

# CASE REPORT

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## ACROKERATOSES VERRUCIFORMIS OF HOPF: AN ASSOCIATION WITH POLYMORPHIC LIGHT ERUPTION

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**ABSTRACT:** Acrokeratoses verruciformis of HOPF (AKV) is a rare Genodermatoses presenting as multiple plane wart-like lesions symmetrically distributed on dorsum of hands and feet. Its pathogenesis is unknown. A case of 45 years old male is described with warty papules and plaques on dorsa of hands since 3 years with similar lesions on nape of neck and anterior aspect of chest with erythematous plaque in a butterfly distribution over the face. With history of photosensitivity. Histopathology showed changes of AKV. We are reporting unusual association of AKV with polymorphic light eruption (PLE).

**KEY WORDS:** Acrokeratoses verruciformis of HOPF (AKV), Genodermatoses, PLE.

**INTRODUCTION;** Acrokeratoses verruciformis of Hopf (AKV) is a rare autosomal dominant cutaneous disorder described by Hopf in 1931<sup>1</sup>. It is characterized by multiple flat-topped skin colored or fleshy, dull red to brown colour keratotic lesions resembling plane warts mainly observed on the dorsum of hands and feet. Lesions are usually present at birth but may appear late in infancy or at puberty. In some cases, onset may be delayed until the fifth decade of life. It has unknown aetiology, affecting both sexes but is more common in males as compared to females with a ratio of 5: 1.3<sup>2</sup>. It can occur as isolated autosomal dominant trait, with mutations in ATPA2, the gene implicated in Darier's disease is found to be involved. Occurrence of similar lesions in other conditions suggest multiple aetiologies.

A variety of skin disorders not actually caused by UVR may sometimes be worsened by it like Darier's disease, transient acantholytic dermatosis (Grover's disease)<sup>3</sup>. Photosensitivity and polymorphic light eruption appears to have a genetic basis<sup>4, 5</sup>. Recent gene microarray studies have further suggested that an inherited deficiency of apoptotic cell clearance genes contribute, such clearance apparently being necessary to permit immune suppression in polymorphic light eruption<sup>6</sup>.

**CASE REPORT:** A 45 year old male presented to skin opd with warty lesions over the dorsa of hands since 3 years. Initially, few lesions started over the dorsum of hands, which gradually increased in number close to earlier lesions and later lesions started over the nape of the neck and anterior part of the chest. History of involvement of butterfly area of the face present associated increase in reddening and burning sensation of the face lesions on exposure to sunlight. No previous history suggestive of photosensitivity present before 3 years. No history of similar lesions in the family. No history of any complaints in the form of itching, or pain present. No history of other systemic complaints present.

On examination hyperkeratotic skin coloured or slightly hyper pigmented flat verrucous papules on the dorsal aspect of the hands, forearms, anterior aspect of chest and few lesions on nape of the neck. The papules closely resembled flat warts. They are closely grouped but not confluent.

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Erythematous plaque present over bridge of the nose and malar areas present. Nails and oral cavity were normal.

Biopsy taken from dorsum of the hand showed marked hyperkeratosis, hypergranulosis, acanthosis and papillomatosis. The papillomatosis is associated with circumscribed elevation of the overlying epidermis resembling church spires.

**DISCUSSION:** AKV is a disorder of keratinisation, a Genodermatoses with unknown aetiology. Acrokeratoses verruciformis of Hopf has an autosomal dominant mode of transmission. This has been suggested since 1962, in a large follow-up series by Niedleman and McKusick that described 24 cases in 4 generations in the same family<sup>7</sup>. Sporadic cases can also occur<sup>8</sup>.

It is a rare heritable hyperkeratotic dermatosis characterised by multiple localized symmetrical flat skin colored wart-like lesions mainly on dorsum of hands and feet<sup>9</sup>. Isolated papules may develop on knees, elbows, forearm and other parts of body. Forehead, scalp, flexures and oral mucosa are never affected as reported by Panja<sup>2</sup>. Friction of lesion may cause blister formation.

Lesions may be finely granular to lichenified papules. Interruption of dermal ridges in finger pads and palms are seen with punctate pits. Nails show whitish discoloration, thickening and may have longitudinal ridges breaking at distal ends.

Histopathology, classically shows hyperkeratosis without parakeratosis, acanthosis, papillomatosis without vacuolization which resembles church spire and helps in ruling out other dermatosis.

Clinically, this case was confused with flat warts, seborrhoic keratosis, epidermodysplasia verruciformis, and Darier's disease, which were ruled out by histopathology. Histopathology is indicated in chronic lesions to validate the diagnosis in AKV<sup>10</sup>.

As mentioned above, a variety of skin disorders not actually caused by UVR may sometimes be worsened by it like Darier's disease, transient acantholytic dermatosis (Grover's disease)<sup>3</sup>. Our case had a history of photosensitivity and polymorphic light eruption like lesion. To our knowledge association of photosensitivity with AKV was not reported and upon reviewing the literature association was found between Darier's disease and photosensitivity. Acrokeratoses verruciformis and Darier's disease are allelic disorders. ATP2A2 encoding the SERCA2 pump has been identified as the defective gene in Darier's disease. In 2003, Dhitavat et al identified a heterozygous P602L mutation in the ATP2A2 gene in a family affected with Acrokeratoses verruciformis for 6 generations<sup>11</sup>. This mutation predicts a nonconservative amino acid substitution in the ATP-binding domain of the molecule. The mutation segregates with the disease phenotype in the family and was not found in 50 controls. Moreover, functional analysis of the P602L mutant showed that it has lost its ability to transport Ca<sup>2+</sup>. This result demonstrates loss of function of the SERCA2 mutant in Acrokeratoses verruciformis, thus providing evidence that Acrokeratoses verruciformis and Darier's disease are allelic disorders.

The effective treatment option was superficial ablation, oral retinoids, Shave excision, liquid nitrogen therapy and CO<sub>2</sub> laser ablation with use of sunscreens and protective clothing.

In our case the patient was treated with superficial ablation.

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Appearance of disease in middle age and absent family history was also unusual findings. Nonfamilial cases like this case have been reported<sup>12</sup>. The age of onset of classical AKV is during childhood; however, the onset age of sporadic AKV is much later<sup>13</sup>.

Unusual findings such as association with polymorphic light eruption, appearance of disease in middle age, and absent family history are noteworthy and interesting atypical presentation of the disease which requires to be notified.

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## FIGURES:



Fig 1: Warty papules over the dorsum of hands



Fig 2: Papules over the anterior part of the chest



Fig 3: Papules over the nape of the neck



Fig 4: Erythematous plaque over butterfly distribution

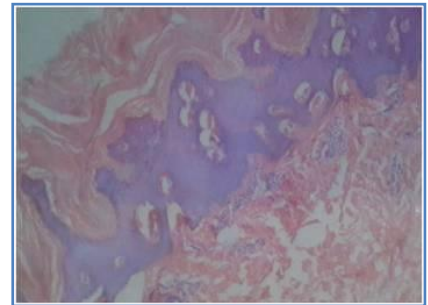
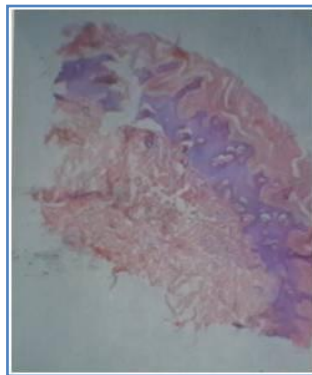
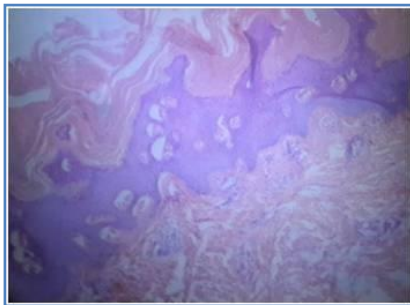


Fig. 5, 6, 7: Histopathology showing hyperkeratosis, hypergranulosis, acanthosis and papillomatosis. The papillomatosis is associated with circumscribed elevation of the overlying epidermis resembling “church spires”.

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