CASE REPORT

Diagnostic Challenges of Pityriasis Lichenoides et Varioliformis Acuta (PLEVA).

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Abstract

Pityriasis Lichenoides et Varioliformis Acuta (PLEVA) is an uncommon, inflammatory skin illness typified by erythematous papules that can mimic various dermatological and systemic conditions, making diagnosis difficult. We describe a case of a young adult who presented with generalized erythematous macules and crusted patches across the trunk that progressed to the extremities and scalp. A provisional diagnosis of guttate psoriasis with PLEVA as the differential diagnosis was made after a thorough history and examination. Further evaluation and skin biopsy confirmed the diagnosis of PLEVA through histopathological examination. The lesions subsided after two months of treatment with oral and topical corticosteroids.

Keywords: *Pityriasis lichenoides et varioliformis acuta, lichenoid dermatitis, primary care.*

Introduction

Mucha-Habermann disease or Pityriasis lichenoides et varioliformis acuta (PLEVA) is a rare benign dermatosis with uncertain etiopathogenesis. It can affect individuals of any age but is most prevalent in children and young adults. PLEVA's diverse morphology and clinical presentation, which may resemble other dermatological conditions, often pose diagnostic challenges, and lead to delays in appropriate management. Here, we present the case of a patient who was initially diagnosed with guttate psoriasis, due to the similarity to Mucha-Habermann disease. However, the final diagnosis was PLEVA. This case highlights the importance of considering this uncommon condition in the differential diagnosis of dermatological disorders.

Case presentation

A 21-year-old male presented with a vesicular eruption that had persisted for one month. Subsequently, recurrent erythematous macules and crusted patches appeared on his trunk, extended eventually to his extremities and scalp. The lesions were itchy, but sparred his oral mucosa and nails. Initially, the patient thought he had the same condition as his nephew, who had been diagnosed with a varicella zoster infection. However, as the lesions persisted, he sought treatment at multiple clinics and was prescribed both systemic and topical antibiotics. Unfortunately, no improvement was seen despite the treatment.

Upon examination, his vital signs were stable, and he appeared well. Brownish scales were present throughout his skin, particularly on the trunk (Figures 1 and 2) and in all flexural areas. Newer erythematous macules, papules, and dried-up blisters with necrotic and occasionally hemorrhagic centers were observed, especially on the extremities (Figure 3). No alopecia, oral ulcers, joint tenderness, or nail involvement was seen. For laboratory investigations, the results were negative for the anti-nuclear antibody (ANA), syphilis, and viral screenings including Human immunodeficiency virus (HIV), Hepatitis B, and Hepatitis C. Given the clinical diagnosis of guttate psoriasis and the need to rule out PLEVA, he was referred to the dermatology department. Histopathological examination (HPE) of his skin biopsy revealed subepidermal blisters with necrotic changes, ischemic epidermis, and marked lichenoid change with interface dermatitis (Figure 4). Spongiosis was also seen in viable epidermis, and focal epidermotropism was apparent (Figure 5). Despite all the immunofluorescence studies of the skin biopsy (IgG, IgA, IgM, and C3) negative for epidermal-dermal junction and blood vessels, PLEVA was still the diagnosis. The lesions resolved after two months of treatment with tapering doses of oral and topical corticosteroids.

Discussion

Pityriasis lichenoides et varioliformis acuta (PLEVA), also known as Mucha-Habermann disease (MHD), is a rare cutaneous disorder that commonly occurs in children and young adults, with a slightly higher occurrence in males (56%) [1]. Although the exact origin of PLEVA is unknown, it is believed to be associated with an inflammatory response that is triggered by infectious agents [2]. PLEVA has been reported in cases following infections with pathogens like Epstein-Barr virus, HIV, varicella-zoster virus, herpes simplex virus type 2, Toxoplasma gondii, and Group A streptococcus [2]. Apart from that, PLEVA has been linked to certain medications such as antidepressants, statins, anti-tumor necrosis factor (anti-TNF), and various vaccines [2-5]. Moreover, T-cell dyscrasia-induced inflammation or immune complex-mediated hypersensitivity were suggested as potential causes of PLEVA [2]. In this case, the first theory is more plausible because he had exposure to varicella zoster infection from his nephew.

The hallmark features of PLEVA are characterized by the acute development of inflammatory papules and papulovesicular with hemorrhagic or necrotic crusts on the skin, usually on the trunk and flexural areas of the

extremities(1). While individual lesions may disappear within a few weeks, new groups of lesions frequently appear, leading to lesions at various stages of development. This can make diagnosing PLEVA difficult in a primary care setting. Rarely, patients have the severe variation of PLEVA known as febrile ulceronecrotic Mucha-Habermann disease (FUMHD) that may involve mucosal membranes, high fevers, and systemic complications like sepsis, splenomegaly, cardiomyopathy, and pulmonary involvement [2].
The present case report highlights two important points. Firstly, it underscores the challenges in diagnosing PLEVA in a primary care setting. PLEVA's clinical resemblance to other conditions and its rarity led to the initial misdiagnosis of diseases like guttate psoriasis, varicella zoster, pityriasis rosea, and secondary syphilis. Table 1 lists the clues that can help distinguish other diseases from PLEVA.
Secondly, there are challenges in delivering appropriate management for patients with PLEVA. Even though, PLEVA is usually a self-limiting disease, many patients experience itching, discomfort, and recurrence of their skin lesions, which can impact their quality of life. A recent systematic review recommended narrow-band ultraviolet B (UVB) phototherapy as the first-line therapy due to its high rate of complete remission. Oral erythromycin with or without topical corticosteroids, and low-dose methotrexate are recommended as second-line therapy [10]. In addition, methotrexate plays an important role in treating refractory PLEVA and FUMHD [2,4]. Acitretin, dapsone, and cyclosporine may help as additional therapies for refractory PLEVA whereas for cases of FUMHD, systemic immunomodulators may be beneficial [2].

Conclusion

This case highlights the diagnostic dilemma posed by PLEVA and its mimickers, where clinical presentation alone may not be sufficient for an accurate diagnosis. A high index of suspicion, coupled with histopathological examination, is essential for distinguishing PLEVA from other similar conditions and guiding appropriate management.

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Conflict of interest
All authors declare no conflicts of interest.

Authors contribution:
1) Mohd Aizuddin Abd Rahman: Writing, Editing, Literature Review, Supervision.
2) Nurjasmine Aida Jamani: Writing, Editing, Literature Review, Supervision.
3) Sarah Abdul Halim: Writing, Editing, Literature Review, Supervision.
4) Norsafina Zainun: Writing, Editing, Literature Review.

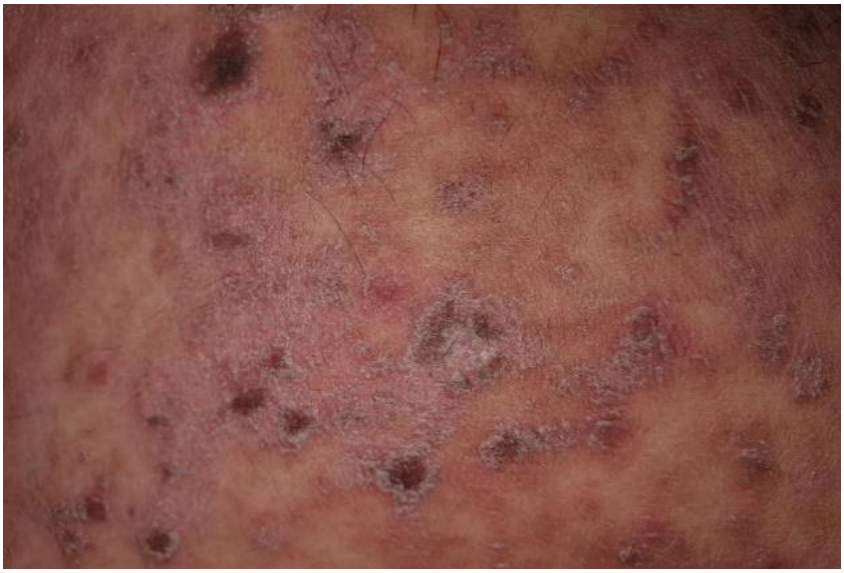

Figure 1. Generalized brownish scales with patches.

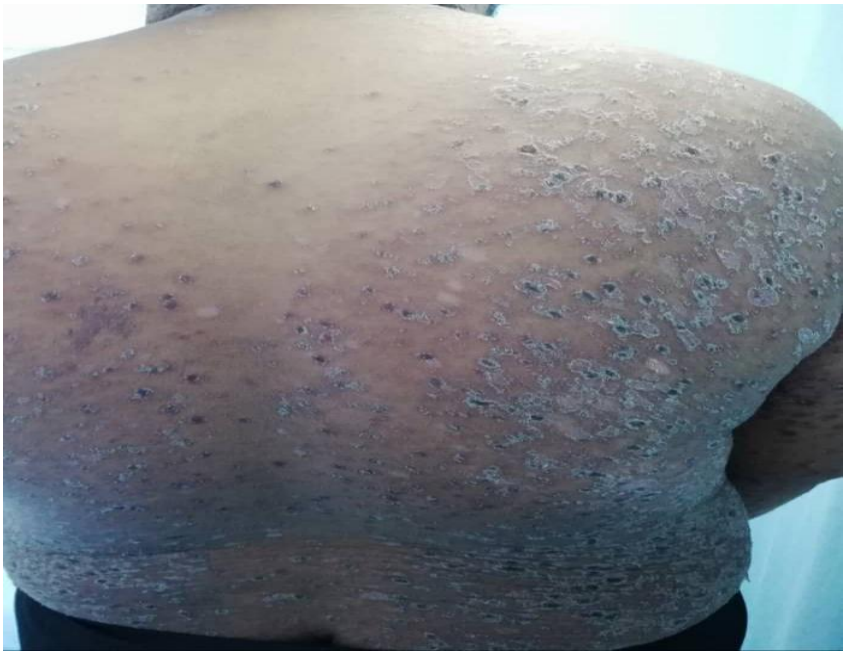

Figure 2. Generalized crusted lesions with central necrosis.

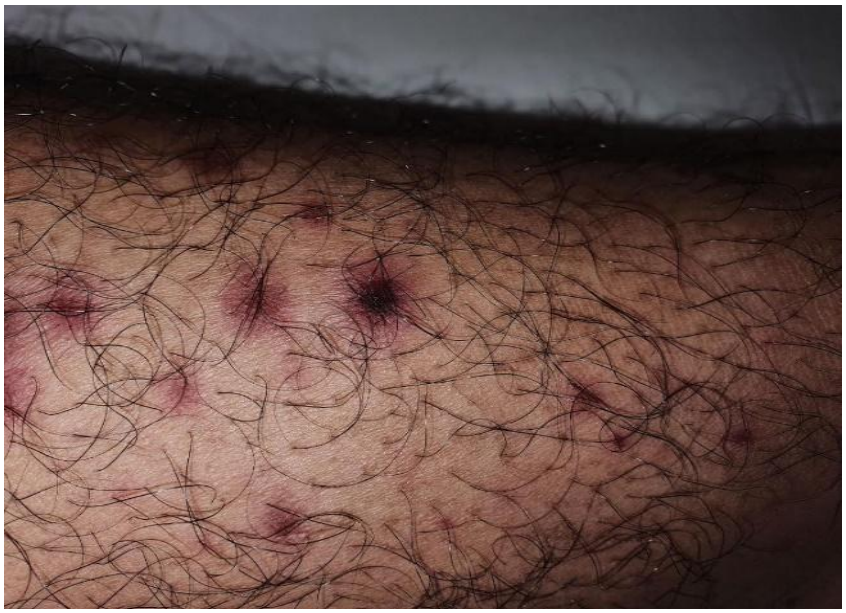

Figure 3. Erythematous macules and papules with dried-up blisters seen together with necrotic and a few hemorrhagic at the center of the crust.

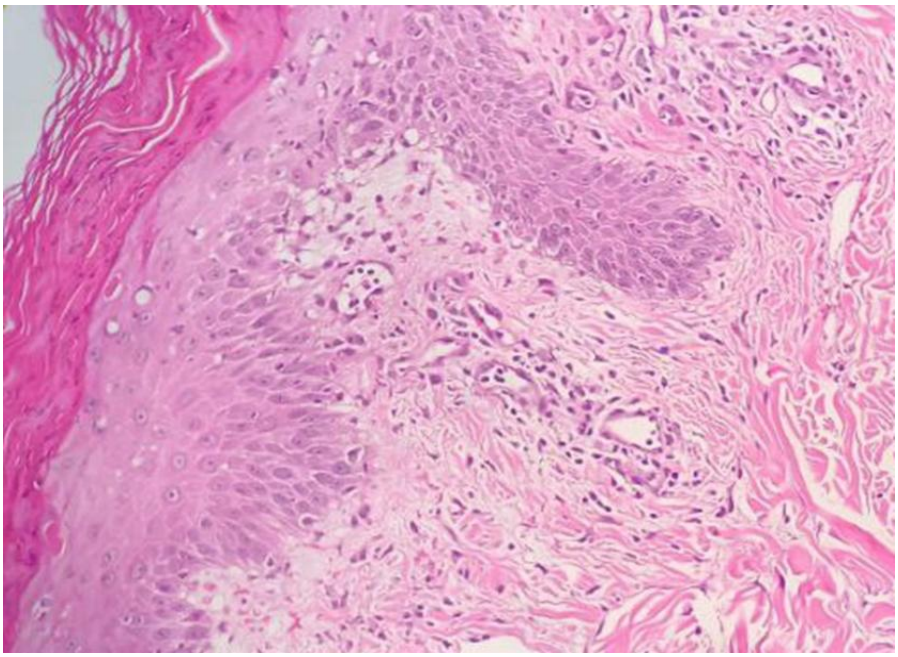

Figure 4. The section shows subepidermal blisters with lichenoid change and interface dermatitis. (Haematoxylin & eosin x100)

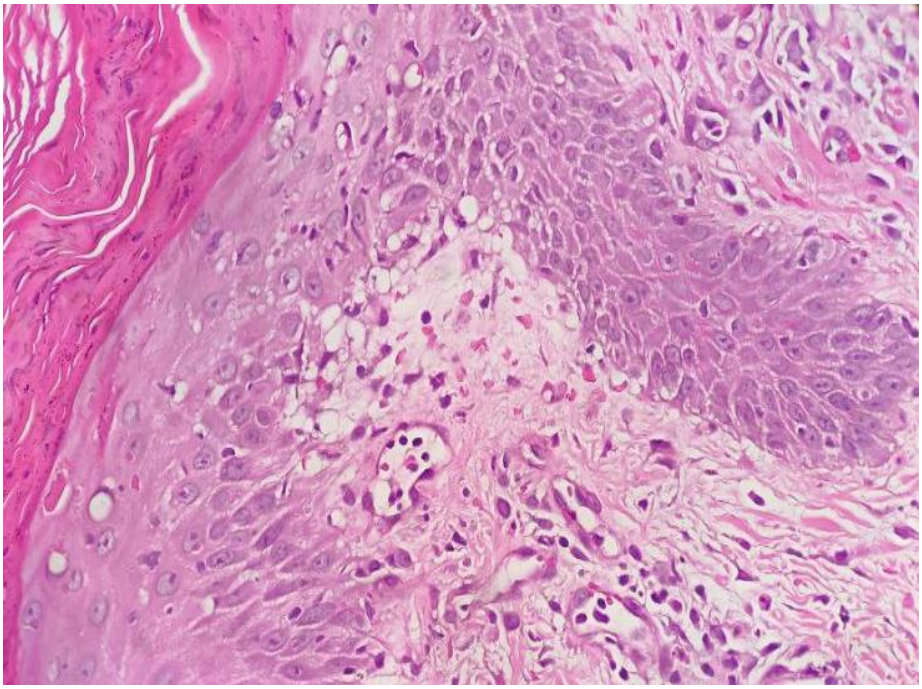

Figure 5: Focal epidermotropism is present. (Haematoxylin & eosin x 200)

Table 1. Clues that help distinguish other diseases from PLEVA.

Disease	**Clinical Findings on Skin**	**Diagnostic test**	**Treatment strategies**
Guttate psoriasis	Guttate psoriasis appears as tiny, erythematous papules and plaques, similar to PLEVA, but may also involve nails (6).	Clinical diagnosis	a) Narrowband UVB Phototherapy b) Topical corticosteroids as adjunct
Varicella	Varicella is frequently likened to a dewdrop on a rose petal and typically manifests as crusted lesions on the sixth day of the illness, resembling PLEVA. Nevertheless, the duration of the disease is briefer than PLEVA and normally lasts for two weeks (7).		a) Self-limited disease b) Symptomatic treatment c) Intravenous antiviral therapy is indicated in complicated cases of varicella.
Pityriasis rosea	The herald patch, "Christmas tree" pattern distribution, and collarette of scale are key features that distinguish pityriasis rosea from PLEVA. (8).		a) Self-limited disease b) Symptomatic treatment
Secondary syphilis	Classically, the rash is a generalized, non-itchy, and symmetrical macular or papular eruption that affects the entire body, including the palms and soles (9).	Specific treponemal tests (TPHA/TPPA/EIA)	a) Intramuscular Benzathine penicillin 2.4 mega units in a single dose

References

[1]. Ersoy-Evans S, Greco MF, Mancini AJ, Subaşı N, Paller AS. Pityriasis lichenoides in childhood: A retrospective review of 124 patients. J Am Acad Dermatol [Internet]. 2007 Feb;56(2):205–10. Available from: https://linkinghub.elsevier.com/retrieve/pii/S0190962206022663

[2]. Fayven Teklehaimanot; Anita Gade; Richard Rubenstein. Pityriasis Lichenoides Et Varioliformis Acuta (PLEVA). Last Update: January 2, 2023. NIH https://www.ncbi.nlm.nih.gov/books/NBK585135/

[3]. Jastrząb BA, Stefaniak AA, Hryncewicz-Gwóźdź A, Nockowski P, Szepietowski JC. Pityriasis Lichenoides et Varioliformis Acuta Triggered by Human Papillomavirus Vaccine: A Case Report and Literature Review. Acta Derm Venereol. 2021 Sep 17;101(9):adv00552.

[4]. Merlotto MR, Bicudo NP, Marques MEA, Marques SA. Pityriasis lichenoides et varioliformis acuta following anti-tetanus and diphtheria adult vaccine. An Bras Dermatol. 2020 Mar-Apr;95(2):259-260.

[5]. Moriwaki, M., Tanaka, A., Tanaka, M., Takahagi, S. and Hide, M. (2018), Pityriasis lichenoides et varioliformis acuta after vaccination. J Cutan Immunol Allergy, 1: 37-38. https://doi.org/10.1002/cia2.12002

[6]. Saleh D, Tanner LS. Guttate Psoriasis. [Updated 2023 Jul 31]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK482498/

[7]. Ayoade F, Kumar S. Varicella-Zoster Virus (Chickenpox) [Updated 2022 Oct 15]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK448191/

[8]. Gay JT, Huq ME, Saleh HM, et al. Herald Patch. [Updated 2024 Feb 15]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK513301/

[9]. Whiting C, Schwartzman G, Khachemoune A. Syphilis in Dermatology: Recognition and Management. Am J Clin Dermatol. 2023 Mar;24(2):287-297. doi: 10.1007/s40257-022-00755-3. Epub 2023 Jan 23. PMID: 36689103; PMCID: PMC9869822.

[10]. Bellinato F, Maurelli M, Gisondi P, Girolomoni G. A systematic review of treatments for pityriasis lichenoides. J Eur Acad Dermatol Venereol. 2019;33(11):2039–49. doi:10.1111/jdv.15813.