



## Verrucous Epidermal Nevus

Verrucous epidermal nevus is also known as linear verrucous epidermal nevus or linear epidermal nevus.

### CLINICAL FEATURES

Verrucous epidermal nevi are characterized by localized or diffuse, closely set, skin-colored, brown, or gray-brown verrucous papules, which may coalesce to form well-demarcated papillomatous plaques . Linear configurations are common on the limbs as is distribution in Blaschko's lines or in relaxed skin tension lines .

Extensive distribution of a verrucous epidermal nevus is termed systemized epidermal nevus. Variants of this type of nevus include nevus unius lateris , epidermal nevi distributed on one-half of the body; and ichthyosis hystrix, epidermal nevi distributed bilaterally. Commonly, systematized nevi take on a transverse configuration on the trunk and linear configuration on the limbs.

An epidermal nevus presenting with pruritus, erythema, and scaling is likely a variant of the epidermal nevus termed an inflammatory linear verrucous epidermal nevus (ILVEN) . These lesions are found most commonly on the buttocks and lower extremities and may resemble linear psoriasis.

## COURSE AND COMPLICATIONS

Linear epidermal nevi tend to appear between birth and adolescence. Although congenital lesions tend not to expand significantly, lesions that present after birth

may expand during childhood, stabilizing in size at or around puberty. Although intertriginous lesions may become macerated and secondarily infected, the majority of epidermal nevi remain quiescent after adolescence. Rare cases of basal cell carcinoma and squamous cell carcinoma arising within epidermal nevi has been reported. This malignant transformation is most common in middle-aged or elderly individuals, though the youngest reported case occurred in a 17-year-old woman.

Epidermal nevi may present in conjunction with other epidermal lesions such as café-au-lait macules, congenital hypopigmented macules, and congenital nevocellular nevi. Extensive lesions may be associated with abnormalities in other systems .

Rarely, patients with epidermal nevi have offspring with epidermolytic hyperkeratosis (EHK), a condition resulting from a mutation in keratin 10 (K10). Paller et al. investigated three families with this occurrence. The analysis of skin samples of parents and offspring with EHK demonstrated a parental mutation in one of the two K10 alleles within the epidermal nevus; non-lesional skin showed no mutation. Offspring showed the same K10 mutation as their parents.<sup>35</sup> The presence of two genetically distinct cell lines in the parents, also known as mosaicism, is a result of postzygotic mutation during embryogenesis. If histopathologic evaluation of an epidermal nevus reveals findings consistent with EHK, the patient is at risk of having a child with EHK. Prenatal counseling may be very important for these patients.

## PATHOLOGY

There are 10 histologic variants of the epidermal nevus, with over 60 percent of lesions

displaying acanthosis, papillomatosis, and hyperkeratosis. Rare variants may have features similar to SKs, with thin, elongated rete ridges; or EHK, with compact orthokeratosis, vacuolization of the granular layer of the epidermis, and large keratohyaline granules within or outside cells.<sup>37</sup> Epidermal hyperkeratosis may be a more common finding in ichthyosis hystrix. ILVEN is a histologically distinct variant of the epidermal nevus that displays a chronic dermal inflammatory infiltrate, psoriasiform epidermal hyperplasia, and alternating bands of ortho- and parakeratosis. In this variant, the granular layer is absent underlying the areas of parakeratosis.

Lichen striatus, linear Darier disease, linear porokeratosis, linear lichen planus, linear psoriasis, and the verrucous stage of incontinentia pigmenti may all have similar clinical presentations as the linear verrucous epidermal nevus. Lichen striatus may mimic ILVEN clinically, but is self-limited and pruritic as compared with epidermal nevi. Histology may be useful in differentiating these entities. Linear Darier disease and linear porokeratosis can be differentiated pathologically with linear Darier disease having the distinct pathologic findings of acantholytic dyskeratosis, and linear porokeratosis having coronoid lamellae. Although some consider linear lichen planus and linear psoriasis to be variants of the ILVEN, the genetics and the immunology has yet to be fully characterized. Incontinentia pigmenti can be distinguished clinically based on the transient nature of this phase and the preceding verrucous phase. Histologically, this entity can be distinguished by its dyskeratosis, pigment incontinence, eosinophilic exocytosis, and basal layer vacuolization.

## TREATMENT

Complete excision of an epidermal nevus to the level of the deep dermis is necessary to prevent recurrences. Based on the size and distribution of the lesion, however, excision may not be an appropriate treatment option. Multiple other surgical and medical treatments are available to treat or destroy these lesions. Laser ablation, electrofulguration, cryotherapy, and medium to full-depth chemical peels may offer partial or full destruction of lesions. Although topical retinoids and calcipotriene offer little relief, these medications can be used as an adjunctive therapy to increase the efficacy of the surgical intervention. Systemic retinoids and anti-psoriatic agents may offer some clinical improvement. There are reports of successful treatment of ILVEN with etanercept. If malignant transformation is confirmed within an epidermal nevus, the lesion should be completely excised.

## Differential Diagnosis of Linear Epidermal Nevus

- • Lichen striatus
- • Linear Darier disease
- • Linear porokeratosis
- • Linear lichen planus
- • Linear psoriasis
- • Incontinentia pigmenti