

What Is This Asymptomatic Linear Eruption Behind a Girl's Right Ear?

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An 8-year-old girl presented with an asymptomatic linear eruption behind her right ear. The eruption was present at birth and had increased in size as she grew older. The lesion had become bumpier in the past 2 years. The patient was otherwise healthy, and the family history was noncontributory.

Physical examination revealed multiple, well-circumscribed, discrete, flesh-colored, smooth-surfaced papules arranged in a linear fashion on the posterior aspect of the right ear and scalp.

The papules were nontender and measured 2 to 3 mm in diameter. The entire lesion measured 2 to 3 cm long, and there was no evidence of erythema or scaling. The rest of the physical examination findings were unremarkable.

What's your diagnosis?



Answer: Linear Epidermal Nevus

Linear epidermal nevus (LEN), also known as linear verrucous epidermal nevus or keratinocytic epidermal nevus, is a hamartoma composed primarily of keratinocytes.

EPIDEMIOLOGY

The prevalence of LEN is approximately 1 in 1000 live births.^{1,2} The sex ratio is approximately equal.^{3,4} The majority of cases occur sporadically, but they also may occur in association with a variety of developmental abnormalities. Familial cases also have been reported.⁴

ETIOPATHOGENESIS

The exact pathogenesis is not known. An epidermal nevus is a hamartoma arising from pluripotent germinative cells in the basal layer of the embryonic epidermis that differentiate into keratinocytes, apocrine glands, eccrine glands, hair follicles, and sebaceous glands.⁵ LEN is due to an abnormal keratinocytic proliferation, resulting in hamartomatous over-

growths of just the epidermis.⁴ The nevus appears to represent populations of ectodermal cells displaying somatic mosaicism that follows the lines of Blaschko. Activating mutations in the fibroblast growth factor receptor 3 gene (*FGFR3*) and in the *PIK3CA* and *HRAS* oncogenes have been identified in some epidermal nevi.^{6,7}

HISTOPATHOLOGY

Histologic findings include epidermal hyperplasia with hyperkeratosis, papillomatosis, and acanthosis with elongation of the rete ridges.⁸

CLINICAL MANIFESTATIONS

Classically, LEN presents as a linear plaque consisting of well-circumscribed, discrete or confluent, pebbly, skin-colored, yellow or brown, closely set or coalescing, noninflammatory papules.³ The lesion usually is present at birth but might arise during the first year of life.^{3,4} Occasionally, it may not be recognized until later in childhood.¹ The nevus may have a white and macerated appearance at birth. The lesion tends to become



thicker, verrucous, and hyperpigmented over time, particularly around puberty.^{3,8} Rarely, it remains hypopigmented. The lesion grows slowly during childhood but usually stops growing and does not extend any further by adolescence.⁹

LEN usually occurs on the trunk, neck, or an extremity. It tends to follow the lines of Blaschko but can occur anywhere on the skin or oral mucosa.^{1,4,10} Typically, the lesions are single and unilateral (nevus unius lateris), although multiple unilateral or bilateral lesions may occur.^{3,11} Extensive bilateral lesions are referred to as *ichthyosis hystrix*.⁹

When LEN occurs in conjunction with extracutaneous (eg, neurologic, musculoskeletal, ocular, renal, cardiovascular) abnormalities, the term *epidermal nevus syndrome* is used. Syndromes associated with LEN include Proteus syndrome, type 2 (segmental) Cowden syndrome, *FGFR3* nevus syndrome, and congenital hemidysplasia with ichthyosiform nevus and limb defects (CHILD) syndrome.⁸

DIAGNOSIS

The diagnosis is mainly clinical, based on the finding of a characteristic linear plaque. A skin biopsy or referral to a dermatologist may be considered if the diagnosis is in doubt.

The differential diagnosis includes inflammatory linear verrucous epidermal nevus, linear nevus sebaceous of Jadassohn, linear nevus comedonicus, incontinentia pigmenti, lichen striatus, linear lichen planus, linear psoriasis, linear porokeratosis, linear Darier disease, allergic contact dermatitis, phytophotodermatitis, and Becker nevus.¹²

COMPLICATIONS AND PROGNOSIS

LEN is cosmetically unsightly and may adversely affect quality of life. Although LEN has no malignant potential per se, there are rare reports of the development of basal cell carcinoma, keratoacanthoma, Bowen disease, squamous cell carcinoma, eccrine poroma, and eccrine porocarcinoma in some of these lesions.^{2,5,13}

The prognosis is good, because the lesions tend to regress or become less noticeable at approximately 40 years of age and thereafter.³

MANAGEMENT

Treatment is not necessary unless for cosmetic reasons. For those who desire treatment for cosmetic purposes, options include topical calcipotriol, dermabrasion, cryotherapy, carbon dioxide laser therapy, erbium:yttrium-aluminum-garnet laser therapy, electrosurgery, and surgical excision.^{1,4} Full-thickness excision provides definitive treatment for small lesions.³ ■

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