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Case Report

Acrokeratosis verruciformis of Hopf: A rare case report

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ABSTRACT

Acrokeratosis verruciformis of Hopf (AKV) is a rare cutaneous autosomal dominant genodermatosis described by Hopf in 1931. It usually presents at birth or may appear as late as 5^{th} decade of life. It is characterized by multiple hyperkeratotic, verrucous papules/plaques to multiple planar wart like lesions on dorsal aspects of hand and feet. Due to rarity of the case, we present a case of AKV in a 60-year-old male that was diagnosed on histopathology. We the authors have attempted to describe the histological features of AKV with differential diagnosis of these lesions through review of literature.

1. Introduction

Acrokeratosis verruciformis of Hopf (AKV) is a rare cutaneous autosomal dominant genodermatosis (inherited genetic skin disorder) described by Hopf in 1931.[1] It affects both sexes equally with no gender predilection.[2] There are two types of AKV based on the age of onset. They are classical AKV which is seen in childhood and sporadic AKV seen in older age group around 5^{th} decade.[3] Clinical presentation may vary from multiple hyperkeratotic skin coloured verucous papules/plaques of hand and feet to multiple planar wart like lesions on hands and feet that may extend to knees, elbows and forearm.[1,4]

2. Case Report

A 60-year-old male presented to Dermatology department with chief complaints of itchy hyperkeratotic papular/plaque lesions around the right ankle since 8 years. The lesions were hyperpigmented and had exacerbations on and off (Figure 1 a, b). Leukonychia, thickened nail plates and longitudinal nail ridges and nicks are other typical nail changes that are frequently reported in association with AKV.[5,6] Our case showed nail plate thickening with longitudinal nail ridges. (Figure 1 c) There was no history of similar complaints in the family. Histopathology of the lesion showed epidermis showing hyperkeratosis, acanthosis, hypergranulosis with papillomatous elevation resembling "church spires". The reteridges are elongated and extend to a uniform level with dermis appearing unremarkable with normal fibrocollagenous tissues and adnexal structures (Figure 2 a,b,c). Thus corroborating the clinical presentation and characteristics histopathology findings diagnosis of Acrokeratitis Veruciformis of Hopf was rendered.

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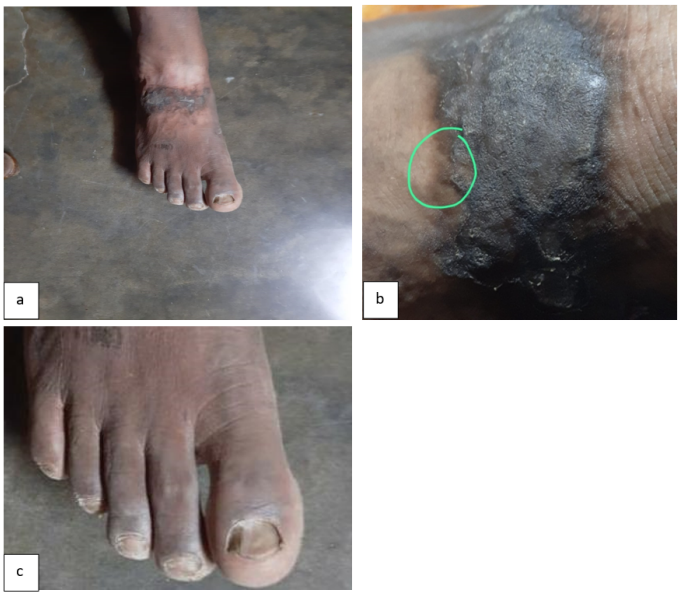

Fig. 1: a,b): Multiple hyperkeratotic, verrucous papules/plaque like lesions on dorsal aspects of hand and feet; **c**): Thickening of the nail plate with longitudinal ridges seen in the big toe

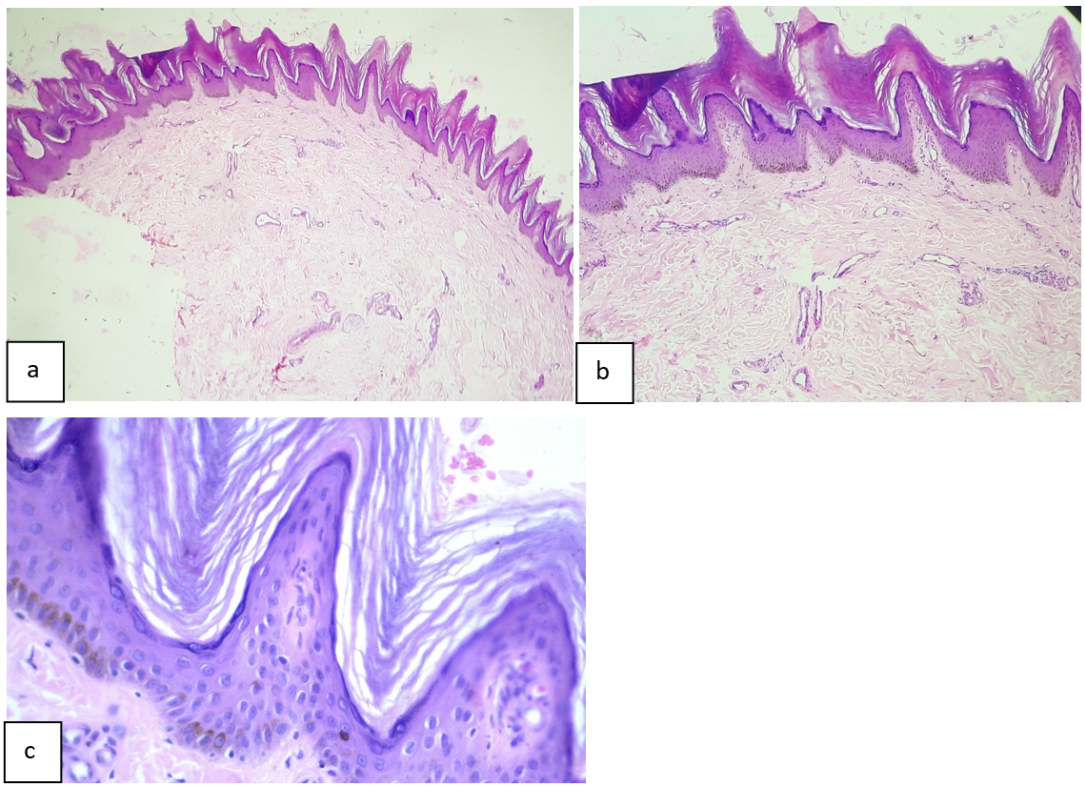

Fig. 2: a): Scanner view: 40x Showing Hyperkeratosis, acanthosis and Papillomatosis (church spire like elevation); **b**): Low powe view 100X- Showing acanthosis, hyperkeratosis, hypergranulosis and papillomatosis Dermis appearing unremarkable; **c):** High power 400x- showing characteristics papillomatosis (Church spire like elavations)

3. Discussion

Acrokeratitis veruciformis of Hopf (AKV) is an autosomal dominant genetic disorder of unknown etiology. The disease follows a chronic course with no spontaneous remission. It has an autosomal dominant pattern of inheritance with incomplete penetrance that explains the diificulty of finding similar cases in the family. Clinically AKV manifests as flat topped, polygonal, papules and verrucous plaques and this clinical presentation led us to consider verruca vulgaris, Darriers disease, seborric keratosis and epidermodysplasia veruciformis as differential diagnosis . The disease doesn't affect the sebaceous areas –frontal scalp,flexures or oral mucosa.[5] Diagnosis of AKV however is clinched by characteristic histopathological features that include acanthosis, hyperkeratosis, hypergranulosis without parakeratosis and papillomatosis (circumscribed epidermal elavations known as church spires). Other differentials were ruled out on basis of their histopathological findings that was not seen in our case. Darrier's disease shows a suprabasal acantholysis with corps ronds,[7] veruca vulgaris shows papillomatous epidermis, hypergranulosis, inward bending of rete ridges with coarse keratohyaline granules and koilocytes,[8] Epidermodysplasia verruciformis shows hyperkeratosis and distinct intracytoplasmic inclusion bodies in the epidermis.[9] Absence of horn cyst and basaloid cells ruled out the possibility of seborrheic keratosis.[3]

Review of literatures have reported that AKV as a variant of Darriers disease due to similarities in clinical presentation and inheritance pattern .Although the exact etiology is not known it is postulated that P602L mutation in the ATP2A2 gene that is alleic to Darriers disease on chromosome 12q24 can cause AKV.[10] The mutation restricts the calcium in the sarcoplasmic reticulum calcium ATPase and cause hyperkeratinision that leads to genodermatosis.[11] Other studies postulated that rather than ATP2A2 mutation, missense mutation in other genes were responsible for AKV. It was also stated that both occur in the same patient,[12,13] however Darriers disease shows dyskeratotic cells, has a predilection for sebaceous areas and affects oral mucosa which goes against the diagnosis of AKV.

There are various treatment modalities for AKV as described in literatures, however no intervention is required until the patient is symptomatic. Although superficial ablation is effective to treat AKV, it is discouraged due to high recurrence rate. Certain studies have reported no improvement with the use of keratolytics such as salicylic acid and topical corticosteroids with cryotherapy.[14,15] However, with the Use of oral retinoids, acitretin showed marked improvement.[16] Our patient was given 0.05% tritenoin that showed improvement and is on continous follow up with no recurance since 2 months.

4. Conclusion

Very few cases of AKV have been reported in Indian context, and the disease is reported because of its rarity. AKV of Hopf may transform to squamous cell carcinoma as reported in few literatures, therefore dermatologist should educate the patient as well as monitor the patients routinely from time to time. Additional case reports of AKV will help to recognize the risk factors in this condition and genetic studies of sporadic cases of AKV should be conducted for better management of the patient.

5. Source of Funding

None.

6. Conflict of Interest

None.

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