



ACROKERATOSIS VERRUCIFORMIS OF HOPF ASSOCIATED WITH DIABETES MELLITUS – A CASE REPORT

Dermatology

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KEYWORDS

INTRODUCTION:

Acrokeratosis verruciformis of Hopf is a rare genodermatosis characterized by keratotic lesions on the dorsum of the hands and feet. Lesions are usually present from birth or early childhood, but some cases in adult life are reported. It affects both sexes. It usually presents at birth or appears in early childhood; however the onset may be delayed until fifth decade. Patient will present with complaints of hyperkeratotic, skin colored, flat, verrucous papules appear on the dorsal aspect of hands and feet, and sometimes on the forearm, elbows, knees, and face as well.

CASE REPORT:

A 49 years old male came to skin OPD with the complaints of pigmented skin lesions over dorsum of hand and feet for past two years. Initially lesion started on the dorsum aspect of the hand and gradually progressed to involve dorsum aspect of feet and forehead. No history of any complaints like itching, burning, or pain present. No history of similar complaints in any of the family members. No seasonal variation or any other systemic complaints present. He is a known case of diabetes mellitus for past one year and on treatment. He is also a known case of seizure disorder for past 10 years and on medication. Dermatological examination showed multiple brownish flat-topped warty plaques and papules seen over B/L dorsum aspect of the hands and feet, and forehead. Palms, soles, and oral mucosa were normal. Acanthosis nigricans present over the neck. 4mm punch biopsy was taken from dorsal aspect of left hand. Histopathology showed hyperkeratosis, acanthosis, and papillomatosis. The papillomatosis is associated with circumscribed elevation of the overlying epidermis resembling "church spire".

DISCUSSION:

Acrokeratosis verruciformis of Hopf (AVH) was first described by Hopf in 1931, is a rare genetic disease transmitted as an autosomal dominant trait. A mutation in the ATP2A2 gene has been shown to be associated with this disorder as in Darier's disease. Many patients with Darier's disease have skin lesions indistinguishable from AVH both clinically and histologically. Such lesions show dyskeratosis and lacunae as in Darier's disease, but these changes are not seen when they occur independent of Darier's disease.

Patients with AVH usually present to skin OPD with hyperkeratotic, skin colored, flat, verrucous papules seen over the dorsal aspect of hand and feet. They resemble a plane wart. Punctate keratoses are present over the palms and soles.

Transformation to SCC has been reported. AVH can also be associated with Haily-Haily disease, multiple keratoacanthoma, congenital poikiloderma, steatocystoma, naevoid basal cell carcinoma syndrome, and hypertrophic lichen planus.

Histopathology show considerable hyperkeratosis, an increased in thickness of granular layer, and acanthosis. In addition, there is slight papillomatosis associated with circumscribed elevations of the epidermis resembling church spires.

Differential diagnosis includes plane wart, epidermodysplasia verruciformis, seborrheic keratosis, Darier's disease, stucco keratosis. Treatment includes topical tretinoin, co, laser ablation, shave excision, liquid nitrogen therapy. But the lesions are likely to recur.

CONCLUSION:

This case has been diagnosed as AVH both clinically and histologically. This case has been reported for its rare incidence.

FIGURE 1:



FIGURE 2:

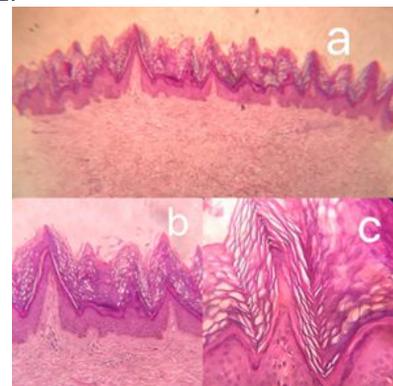


Figure 1: Multiple pigmented flat-topped warty plaques and papules seen over B/L dorsum aspect of the hands and feet, and forehead.

Figure 2a: H&E stained section of skin 10x showing epithelial hyperplasia, hyperkeratosis and papillomatosis.

Figure 2b: 20x view showing epithelial hyperplasia, exuberant hyperkeratosis and papillomatosis.

Figure 2c: 40x view showing Circumscribed Elevation of Epidermis Resembling 'Church Spires'

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