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A Rare Case Report of Non-Familial Acrokeratosis Verruciformis of Hopf

Dr. Shaik Humera[1], Dr. Deepak Tony Raj[2]

MBBS, Post Graduate, Department of DVL, Fathima Institute of Medical Sciences, Kadapa

MBBS, MD, Assistant Professor, Department of DVL, Fathima Institute of Medical Sciences, Kadapa

Abstract: ***Acrokeratosis verruciformis of Hopf is a rare genodermatosis with an autosomal dominant inheritance. It is a disorder of keratinization, characterized by multiple, flat-topped, skin-colored keratotic lesions resembling plane warts typically observed on the dorsum of the hands and feet. Histopathologically, the lesion shows hyperkeratosis, acanthosis, and papillomatosis, mimicking a "church spire", and a thickened granular layer. It arises in early life, often at birth or infancy. Herein, we report on a rare sporadic case of acrokeratosis verruciformis of Hopf. A 50 year old male presented with asymptomatic skin lesions on the extremities which are spreading bilaterally and symmetrically since six years multiple hyper-pigmented, hyperkeratotic discrete papules on the dorsum of hands and feet extending on to lower one third of tibia were present. His past and personal history is not significant. Family history is unremarkable. Histopathological examination showed typical findings of acrokeratosis verruciformis of Hopf. Our case is unique in that the patient had no familial history of similar skin lesions.[2]***

Keywords: Genodermatosis**,** Acrokeratosis verruciformis of Hopf, Non-familial acrokeratosis verruciformis ,keratotic lesions, Church spire

1. Introduction

Acrokeratosis verruciformis (AKV) is a rare hyperkeratotic genodermatosis, which was first described by Hopf in 1931[1]. AKV is a rare autosomal dominant genodermatosis. characterized by keratotic lesions on the dorsum of hands and feet. It presents at birth or appears in early childhood in familial cases but the onset may be delayed until fifth decade in sporadic cases.[3]These lesions are multiple flat topped papules on dorsum of hands and feet but may extend on to knees, elbows, forearms and other parts of body. Palms show thickening with punctate pits. Nails are thickened with whitish discolouration. Transformation of skin lessons to SCC has been reported.

2. Case Report

A 50 year old male presented with asymptomatic skin lesions on the extremities which are spreading bilaterally and symmetrically since six years. The lesions started as skin coloured, slowly enlarging with dark discolouration. His past and personal history is not significant. Family history is unremarkable.

Systemic examination did not reveal any abnormality.

On local examination multiple hyper-pigmented, hyperkeratotic discrete papules on the dorsum of hands and feet extending on to lower one third of tibia were present. Scalp, forehead, oral mucosa, thorax, axillae , palms, nails and soles were normal.

Investigations: Hb, RBS, viral serology were normal. Punch biopsy was done from lesion of lower third of tibia and sent for HPE.

HPE: Features of AKV as shown below.

He was started on Tretinoin 0.025% cream for lesions on hands and feet and with mometasone furoate cream and liquid paraffin for thickened patches on feet . He showed mild decrease in thickness of lesions after two weeks and is under follow up.

3. Discussion

AKV is a rare heritable, hyperkeratotic genodermatosis of unknown etiology. Mutation in ATP2A2 gene[4] is associated with this disorder as in Darriers disease. Both are allelic disorders. AKV is characterized by multiple, symmetrical, localized, flat, skin coloured wart like lesions on dorsum of hands and feet. Forehead, scalp, oral mucosa and flexures are never affected. Lesions may be finely granular to lichenified papules. Interruption of dermal ridges with punctate pits may be seen in finger pads and palms. Nails show whitish discoloration, thickening and longitudinal ridges with breakage at distal ends. There are 2 types of AKV. Classical and Sporadic types. In classical AKV age of onset is during childhood while it is later in sporadic AKV.[2] Palmar and plantar keratosis has been reported in classical AKV but not in sporadic type.

Dhitava et al.,[4] reported a novel P602 L mutation within the ATP binding domain of ATP2A2 in classical AKV.

Berk et al.,[5] reported an A698V codon Change in ATP2A2 in sporadic cases.

Treatment
Superficial ablation of skin lesions.
Application of retinoic acid, cryotherapy or destructive lasers such as Co2 or Nd:YAG have also been tried .

4. Conclusion

The case is reported due to its rare and sporadic occurrence.[3,5]

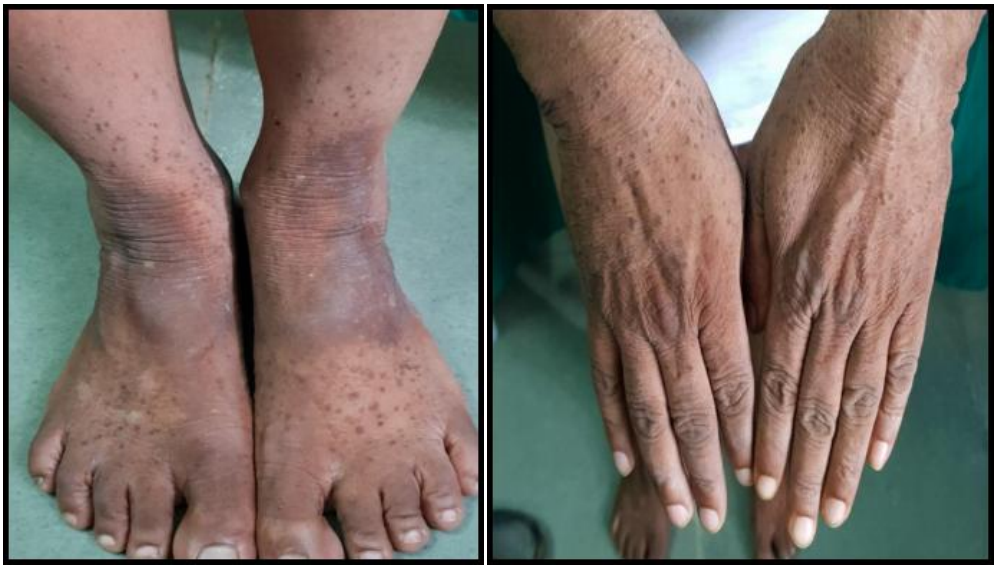

Figure 1 & 2: Multiple hyper-pigmented, hyperkeratotic discrete papules on the dorsum of hands and feet extending on to lower one third of tibia were present.

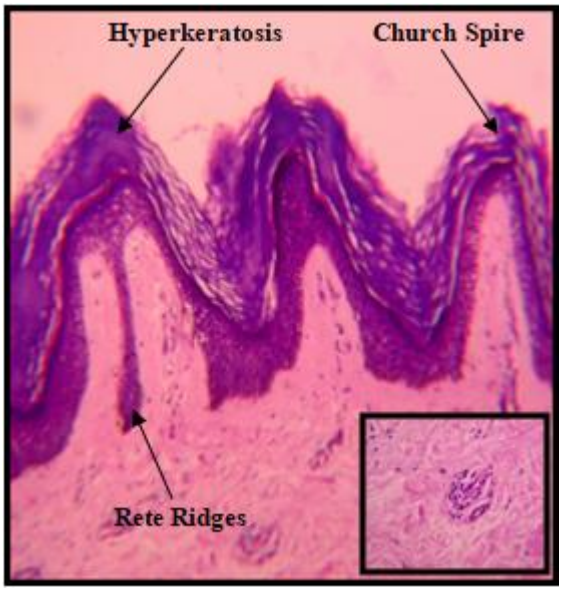

Image 1: Microphotograph of tissue section of skin showing hyperkeratosis, papillomatosis, irregular acanthosis, elongated rete ridges & church spire morphology.

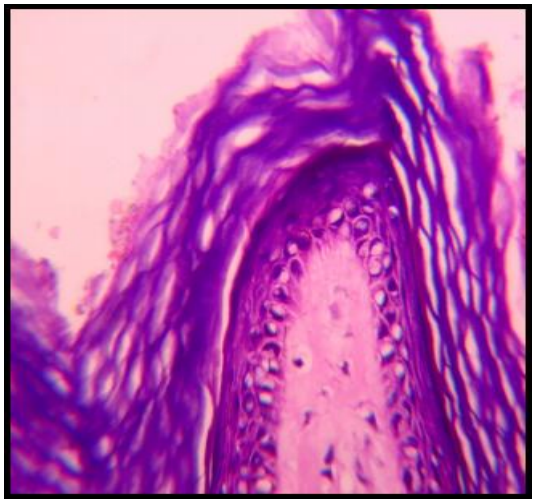

Image 2: Tissue section of skin showing hyperkeratosis, papillomatosis & church spire morphology

References

[1] Hopf G. Über eine bisher nicht beschriebene disseminierte. Keratose (Akrokeratosis verruciformis) *Dermatologische Zeitschrift.* 1931;60:227–250.
[2] 2.Bang CH, Kim HS, Park YM, Kim HO, Lee JY. Nonfamilial Acrokeratosis Verruciformis of Hopf. Ann Dermatol 2011; 23: S1. S61-3.
[3] Alain H. Acantholytic disorders of the skin: Darier-White disease, acrokeratosis verruciformis, Grover disease, and Hailey-hailey disease. In: Wolff K, Goldsmith LA, Katz SI, Glichrest BA, Paller AS, Leffell DJ, editors. *Fitzpatrick's dermatology in general medicine.* 7th ed. New York: McGraw-Hill; 2006. p. 437.
[4] Dhitavat J, Macfarlane S, Dode L, Leslie N, Sakuntabhai A, MacSween R, Saihan E, Hovnanian A. Acrokeratosis verruciformis of Hopf is caused by mutation in ATP2A2: evidence that it is allelic to Darier'sdisease. J Invest Dermatol 2003; 120:229-32.
[5] Berk DR, Taube JM, Bruckner AL, Lane AT. A sporadic patient with acrokeratosis verruciformis of Hopf and a novel ATP2A2 mutation. *Br J Dermatol.* 2010;163:653–654.