ejpmr, 2015,2(5), 1277-1282.

SJIF Impact Factor 2.026

EUROPEAN JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.ejpmr.com

Case Report

ISSN 3294-3211

EJPMR

INFLAMMATORY LINEAR VERRUCOUS EPIDERMAL NEVUS IN THE PERIANAL AND VULVAL REGION: A CASE REPORT AND SHORT REVIEW OF THE LITERATURE.

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Article Received on 30/07/2015 **Article Revised on 21/08/2015** **Article Accepted on 13/09/2015**

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ABSTRACT

Inflammatory linear verrucous epidermal nevus (ILVEN) is a unilateral, persistent, linear, pruritic eruptions that usually appear on an extremity in infancy or childhood. These lesions are resistant to therapy. It is commonly found over buttocks and lower limbs. ILVEN in perineum and vulva is very rare. ILVEN may be an isolated finding or may be associated with other abnormalities. The diagnosis may sometimes be difficult and necessitate biopsy and advanced immunohistochemical analysis. The management usually is only symptomatic and often unsatisfactory. ILVEN occasionally presents in the inguinogenital region and in this area may, like many vulval naevi, be misdiagnosed as vulvitis, psoriasis, genital warts or sexual abuse. We present a case of ILVEN in a 26-year-old female in perivulval and perianal region and provide a short review of the literature, with emphasis on our current understanding of the etiology, clinical presentation, histopathological features and differentials along with its management.

KEYWORDS: Inflammatory linear verrucous epidermal nevus, pruritic eruptions, perineum, perianal, vulva.

INTRODUCTION

Inflammatory linear verrucous epidermal nevus (ILVEN) is unilateral, pruriginous dermatosis manifested as erythematous, inflammatory, linear papules and plaques. It usually presents in infancy or early childhood as an itchy linear plaque resistant to treatment. It is commonly found over buttocks and lower limbs. ILVEN in perineum and vulva is very rare.[1] We hereby report case of ILVEN, over the perivulval and perianal region of a 26-year-old

female for its rarity along with its clinical and pathological features with differential diagnosis.

CASE REPORT

A 26-year-old female presented with intense pruritic erythematous plaque in a linear fashion localized mostly over left perianal and perivulval region since the age of 5 years with intermittent flare-up and remission. The lesion extended from the perianal region to the perivulval region in a circumferential fashion.[Fig.1] Family history was not significant. Results of a physical examination revealed multiple erythematous plaques coalesced to form a linear, hyperkeratotic and excoriated, hyperpigmented plaque. The plaque gradually extended to the groin, perineum, and gluteal region on the same side. The rest of the physical examination results were unremarkable, and no associated physical anomaly was found. The eruption and the associated pruritus did not respond to either oral antihistamines or topical high potency steroids.

Biopsy specimen taken from the lesion revealed presence of hyperkeratosis with foci of parakeratosis, moderate acanthosis and elongation of rete ridges.[Fig.2] Alternate areas of parakeratosis without a granular layer and hyperkeratotic areas with distinct granular layer were seen.[Fig.3] A moderate mononuclear infiltrate of lymphocytes was evident in the upper dermis. A diagnosis of inflammatory linear verrucous epidermal nevus (ILVEN) was made based on clinical and histopathologic grounds. Topical mupiricin ointment in combination with methylprednisolone 0.1% cream was prescribed for symptomatic relief and a surgical referral was made for discussion of the viability of excision in this location.

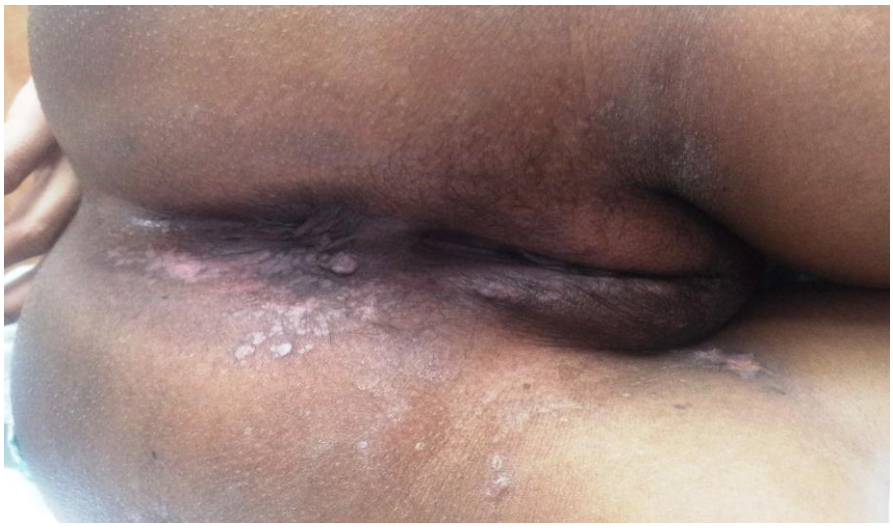

Figure – 1

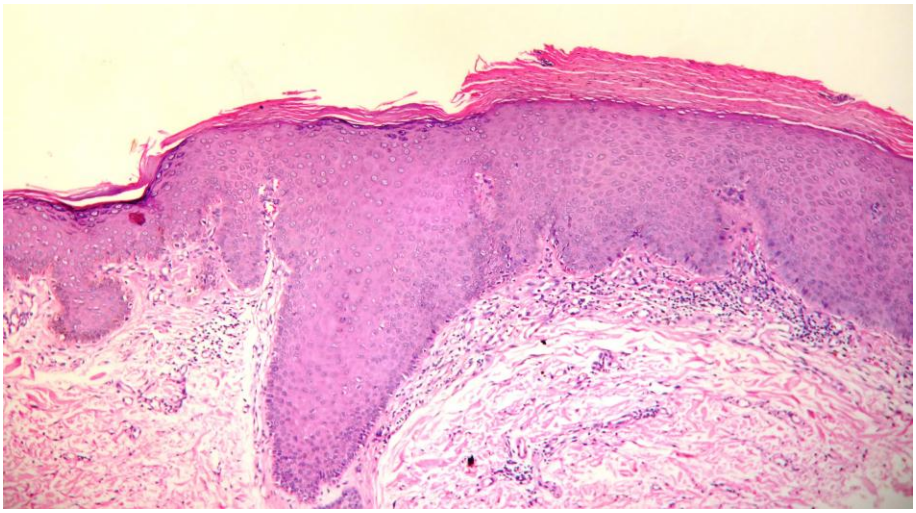

Figure – 2

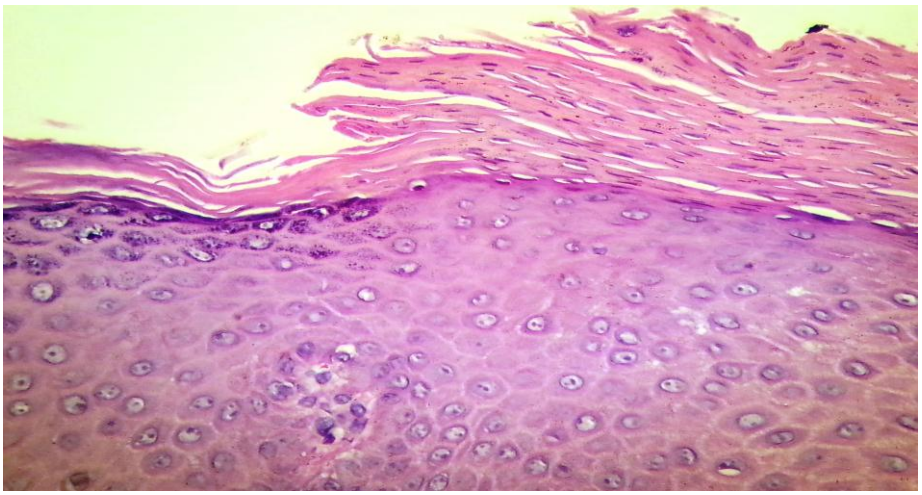

Figure – 3

DISCUSSION

ILVEN is characterized by erythematous pruritic inflammatory lesions that occur as linear bands following Blaschko's line was first described by Altman & Mehregan in 1971 as distinct entity.[2,3] It is considered a variant of keratinocytic epidermal nevus. The definitive etiology of ILVEN is not yet clear, but is considered to be related to the following aspects: 1) alteration of involucrin, which was increased in orthokeratotic area and decreased in parakeratotic area; 2) alteration of cytokine like interleukins 1 and 6, tumor necrosis factor α, and intercellular adhesions; 3) clonal deregulation of growth of keratinocytes and 4) somatic mosaicism.[4]

ILVEN usually appears within 6 months of life or early childhood but may be present at birth. The condition very rarely starts in adulthood. ILVEN occurs predominantly in females (male to female ratio is 1:4), and no racial predominance has been noted. The distribution is

almost exclusively on the lower half of the body, and the buttock is the most frequently involved area. Only 16% of lesions were found on the upper half of the body, including the axillae, arm and hand and common left side of body.[5] In a series of 233 patients with epidermal naevi, there were 15 with ILVEN, three of whom had inguinogenital lesions over scrotum and penis.[1]

Altman and Mehregan described 6 characteristic features of ILVEN: (1) early age of onset, (2) predominance in females (4:1 female-male ratio), (3) frequent involvement of the left leg, (4) pruritus, (5) marked refractoriness to therapy, and (6) a distinctive psoriasiform and inflammatory histologic appearance.[2]

There are two major histopathological features of ILVEN: 1) psoriasiform changes, these included hyperkeratosis, parakeratosis, acanthosis with thinned suprapapillary plates, and mononuclear cell infiltrates; 2) sharply demarcated alternated areas of parakeratosis and orthokeratosis. The parakeratotic areas are slightly raised, with agranulosis, whereas the orthokeratotic areas are slightly depressed with hypergranulosis. Microabscesses of Munro, typically found in psoriasis, can be found in ILVEN as well.[6] Ki-67 index is low in ILVEN. Keratin and HLA-DR expression tend to be higher in patients with ILVEN. The density of CD8+, CD45RO+, CD2+, CD94, and CD161 also showed a marked difference between ILVEN and psoriasis.[7]

ILVEN has been found to occur in association with musculoskeletal or other abnormalities, including supernumerary digits and strabismus, congenital bony anomalies, Fallot tetralogy of the heart, autoimmune thyroiditis, lichen amyloidosis, nevus depigmentosus, arthritis.[8]
The verrucous nature of ILVEN and the occurrence in the genital area may raise the question of sexually transmitted disease and sexual abuse especially in children.

Clinically, ILVEN should be distinguished from linear psoriasis, other epidermal nevi, and lichen striatus. The absence of pruritus and spontaneous regression are pathognomonic of lichen striatus.

ILVEN is often difficult to differentiate from linear psoriasis. Psoriasis can occur in a nevoid form following Blaschko's lines or Koebnerize and become superimposed on an epidermal nevus. Early onset in ILVEN and the positive family history in psoriasis may help to differentiate the two. There may be scattered lesions elsewhere on the skin in psoriasis.

Quantitative IHC studies with T-cell markers showed different patterns in these two conditions.[9]

Epidermal nevi (Nevus verrucosus) are localized lesions may occur almost anywhere on the head, neck, legs, or trunk. Clinically, there is no erythema or pruritus. IHC studies further help differentiate ILVEN from other noninflammatory linear epidermal nevi.

Linear lichen planus mainly affects children and is characterized by discrete pruritic, polygonal, violaceous papules arranged in a linear fashion, usually along an entire extremity; however, pathological pattern of lichen planus is that of lichenoid dermatitis.

Typically, ILVEN is resistant to treatment. Therapeutic options include intralesional or topical corticosteroids, calcipotriol, 5-fluorouracil, tretinoin, dermabrasion, cryotherapy, laser, and partial thickness excision. The CO_2 laser at low fluence has been used to treat patient with disfiguring ILVEN in vulvar region. However, the clinical appearance and associated intense pruritus usually are refractory to treatment.[10]

CONCLUSION

The diagnosis of ILVEN may sometimes be difficult and necessitate biopsy and advanced immunohistochemical analysis in few cases. The management usually is only symptomatic Clarification of various differential diagnosis is of practical importance for these are different in aetiopathogenesis, management and outcome.

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