

Acrokeratosis Verruciformis of Hopf (Hopf Disease)

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Abstract

Acrokeratosis verruciformis of Hopf is an autosomal dominant genodermatosis usually presenting with multiple planar wart-like lesions, typically observed on the dorsum of the hands, feet, elbows, and knees. The disease is very rare and the pathogenesis remains unknown. Considerable controversy surrounds the nature and relationship of acrokeratosis and Darier disease and whether they are manifestations of one genetic abnormality. We describe the case of a 40-year-old man seen in our clinic with skin-coloured, flat, warty papules localized to the dorsum of the hands and feet. Both clinical and histological findings were compatible with acrokeratosis verruciformis. We also review the disease, particularly its relation with Darier Disease and therapeutical options.

Keywords: Acrokeratosis verruciformis; Hopf disease; Church spire; Darier's disease

Literature Review

Introduction

Acrokeratosis verruciformis is a very rare, inheritable Hyperkeratotic Dermatitis that was originally described by Hopf [1]. It is characterized by multiple, localized, symmetrical, flat, skin-colored, wart-like lesions, typically observed on the dorsum of the hands and feet.

Definition & clinical picture

Two distinct entities have been described in the literature under this title: true acrokeratosis verruciformis and acral Darier's disease (differentiation is made by histopathologically – clefts) [2,3]. Acrokeratosis verruciformis is a genodermatosis inherited as autosomal dominant trait disorder [1-4]. It affects both sexes and is usually present at birth or appears in early childhood. However, the onset may be delayed until the fifth decade [3,4]. Numerous flat, hyperkeratotic occasionally verrucous papules are present on the distal part of the extremities, predominantly on the dorsa of the hands and feet [5]. Small groups or isolated papules may develop on the knees, elbows, forearms, and also on other parts of the body. The forehead, scalp, flexures, and the oral mucosa are never affected.

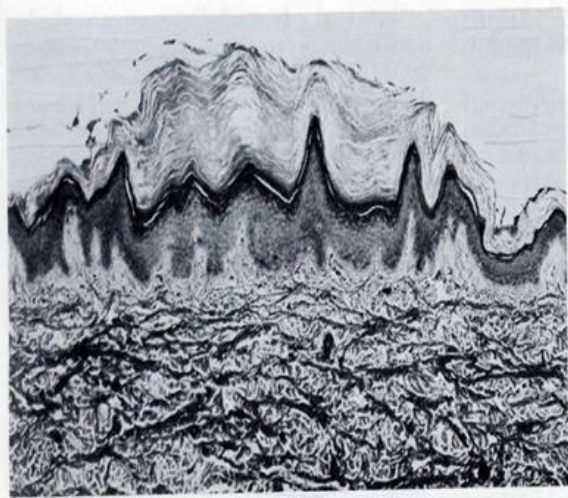


Figure 1: Acrokeratosis Verruciformis of Hopf [8].

Histopathology

The papules show considerable hyperkeratosis, an increase in thickness of the granular layer, and acanthosis. In addition, there is frequent slight papillomatosis. There is elevation of epidermis resembling church spire appearance. The rete ridges are slightly elongated (Figure 1).

Course & prognosis [1-4]

- After treatment, recurrence is expected.
- Transformation to S.C.C has been reported.

Treatment [1,6-8]

- Superficial destruction using liquid nitrogen, CO₂ laser or shave excision.
- Topical retinoic acid has been helpful in some individual.

Summary

The disease is inherited as autosomal dominant Hyperkeratotic Dermatitis, was first described by Hopf [1]. Our patient was 40 years old with Lesions on hands and feet elbows and knees. Family history is negative for similar condition but parents are cousins

Clinical synopsis

In mid-July 2013 a middle aged (40 years) man stepped in O.P.D in KDVD hospital. The patient was anxious with a frown face. He was reluctant to shake hand & could hardly uttered (salam). He complained that he had itchy rough keratotic skin eruptions on hands, feet, elbows & knees for (1) year. The lesions are also disfiguring & embarrassing; the lesions are slow in course

Race

- Rofa'a tribe (Aljazera state)

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Figure 2: Lesions on hands and feet.



Figure 3: Lesions on dorsa of hands.



Figure 4: Lesions on knee.

Profession:

- Merchant

P.H

- PH of atopy, rhinitis. This condition was diagnosed as eczema abroad, In the Sudan he was diagnosed as psoriasis

Family history

- Married (wife is a cousin), he has [3] kids who are not affected

Clinical examination

- There is no similar condition in his small family or grand family.
- The parents of the patient are also cousins.
- The lesions are distributed mostly on the dorsum of hands, feet & interdigital spaces in the form of papules, nodules having skin-color appearance with rough keratotic surface (Figure 2,3), the lesions on the knees & elbows are in the form of papules, nodules & others co-alesced to form plaques (Figure 4,5).
- The scalp, hair, nails & mucous membranes are free
- No specific hobbies or habits
- Differential Diagnosis [1-7].

Thought of

- Darier's disease
- Ackrokeratosis verruciformis of Hopf
- Common warts
- Ackrokerato-elastodoidosis
- Epidermodysplasia verruciformis
- Reiter's disease
- Erythema elevatum diutinum
- Psoriasis

Histopathology (Biopsy)

- Epidermal findings: - Hyperkeratosis prominent granular layer, acanthosis and church spires papillomatosis elongated rete ridges (Figure 6).
- Other positive findings: High total immunoglobulin (IgE).

Final diagnosis & management [1,5,9-12]

- Ackrokeratosis verruciformis of Hopf+- Atopic dermatitis (coincidence), (syndrome)



Figure 5: Lesions on elbows.

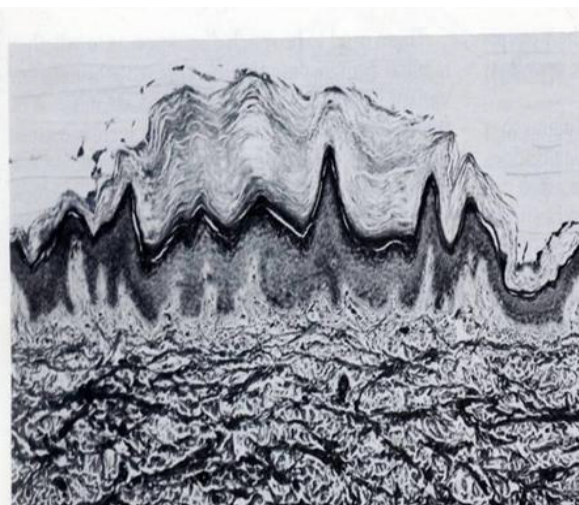


Figure 6: Biopsy of Acrokeratosis verruciformis of Hopf.

Management [5,8,12]

- Educational explanation to the patient of the disease nature and the outcome

Treatment

- Topical applications (keratolytics), e mild improvement
- Systemic vitamin-A acid orally after doing required investigations.
- PUVA was irritant (stopped).
- The patient is to take other modalities (eg. Cryo ,laser)

Conclusions & Recommendations

Rare problems in dermatology but can be encountered.

Dermatology is specialty of differential diagnosis. Since many diseases are not liable for radical therapy, approach should include management [11,13]. Clinical examination should be thoroughly done, in a naked patient (full privacy) in a day light, or a day-lit room (keep it as personal motto) [11].

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