Case report

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Late-onset non-familial acrokeratosis verruciformis of hopf: a case report

V. Faina[1], F. Magri[1], A. Sernicola[1], C. Chello[1], N. Gagliostro[1], T. Grieco[1*]

[1]*Department of Dermatology, Policlinico Umberto I, "Sapienza" University of Rome, Italy*

Abstract

Acrokeratosis verruciformis of Hopf is a rare heritable autosomal dominant genodermatosis and keratiniziation disorder, first described in 1931 by Hopf. It presents with multiple flat skin colored papules, mainly localized at the dorsal region of the hands and feet. AKV is an allelic disorder, associated with heterozygous missense mutation in ATP2A2 gene, which is involved in Darier's disease. Usually, it arises in early life but rare sporadic cases with adult onset have been reported. We report a case of late-onset non-familial AKV in a 52-year old patient.
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Key words: *Acrokeratosis verruciformis of Hopf, keratosis, skin diseases*

Introduction

Acrokeratosis verruciformis of Hopf (AKV) is a rare heritable autosomal dominant genodermatosis and keratiniziation disorder, first described in 1931 by Hopf (1).

It affects both sexes (2) and is usually chronic with no spontaneous resolution.

AKV presents with multiple flat skin colored papules, mainly localized at the dorsal region of the hands and feet (3). Lesions can be located at the knees, elbows, and forearms, while forehead, scalp, flexor areas, and oral mucosa are usually not involved (4).

Other inconstant signs are palmoplantar pitting and nail dystrophy.

AKV is an allelic disorder, due to its association with heterozygous missense mutation in ATP2A2 gene, also involved in Darier's disease (DD) (5).

As an allelic disorder, AKV arises in early life but some sporadic cases with adult onset have been reported, as well as cases with no familial history (3,6).

We report a case of late-onset non-familial AKV in a 52-year old patient.

Case report

Patient information. A 52-year old woman came to visit for a papular hand dermatitis, present for 10 years and resistant to topical steroids. Family and medical history were negative for skin and systemic diseases.

Clinical findings. On examination, multiple skin-colored papules were present at the dorsal surface of both hands, extending to the wrist like a "glove"(Figure 1a, 1b); palms and other sites were spared.

Diagnostic assessment. To rule out allergic contact dermatitis, patch and prick tests were performed, with negative result.

A skin biopsy of a papule revealed ortho-keratotic hyperkeratosis and mild hyperplasia with wavy elevations of the epidermis, with a "church spire" pattern and no dermal alterations (Figure 2). Therefore, a diagnosis of AKV was made.

Therapeutic intervention. The patient started oral alitretinoin 10 mg/day, according to current guidelines of chronic hand eczema, with moderate improvement.

Discussion

The diagnosis of AKV can be a challenge. In fact, this rare disease may resemble several common conditions, including warts, lichen planus, and seborrheic keratosis. Other differential diagnosis are DD and epidermodysplasia verruciformis (3,7).

Indeed, skin biopsy is mandatory for the diagnosis. It reveals hypergranulosis, acanthosis and hyperkeratosis, with a distinctive "church spire" pattern (8,9), as described in our patient.

Correspondence: Teresa Grieco, Department of Dermatology, Policlinico Umberto I, "Sapienza" University of Rome, Italy
Viale del Policlinico 155, Rome, Italy. Phone n°: +39 338 3276733. E-mail: teresa.grieco@uniroma1.it.

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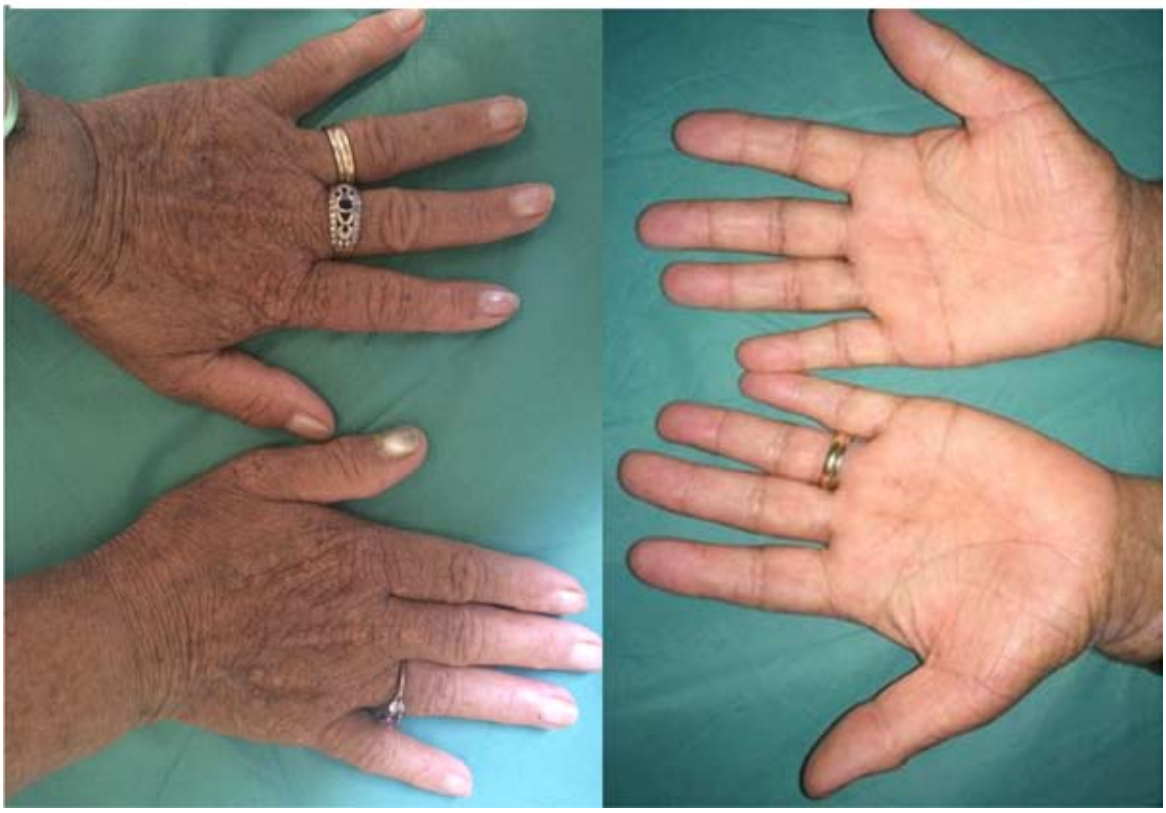

Fig. 1. Multiple papules of various sizes, of normal skin color, present bilaterally on the back of the hands; no lesions at the palmar region.

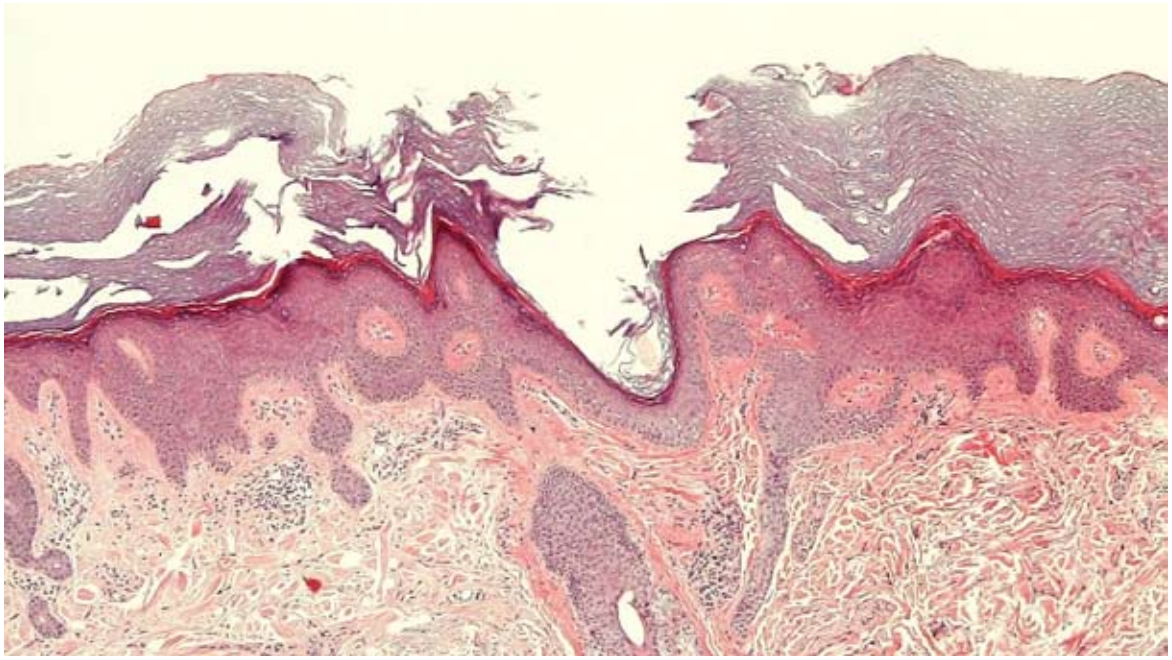

Fig. 2. Hematoxylin and Eosin stain, 10x. Compact orthokeratotic hyperkeratosis, mild epidermal hyperplasia with a "church-like" aspect. No changes or inflammatory infiltrates are present in the dermis.

AKV is a familial condition, which commonly arises at birth or during infancy.

However, our patient showed a late onset and her familial history was negative for similar conditions.

Consequently, the late onset and the clinical and histological features, guided us to diagnosing "Late-onset non-familial AKV", which is rarely reported in literature.

In 2011, Bang *et al.* described one case of late-onset non-familial AKV and collected six other cases from the literature (2).

Familial and non-familial forms do not show clinical or histological differences. However, Bang et al. observed that keratosis punctata, which is found in the familial form, has been described only in one of the seven studied patients (2). Anyway, in our case palms were spared from lesions.

Patel *et al.* described another similar case in a 41-year old patient presenting an unusual involvement of forearm flexor and leg extensor surfaces (10).

Because of the rarity of AKV, there are currently no therapeutic guidelines. Main treatments are cryotherapy, laser therapy, surgical excision (11), keratolytic preparations and acitretin (7,12).

In our case, we started oral alitretinoin 10 mg per day with moderate improvement of the lesions.

Conclusion

We presented this case because of its rarity, and to improve the number of non-familial AKV cases in literature. In our opinion, this condition is underestimated because of its clinical similarity to other more frequent conditions.

An accurate anamnesis and histopathologic evaluation are essential to formulate a correct diagnosis.

Informed consent

Written informed consent for publication of their clinical details and clinical images was obtained from the patient.

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