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Verrucous epidermal nevus

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Case Presentation

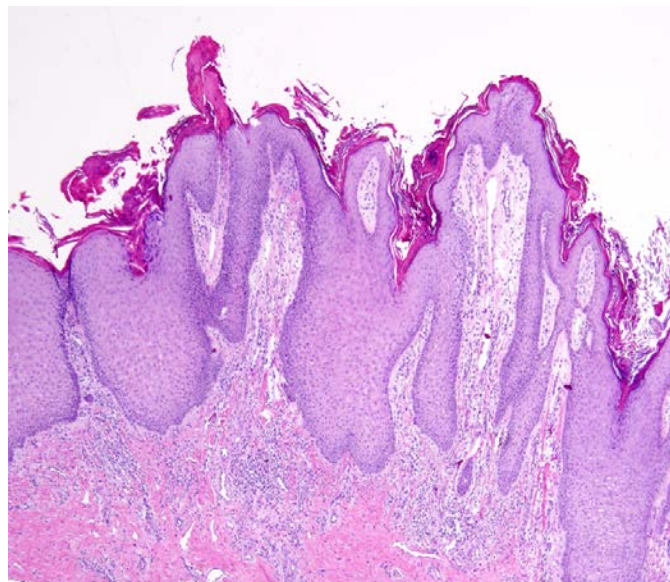
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Abstract

A 64-year-old man presented with a three-year history of an enlarging, pruritic, linear, verrucous plaque on his left lower extremity. Histopathologic examination was consistent with a verrucous epidermal nevus, which is a benign epidermal hamartoma, most commonly observed in the pediatric population. Verrucous epidermal nevi are often refractory to treatment and have high rates of recurrences, causing them to be therapeutic challenges. We review the treatment modalities reported to be effective in verrucous epidermal nevi.



Case synopsis

A 64-year-old man presented to Bellevue Hospital Center in December 2012, for evaluation of verrucous, linear plaques on his left lower extremity. The lesions began three years prior to presentation and had since been enlarging. The lesions were pruritic but non-tender. He was otherwise healthy with a past medical history of hypertension.

Physical Examination: On the left lateral lower extremity, there were discrete, linear, verrucous, pink-to-flesh-colored plaques. The most distal plaque was located on the dorsum of the foot and ankle. This scaly erythematous plaque was excoriated and fissured, with superimposed thick, verrucous, cerebriform, surface change.

Histopathology: There is papillated, epidermal hyperplasia with hypergranulosis, compact orthokeratosis, focal scale-crust, dilated blood vessels in the papillary dermis, and an inflammatory infiltrate of neutrophils, lymphocytes, and eosinophils.

Diagnosis: Adult onset verrucous epidermal nevus.

Discussion

Epidermal nevi represent Blaschkoid hamartomas of the skin that result from mosaic post-zygotic mutations [1]. They are a heterogeneous group of lesions of which verrucous epidermal nevi (VEN) are the most common [2]. VEN can exist as single or multiple lesions, may be of any size, and may occur on any site [3]. Clinically, they appear as skin-colored-to-brown, sharply demarcated, papillomatous papules that coalesce into plaques. The majority of epidermal nevi are either present at birth or occur within the first year of life [2]. Adult onset verrucous epidermal nevi are exceedingly rare, with the previously oldest reported case developing at age 60 years [4].

Discomfort, pruritus, and the undesirable cosmetic appearance of VEN can prompt patients to seek treatment. VEN pose a therapeutic challenge because superficial destruction may result in recurrence, whereas deeper modalities, although more reliable, are limited by the extent of affected areas and a higher risk of post-procedural scars [5]. Various light-based treatments, which include Argon [6], PDT [7], ruby laser, [8] Erbium:YAG [9], and CO₂ laser [6, 10-12] have been reported to be variably effective in VEN. A case series evaluating Erbium:YAG ablation of VEN in 20 patients led to clearance in 15 patients, with a two-year follow-up and recurrent lesions in the remaining five. No scars were noted in any patient and post-procedural dyspigmentation resolved within three months [9]. The extent of lesional hyperkeratosis may be a predictor of response to laser therapy. For hard or keratotic VEN, the CO₂ laser was shown to be more effective than the Argon laser [6]. In a study of 25 patients with epidermal nevi (24 patients with VEN), CO₂ laser vaporization led to complete or almost complete resolution in 92% of patients with soft flattened nevi but only in 33% of patients with keratotic nevi. The majority of recurrent lesions occurred in patients with keratotic nevi. Most patients required a mean number of four sessions. Of those treated, 12.5% had hypertrophic scars [12].

For extensive lesions not amenable to surgery, nonsurgical treatments have included cryotherapy [13], topical tretinoin, topical 5% 5-fluorouracil (5-FU) [14], topical and intralesional glucocorticoids [15], tar preparations, and antihistamines [16]. Topical and intralesional glucocorticoids have not been effective, although they may alleviate erythema and elevation [15]. Combination treatment with topical 0.1% tretinoin and 5% 5-FU under occlusion twice daily for three months followed by daily 5% 5-FU and twice daily 0.1% tretinoin for six months led to appreciable improvement in an extensive inflammatory linear verrucous epidermal nevus (ILVEN) in a 9-year-old boy. However, the lesion recurred after three to four weeks and the patient was maintained on treatment two-to-three times a month [14]. Cryotherapy has been successful in treating small lesions in a series of 11 patients; this treatment resulted in no scars, one recurrence, and one transient post-procedural hypopigmentation [13].

Verrucous epidermal nevi continue to be a therapeutic challenge in dermatology. In our patient, the extent, location, and degree of hyperkeratosis must be considered when pursuing appropriate treatment approaches. He is currently being treated symptomatically with topical glucocorticoids and antihistamines for his pruritus with improvement and is undergoing evaluation for more definitive therapy.

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