Genital/Perigenital Inflammatory Linear Verrucous Epidermal Nevus: A Case Series

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Abstract

Background: Inflammatory linear verrucous epidermal nevus (ILVEN) is a distinct variety of keratinocytic epidermal naevus. In contrast to non-inflammatory epidermal naevi, ILVEN are far less common, usually erythematous and intractably pruritic. ILVEN usually appears at birth or early childhood and has a linear distribution following the Blaschko lines. Genital/perigenital involvement is relatively rare. Objectives: To describe the clinical features of 9 children with ILVEN localized to the genital and perigenital areas. Method: A retrospective study of 9 children with ILVEN presenting to a tertiary care Dermatology Clinic between 2007 and 2014 was undertaken. The clinical and histopathological features were reviewed. Results: Nine children (6 females, 3 males) were included in the study based on their characteristic clinicopathological features. The lesions were associated with severe itching in all cases. The mean age at presentation was 4 years (range 1-11 years). Onset of lesions was before 6 months of age in 8 patients. Left sided involvement was twice as common as the right sided one. Male patients had penoscrotal and groin involvement while all the female children had vulvar lesions. None of the children had any extracutaneous abnormalities. The children were treated with topical agents with variable relief or symptoms. Conclusions: The possibility of ILVEN should be considered in every linear genital lesion in children. We have presented the largest series of perigenital ILVEN reported in English literature.

Key Words: Genital, inflammatory, perigenital, and verrucous epidermal nevus

Introduction

Inflammatory linear verrucous epidermal nevus (ILVEN), also known as dermatitic epidermal nevus, is a pruritic nevoid condition occurring along Blaschko’s lines. Most cases of ILVEN appear within the first 5 years of life although adult onset of the nevus had been described. It is 4 times more common in girls. The lesions are pruritic, linear, most commonly seen on a limb, and are eczematous or psoriasiform in morphology. It can be of any length, occasionally involving the whole limb and as with other epidermal nevi, nail dystrophy may occur. Genital and perigenital location of ILVEN is relatively rare, and may be mistaken for sexual abuse, genital warts, or other inflammatory dermatoses. In this series, we are reporting nine cases of genital and perigenital ILVEN, all of them had presented to the Dermatology Outpatient Department of a tertiary care center.

Methods

A retrospective study was performed on nine children who had presented to us between 2009 and 2014 and were diagnosed to have ILVEN located over the genital and perigenital areas. The diagnosis was based on the characteristic clinical features (pruritic, linear verrucous lesions along Blaschko’s lines with poor response to treatment) and histopathology of punch biopsy specimens. We reviewed the clinical details including the age and sex, symptoms, family history, age of onset, site of onset, spread of lesions, and location and extent of involvement, any other significant cutaneous/systemic features, and histopathological findings.

Results

The clinical profiles of the patients are summarized in Table 1.
Out of 34 patients of ILVEN diagnosed during the study period, 9 (26.5%) had genital/perigenital involvement. Moderate to severe pruritus was the chief complaint in all. In addition, the parents of two older children complained of social embarrassment owing to the scratching of the genital area. Females outnumbered males in a 2:1 ratio (6 females, 3 males). The age of the children ranged from 1 to 11 years with a mean age of presentation of 4 years. The onset of the lesions was before 6 months of age in eight patients (at birth in 6, 2 months in 1, 6 months in 1). The disease started at the age of 8 years in a girl. All the children were otherwise healthy, and no significant past history was obtained in any of them. None of the children had any family history of similar lesions. All the patients had a history of applications of various topical medications (steroid, salicylic acid, and tacrolimus) without any appreciable benefit. All the patients had linear bands of papular and occasionally vesicular lesions with the varying amount of scales along the lines of Blaschko. Left sided involvement was twice as common as the right sided one.

In the male children, the sites of involvement were the penis, scrotum, and inguinal region. One patient had penoscrotal involvement in addition to the facial involvement. All the male patients had left-sided involvement [Figures 1 and 2].

In case of females, vulva remained the commonest and constant site of involvement. In two of them the lesions were localized to vulva only, whereas in others, it had extended beyond the vulva to involve the groin, upper thigh and in one case, up to the knee [Figures 3-6].

None of the children had any other mucocutaneous or systemic abnormalities. Routine hemogram and biochemistry panel were normal in all. Histopathologic findings were more or less similar in all cases with psoriasiform hyperplasia of the epidermis with the varying amount of spongiosis, parakeratosis with hypogranulosis alternating with orthokeratosis with normal or hypergranulosis, and variable upper dermal perivascular lymphocytic infiltrate [Figure 7].

The children were treated symptomatically with oral antihistamines, topical steroids, or topical calcipotriol with variable relief or symptoms.

Discussion
ILVEN is a distinct variety of keratinocytic epidermal nevus that is clinically inflammatory and like other epidermal nevi reflects a phenomenon of mosaicism. ILVEN is usually sporadic in nature although the familial cases had also been reported.[4] Cutaneous lesions of ILVEN follow Blaschko’s lines, usually on the limbs and may extend to a variable length, sometimes to the whole limb. Pruritus is universal and the main concern for the patients. Occasional report of lib reduction defects had led to the suggestion that ILVEN represents a limited form of congenital hemidysplasia with ipsilateral limb defect (CHILD) nevus, and that ILVEN and CHILD would more appropriately be named, respectively, PEN and PENCIL, indicating psoriasiform epidermal nevus with or without congenital ipsilateral limb defects.[7] The following characteristics had been suggested as diagnostic aids: (i) Typical clinical, morphologic appearance accompanied by intense pruritus, (ii) early age onset of lesions; in 50% of patients before age 6 months, in 75% before age 5 years, (iii) a male-female ratio of 1:4, (iv) greater frequency of lesions on the left side of the body, mainly left lower extremity, (v) histologic findings consisting of inflammatory as well as psoriatic elements (psoriasiform chronic dermatitis), and, (vi) resistance to therapy.[5]

Our diagnosis of ILVEN in all the cases was based on the pathognomonic clinical signs and compatible histological picture. Although the lower limb is the most common reported site of affection, genital/perigenital involvement is relatively rare. In a series of 233 cases of epidermal nevi, 15 had ILVEN of whom three had lesions on the inguinal genital areas.[6] In another series of seven cases,[1] one child had a scrotal lesion. In our series, however, perigenital involvement was present in

<table>
<thead>
<tr>
<th>Case number</th>
<th>Age (years)</th>
<th>Gender</th>
<th>Onset</th>
<th>Side of involvement</th>
<th>Site</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>1</td>
<td>Female</td>
<td>Since birth</td>
<td>Right</td>
<td>Vulva, groin, upper thigh</td>
</tr>
<tr>
<td>2</td>
<td>11</td>
<td>Female</td>
<td>8 years</td>
<td>Right</td>
<td>Vulva, groin</td>
</tr>
<tr>
<td>3</td>
<td>4</td>
<td>Female</td>
<td>6 months</td>
<td>Right</td>
<td>Vulva, groin, extending to thigh up to knee</td>
</tr>
<tr>
<td>4</td>
<td>4</td>
<td>Male</td>
<td>Since birth</td>
<td>Left</td>
<td>Scrotum, inguinal region, left side of face</td>
</tr>
<tr>
<td>5</td>
<td>3</td>
<td>Male</td>
<td>Since birth</td>
<td>Left</td>
<td>Undersurface of penis and scrotum</td>
</tr>
<tr>
<td>6</td>
<td>2</td>
<td>Female</td>
<td>Since birth</td>
<td>Left</td>
<td>Vulva and groin</td>
</tr>
<tr>
<td>7</td>
<td>5</td>
<td>Male</td>
<td>Since birth</td>
<td>Left</td>
<td>Inguinal region</td>
</tr>
<tr>
<td>8</td>
<td>4</td>
<td>Female</td>
<td>2 months</td>
<td>Left</td>
<td>Vulva</td>
</tr>
<tr>
<td>9</td>
<td>1</td>
<td>Female</td>
<td>Since birth</td>
<td>Left</td>
<td>Vulva</td>
</tr>
</tbody>
</table>

ILVEN: Inflammatory linear verrucous epidermal nevus
more than one-fourth of the cases thus making this area a common site of involvement. Other previous reports of perigenital involvement includes penile and scrotal lesions occurring in a 21-year-old man who had linear, verrucous plaques on the left side of the body,\[9\] a familial case of 27-year man with ILVEN on his right side of scrotum and his daughter having lesion in her right axilla,\[10\] involvement of right inguinal region extending to the labia majora and perianal region,\[11\]
and two cases with lesions over the left perineum and vulva.\textsuperscript{[12]}

Clinically, the lesions in all the cases of this series were similar: A hyperpigmented or erythematous linear plaque consisting of hyperkeratotic papules with scaling located over the genital or perigenital areas. We considered and excluded the differentials of ILVEN including lichen striatus, verrucous epidermal nevus, psoriasis, and linear lichen planus. ILVEN differs from true nevus psoriasis clinically by pruritus and lack of response to antipsoriatic treatments. Clinicopathological correlation enabled us to correctly diagnose the cases. Considering the persistent and nonhealing course, the diagnosis of genital ILVEN can be possible in adults also; here the development of malignancy over the nevoid condition can be a serious complication.\textsuperscript{[12]}

Treatment of ILVEN is frustrating. Topical treatments such as topical corticosteroids with or without occlusion, intralesional steroid injections, and topical retinoids are rarely beneficial. Vitamin D analogs were found to be helpful in some cases.\textsuperscript{[14,15]} Among the physical modalities, cryotherapy, photodynamic therapy,\textsuperscript{[16]} and carbon dioxide laser therapy\textsuperscript{[17]} had been tried successfully. Surgical excision tends to be followed by rapid recurrence and requires a generous depth of underlying dermis to be removed.\textsuperscript{[18]} Our patients had a variable response to topical potent steroids combined with calcipotriol. All of them were referred to plastic surgery department, but considering the potential for anatomical deformity of the area, none of them opted for this modality of treatment.

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Conflicts of interest
There are no conflicts of interest.

References