Case presentation: An 18-year-old male patient, a known case of atopic dermatitis since childhood, consulted us for a new onset of upper limb papules of 2 years’ duration. Examination showed multiple flesh-coloured flat-topped papules on the hands, forearms, and knees. Biopsy of the lesions revealed hyperkeratosis, epidermal hyperplasia and papillomatosis, histologically consistent with acrokeratosis verruciformis of Hopf. Clinical improvement was observed with application of topical tretinoin.

Discussion: Acrokeratosis verruciformis of Hopf (AKV) is a rare genodermatosis allelic to Darier’s disease, arising from an autosomal dominant mutation in the ATP2A2 gene, which encodes for the sarcoplasmic reticulum calcium-ATPase 2 (SERCA2) pump. We report this case to highlight the importance of clinical and histopathological examination in differentiating AKV from conditions such as Darier’s disease, lichen nitidus, plane warts, and epidermodyplasia verruciformis.