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CASE REPORTS

Palmar Filiform Parakeratotic Hyperkeratosis Without Underlying Malignancy

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Abstract. Filiform hyperkeratosis (FH) is a rare entity clinically characterized by keratotic spicules on the palms, soles or other areas of the body surface. Its association with several diseases, including neoplasms, has been extensively discussed but currently it is not considered a well-defined paraneoplastic disorder. We report a 72-year-old patient that referred lesions on both palms of three months duration. The rest of the body surface did not show similar lesions. Complementary exams did not reveal any abnormal findings. The histopathological exam showed parakeratotic columns with a slightly decreased granular layer and a mild dermal inflammatory infiltrate. In the last years different terms have been employed to refer to these hyperkeratotic lesions contributing to the lack of clarity that currently persists. Although FH might be classified close to the clinical spectrum of porokeratoses, it presents particular clinical and histological findings that allow it to be considered a separate entity. We report a new case of this rare disorder with exclusive palmar involvement in a patient without underlying malignancy and review the main characteristics of similar cases reported in the literature. Despite the obscure association between this entity and neoplasms, the majority of authors deem it necessary to rule out underlying diseases.

Key words: filiform hyperkeratosis, spiny keratoderma, porokeratosis punctata, cornoid lamella.

HIPERQUERATOSIS FILIFORME PARAQUERATÓSICA PALMAR SIN MALIGNIDAD SUBYACENTE

Resumen. La hiperqueratosis filiforme (HF) es una entidad infrecuente clínicamente caracterizada por la presencia de espículas queratósicas en las palmas, plantas u otras zonas de la superficie corporal. Su asociación con diversas enfermedades, incluyendo neoplasias, ha sido ampliamente discutida, sin que en la actualidad se considere un cuadro paraneoplásico bien definido.

Se presenta el caso de un paciente de 72 años que consultó por lesiones de tres meses de evolución en las palmas de ambas manos. El resto de la superficie corporal no mostraba lesiones similares. Los estudios complementarios realizados no revelaron hallazgos patológicos de interés. El examen histopatológico mostró la presencia de columnas paraqueratósicas con una capa granulosa ligeramente disminuida y un leve infiltrado inflamatorio dérmico.

Diferentes términos han sido empleados en los últimos años para hacer referencia a estas lesiones hiperqueratósicas, contribuyendo a crear un ambiente de confusión que perdura actualmente. A pesar de que la HF podría ser clasificada en la proximidad del espectro clínico de las poroqueratosis, presenta hallazgos clínicos e histopatológicos particulares que permitirían considerarla una entidad aparte. Describimos un nuevo caso de esta infrecuente entidad con afectación exclusiva palmar en un paciente sin malignidad oculta y revisamos las principales características de los casos similares descritos en la literatura. A pesar de la oscura asociación de este cuadro con neoplasias, la mayoría de los autores consideran necesario el despistaje de patología subyacente.

Palabras clave: hiperqueratosis filiforme, queratodermia espinosa, poroqueratosis *punctata*, lámela cornoide.

Introduction

Filiform hyperkeratosis is a rare dermatosis described in the literature using a number of different terms,[1] and its association with underlying disease is still under discussion. We report a case of filiform hyperkeratosis with no underlying malignancy and review existing case reports

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from the literature describing parakeratotic columns and palmar and/or plantar lesions. According to the classification of the disease established by Zarour et al[2] and later modified by McGovern and Gentry,[1] these clinical and histological findings are characteristic of type Ia filiform hyperkeratosis (palmoplantar filiform parakeratotic hyperkeratosis).

Case Description

A 72-year-old man with a history of spondylolisthesis consulted for cutaneous lesions that had been present for 3 months with no other symptoms. He described the lesions as spikes on the fingers of both hands. They were slightly painful to the touch and were not easily detached from their base. The patient did not report altered sweating, a history of exposure to arsenic, or a family history of similar lesions.

Examination of the underside of the fingers on both hands revealed multiple irregularly distributed filiform hyperkeratotic lesions. These were raised by 1-2 mm, yellowish in color, and some rested on thin erythematous, edematous macules (Figure 1). No significant abnormalities were observed on the soles of the feet, hair, or nails.

The results of complementary studies (blood workup and urinalysis; protein profile; thyroid hormones; autoantibody and biomarker study; syphilis serology; peripheral blood smear; endoscopy of the upper and lower digestive tract; electromyogram; computed tomography scan of the head, thorax, abdomen, and pelvis; abdominal ultrasound scan; and simple X-ray of the chest, spinal column, and sacroiliac joints) were normal and a possible occult neoplasm was ruled out.

Histological examination of the skin biopsy revealed well-defined parakeratotic columns with clear limits and a normal adjacent orthokeratotic epidermis. Some of the columns emerged from a moderately reduced stratum granulosum (Figure 2). A mild inflammatory infiltrate and dilatated capillaries were observed in the upper dermis. There was no vacuolization of keratinocytes or dyskeratosis.

The application of emollients containing urea 3 times daily led to an improvement in the lesions, although many persisted. Subsequent examinations did not reveal new lesions.

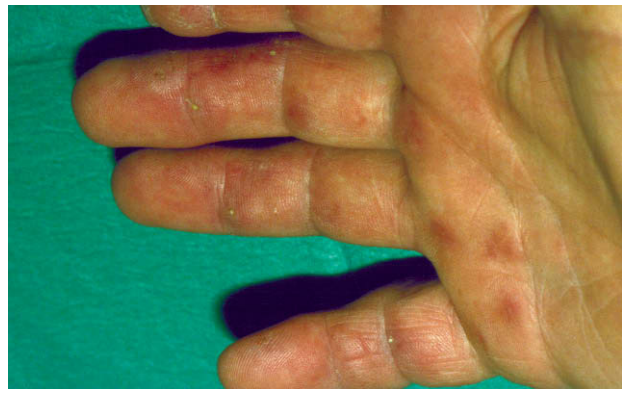

Figure 1. Hyperkeratotic spicules on the palmar surface of the fingers.

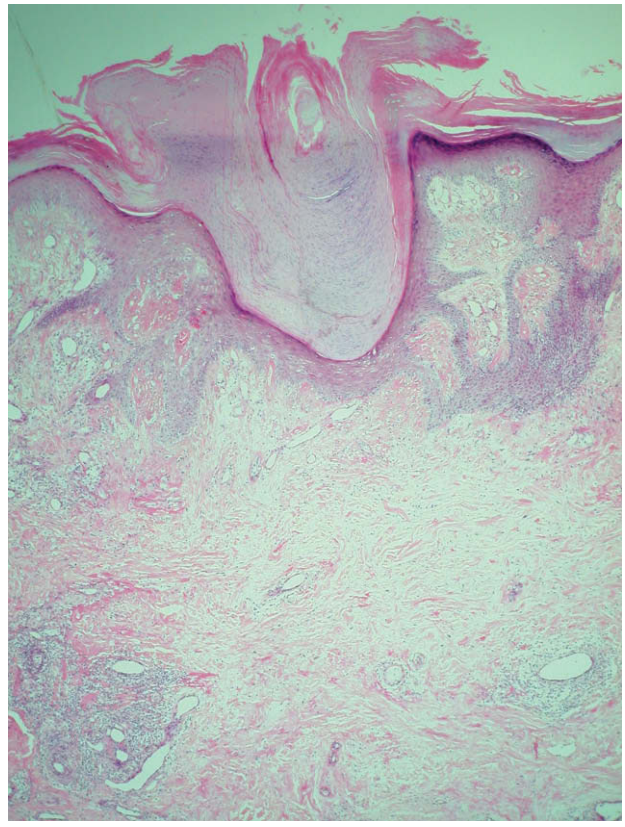

Figure 2. Parakeratotic column resting on a thinned stratum granulosum (hematoxylin-eosin, ×40).

Discussion

Since Goldstein[3] reported the first case of digitate hyperkeratotic lesions in 1967, several authors have described similar cases with hyperkeratotic spikes on the palms, soles, and other skin surfaces. Different terms have been coined to describe these lesions, including punctate keratoderma,[4] punctate porokeratotic keratoderma,[5,6] punctate porokeratosis,[7] punctate porokeratosis palmaris et plantaris,[8] filiform palmoplantar hyperkeratosis,[9,10] spiny keratoderma,[1,11,12] palmar filiform hyperkeratosis,[13] and multiple minute digitate hyperkeratosis.[14] The fact that similar entities have different names and the same term is used to describe different conditions illustrates the confusion still surrounding this dermatosis. Although some authors have made serious attempts to group and classify the cases

Table 1. Classification of Filiform Hyperkeratosis According to Zarour et al[2] and McGovern et al[1]

Type		*Localization*	*Histology Findings*
I	a: Palmar parakeratotic FH	Palms, soles	Parakeratosis
	b: Disseminated parakeratotic FH	Generalized, with sparing of palms and soles	Parakeratosis
II	a: Palmoplantar orthokeratotic FH	Palms, soles	Orthokeratosis
	b: Disseminated orthokeratotic FH	Generalized, with sparing of palms and soles	Orthokeratosis
III	FH in eccrine hamartoma	Linear appearance; any skin surface	Parakeratosis

Abbreviation: FH, filiform hyperkeratosis.

reported to date, there is still a lack of consensus on the correct name for this entity.

The main differences between the cases reported in the literature are found in the location of the lesions and the associated histological findings, given that the clinical appearance is very similar in all of them. These observations led Zarour et al[2] to divide the hyperkeratotic lesions into 3 groups (I, II, and III) and 2 subgroups (a and b) based on the histological findings and the location of the lesions (Table 1). McGovern et al[1] later proposed a slight modification to the classification and introduced the term spiny keratoderma and the subtypes parakeratotic and orthokeratotic. Our case meets the criteria for type Ia filiform hyperkeratosis, given that the lesions were found only on the palms of both hands and the main histological finding was the presence of well-defined parakeratotic columns.

Based on a PubMed search using the key words "filiform hyperkeratosis," "palmar filiform hyperkeratosis," and "spiny keratoderma," we identified 29 cases (including the present case) that could also be classified as type Ia filiform hyperkeratosis (Table 2). The prevalence seems to be higher in men (20 of the 29 cases) and the lesions appear mainly in older patients (mean, 62.48 years; range, 20-85 years). The palms and soles were simultaneously involved in 17 of the 29 cases and the palms alone in 12 patients, including ours.[1,11,13-17] An underlying tumor was detected in only 9 patients: breast cancer,[18] lung cancer,[6,19] melanoma,[16] sigmoid colon cancer,[10] chronic lymphatic leukemia,[11] myelofibrosis,[13] renal cancer,[9] and esophageal cancer.[15] In other cases the lesions were associated with different processes such as polycystic kidney disease[10,20] or bronchial asthma.[15]

Filiform hyperkeratotic lesions are usually asymptomatic, raised by 1 to 3 mm, whitish-yellow in color, and emerge from a noninflamed base from which they are not easily detached. Nevertheless, in some cases the lesions sit on erythematous areas (5, present case).

The main histological finding of parakeratotic filiform hyperkeratosis is a well-defined column of parakeratotic cells that is reminiscent of the stratum corneum observed in porokeratosis. To avoid confusion, Lestrigant and Berge[8] coined the term columnar parakeratosis to describe this finding. The parakeratotic column generally rests on a thinned, or even absent, stratum granulosum and a slightly thinned Malpighian layer. It presents very well-defined limits in relation to the adjacent normal epidermis, and it can occasionally be observed in association with hair follicles[5,16] and the acrosyringium,[6,17,19] similar to eccrine porokeratotic dermal duct nevus.[21] Mehta et al[13] described a case involving both orthokeratotic and parakeratotic columns. Vacuolization of keratinocytes is rarely reported.[22] In these cases, it is more difficult to reject the hypothesis of an association between this entity and porokeratosis; however, in parakeratotic filiform hyperkeratosis, no dyskeratosis or angularity in the column is observed. The underlying dermis does not show significant abnormalities in most cases, although a slight inflammatory infiltrate[15] and dilatation of capillaries can be observed.[5,6]

No cases of filiform hyperkeratosis have been reported to have improved or resolved spontaneously.[12] Varied results have been obtained with a number of therapeutic options, including topical[20] and oral retinoids,[13] and emollients containing salicylic acid,[1,13] urea,[13] ammonium lactate,[20] propylene-glycol,[13] and 5-fluouracil.[1,12]

Given the various clinical presentations and histopathology findings in palmoplantar hyperkeratosis, we propose that, rather than attempting to define specific conditions, we consider this entity as a spectrum of conditions, whose medical relevance lies in its possible association (doubtful in some cases, but clearly defined in others) with underlying cancer.

Despite the fact that porokeratosis and parakeratotic filiform hyperkeratosis show clear clinical and histopathological differences, both are keratinization disorders that display similarities in terms of morphology, genetic basis, and disease course, and can present before, with, or after a tumor. Therefore, a detailed study at diagnosis can prove particularly important in these patients in order to rule out systemic diseases. Equally important is long-term follow-up, which can avoid late diagnoses of tumors that could have benefited from therapy.

At present, reports in the literature suggest that it is sufficient to rule out gastrointestinal, lung, and breast tumors.

Table 2. Main Characteristics of Cases of Filiform Hyperkeratosis Reported in the Literature

Source	*Location*	*Sex*	*Age, y*	*Duration*	*Name*	*Observations*
Brown, 1971[4]	P, S	M	20	1 y	*Punctate keratoderma*	Familial
Herman, 1973[19]	P, S	M	50	10 y	*Punctate porokeratotic keratoderma*	Lung cancer
Beylot et al, 1982[9]	P, S	M	54	10 y	*Palmoplantar filiform hyperkeratosis*	Renal cancer
Himmelstein et al, 1984[7]	P, S	M	26	3 y	*Punctate porokeratosis*	–
Sakas et al, 1985[22]	P, S	M	60	15 y	*Porokeratosis punctata palmaris et plantaris*	–
Friedman et al, 1988[5]	P, S P, S	M F	72 55	13 y 11 y	*Punctate porokeratotic keratoderma*	Familial
Lestrigant et al 1989[8]	P, S	M	75	?	*Porokeratosis punctata palmaris et plantaris*	Familial
Hillion et al, 1990[18]	P, S	M	71	>30 y	*Palmoplantar filiform hyperkeratosis*	Breast cancer
Kondo et al, 1990	P, S	M	82	20 y	*Punctate porokeratotic keratoderma*	–
Osman et al, 1992[12]	P, S	M	72	2 y	*Spiny keratoderma of the palms and soles*	–
McGovern et al, 1994[1]	P	M	62	10 y	*Spiny keratoderma*	Familial
Bianchi et al, 1994[6]	P, S	M	60	3 mo	*Punctate porokeratotic keratoderma*	Bronchial carcinoma
Kaddu et al, 1995[16]	P	M	70	2 y	*Palmar filiform hyperkeratosis*	Nodular melanoma
Anderson et al, 1996[20]	P, S	F	46	2 y	*Spiny keratoderma*	Polycystic liver and kidney disease
Rault et al, 1997[10]	P, S	M	79	1 y	*Palmoplantar filiform parakeratotic hyperkeratosis*	Sigmoid colon cancer
Horton et al, 1998	P	M	64	1 y	*Spiny keratoderma*	–
	P	M	54	1 y		Simvastatin
	P	M	49	5 mo		–
	P	M	80	?		Simvastatin
	P, S	F	85	1 y		Simvastatin
	P	M	40	?		–
Bernal et al, 2000[11]	P	M	70	30 y		CLL
Handa et al, 2000[15]	P, S	F	85	?	*Spiny keratoderma of the palms and soles*	Esophageal cancer
	P	M	56	7-10 d		Asthma attack
Mehta et al, 2002[13]	P	M	73	1 y	*Palmar filiform hyperkeratosis*	Myelofibrosis
	P, S	F	70	11 y: hands 7 y: feet		– –
Guhl et al, 2005[17]	P	F	60	7 y	*Spiny keratoderma of the palms*	–
Present case	P	M	72	3 mo	*Palmar filiform parakeratotic hyperkeratosis*	–

Abbreviations: P, palms; S, soles; M, male; F, female; CLL: chronic lymphatic leukemia.

Symptoms and physical findings after rigorous examination can provide us with valuable information to direct our search in some cases.

Conflicts of Interest
The authors declare no conflicts of interest.

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