calculations as it was an outlier). Most lesions were located on the right palm, mainly affecting the thenar region (44 of 79 lesions). Lesions affecting the digits have been detected in isolated cases,[2,11,16,24,27,31,34,38,41] most commonly on the thumb,[2,11,16,27,31,34] but in 1 case on the dorsum of the index finger.[24] Lesions have also been reported on the palm,[26] on the dorsum or border of the feet,[24,34,44] and on the sole.[2,7,21,22,26] The mean duration of lesions was 12.8 years, with an approximate range of 1 to 50 years; this could not be exact as 1 patient of 64 years of age[2] had developed lesions as a child and these were arbitrarily determined to have appeared at 14 years of age; the range was calculated on the basis only of those patients who stated the duration of the lesions in years or in whom this could be estimated.

It is interesting to observe that the lesions had been present for a long period before consultation in the large majority of patients, possibly because they were considered banal by the patients themselves. The onset of lesions occurred most commonly between 61 and 70 years of age (13 cases), followed in decreasing frequency by the intervals of 51 to 60 years (12 cases), 41 to 50 years (8 cases), and 31 to 40 years (6 cases); these calculations were based only on the 47 cases in which the figures were known or could be accurately estimated. Some patients reported a history of trauma to the area, in particular burns,[5,21,41] but also trauma from gardening or frequent writing,[11] a bird peck,[17] 1 minor trauma,[19] or long-standing practice of embroidery,[32] as a possible trigger, but the large majority of patients reported no history of trauma and were unaware what could have triggered the lesions.

The clinical appearance of the lesions was almost identical in all cases, with a well-defined, round or oval depression on the palm (Fig. 1) or sole (Fig. 2), with reddish discoloration, and a slightly scaly border. The size of the lesions varied between a few millimeters[11,21,37,43] and 5.5 cm,[10] but mostly measured around 1.5 cm. The lesions were typically asymptomatic, although some patients described the skin as being more delicate and sensitive in the area of the lesion.