WORLD JOURNAL OF PHARMACEUTICAL RESEARCH
SJIF Impact Factor 8.453
Volume 14, Issue 9, 845-852. Case Study ISSN 2277–7105

CASE REPORT ON PITYRIASIS LICHENOIDES ET VARIOLIFORMIS ACUTA

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Article Received on
01 April 2025,

Revised on 22 April 2025,
Accepted on 12 May 2025

DOI: 10.20959/wjpr20259-36503

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ABSTRACT

Background: Pityriasis Lichenoides et Varioliformis Acuta (PLEVA) is a rare inflammatory skin disorder characterized by the sudden onset of erythematous papules that may undergo necrosis or ulceration. While generally self-limiting, severe cases can present with systemic involvement, posing diagnostic challenges and requiring a multidisciplinary approach. **Case Presentation:** A male patient presented with a one-month history of painful, erythematous, and black-colored lesions over the bilateral thighs, upper and lower limbs, trunk, scalp, and ankles, associated with mild itching. Lesions began on the left shin and progressively spread to other areas. Some lesions developed central necrosis within two days. The patient also reported bilateral pedal edema and severe pain in the lower limbs. There was no history of fever, recent drug intake, or systemic prodrome. A punch biopsy from the left thigh was performed. **Key Findings:** Biopsy confirmed the diagnosis of PLEVA. Blood pressure was elevated and managed with antihypertensives including Amlodipine, Cilacar, Nebivolol, and supportive medications. Cardiology evaluation showed good biventricular systolic function with no RWMA. Ophthalmologic examination revealed a single dot hemorrhage, likely secondary to hypertension. Surgery consultation was sought for severe lower limb pain, edema, and suspected PVOD. The patient was treated with systemic corticosteroids (Inj. Betnesol), antibiotics (Ciplox TZ, Metrogyl), antiplatelets (Ecosprin), vasodilators (Isordil), and Pentoxiphyllin. **Conclusion:** This case highlights an unusual, extensive presentation of PLEVA with systemic symptoms requiring a multidisciplinary approach. Early diagnosis through biopsy and prompt supportive care are essential to prevent complications and ensure optimal patient outcomes.

KEYWORDS: Pityriasis Lichenoides et Varioliformis Acuta, necrotic skin lesions, punch biopsy, systemic symptoms.

INTRODUCTION

Pityriasis Lichenoides et Varioliformis Acuta (PLEVA), also referred to as Mucha-Habermann disease, is a rare, self-limiting inflammatory dermatosis that belongs to a spectrum of pityriasis lichenoides disorders. It primarily affects children and young adults and is characterized by the abrupt onset of papular, vesicular, and necrotic skin lesions that may ulcerate and crust.[1] PLEVA represents the acute end of the spectrum, with pityriasis lichenoides chronica (PLC) representing the more indolent, chronic form.[1]

PLEVA typically presents with a rapid onset of lesions, which can be varied in appearance. The clinical course is often self-limiting, with a duration of several weeks to months, though some patients may experience recurrent episodes or chronic symptoms. The condition may resolve spontaneously in some cases, while in others, it may progress to chronic forms or recur periodically.[1,2]

While PLEVA can affect individuals of all ages, it is most commonly observed in children and young adults, particularly between the ages of 10 and 30 years. There is no significant gender predilection, although some studies suggest a slight male predominance.[1,3]

The etiology of PLEVA is not completely understood. However, proposed mechanisms include immune dysregulation, hypersensitivity reactions to infectious agents, and T-cell lymphoproliferative processes.[1,2] Various pathogens, such as Epstein-Barr virus, cytomegalovirus, and parvovirus B19, have been implicated in triggering the condition, though a definitive link remains elusive.[3] PLEVA has been associated with several environmental and infectious triggers, including viral infections like Epstein-Barr virus (EBV) and cytomegalovirus (CMV). In some cases, vaccinations such as the measles-rubella vaccine have been linked to the onset of the disease, suggesting a potential role for immune modulation and hypersensitivity reactions.[2,3]

Clinically, patients present with crops of erythematous macules and papules, some of which develop into vesicles or pustules, and subsequently crust or ulcerate. Lesions are often polymorphic and distributed primarily over the trunk and extremities.[4]

Histologically, PLEVA is characterized by parakeratosis, necrotic keratinocytes, red blood

cell extravasation, and a dense superficial perivascular lymphocytic infiltrate, typically composed of CD8+ T cells.[1,5] The condition can resemble several other dermatologic and infectious diseases, necessitating histopathologic confirmation for accurate diagnosis.

Treatment options are empirical and based on case reports due to the rarity of the disease. Although PLEVA is typically self-limiting, it can have a chronic or relapsing course in some individuals. Treatment options vary and are often tailored to the severity of the disease, with the primary goal being the reduction of inflammation and the prevention of scarring or secondary infections. Relapse or chronicity is more common in cases with extensive involvement or in patients who have not received appropriate treatment.[1,6]

Antibiotics such as erythromycin and tetracyclines, systemic corticosteroids, phototherapy, and immunosuppressants like methotrexate have all been used with variable success.[1,6] The prognosis is generally good, although relapses can occur.

CASE PRESENTATION

Basic information of the patient

Name-XYZ

Age-60

Gender-Male

Socioeconomic status-Middle class

Chief Complaints

History of present illness: C/O Multiple lesions – Erythematous, painful and black coloured over b/l thighs, lower limbs, upper limbs, front and back of trunk associated with pan and mild itching* 1 month. Complaints of multiple reddish and black coloured painful lesions over scalp, bl upper limbs, front of trunk, right palm, b/l lower limbs for past one month. Lesions started as reddish papules and enlarged in size. Some of the lesions underwent central necrosis after 2 days. Initially lesions started on the left shin, which progressed to involve entire lower limb and other sites. Patient c/o pain over each lesion over b/l ankles.

C/O bilateral pedal edema for past 3 weeks

Family History- Nil

Allergy- Ibuprofen

On Examination

Patient conscious and oriented

Vitals

BP:130/90 mmHg

PR:75/min

RR:16/min

SYSTEMIC EXAMINATION

CVS: S1 and S2 heard normal, no murmurs

Resp: Air entry bilaterally equal. Clear, NVBS

CNS: Higher mental functions normal, no focal neurologic deficits

P/A: Soft, non-tender, bowel sounds present

DERMATOLOGIC EXAMINATION

Skin

Multiple erythematous discrete papules, nodules and plaques present over bilateral upper limb, front of trunk, bilateral, lower limb, right palm and scalp

Multiple discrete erythematous plaques and nodules with necrotic lesion present over upper limb, front of trunk, bilateral lower limb

Mucosa

Oral cavity: normal

Ocular mucosa: normal

Genital mucosa: normal

Scalp and hair- single excoriated papule

Palm- Right palm: a single erythematous papule present

Soles- Normal

Nails- Normal

External genitalia- Normal

Lower limb – B/L Pitting pedal edema present

INVESTIGATIONS

Abnormal ranges shown

HB	12.3
PCV	34
TOTAL WBC	11200
DC POLY	89
DC LYMPH	8
DC EOSINOPHILS	0

Punch biopsy from lesion skin (27/12/2024)- Histological features favours Pityriasis lichenoides et varioliformis acuta

HISTOPATHOLOGY REPORT

Epidermis shows mild hyperkeratosis, parakeratosis with blister formation in cornified layer.

DEJ shows vacuolar alteration of basal layer and exocytosis of lymphocytes.

Papillary dermis shows edema and extravasated erythrocyte.

Dermis shows extensive perivascular lymph histiocytic infiltrate admixed with neutrophils extending to reticular dermis.

IMP- PLEVA

TREATMENT

NAME OF MEDICINE	DOSE AND FREQUENCY
Inj. Ceftriaxone	1gm , 1-0-1
Tab. Atarax	10mg, 0-0-1
Tab. Lyser Forte	10mg, 1-0-1
Tab.Pletoz	50mg, 0-1/2-0
Tab. Limcee	500mg, 1-0-0
Tab.Clopilet A	75/150mg, 1-0-0
Tab.Paracetamol	650mg, sos
Tab. Pantop	40mg, 1-0-0
Tab. Dolo	650mg, 1-1-1
Tab. Ciplox TZ	500/600mg, 1-0-1
Tab. Metrogyl	400mg, 1-1-1
Tab. Ecospirin	75mg, 0-1-0
Tab. Pentoxiphylline	400mg, 1-0-1
Tab. Cilacar	5mg, 1-0-0
Tab. Atorva	10mg, 0-0-1
Tab. Ciplox	500mg, 1-0-1
Tab. Nebi	2.5mg, 1-0-0

Inj. Betnesol	8mg, 1-1-1
Betnovate GM cream	L/A ,0-0-1
Dermasoft lotion	L/A, 1-0-1

DISCUSSION

PLEVA presents both diagnostic and therapeutic challenges. Given its rarity and overlap with other dermatological conditions, timely recognition and diagnosis are essential to initiate appropriate management and rule out more serious disorders.

Etiology and Pathogenesis

The etiology of PLEVA is multifactorial and still under investigation. Theories suggest a hypersensitivity reaction to infectious agents, including viruses such as EBV and CMV, or a possible lymphoproliferative origin due to the predominance of CD8+ T cells in histological samples.[1,2] It has also been reported post-vaccination, such as following the measles-rubella vaccine, further supporting the hypothesis of an immune-mediated trigger.[2]

The immune response likely targets altered keratinocytes or viral antigens present in the skin, resulting in necrosis and inflammatory infiltrates. The presence of apoptotic keratinocytes, vascular damage, and extravasation of erythrocytes further supports the immune-mediated destruction seen in histopathology.[1,5]

Clinical Manifestations and Diagnosis

PLEVA typically manifests as crops of erythematous macules and papules, which may become vesicular or necrotic and develop into crusted or ulcerated lesions. The lesions are often in different stages of evolution and primarily appear on the trunk and proximal limbs. Pruritus may be present, though it is not always a dominant symptom.[3,4]

Histologically, key features include focal parakeratosis, necrotic keratinocytes, a superficial perivascular lymphocytic infiltrate, and occasional vasculitis. Immunohistochemistry reveals a CD8+ T-cell predominance.[1] Differential diagnoses include lymphomatoid papulosis (which features CD30+ cells), varicella, scabies, secondary syphilis, and drug eruptions.[5] Accurate diagnosis requires a combination of clinical examination and skin biopsy.

Treatment Approaches

There are no universally accepted treatment guidelines for PLEVA due to its rarity and variability in clinical presentation. Treatments are primarily supportive and aimed at

symptom control. Antibiotics like erythromycin and tetracycline have been effective in mild to moderate cases, possibly due to their anti-inflammatory properties.[1,6]

Systemic corticosteroids are often used in severe or widespread disease to suppress inflammation and promote lesion resolution. Phototherapy, especially narrow-band UVB (NB-UVB), has shown efficacy in inducing remission in chronic and recurrent cases.[1,6]

In refractory or severe cases, methotrexate and cyclosporine have been used successfully. Immunomodulatory agents may be considered in cases where conventional treatments fail or in cases that suggest an underlying immune or lymphoproliferative disorder.[6]

Prognosis and Follow-Up

The prognosis of PLEVA is generally favorable. Many patients experience spontaneous resolution, although recurrence is not uncommon. Chronic or recurrent cases may require long-term follow-up and management. Importantly, PLEVA rarely progresses to more serious lymphoproliferative diseases, but vigilance is required, especially in cases with atypical features or poor response to treatment.[4]

Regular follow-up ensures monitoring for recurrence, treatment efficacy, and any signs suggestive of disease progression. Patient education is essential to manage expectations and ensure adherence to treatment regimens.

CONCLUSION

This case highlights the importance of early dermatological evaluation in chronic, painful skin lesions with ulceration by demonstrating an atypical manifestation of PLEVA in a 60-year-old guy. The intricacy of treating cutaneous inflammatory illnesses in the elderly is underscored by the lesions' development, systemic comorbidities such as hypertension, and the requirement for multispecialty teamwork. The diagnosis was made easier by a timely biopsy, and stabilization was guaranteed by supportive and focused systemic medication. To control underlying vascular insufficiency, monitor recurrence, and avoid complications, ongoing follow-up is crucial. This case emphasizes how crucial customized and integrated care is for uncommon dermatological disorders, especially in older adults.

ACKNOWLEDGEMENT

We would like to express our gratitude and regards to everyone who guided in the publish of this work.

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