Inflammatory Linear Verrucous Epidermal Nevus Complicated by Jacquet Erosive Dermatitis: A Rare Overlap in Dermatology

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ABSTRACT:
Inflammatory Linear Verrucous Epidermal Nevus (ILVEN) and Jacquet's erosive dermatitis are uncommon dermatological disorders reported in young age groups. Their coexistence of these conditions has not been previously reported. Herein we report a 12-year-old boy concerned about the macerated patches arising on his innate plaque on his right thigh. Skin biopsies were obtained from these 2 different morphologies of lesion which later confirmed the diagnosis of Jacquet erosive dermatitis arising on ILVEN. To the best of our knowledge, this is the first report case of ILVEN occurring concurrently with Jacquet erosive dermatitis on the same lesion in the medical literature. Further research is essential to explore the pathophysiology between these 2 conditions.
Key words: Inflammatory Linear Verrucous Epidermal Nevus (ILVEN), Jacquet Erosive Dermatitis

Introduction

Inflammatory linear verrucous epidermal nevus (ILVEN) is a chronic inflammatory epidermal nevus following Blaschko lines and typically manifests in early childhood. Jacquet's erosive dermatitis (JED) is an uncommon but severe form of irritant dermatitis characterized by erosive verrucous papulonodules in areas subjected to chronic maceration and friction. To date, coexistence of ILVEN with JED on the same anatomical site has not been reported. We present a rare case of ILVEN complicated by JED, confirmed histopathologically, and discuss potential pathogenetic links between these two conditions.

Case presentation

Here, we report a case of a 12-year-old boy presenting with non-itchy erythematous rashes on the right groin and inner right thigh, present since birth. The lesion initially appeared on the right groin and progressively extended downward in a linear pattern to the inner thigh. Previous treatment with a topical antifungal was ineffective. The patient reported no systemic symptoms.

Physical examination revealed linear, well-demarcated erythematous to brownish hyperkeratotic papules and plaques along Blaschko lines on the medial right thigh, interspersed with macerated erythematous verrucous plaques in the right groin area (Figure 1). Two tissue samples were taken from the lesion. The first one was from the macerated area and the other is from the hyperkeratotic lesion.

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Figure 1 (a and b) A linear, well-demarcated erythematous to brownish hyperkeratotic papules and plaques along Blaschko lines on the medial right thigh, interspersed with macerated erythematous verrucous plaques in the right groin area

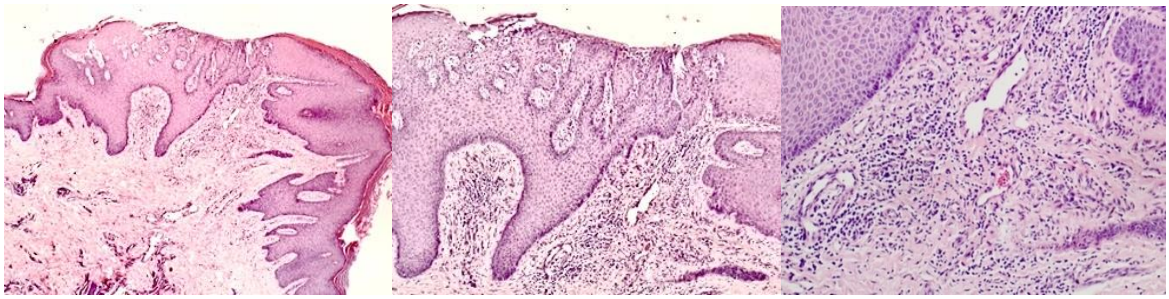

Figure 2 (a, b, and c) Epidermal erosion with parakeratosis and irregular acanthosis. The upper dermis demonstrated a mixed inflammatory cell infiltrate, predominantly lymphocytes

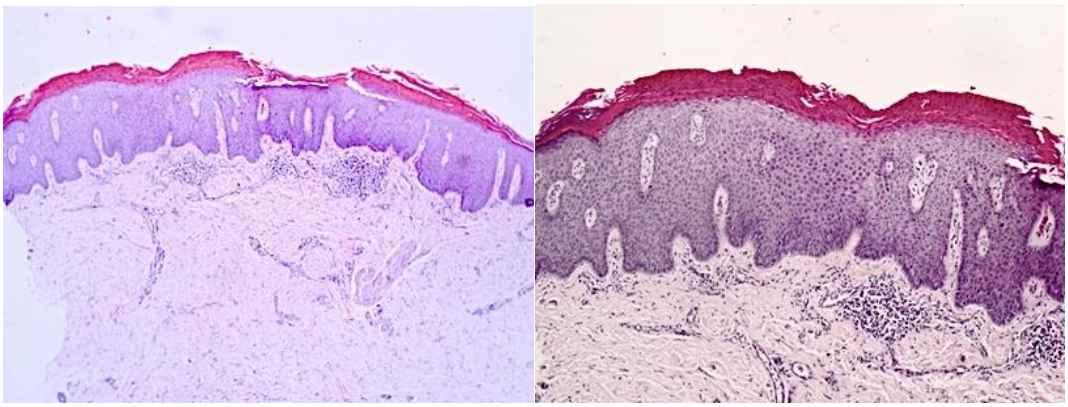

Figure 3 (a and b) Epidermal hyperplasia with hyperkeratosis alternating sharply with regions of orthokeratosis. The dermis showed mild inflammation, and the lesion extended the specimen

Histopathological analysis of the macerated area revealed epidermal erosion with parakeratosis and irregular acanthosis. The upper dermis showed a mixed inflammatory cell infiltrate, predominantly lymphocytes, consistent with Jacquet erosive dermatitis (Figure 2). The second specimen demonstrated epidermal hyperplasia with pronounced hyperkeratosis alternating sharply with regions of orthokeratosis. The dermis displayed mild inflammation, and the lesion extended to the resection margins in the examined sections which is the findings of ILVEN (Figure 3). Histopathological examination of both biopsy sites confirms the diagnosis of Jacquet erosive dermatitis occurring on ILVEN. To our knowledge, this is the first reported case of Jacquet erosive dermatitis (JED) occurring in association with an inflammatory linear verrucous epidermal nevus (ILVEN). This case underscores the possibility that JED may arise as a complication or coexist with preexisting dermatoses, such as ILVEN.

Discussion

Jacquet erosive dermatitis (JED), initially identified by Jacquet in 1905, is an uncommon form of ulcerating severe irritant dermatitis mostly affecting the genital and perianal skin of infants older than six months, though it can also occur in adults. Characteristic of the lesion are well-demarcated papules and nodules with erosion or ulcer. Heal-up borders can also be observed[1]. Its development is influenced by multiple factors, with skin irritation playing a pivotal role. Major risk factors include urinary and fecal incontinence, further aggravated by diaper friction and residual detergent exposure. Other forms of irritant dermatitis that can also occur on these areas such as perianal pseudoverrucous papules and nodules and granuloma gluteale infantum/adultorum which have the clinical feature of erythematous perianal plaque comprising multiple small, shiny, flattop, round, smooth moist papules and nodules[2]. Jacquet's erosive dermatitis is primarily a clinical diagnosis; however, histopathology is often supportive. Although JED is a severe variant of irritant dermatitis, it can be differentiated from simple irritant contact dermatitis by its sharply demarcated papulonodular lesions, deeper erosions, and pseudoverrucous appearance, while classic irritant contact dermatitis more often presents with diffuse erythema, scaling, and shallow erosions without nodularity.

Inflammatory linear verrucous epidermal nevus (ILVEN) is an uncommon skin disorder first described by Altman and Mehregan in 1971. It is a distinct variety of keratinocytic epidermal nevus that is clinically inflammatory and usually psoriasiform. The condition predominantly affects females, with a 4:1 ratio compared to males[3]. Although the exact cause remains unknown, the disease has been linked to a postzygotic mutation in the GJA1 gene, which encodes connexin 43[4]. Additionally, a mutation in the CARD14 gene has been associated with ILVEN[5]. Germline mutations in CARD14 can lead to CARD14-associated papulosquamous eruption (CAPE), a condition that exhibits features of both psoriasis and pityriasis rubra pilaris. ILVEN is characterized by erythematous papules that merge into linear plaques following Blaschko lines. It typically affects one side of a limb and occasionally involves the trunk. Although primarily unilateral, rare cases of bilateral lesions have been reported. The condition usually appears within the first six months of life, with about 75% of cases diagnosed before the age of five[6]. Adult onset and familial occurrence have also been documented[1, 7]. Significant pruritus is a common symptom. If untreated, the lesions often persist lifelong. ILVEN has been linked to arthritis, which can lead to complications such as cartilage damage, growth abnormalities, and joint contractures[8]. Rarely, ipsilateral skeletal anomalies may be present, requiring differentiation from congenital hemidysplasia with ichthyosiform nevus and limb defects

(CHILD) syndrome, an X-linked dominant condition characterized by epidermal nevi[9].

ILVEN is notoriously difficult to treat, with various approaches documented in the literature[8]. Surgical excision may be effective, though patients should be advised of the risk of scarring. The 585 nm pulsed dye laser, which works by inducing mast cell degranulation and destroying blood vessels within the lesion, has shown success in certain cases[10]. Topical therapies, such as a combination of 0.1% tretinoin cream and 5% 5-fluorouracil, have demonstrated promising outcomes[11]. Additional treatments include crisaborole ointment, intralesional steroids, podophyllin ointment, TNF inhibitors, and laser-based therapies such as excimer laser, fractional CO2 laser, and ablative CO2 laser. Treating JED is similarly challenging and typically involves topical antibiotics, miconazole, zinc oxide, and nonsteroidal anti-inflammatory drugs (NSAIDs). Topical sucralfate has shown some success in managing the condition; however, recurrence is common once treatment is discontinued[12]. Our patient was counseled and chose treatment through serial excisions to reduce the risk of scarring. During follow-up, no significant complications were noted, and the disease has not relapsed (Figure 4).

Figure 4 (a and b) Patient was treated with serial excision

A review of published case reports indicates that Jacquet erosive dermatitis is most commonly associated with frequent or prolonged irritation, such as exposure to urine, feces, or occlusive diaper use. It has also been described in conditions like Hirschsprung disease and COVID-19, where recurrent defecation contributes to chronic perianal irritation. In our patient, the diagnosis of JED was supported by the presence of characteristic macerated verrucous erosions confined to the groin, the absence of other potential irritant exposures, and histopathologic findings fully consistent with JED. The development of JED in this case is most plausibly explained by the underlying ILVEN. The elevated, hyperkeratotic surface of the ILVEN plaque likely increased local susceptibility to friction and maceration, while the preexisting inflammatory infiltrate within ILVEN may have further lowered the irritant threshold. Chronic friction and moisture accumulation over this raised lesion therefore created a persistently vulnerable microenvironment that favored erosive dermatitis. This is consistent with the observation that the contralateral limb,

which does not have ILVEN, showed no evidence of JED.

Conclusion

In conclusion, this is the first reported case of Jacquet erosive dermatitis occurring on ILVEN. As previously established, Jacquet erosive dermatitis typically results from prolonged irritation. In this case, we hypothesize that its occurrence on ILVEN is due to repeated external irritation from its raised lesion. Therefore, it is important to assess patients with ILVEN for the presence of associated erosions or ulcerations. Further studies are needed to clarify the precise pathophysiology linking these two conditions and to establish appropriate management strategies.

Author Contributions

Conceptualization: K.O., P.C.; Investigation: K.O.; Writing – original draft preparation: K.O.; Writing – review and editing: P.C.; Supervision: P.C. All authors have read and agreed to the published version of the manuscript.

Ethics Statements

This case report was conducted in accordance with the Declaration of Helsinki. Written informed consent was obtained from the patient's parent/legal guardian for the publication of this case report, including potentially identifiable clinical images.

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Conflict of Interest

The authors declare that there are no conflicts of interest regarding the publication of this paper.

AI Declaration

During the preparation of this work, the authors used ChatGPT (GPT-4o; OpenAI) to check grammar and improve language readability. After using this tool, the authors reviewed and edited the content as needed and take full responsibility for the content of the published article.

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