International Journal of Health Sciences and Research
Volume 14; Issue: 12; December 2024
Website: www.ijhsr.org
Case Report *ISSN: 2249-9571*

Mucha-Habermann Disease Complicated by Post-Treatment Scabies Pruritus: A Case Report

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DOI: https://doi.org/10.52403/ijhsr.20241229

ABSTRACT

Mucha-Habermann disease, or Pityriasis Lichenoides et Varioliformis Acuta (PLEVA), is a rare dermatological illness marked by the abrupt emergence of reddish-brown papules, some of which may develop into necrotic lesions. Misdiagnosis of PLEVA can lead to unnecessary treatments and adverse symptoms.

A 13-year-old boy was receiving ongoing scabies treatment. He came to the hospital with worsening skin lesions and systemic symptoms like headaches, weakness, and a burning sensation in the affected areas. Examining his arms, thighs, and trunk revealed lesions at different phases of development, such as tiny papules, vesicles, and necrotic-crusted patches. The diagnosis was confirmed by a skin biopsy that showed perivascular lymphocytic inflammation, spongiosis, and parakeratosis—all of which are characteristic characteristics of PLEVA. The patient's condition was made worse by the misdiagnosis and prolonged scabies treatment, which also resulted in excruciating itchiness. Erythromycin, triamcinolone, antihistamines, and topical steroids were provided to the patient. Significant progress was seen after two weeks, and the active lesions disappeared in a month, leaving behind varioliform scars and patches with changed pigmentation.

This case underscores how crucial it is to make an appropriate diagnosis when a patient presents with a chronic or uncommon dermatological illness. In addition to delaying required treatment, misdiagnosing PLEVA as a more prevalent illness, like scabies, can cause needless difficulties and patient suffering.

Keywords: Pityriasis Lichenoides et Varioliformis Acuta, Mucha-Habermann disease, post-treatment scabies pruritus, Pityriasis Lichenoides Chronica.

INTRODUCTION

Mucha-Habermann disease (MHD), also known as Pityriasis lichenoides et varioliformis acuta (PLEVA), is a dermatological condition characterized by erythematous papules and macules, typically on the trunk and flexural portions of the limbs. [1] It is a benign condition of unknown etiopathogenesis. It is more common in paediatric patients and young adults. PLEVA may be an immune complex-mediated hypersensitivity, an inflammatory response secondary to T-cell dyscrasia, or an inflammatory reaction brought on by specific infectious pathogens. Pityriasis lichenoides chronica could be interpreted as a drug reaction or a paraneoplastic syndrome. [2, 3] Three

subtypes - pityriasis lichenoides et varioliformis acuta, pityriasis lichenoides chronica (PLC), and febrile ulceronecrotic Mucha-Habermann disease - are used to categorize the range of appearances. [4]

The hallmark of PLEVA is the initial appearance of 3-5 mm diameter papulosquamous lesions or erythematous-squamous plaques, which are typically covered in fine scales and may combine to create plaques that afflict the trunk and extremities. Vesicles or pustules develop over the papules as the condition worsens. These umbilicate and develop into haemorrhagic necrosis, which has purpuric and crusty patches that, when removed, expose necrotic ulcers. In addition to leaving a varioliform scar and healing in a few weeks, necrotic lesions can occasionally cause pruritus alone. Dermatitis and perivascular lymphocytic infiltrates, together with bleeding, are hallmarks of PLEVA histology. Although lymphocytic vasculitis (LV) is known to exist, it has seldom been shown. The main characteristic of LV is fibrinoid necrosis of the vessel walls with a preponderance of lymphocytic infiltration.

The objective of this case report is to discuss although a skin biopsy specimen's histologic analysis is the gold standard for identifying PLEVA, a conclusive diagnosis can be challenging as Pityriasis Lichenoides (PL) is easily mistaken for other disorders, so it is important to appropriately identify it and recognise the types that exist using the current categorization; so we can guide a diagnosis by defining its clinical presentations. The primary goal of treatment is to reduce pruritus symptoms. The majority of available treatments only have a temporary or restricted impact. Phototherapy and topical corticosteroids are the cornerstones of treatment. Antibiotics, topical tacrolimus, methotrexate, and cyclosporine are further therapies. [4-7]

CASE PRESENTATION

A 13-year-old Caucasian male was initially being treated for scabies with permethrin for one month. His skin problem worsened over time despite treatment, so his parents sought additional medical care. In addition to systemic symptoms like headaches, weakness, and a burning feeling in the affected areas, the patient arrived at the hospital with worsening skin lesions. Upon physical examination, the patient had many lesions on the arms, thighs, and trunk that were in various stages of growth. Reddish-brown papules, vesicles, and necrotic materials—many of which had brown crusts on them—were among the lesions. (Fig. 1). These results raised the possibility of a different diagnosis, which prompted more research. Laboratory testing showed that the liver and kidneys were functioning well, and the total blood count was within normal limits. Nonetheless, there remained underlying inflammation shown by the high erythrocyte sedimentation rate (ESR) of 29 mm/hr. The diagnosis of Pityriasis Lichenoides et Varioliformis Acuta (PLEVA) was confirmed by histological investigation following a skin biopsy. Parakeratosis, spongiosis, dyskeratosis, and lymphocytic infiltration in the dermis were among the distinctive observations. Interestingly, pruritus from previous scabies management was also seen after therapy (Figure 2). Following hospitalization, the patient was prescribed 200 mg of erythromycin four times a day, 16 mg of triamcinolone daily, antihistamines, and topical steroids. After just two weeks of treatment, there was a noticeable improvement. Over the course of the next month, the dosage was progressively reduced. The patient had varioliform scars and patches of hyperpigmentation and hypopigmentation when the active lesions resolved (Figure 3). The patient had no active lesions and showed no signs of recurrence at his one-month follow-up. He still has routine follow-up appointments for ongoing observation. This instance emphasizes how difficult it can be to distinguish between dermatological disorders that share clinical characteristics and how crucial a comprehensive diagnostic

diagnosis is to directing successful treatment.

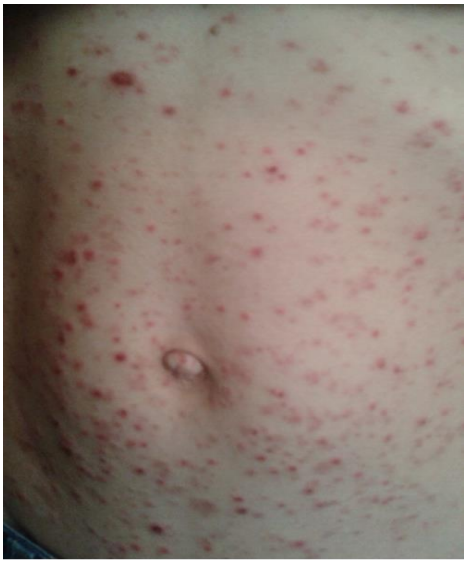

Figure 1: Pre-Treatment Skin Lesions.

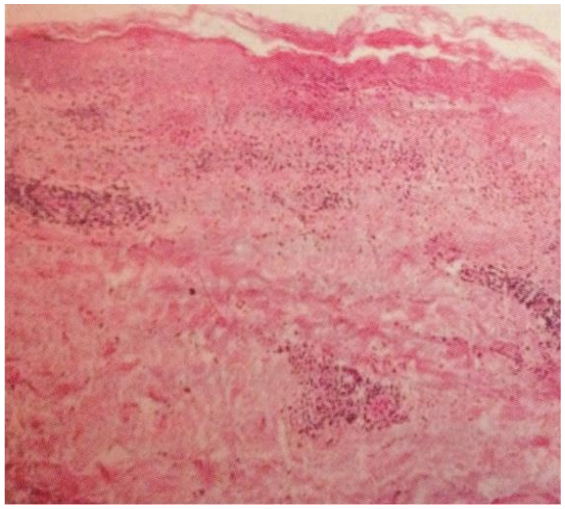

Figure 2: Histopathological findings of PLEVA revealing parakeratosis, spongiosis, and perivascular lymphocytic infiltrations.

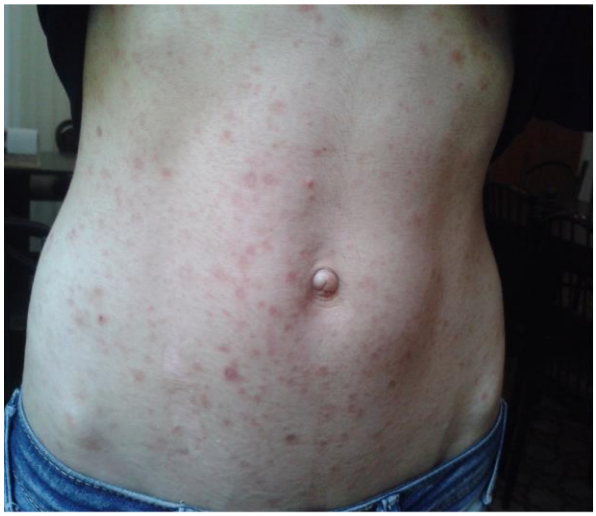

Figure 3: Post-Treatment Skin Condition.

DISCUSSION

We present a case report demonstrating the complexities involved in diagnosing and managing Pityriasis Lichenoides et Varioliformis Acuta (PLEVA) in paediatric patients, particularly when initial clinical presentations may resemble those of more prevalent dermatological conditions. The 13-year-old male patient initially demonstrated symptoms including pruritic, and inflammatory skin lesions, before being diagnosed with scabies due to overlapping symptoms such as erythematous papules and itching. Scabies, caused by Sarcoptes scabiei mites, is characterized by pruritic papules, pustules, vesicles, and linear burrows that worsen at night. [8] The patient received treatment with permethrin, a topical antiparasitic drug frequently used to treat scabies. However, the absence of linear burrows, combined with a lack of improvement and worsening of symptoms following therapy, necessitated a re-evaluation and consideration of differential diagnosis. [9] Histopathological analysis and clinical evaluation ultimately led to the diagnosis of PLEVA, also known as Mucha-Habermann disease. As highlighted in Table 1, the distinguishing clinical and histopathological features of PLEVA, such as reddish-brown papules, vesicles, and necrotic material with a burning sensation and central necrosis, and histopathology findings of parakeratosis, lymphohistiocytic infiltrates, and epidermal necrosis, ultimately guided the diagnosis. [9, 10]

Table 1: Comparison of Clinical, Histopathological, and Management Features of Scabies and PLEVA. [9, 10]

FEATURE	SCABIES	PLEVA (Pityriasis Lichenoides et Varioliformis Acuta)
Etiology	Caused by Sarcoptes scabiei mites (parasitic infestation).	Likely immune-mediated, potentially triggered by infections or hypersensitivity reactions.
Age Group	All age groups, more common in children.	Late childhood and early adulthood. Slight male predominance.
Clinical Presentation	Intense itching (worse at night), erythematous papules, and linear burrows.	Reddish-brown papules, vesicles with necrotic centers, and burning sensation.
Histopathology	Burrows containing mites, eggs, and feces; inflammatory infiltrates.	Parakeratosis, lymphohistiocytic infiltrates, and epidermal necrosis.
Treatment	Topical antiparasitic agents (e.g., permethrin).	Oral antibiotics (e.g., erythromycin), topical corticosteroids, and antihistamines.
Prognosis	Resolves quickly with appropriate treatment.	Chronic course; scarring and pigmentation changes may persist despite improvement.

PLEVA, a rare inflammatory skin condition of unknown etiology, is predominantly observed in late childhood and early adulthood, with a slight male prevalence. [11] It manifests in 3 forms: pityriasis lichenoides chronica (PLC), pityriasis lichenoides et varioliformis acuta (PLEVA), and the febrile ulcero-necrotic Mucha-Habermann disease (FUMHD) variant of PLEVA, which can develop from one another or exist independently. The pathogenesis of PLE

VA is believed to involve an atypical hypersensitivity reaction triggered by a multiplicity of infections, including Epstein-Barr Virus(EBV), human immunodeficiency virus (HIV), varicella-zoster virus (VZV), Toxoplasma gondii, Group A streptococcus, Herpes simplex (HSV) type 2, hepatitis B. [9-14] Additionally, certain medications including Monoclonal antibodies like atezolizumab/pembrolizumab, anti-TNF inhibitors, statins, antidepressants–and vaccines like HPV, influenza, tetanus, MMR, & COVID-19 have also been implicated. [9, 11, 12, 13, 14]

Studies have revealed elevated immune complex deposition, particularly IgM and C3, within the skin lesions, suggesting a potential link to hypersensitivity or immune complex-mediated process.[15] Furthermore, some theories propose that T-cell dyscrasia,

an abnormality or dysfunction in T-cells, may contribute to the pathogenesis of PLEVA, potentially increasing its malignant potential. [16, 17] CD8+ cytotoxic/ suppressor T cells are thought to cause significant epidermal damage and necrosis. [16, 17]

The management of PLEVA necessitates a multidisciplinary approach to control the condition effectively. Treatment aims to prevent lesion formation and control inflammation. First-line therapies include topical corticosteroids and oral antibiotics like erythromycin, with Azithromycin as an alternative in resistant cases [18, 19] Phototherapy, such as narrowband UVB, has shown efficacy in paediatric cases. [19] In this clinical case, the patient exhibited a favorable response to a treatment regimen comprising erythromycin in combination with triamcinolone (a systemic corticosteroid), antihistamines, and topical corticosteroid ointments, leading to significant improvement within 14 days, followed by a gradual dose reduction over the following month. Continued monitoring is recommended due to rare cases of progression. Post-treatment observations revealed residual varioliform scarring and areas of hyper- and hypopigmentation, consistent with PLEVA's chronic nature and potential for recurrence. PLEVA generally has a good prognosis and may resolve spontaneously. Monthly follow-ups have been scheduled to monitor for any signs of relapse or persistent inflammation. Although the patient's condition is stable with no new lesions, ongoing medical supervision remains critical to ensure effective long-term disease management and timely intervention if necessary.

This case highlights the importance of early and accurate diagnosis, timely intervention, and diligent follow-up in managing PLEVA. Prompt recognition and appropriate treatment prevent complications and alleviate the physical and psychological burden on patients. It emphasizes the need for dermatologists to remain vigilant and consider rare conditions like PLEVA in atypical or refractory presentations.

Limitations of the case report include the lack of documentation regarding the patient's past medical and family history, which could have revealed potential predispositions to PLEVA. Details about the diagnostic methods used to confirm scabies and the exact permethrin treatment regimen were also absent, raising concerns about the accuracy of the initial diagnosis and its management.

CONCLUSION

The case report discusses the dermatological condition Pityriasis Lichenoides, which primarily affects children. PLEVA frequently has a short time course, whereas PLC has a longer time course and may be characterized by a smoldering phase with pigmentary abnormalities in the absence of other indications of active inflammation. Compared to the general paediatric population, our sample of children with PL did not exhibit a greater prevalence of malignancies. Acute and chronic lesions in the same individual have led to speculation that they are related to and may be a component of a continuous spectrum of illnesses. [20] More studies are needed to assess treatment outcomes over a longer follow-up period, and there is presently not enough evidence to create a strategy for the treatment of PLC. In this case, cyclosporine and NB-UVB phototherapy might work well together to treat PLC. Further research with longer follow-ups is required to confirm the safety and efficacy of this combo medication.

Declaration by Authors

Human subjects: Consent was obtained or waived by all participants in this study. Conflicts of interest: In compliance with the ICMJE uniform disclosure form, all authors declare the following: Payment/services info: All authors have declared that no financial support was received from any organization for the submitted work. Financial relationships: All authors have declared that they have no financial relationships at present or within the

previous three years with any organizations that might have an interest in the submitted work. Other relationships: All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

Acknowledgement: None

Source of Funding: None

Conflict of Interest: The authors declare no conflict of interest.

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How to cite this article: Ranasinghe Arachchige Dona Kashmira Nawodi Weerasekara, Jawad Mchaik, Madihah Osman, Shaan Sinojia Kiritkumar. Mucha-Habermann disease complicated by post-treatment scabies pruritus: a case report. *Int J Health Sci Res.* 2024; 14(12):236-242. DOI: *10.52403/ijhsr.20241229*

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