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Case Report

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Acrokeratosis Verruciformis of Hopf: Report of a Case of Onset in Adulthood and Review of Literature

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Abstract

Acrokeratosis verruciformis (AKV) of Hopf is a rare genodermatosis of autosomal dominant inheritance that is characterized by the presence of keratotic papules of symmetrical arrangement in rubbing areas on the back of the hands and/or feet. There are few cases of sporadic presentation in adulthood. The histopathological study shows the typical "church capitals". Due to its benign evolution, treatment is indicated at the request of patients for aesthetic reasons. We present a 76-year-old patient with sporadic Hopf's verruciform acrokeratosis of onset in adulthood.

Keywords: Acrokeratosis, Verruciform acrokeratosis, Hopf, Church capitals, Keratotic papules

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Introduction

Hopf's verruciform acrokeratosis is a rare genodermatosis characterized by the appearance of keratotic papules on the dorsum of the hands and feet. Described by Hopf in 1931, it has a worldwide distribution and affects all ethnicities, with a higher prevalence in males. It is inherited in an autosomal dominant pattern, so it is common to find several members of a family with the same dermatosis, which is usually presents in childhood. However, cases of sporadic onset in adulthood have been described, without a family history [1-3].

We present the case of a 76-year-old patient with sporadic Hopf's verruciform acrokeratosis, with onset in adulthood.

Case Report

A 76-year-old man, with no significant personal or family medical history, presented with an asymptomatic dermatosis located solely on the dorsum of both hands, of one year's duration.

Physical examination revealed multiple keratotic papules of varying sizes, some nodular in appearance, euchromic, and of firm, elastic consistency, located on the dorsum of both hands, predominantly in areas of friction (Figure 1 and figure 2). Dermatoscopic examination revealed multiple whitish patches on an erythematous-brownish background (Figure 3). Histopathological examination reported: epidermis with hyperorthokeratosis, mild acanthosis, slight papillomatosis, and isolated perivascular lymphocytes in the superficial dermis.

Based on clinical findings, dermatoscopy, and histopathology, a diagnosis of Hopf's verruciform acrokeratosis was made, and topical treatment with 20% urea cream was prescribed. The patient's condition improved with the lesions flattening one month after starting treatment.

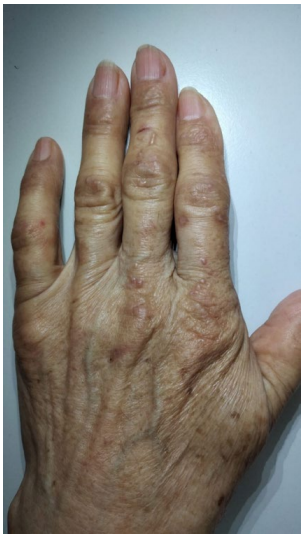

Figure 1: Euchromic or slightly erythematous hyperkeratotic papules in friction areas of the back of the left hand.

Comments

Hopf's AKV is a rare genodermatosis characterized by the appearance of keratotic papules on the dorsum of the hands and feet [4]. The keratotic papules are euchromic or slightly brownish

and are usually located on the dorsum of the hands and/or feet with a symmetrical distribution, although they can affect other areas of friction, such as forearms, elbows, knees, or even the face. They may remain isolated, maintaining their individuality, or group together in plaques, forming mosaics [2]. To confirm the diagnosis, a biopsy and histopathological examination are necessary, which reveal the presence of hyperkeratosis, acanthosis, and papillomatosis that form, although not always, circumscribed elevations in the epidermis described as "church steeples." Dyskeratosis and acantholysis are not observed [5, 6].

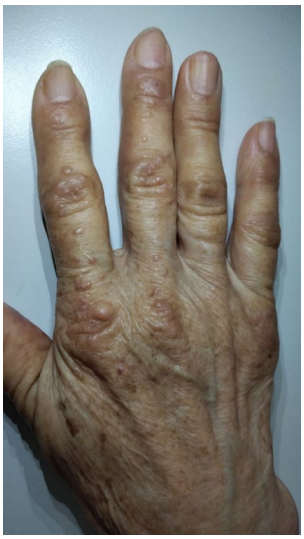

Figure 2: Hyperkeratotic papules, some larger, in friction areas on the back of the right hand.

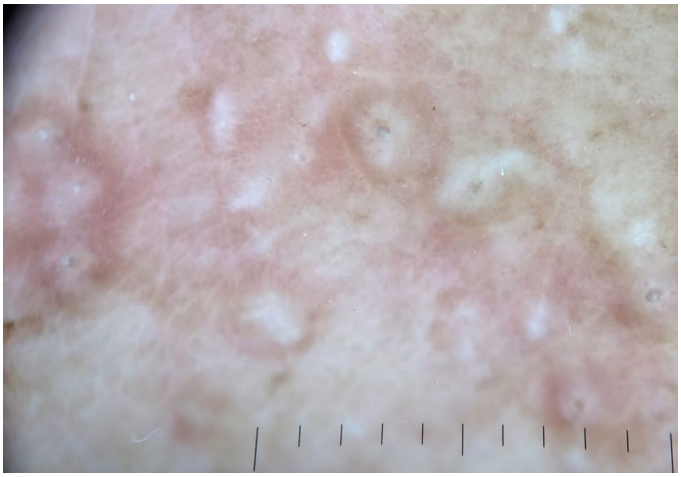

Figure 3: Dermatoscopy: whitish patches on an erythematous-brownish background.

Dermoscopy is a diagnostic tool that allows the detection of submacroscopic structures, that is, those not visible to the naked eye. In AKV, homogeneous white areas with a cobblestone-like border and brown spots are found, which histologically correlate with hyperkeratosis and acanthosis. A sunburst appearance has been described in large, mature lesions, suggesting a stretching effect that would cause the overlapping of peripheral brown spots. These latter findings have not been reported in other conditions with acral papules and therefore could have significant diagnostic value in differentiating AKV [7-9].

AKV and acral Darier's disease (AD) share similar clinical characteristics and can even be associated, leading several authors to suggest a connection between the two diseases. AD with exclusively acral localization occurs in only 2% of patients with AD. Recently, Dhitavat et al. [10] identified a heterozygous mutation (P602L) in the ATP2A2 gene in patients with vitamin K antagonists. This gene encodes an adenosinetriphosphatase that hydrolyzes adenosine triphosphate, allowing the entry of two calcium ions into the sarcoplasmic reticulum. Calcium plays a fundamental role in the regulation and differentiation of keratinocytes, so this mutation would explain the clinical and histopathological findings of dermatosis. This same mutation has also been found in dermatosis, suggesting the possibility that both diseases are different phenotypes of the same genetic alteration [11].

In addition to AD, the main differential diagnoses for AKV are Costa's acrokeratoelastoidosis and flat warts (Table 1).

AKV is a purely cutaneous and localized condition, not associated with any systemic conditions or involvement. Therefore, it does not require systematic patient evaluation or treatment, except for cosmetic reasons, where topical keratolytic agents such as urea and retinoids can be used; destructive methods such as cryotherapy or laser; or surgical excision. Cases have also been documented in which a satisfactory response is achieved with the use of oral acitretin. In our case, 20% urea cream was prescribed, resulting in a significant reduction in the thickness of skin lesions [13, 14].

Conclusions

Hopf's verruciform acrokeratosis is a rare, autosomal dominant genodermatosis characterized by symmetrically arranged keratotic papules on areas of friction on the dorsum of the hands and/or feet. Sporadic presentations in adulthood are rare, as occurred in our patients.

Table 1: Differential diagnoses of Hopf's verruciform acrokeratosis [12].

Diagnoses	AKV	Acrokeratoelastoidosis	Flat wart	AD variant xcvz
Epidemiology	Childhood/Adulthood	Childhood	First 20 years	First or second decade
Etiopathogenesis	AD, gen ATP2A2	AD (Crom 2)	HPV 3, 10, 26, 29 Y 41	AD, gen ATP2A2
Clinic	Papules in friction areas of the backs of the hands and feet (more rarely on the extensor surfaces of the forearms and legs)	Polygonal, translucent papules on the margins of palms and plants	Smooth, flat papules, yellowish-brown, pink, or euchromic in color	Small papules (red, yellowish, brown) that coalesce into symmetrical plaques with a rough, dry, and crusted surface. 95% nail involvement: leukonychia, longitudinal lines, subungual hyperkeratosis, brittleness, and striation with a distal "V" notch
Histology	Hyperkeratosis, hypergranulosis, acanthosis, mild papillomatosis "in church steeples" and perivascular lymphocytic infiltration	Orthokeratotic hyperkeratosis and acanthosis with elastorrhexis (decrease and fragmentation of elastic fibers)	Acanthosis, papillomatosis, hyperkeratosis, and koilocytes	Acantholysis, suprabasal clefts, dyskeratosis, and abnormal keratinocytes spreading into suprabasal spaces

Knowledge and recognition of this condition are essential for accurate diagnosis, ruling out differential diagnoses, and avoiding unnecessary testing for the patient. Histopathological examination reveals the characteristic "church capitals" appearance. Dermatoscopy is a non-invasive and readily available tool that can facilitate and guide the diagnosis of this rare entity.

Treatment is indicated only if the patient requests it for cosmetic reasons, given that it is a benign condition.

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Conflict of Interest

None.

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