

# Acrokeratosis Verruciformis of Hopf: A Rare Case Report

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## ABSTRACT

Acrokeratosis verruciformis (AKV) of Hopf is a rare autosomal dominant genodermatosis characterised by multiple hyperkeratotic, skin-coloured, verrucous papules on hands and feet. We present a 4-year-old female with itchy lesions around the right ankle since the age of 3 months without any family history of AKV. Clinical investigation showed multiple hyperpigmented, hyperkeratotic grouped papules and histopathology showed hyperkeratosis, acanthosis, papillomatosis with church-spire like elevation of the epidermis without signs of dyskeratosis and mild lymphocytic infiltrate in the dermis. Treatment with 0.05% topical tretinoin showed improvement within two weeks.

**Keywords:** Histopathology, differential diagnosis, follow-up, genodermatosis, Tretinoin, treatment

## INTRODUCTION

Acrokeratosis verruciformis (AKV) of Hopf is a rare autosomal dominant genodermatosis (an inherited genetic skin disorder) initially described by Hopf in 1931. [1] Multiple hyperkeratotic skin-coloured verrucous papules on the dorsum of hands, feet, knees, elbows, and forearms are the characteristic features of AKV. [2,3] Leukonychia, thickened nail plates, and longitudinal nail ridges are other typical features. [4] It usually presents at birth or in early childhood, however in certain cases presentation may be delayed up to fifth decade. Both the sexes are equally affected by AKV without any gender predilection. [3] No intervention is required to treat the

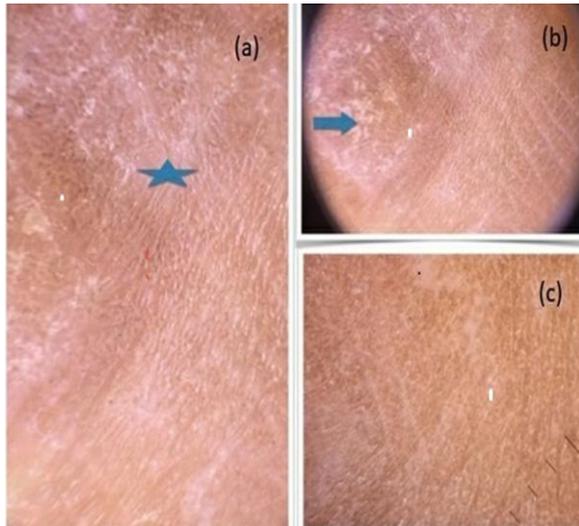
disorder until the patient is symptomatic. Superficial ablation, retinoic acid, cryotherapy, carbon dioxide laser therapy are the treatment modalities when the condition aggravates. [3,4] Here, we present a patient with AKV who was successfully managed with tretinoin.

## CASE REPORT

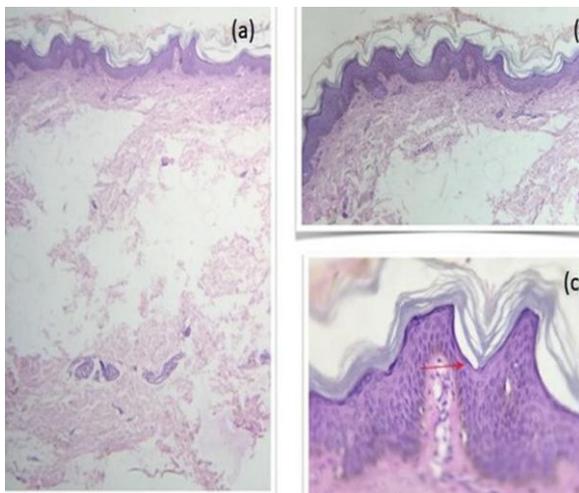
A 4-year-old female child presented to the Department of Dermatology with a chief complaint of itchy lesions around the right ankle since the age of three months with no history of similar complaints in the family.

The dermatological investigation found multiple hyper-pigmented, hyperkeratotic grouped papules on the dorsum of the right foot extending to the lateral and posterior aspect of the ankle (Figure 1 and 3a). Other typical findings include multiple, tiny skin-coloured papules on the extensor aspect of both forearms and fish-like scales on extensor aspect of both legs suggestive of lichen spinulosus and ichthyosis vulgaris. Differentials considered were verruca vulgaris, acral darier's disease, and epidermodysplasia verruciformis.

Skin biopsy was performed and histopathological evaluation showed hyperkeratosis, acanthosis, papillomatosis with church-spire like elevation of the epidermis without signs of dyskeratosis and mild lymphocytic infiltrate in the dermis (Figure 2). Summing up both dermatological and histopathological findings confirmed AKV of Hopf in the patient.



**Figure 1:** a) Clinical photograph showing dermoscopic features of the lesions over the ankle. a) & c) irregular white areas (blue star); b) White network (blue arrow)



**Figure 2:** a) Scanning view & b) Lower power view (10x): Hyperkeratosis, acanthosis, papillomatosis with church-spire like elevation; c) High-power (40x) Magnified view of the epidermis without signs of dyskeratosis and mild lymphocytic infiltrate in the dermis with church-spire like elevation (red arrow)



**Figure 3:** Before treatment a) Clinical photograph showing multiple skin-colored papules on the dorsum of the right foot of the patient extending to the lateral and posterior aspect of the ankle; After treatment b) Clinical photograph showing improvement within two weeks after using topical tretinoin

Our therapeutic attempt with topical 0.05% tretinoin showed a significant improvement within two weeks (Figure 3b). Thus, the patient might have stopped the treatment and not appeared for further follow-up sessions.

## DISCUSSION

AKV is an underlying chronic autosomal dominant mutation without remission. Studies have reported lesions of AKV usually manifests as brown or skin-colored flat-topped papules on the interphalangeal joints of the hands and feet and less often appears on the legs, knees, arms, elbows and other parts of the body. [1,5,6] Similarly, in our case, the disease features as hyperkeratotic grouped papules on the dorsum of right foot.

The clinical resemblance of multiple hyperpigmented hyperkeratotic grouped papules in our patient led us to consider verruca vulgaris, Darier's disease, and epidermodysplasia verruciformis as differential diagnoses. Hence, the histological investigation was carried out for definitive diagnosis. Histopathological investigation of the biopsied lesion found a typical church-spire like pattern- a classical feature of AKV without signs of dyskeratosis and mild lymphocytic infiltrate made us confirm and rule out other mimicking diseases. While Darier's disease shows a suprabasal dyskeratosis with corps rond, [6] verruca vulgaris shows hypergranulosis with coarse keratohyalin granules and koilocytes, [7] epidermodysplasia verruciformis shows hyperkeratosis and distinct intracytoplasmic inclusion bodies in the epidermis [8] as characteristic features on histopathological examination.

Literature also reported that AKV and darier's disease were allelic disorders and has clinical similarities. Although the exact aetiology is not known, it is postulated that P602L mutation in the ATP2A2 gene that is allelic to darier disease on chromosome 12q24 can cause AKV. [4] The mutation restricts the transport of calcium in

the sarcoendoplasmic reticulum calcium ATPase and causes hyperkeratinisation and leads to a genodermatosis. [9] While other study suggested rather than ATP2A2 mutation, missense mutations in other genes were responsible for AKV. It was also stated that both Darier's disease and AKV exist in the same patient. [10,11]

Although superficial ablation is effective to treat AKV, it not recommended due to quite a high recurrence rate. Studies [3,5] opted for keratolytic agents such as salicylic acid and topical corticosteroids with cryotherapy also reported no improvement. Previous studies found varied results in few cases with oral retinoids, however, acitretin showed significant improvement. [12] In our case, 0.05% tretinoin showed a significant improvement within two weeks in the patient; nevertheless, there is no response later on for follow-ups.

## CONCLUSION

In the Indian context a very few cases of AKV are reported, the disorder was therefore presented due to its rarity. The case report highlights that the patient responded to the topical application of tretinoin and improved the condition within two weeks. Furthermore as a future perspective, AKV of Hopf may rarely transform to squamous cell carcinoma. Therefore, dermatology clinical professionals should cautiously monitor and educate patients diagnosed with AKV.

## ACKNOWLEDGEMENTS

The authors thank the patient's family for allowing them to share this case.

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How to cite this article: Chaitanya NT. Acrokeratosis verruciformis of Hopf: a rare case report. International Journal of Research and Review. 2020; 7(7): 94-96.

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