Acrokeratosis Verruciformis of Hopf: An Unusual Presentation

Sir,

A 23-year-old Nepalese female patient presented with complaints of multiple raised warty lesions over bilateral feet, legs, hands, forearm, inguinal region, face, and scalp for 10 years. Similar lesions were present in her sibling, but were few in number and limited to hands and feet. On examination, there were multiple brownish verrucous papules and plaques over the bilateral dorsum of feet, planter aspect of feet, legs, inguinal region, hands, forearm, face, and scalp. Few were discrete and few coalescing [Figure 1a and b]. Toe nails of bilateral foot were dystrophic [Figure 2a], whereas mucous membrane was not involved. A skin biopsy was performed, which revealed hyperorthokeratosis, hypergranulosis, markedly acanthotic epidermis, and papillomatosis with epidermal upgrowths resembling “church spires” [Figure 2b]. Parakeratosis and koilocytes were not found. Dermis showed mild perivascular infiltrate of lymphocytes. Genetic studies for Acrokeratosis verruciformis of Hopf (AKV) and Epidermodysplasia verruciformis (EDV) could not be performed as such facilities are not available in our setting.

Based on clinical and histopathological examination, a diagnosis of AKV was made. Treatment was started with oral Isotretinoin 40 mg/day and topical salicylic acid ointment. Patient was followed up at regular intervals and lesions improved after 3 months of therapy as shown in [Figure 3a and b].

AKV originally described by Hopf in 1931 is a rare autosomal dominant disorder of keratinization.[1] Classical AKV, which commonly occurs during childhood, has typical morphology at dorsum of hands and feet whereas sporadic AKV, which occurs much later than that of classical AKV, can affect other sites like face, scalp, and trunk. In classical AKV, positive family history with palmar pits and nail changes are seen.[2] Our patient had onset at 13 years of age and had a positive family history along with nail changes.

AKV classically presents as dry, rough, skin-colored or reddish-brown, flat-topped, or warty papules resembling flat warts on the dorsum of the hands and feet, on the knees, elbows, forearms, or lower legs.[2] Sebaceous areas, like the frontal scalp, flexural surfaces, and oral mucosa are spared.[3] Previously reported case by Andrade TC et al.[4] also presented with verrucous exuberant lesions on the hand and feet, however there was no involvement of sebaceous areas as seen in our patient, which is a rare presentation. There may be thickening of palmar skin and punctate keratosis, pits, or punctiform breaks in dermatoglyphics.[2] Nail changes include whitish discoloration, thickening and longitudinal ridges, with breakage at the distal ends.[5]

Histopathological features include papillomatosis (circumscribed epidermal elevations known as “church spires”), acanthosis, hyperkeratosis, and hypergranulosis without parakeratosis.[3] Histopathology of our patient revealed hyperorthokeratosis, hypergranulosis, markedly acanthotic epidermis, papillomatosis with epidermal upgrowths resembling “church spires,” which are in favour of AKV. Epidermodysplasia verruciformis was excluded as histopathology didn’t reveal parakeratosis and koilocytes, and nail changes was in favour of AKV. Verruca vulgaris was excluded as histopathology didn’t reveal koilocytes and vertical tiers of parakeratosis. Similarly, Darier disease was ruled out as there are no features of dyskeratosis and acantholysis. Treatment

How to cite this article: Lohanee K, Thapa DP. Acrokeratosis verruciformis of hopf: An unusual presentation. Indian Dermatol Online J 2021;12:928-9.

Received: 27-Oct-2020. Revised: 04-Mar-2021. Accepted: 23-Mar-2021. Published: 22-Nov-2021.
modalities are not completely curative in AKV although topical and systemic retinoids, cryotherapy, or lasers can be used in the treatment. Effective treatment option of AKV is superficial ablation. Our patient was treated with oral Isotretinoin and topical salicylic acid ointment. Counseling was done regarding the necessity of long-term follow-up and biopsy of any atypical lesion as there is a rare chance of transformation to squamous cell carcinoma.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her names and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

**References**

1. Nair PA. Acrokeratosis verruciformis of hopf along lines of blaschko. Indian J Dermatol 2013;58:406.
2. Diwan NG, Jivani NB, Nair PA. Acrokeratosis verruciformis of hopf clinically mimicking epidermodysplasia verruciformis. Indian J Dermatol 2016;61:237.
3. Andrade TC, Silva GV, Silva TM, Pinto AC, Nunes AJ, Martelli AC. Acrokeratosis verruciformis of Hopf - Case report. An Bras Dermatol 2016;91:639-41.
4. Andrade TC, Silva GV, Silva TM, Pinto AC, Nunes AJ, Martelli AC. Acrokeratosis verruciformis of Hopf - Case report. An Bras Dermatol 2016;91:639-41.
5. Patel N, Diwan N, Nair PA. Nonfamilial acrokeratosis verruciformis of Hopf. Indian Dermatol Online J 2015;6:110-2.
6. Serarslan G, Balci DD, Homan S. Acitretin treatment in acrokeratosis verruciformis of Hopf. J Dermatolog Treat 2007;18:123-5.