Inflammatory and linear verrucous epidermal nevus in vulvar region: a differential diagnosis with vulvar condyloma

Nevo epidérmico verrucoso inflamatório e linear na região vulvar: um diagnóstico diferencial com condiloma vulvar

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ABSTRACT

Introduction: Inflammatory linear verrucous epidermal nevus (ILVEN) is a variant of verrucous epidermal nevus. It has a psoriasiform or eczematous and itchy aspect, and has differential diagnosis compared to other more common dermatoses; thus, histological studies are often necessary. It mainly affects women of early age and must be differentiated from condyloma acuminatum. Interestingly, the lower left limb is often involved, but the genital region is rarely affected. Treatment is refractory and the best method is not yet established. **Objective:** We present a case of unusual vulvar involvement known as inflammatory linear verrucous epidermal nevus. **Methods:** This was a clinical case report of a child diagnosed with ILVEN in the vulvar region. **Case report:** An 11-year-old female presented to the gynecology department of the Universidade Federal de Alagoas complaining of pruritic lesions on the large left vulvar lip, perianal and anal regions, and vaginal introitus. The lesions were hypochromic, eroded, and covered by scabs along the Blaschko line with verrucous lesions in the abdomen and upper and lower limbs. These characteristics fit the clinical criteria of Altman and Mehregan, and the histological criteria of Dupre and Christol for diagnosis of ILVEN. The treatment was performed with Vitanol A® and Epidrat Ultra® with partial improvement of the lesions. We chose to excise the lesions to control the condition. **Conclusion:** These lesions are characterized by recurrent inflammatory phenomena including psoriasiform or eczematous aspects in the extremities with genital involvement being rare. Other common dermatoses are often confused with ILVEN and make anatomically pathological analysis extremely important for diagnosis. Despite details on several types of treatment for ILVEN, there are no studies on relative advantages because this lesion is very refractory to the treatment.

Keywords: nevus, pigmented; neoplasms; vulva.

RESUMO

Introdução: Nevo epidérmico verrucoso inflamatório linear (NEVIL) é uma variante do nevo epidérmico verrucoso. Tem aspecto psoriasiforme ou eczematoso e pruriginoso, sendo o diagnóstico diferencial com outras dermatoses mais comuns e o estudo histológico é necessário para diferenciá-las. Afeta, principalmente, mulheres em idade precoce e deve ser diferenciado do condiloma acuminado. Curiosamente, o membro inferior esquerdo está envolvido, sendo a região genital raramente afetada. O tratamento é refratário e ainda não está estabelecido qual o melhor método. **Objetivo:** Apresentar um caso de incomum acometimento vulvar conhecido como nevo epidérmico verrucoso inflamatório linear (NEVIL). **Métodos:** Realizada documentação de caso clínico de criança com diagnóstico de NEVIL em região vulvar. **Relato de caso:** Uma paciente do sexo feminino, 11 anos de idade, procurou o serviço de ginecologia da Universidade Federal de Alagoas queixando-se de lesões pruriginosas vegetantes em grande lábio vulvar esquerdo, regiões perianal e anal e introito vaginal. As lesões eram hipocrômicas, erosadas, recobertas por crostas ao longo da linha de Blaschko com lesões verrucosas no abdome e nos membros superiores e inferiores. Essas características se enquadram nos critérios clínicos de Altman e Mehregan e aos critérios histológicos de Dupre e Christol para diagnóstico de NEVIL. O tratamento foi realizado com Vitanol A® e complexo hidratante (Epidrat Ultra®) com melhora parcial das lesões. Optou-se pela exérese das lesões com controle do quadro. **Conclusão:** Essas lesões se caracterizam por fenômenos inflamatórios recorrentes, incluindo aspecto psoriasiforme ou eczematoso, normalmente em extremidades, sendo raro o acometimento genital. Outras variedades de dermatoses mais comuns são frequentemente confundidas com NEVIL, tornando a análise anatomopatológica de extrema importância para o diagnóstico. Apesar dos detalhes sobre diversos tipos de tratamento para NEVIL, não há estudos sobre vantagens relativas entre eles, uma vez que essa lesão é muito refratária ao tratamento.

Palavras-chave: nevo pigmentado; neoplasias; vulva.

INTRODUCTION

Inflammatory and linear verrucous epidermal nevus (ILVEN) is a rare hamartomatous lesion that histologically consists of hyperplasia of components of the epidermis[1]. The clinical presentation includes erythematous as well as hyperkeratotic and verrucous lesions in a linear arrangement following the Blaschko lines. They are typically pruritic and unilateral lesions, and commonly affect the limbs in a curvilinear pattern. It mostly affects female children[2].

ILVEN has been described since 1896 and corresponds to a linear inflammatory verrucous lesion variant of the verrucous epidermal nevus. It is characterized clinically by recurrent inflammatory phenomena, and may have psoriasiform or eczematous aspects; this necessitates differential diagnosis. This is normally present in one of the extremities; curiously, the left leg is more affected. Its location in the genital region is less common[3]. It is four times more common in women and usually appears within the first four years of life although it may also appear in adulthood[4].

The diagnosis of ILVEN is based on clinical and histological presentation. In 1971 Altman and Mehregan proposed classical clinical criteria for diagnosis. This was updated by Morag and Metzker

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in 1985 to include female sex, young age, common involvement of the left lower limb, itching, psoriasiform histology, and refractory to treatment[1].

Histological changes were described by Dupre and Christol and include alternation of orthokeratosis and parakeratosis as well as the presence or absence of the granular layer although these are not pathognomonic. Other microscopy findings have shown papillomatosis, acanthosis, lymphocytic dermal infiltration, or even Munro microabscesses, but these are non-specific markers[5].

The diagnosis of ILVEN is clinical; however, it can be confused with more common conditions, such as candidiasis or even psoriasis, necessitating anatomopathological studies for differentiation.

There are several treatment options documented. However, there are no established relationships as to the superiority of the treatment mainly because the lesions are extremely refractory to the treatment options.

CASE REPORT

An 11-year-old female patient was taken by her mother to the Dermatology and Gynecology Department at the University Hospital Professor Alberto Antunes (HUPAA) in Maceió, Alagoas. Since 2 years old, the patient had had pruritic lesions with a vegetative appearance on the large left lip, perianal and anal regions, and vaginal introitus. Initially, they were treated with antifungals and antibiotics but without success. The lesions were linearly distributed along the Blaschko lines to the left and characterized by hypochromic eroded areas-some were covered by crusts and verrucous lesions in the left abdomen, axilla, upper limb, and lower limbs (**Figures 1 and 2**). There was no family history of a similar pathology or cancer. There was no history of sexual abuse or HPV vaccine.

A biopsy was performed in 2008 that revealed hyper and parakeratosis, hypergranulosis, presence of neutrophils, acanthosis, and papillomatosis-features all consistent with ILVEN. The patient underwent cryotherapy sessions and then used tretinoin 0.025% cream (vitanol A®) and moisturizing complex (Epidrat Ultra®), with a slight improvement in the appearance of the lesions.

At the beginning of 2015, the proliferative lesions were removed from the perianal region, and the tretinoin dose was increased to 0.05% (**Figure 3**).

In 2016, there was a vegetative lesion on the large left lip and scars on the large right lip and perianal region. A biopsy of the lesion was consistent with ILVEN. The lesion was excised following its path that extended from the pubis to the smaller left lip. The pathology showed a squamous papilloma with important associated inflammatory changes and the absence of dysplastic alterations. The procedure and the postoperative period were uneventful. The patient is currently stable.

DISCUSSION

The case fulfills the clinical criteria (age of early onset, female, pruritus, and refractoriness to the treatment) and histological criteria (squamous papilloma with important associated inflammatory changes and absence of dysplastic alterations) for the diagnosis of ILVEN. The differential diagnosis of ILVEN should be made with

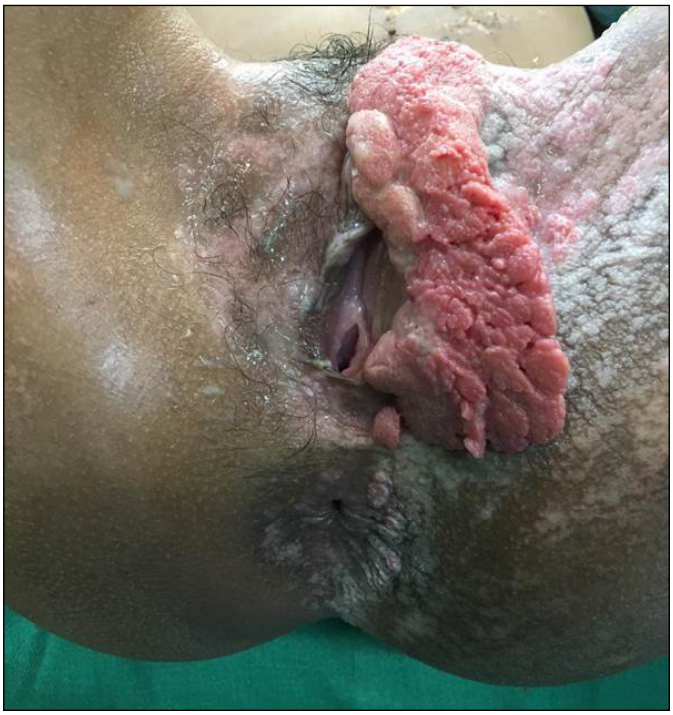

Figure 1 – Vegetative lesion on the large vulvar left lip, perianal and anal regions, and vaginal introitus in case of 11-year-old girl with inflammatory and linear verrucous nevus in the vulva.

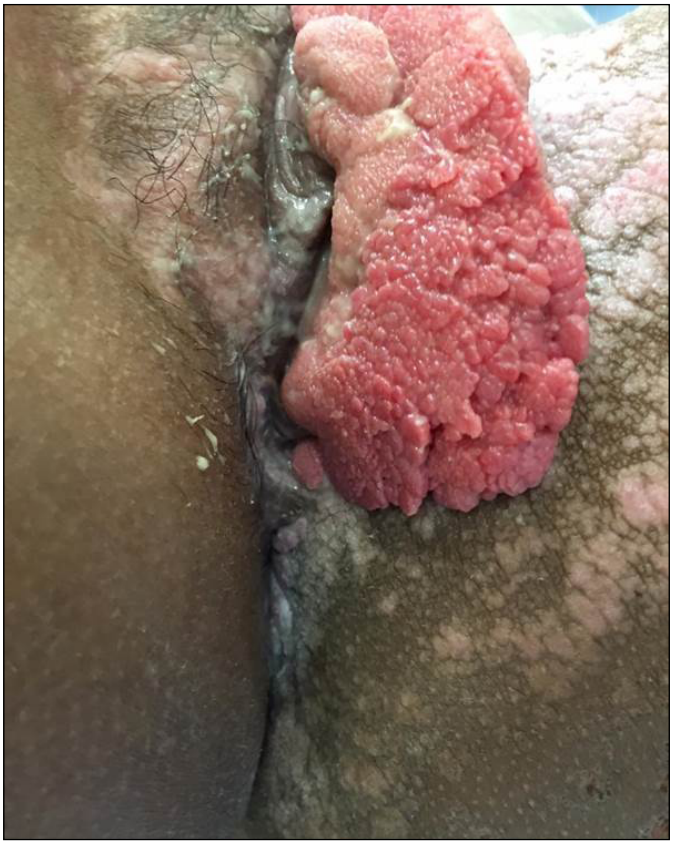

Figure 2 – Details of lesion in the large vulvar left lip of the inflammatory linear verrucous epidermal nevus' patient (11-year-old girl).

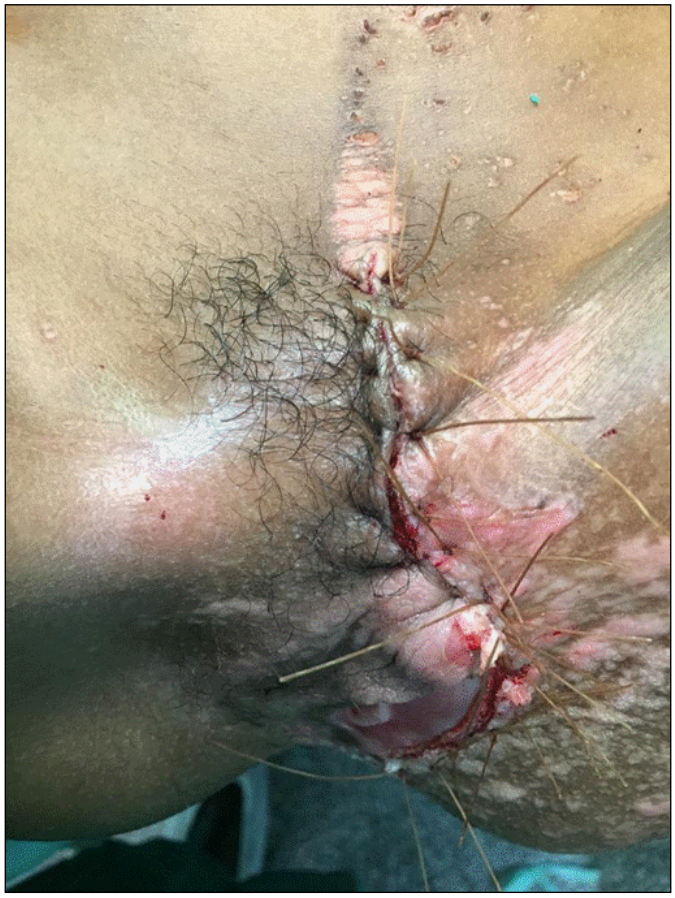

Figure 3 – Immediate postoperative aspect of the patient with inflammatory linear verrucous epidermal nevus (11-year-old girl).

a variety of dermatoses such as other epidermal nevi, linear psoriasis, and striated lichen. It is very important to do differential diagnosis with condyloma acuminatum and, in more severe cases, with Buschke-Lowenstein tumor, both HPV-induced lesions. Other diagnoses are often confused (vulvar candidiasis, for example), and the macerated appearance leads to intense itching[6] that is more severe than psoriasis or even eczema. In children, it is important to exclude the possibility of sexual abuse. When the diagnosis is in doubt, an anatomopathological study should be considered[7,8].

The antifungal and antibiotic treatment was based on a presumptive diagnosis of infection, but there was no response. Thus, a biopsy confirmed the suspicion of ILVEN and the patient was given tretinoin 0.025% cream (Vitanol A®) and a hydrating complex (Epidrat Ultra®). The proliferative lesions were removed from the perianal region followed by an increase in the dose of Vitanol A® to 0.05%.

ILVEN is quite refractory to treatment, making clinical work frustrating. Several modalities in the management of this condition have already been reported; however, no study has yet demonstrated superiority between any of the treatments. Topical treatments such as topical corticosteroids with or without occlusion and intralesional steroids are rarely beneficial. In contrast, vitamin D analogs have proved useful in some cases[9,10]. Other documented options are a combination of 0.1% tretinoin with 5% fluorouracil, anthralin, tar, vitamin D 3 analogs, surgical excision, cryotherapy with nitrogen liquid, and carbon dioxide laser therapy[11-14].

CONCLUSION

We describe a rare case of ILVEN including the differential diagnosis with lesions of infectious origin such as those induced by human papillomavirus (HPV) (condyloma acuminatum and Buschke-Lowenstein tumor), as well as other dermatoses such as epidermal nevi, linear psoriasis, lichen striatus, candidiasis, psoriasis, and eczema. Both children and adults can have ILVEN. It presents in unusual regions such as the vulva and perineum. The investigation must proceed with an anatomic-pathological study to differentiate it from other conditions. The literature describes several treatments although there is no documented superiority among them. Surgical excision is common in refractory cases.

Participation of each author

José Humberto Belmino Chaves followed the case and wrote the article. Julia Espíndola Guimarães followed the case and wrote the article. José Eleutério Jr. wrote the article, reviewed the English version, and formatted the text.

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Conflict of interests

The authors declare no conflicts of interest.

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