International Journal of Science and Research (IJSR)
ISSN: 2319-7064
Impact Factor 2024: 7.101

Coexistence of Pityriasis Lichenoides et Varioliformis Acuta in a Patient with Acrodermatitis Enteropathica: A Rare Case Report

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Abstract: ***Acrodermatitis Enteropathica (AE) is a rare autosomal recessive disorder of zinc metabolism typically presenting in infancy with periorificial dermatitis, alopecia, and diarrhea. Pityriasis Lichenoides et Varioliformis Acuta (PLEVA) is a rare inflammatory skin disorder of unknown etiology, commonly presenting in children and young adults with sudden onset of erythematous macules and papules that may ulcerate. We present a rare case of a 37-year-old male with a history of AE who developed features consistent with PLEVA. Histopathological confirmation and clinical response to zinc, dapsone, and corticosteroids emphasize the overlapping immunological and dermatological features of these two distinct conditions.***

Keywords: Acrodermatitis enteropathica, PLEVA, zinc, papulonecrotic lesions.

1. Introduction

Acrodermatitis Enteropathica (AE) is a genetically inherited disorder of zinc absorption due to mutations in the SLC39A4 gene [1]. Clinical presentation typically includes periorificial dermatitis, alopecia, and chronic diarrhea in early infancy. Zinc supplementation usually results in dramatic clinical improvement.

Pityriasis Lichenoides et Varioliformis Acuta (PLEVA) is a rare, self-limiting, inflammatory skin condition characterized by recurrent crops of erythematous macules and papules, some of which may become necrotic [2]. Though the etiology remains unclear, immune dysregulation has been proposed [3].

We report a rare coexistence of AE and PLEVA in a middle-aged male, highlighting diagnostic challenges and the therapeutic response to zinc and immunomodulatory treatment.

2. Case Report

A 37-year-old male presented with mildly itchy pustular lesions over the body of one month's duration. He had a known diagnosis of AE from infancy, when he initially developed red, scaly lesions around the mouth, irritability, and diarrhea. Low serum zinc levels confirmed the diagnosis, and he was started on zinc supplementation, with significant improvement noted.

The current episode involved development of pustular lesions predominantly over the face, limbs, scalp, and abdomen, without accompanying fever, alopecia, arthralgia, or mucosal involvement [Fig 1 and 2] . There was no history of new drug use or topical applications.

On examination there was diffuse erythema involving the central face, nose, cheeks, and nasolabial folds, with clusters of hypopigmented papules and plaques showing peripheral scaling on the perioral area, limbs, abdomen, and ears. Discrete papules and pustules were also present on the scalp. No mucosal lesions or systemic signs were evident.

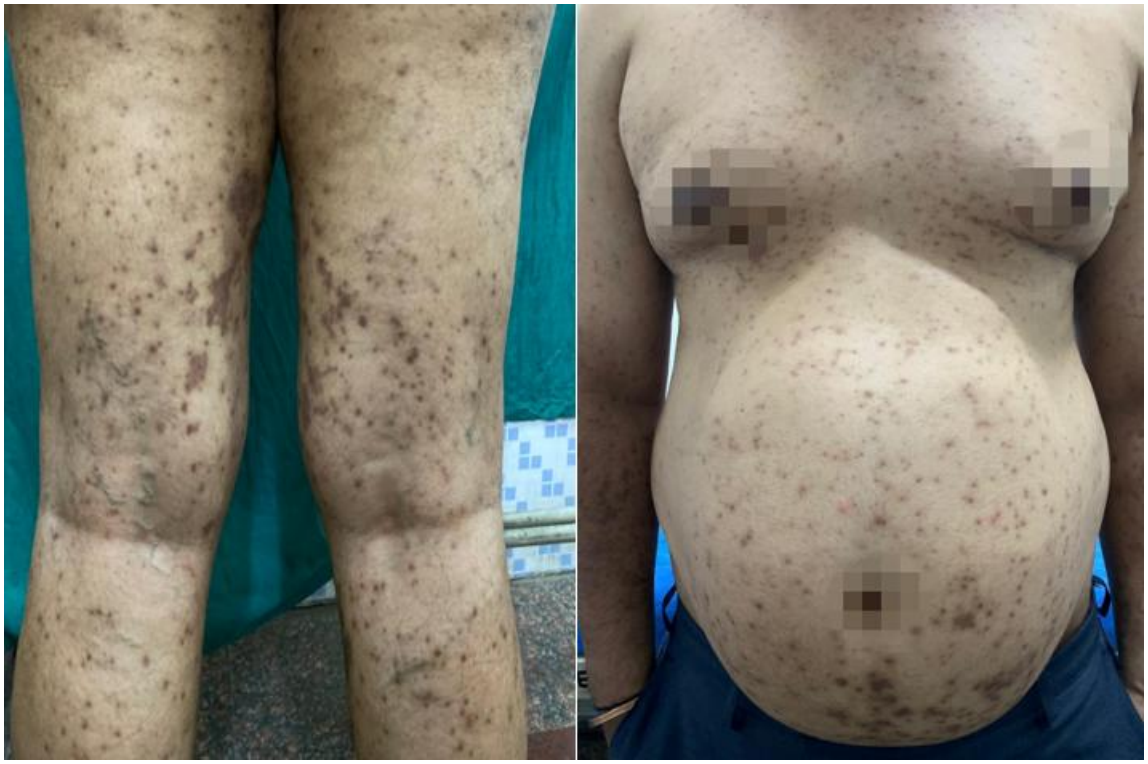

Figure 1: Hyperpigmented papules,plaques over the bilateral Legs and erythematous vesicles and pustules with hyperpigmented papules over abdomen

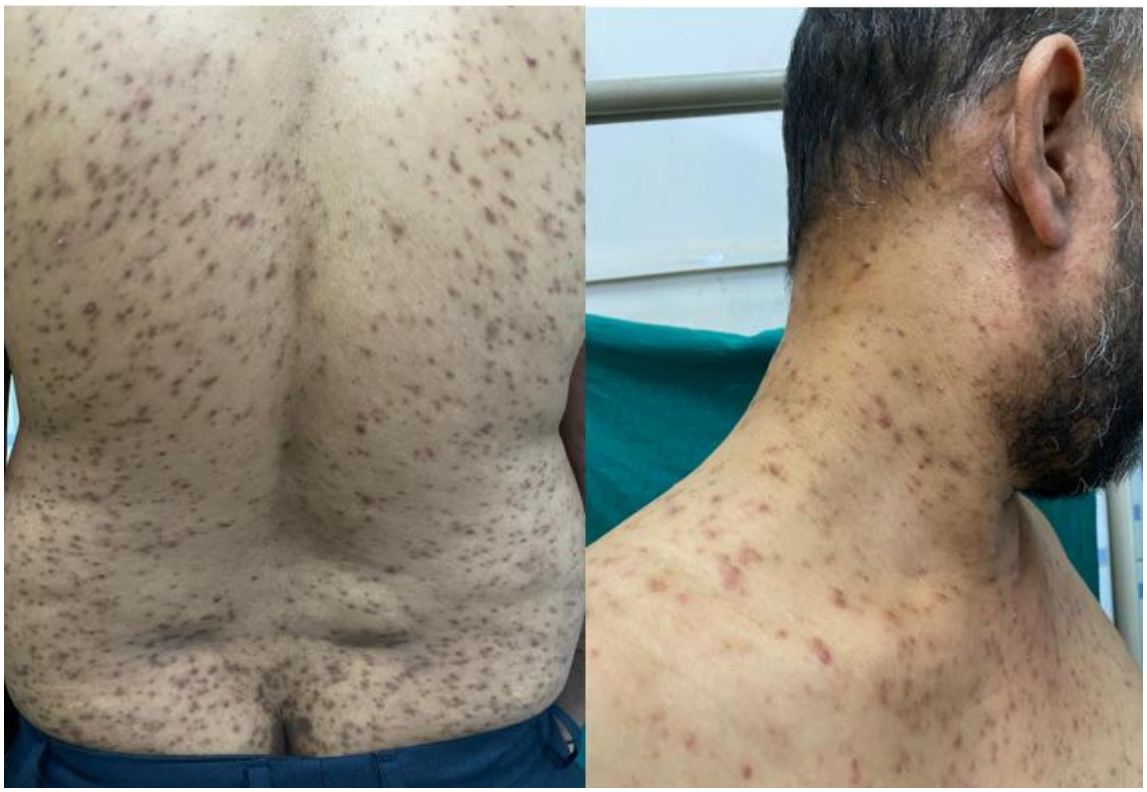

Figure 2: Hyperpigmented papules, plaques over the back and erythematous papules and vesicles over the neck and scalp

Laboratory Investigations: Serum zinc was normal. CBC, LFT, and RFT were within normal limits. HIV and VDRL were non-reactive.

Histopathology: Skin biopsy revealed parakeratosis, focal epidermal necrosis, interface dermatitis with vacuolar degeneration of basal keratinocytes, dermal edema, and a perivascular lymphocytic infiltrate. These findings were consistent with PLEVA.

Treatment: The patient was treated with oral prednisolone (1 mg/kg/day), dapsone (100 mg/day), zinc acetate (50 mg elemental zinc daily), and topical corticosteroids with emollients. A marked improvement was seen within two

weeks, with resolution of pustules and reduction in erythema and scaling.

3. Discussion

AE is a disorder characterized by impaired zinc absorption, leading to cutaneous and systemic manifestations. Zinc is an essential element in the body which has an important role in the human immune system, growth and development, hormones synthesis and activation, and gene regulation[4]. It is also considered a co-enzyme for several enzymes, including ALP, alcohol dehydrogenase, RNA polymerase, and numerous digestive enzymes[5]. The clinical presentation of AE differs according to the age group; however, the classical disease triad includes alopecia, periorificial dermatitis, and diarrhea. Recommended elemental zinc therapy is 3 mg/kg/day as initial treatment followed by 1–2 mg/kg/day as a maintenance dose[6]. Follow-up is required for adjustment of the dose according to the weight and to check for zinc toxicity. Although classical AE responds well to zinc supplementation, cutaneous immune dysregulation may predispose patients to other dermatoses.

PLEVA, first described by Juliusberg in 1899, is part of the pityriasis lichenoides spectrum, ranging from the acute (PLEVA) to the chronic form (PLC) [7]. It is generally accepted that PLEVA and PLC represent two ends of a continuous spectrum, and therefore it is not uncommon to observe both acute and chronic lesions in the same patient, as well as lesions at intermediate stages between PLEVA and PLC [8].It is hypothesized to represent a hypersensitivity reaction to infectious agents or immune dysregulation [9].PLEVA most often occurs on the trunk, extremities, and flexural areas, but diffuse and generalized patterns may also occur. The eruption is polymorphous, as lesions exist in all stages of development, and successive crops of lesions can last indefinitely, from a few weeks to months or years [10]. Histopathological findings include parakeratosis, necrotic keratinocytes, and a lymphocytic infiltrate, consistent with our patient's biopsy.

The coexistence of AE and PLEVA in our patient highlights possible immunological overlap. Zinc plays a role in modulating T-cell function and cytokine production, and its deficiency has been implicated in altered immune responses [11]. Although our patient had normal zinc levels at presentation, his underlying genetic defect in zinc transport may have contributed to cutaneous immune dysregulation.

Therapeutic options for PLEVA include systemic corticosteroids, dapsone, tetracyclines, and phototherapy [12]. In our case, a combination of zinc, corticosteroids, and dapsone resulted in marked clinical improvement.

To the best of our knowledge, this is the first documented case from India reporting coexistence of AE and PLEVA in the same patient, underscoring the need for clinicians to consider overlapping immunological mechanisms in rare dermatological presentations.

4. Conclusion

This case emphasizes the importance of considering immune dysregulation in patients with genetic disorders such as AE when they present with new cutaneous eruptions. The favorable response to a combination of zinc and immunomodulatory therapy suggests potential synergistic effects in managing such cases.

Funding: None

Conflict of Interest: None declared

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