Abstract

Epidermal nevi are congenital hamartomatous lesions that are typically present at birth, though they can occur anytime during childhood and rarely appear in adulthood. Inflammatory linear verrucous epidermal nevus (ILVEN) is a rare variant of epidermal verrucous nevus that is four times more common in females than males. This condition is clinically characterized by the appearance of recurrent inflammatory phenomena, with chronic eczematous and psoriasiform aspects, usually unilateral, with pruritus, and it is often refractory to therapy. We report a case of ILVEN syndrome in a 27-year-old female patient who demonstrated very significant clinical worsening during pregnancy.

Introduction

Inflammatory linear verrucous epidermal nevus (ILVEN) is a benign cutaneous hamartoma that consists of erythematous, pruritic, inflammatory plaques that occur as a linear band along a line of Blaschko. ILVEN is a chronic condition, thus patients typically seek medical attention for relief of discomfort along with cosmetic concerns. A few reported therapeutic approaches include topical agents, dermabrasion, cryotherapy, laser therapy and excision. However, no one treatment has been consistently successful. Therapy is often unsatisfactory.

We report a case of ILVEN in a 27-year-old female patient who demonstrated very significant worsening, determined by an increase in thickness from approximately 1 mm to 7 mm, during her second trimester of pregnancy. The average thickness of ILVEN is 1 mm to 3 mm. In our literature search of ILVEN in pregnancy, we found no other cases reporting marked worsening of ILVEN during pregnancy.

Case Report

A 25-year-old African American female, currently in her second trimester of pregnancy, presented with history of a pruritic scaly eruption on the right lower extremity. The lesions first appeared at the age of 5 and gradually progressed in size, with significantly accelerated rates of growth during pregnancy. Aggravating factors for her included pregnancy and sunlight exposure, which resulted in increased pruritus and crusting. Her past medical history was significant for scalp psoriasis, which was managed with shampoos containing tar. The patient was otherwise healthy.

Physical examination revealed a hyperkeratotic linear plaque overlying a base of friable erythema extending from the right posterior ankle to the popliteal fossa (Figure 1). The anterior right leg showed linear-to-ovoid pink patches with areas of central crusting (Figure 2).

Microscopic Findings

Our patient had multiple biopsies in the past consistent with ILVEN. We performed a skin biopsy of the right calf (Figures 3a, 3b), which demonstrated marked psoriasiform epidermal hyperplasia with slight papillomatosis. There were areas of alternating orthokeratosis with a thickened granular layer, and parakeratosis with loss of the granular layer. Collections of neutrophils with scale and crust were seen in the cornified layers of the epidermis. PAS stain was negative for fungi. The psoriasiform process resembled some changes seen in psoriasis, but the histopathological findings were more consistent with ILVEN. Clinically, the lesion had been present for 22 years, confirming the diagnosis of ILVEN.

Our patient did not see improvement with trials of clobetasol ointment, topical calcipotriene, entanercept, adalimumab, methotrexate, or betamethasone/calcipotriene prior to her pregnancy. Topical gentamicin 1% ointment was prescribed for potential secondary infection. Application of the ointment three times per day did not result in significant clinical improvement but was successful in eliminating the patient’s pruritus.

Figure 1. Hyperkeratotic papules coalescing into a large, linear plaque overlying a base of friable erythema extending from the right posterior ankle to the popliteal fossa (pre-pregnancy, Jan. 2014).

Figure 2. Brown verrucous papules coalescing into a linear plaque with overlying crust on a base of erythema on the right leg. Worsening of ILVEN with increase in overlying crust at 25 weeks pregnant (Sept. 2015).
Discussion
ILVEN is a rare form of epidermal nevus caused by somatic mutations, reflecting genetic mosaicism, though the exact physiopathology remains uncertain. It may be associated with an increase in the production of interleukin 1 and interleukin 6, along with tumor necrosis factor-alpha and intercellular adhesion molecule 1. ILVEN is more common among females and children and is sometimes familial.

Altman and Mehregan established the classic ILVEN diagnostic criteria in 1971. Morag and Metzker modified the criteria in 1985 to include unilateral, linear verrucous eruption (most frequently involving the left leg), severe pruritus, unilateral, linear verrucous eruption (most frequently involving the left leg), severe pruritus, early age of onset, and resistance to therapy. Prior research has shown that ILVEN is often resistant to topical steroids, 5-fluorouracil cream, tretinoin cream, podophyllin ointment and tar. Treatment is further limited in patients with ILVEN during pregnancy. We hypothesize the clinical worsening of ILVEN in our patient during her second trimester of pregnancy is due to an imbalance of hormone ratios. Studies suggest the increase in sex hormones during pregnancy, a natural state of immunomodulation, may play a potential role in the exacerbation of various inflammatory dermatological diseases.

Our patient declined further treatment until after her pregnancy. Due to unsuccessful therapies in the past, laser ablation will be the next treatment modality. Resurfacing lasers, such as Er:YAG and CO2 lasers, have proven to be effective at decreasing the thickness of ILVEN. Er:YAG laser treatment has been successful in the treatment of superficial, discrete ILVEN lesions. Recent CO2 laser clinical trials have demonstrated greater than 50% reduction in 50% of ILVEN patients treated with CO2 laser ablation, and greater than 75% reduction in 30%. Minor adverse effects consisted of scarring and hyperpigmentation, which was seen in 20% and 25%, respectively.

References


Correspondence: Brianna McDaniel, DO; Sampson Regional Medical Center, Clinton, NC; mcdaniel.brianna@gmail.com