

A CASE OF PITYRIASIS LICHENOIDES ET VARIOLIFORMIS ACUTA (PLEVA) LASTING FOR 22 YEARS-A SUBACUTE VARIANT?

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Sir,

Pityriasis Lichenoides et Varioliformis acuta (PLEVA) is an acute variant of Pityriasis Lichenoides which encompasses a spectrum of clinical presentations ranging from rapidly progressing severe ulceronecrotic variant with systemic manifestations and a high mortality rate (Febrile ulceronecrotic Mucha Haberman Disease- FUMHD) to a more chronic manifestation consisting of benign-appearing scaly papules of Pityriasis Lichenoides Chronica (PLC).¹

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. The maximum duration of PLEVA lasting in any person has been reported as long as 08 years.² Herein we report a case where it lasted for nearly 22 years.

A 25-year-old female presented to our center with complaints of recurrent crops of red raised lesions over her body, occurring at a frequency of once a week and then resolving spontaneously over a period of 10 to 15 days since the age of three years. There was no history of any pain or itching over the lesions. No history of fever or malaise associated with the occurrence of lesions. There was no history of weight loss, evening rise of temperature or night sweats. The patient denied any similar lesions in family members.

General and systemic examination was essentially normal. Dermatological examination revealed the involvement of face, trunk, and lower limbs in the form of few discrete erythematous papules, few vesiculopustules, with some papules showing a central necrotic eschar or crust, measuring 0.5 cm – 2 cm in diameter [Figures 1 & 2]. Examination of mucosa, palms, and soles was within normal limits.

Relevant hematological and biochemical parameters were within normal limits.

Histopathology showed characteristic features of necrotic keratinocytes with extravasation of RBC's into the epidermis. Papillary dermis was edematous and the capillaries were congested with RBCs. Superficial and the deep dermis showed endothelial swelling with foci of fibrinoid necrosis within the capillaries [Figure 3]. Perivascular chronic inflammatory infiltrates of lymphocytes was seen. Immunohistochemistry was



Figure 1 : Presence of few discrete erythematous papules, few with necrotic centre over the thigh

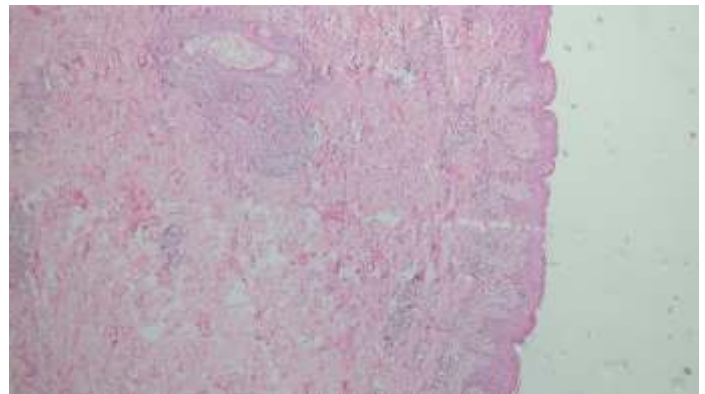


Figure 2 : H&E(100x)- scanner view showing predominant interface dermatitis with vacuolar degeneration and edematous papillary dermis

positive for CD3+, CD7+ and CD8+ and CD 30- [Figure 4]. Based on the above clinical, histopathological and immunohistochemistry a diagnosis of PLEVA was made.

The patient was managed with Cap Doxycycline 100mg twice daily along with 0.1% mometasone cream applied twice daily with which she has shown an excellent response in the form of no new lesions and regression of older lesions.

PLEVA is a disease of young people and children with inflammatory and usually diffuse erythematous papules of rapid onset which

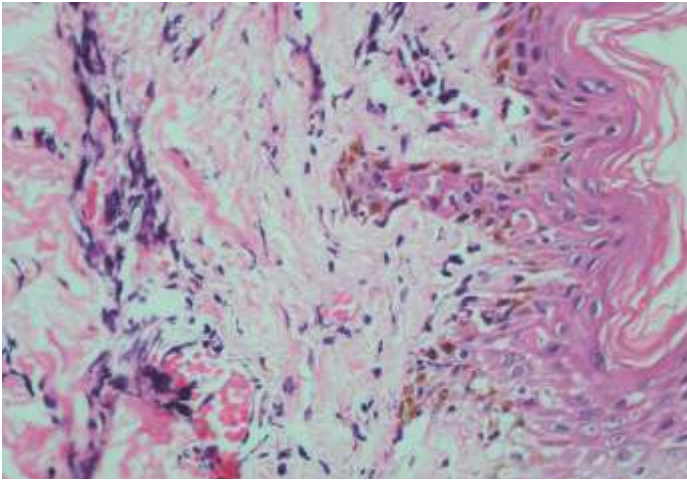


Figure 3 : H & E (200x) Necrotic keratinocytes with extravasation of RBCs into the epidermis with superficial and deep dermis showing endothelial swelling with foci of fibrinoid necrosis within the capillaries. Papillary dermis was edematous and capillaries were congested with RBCs.

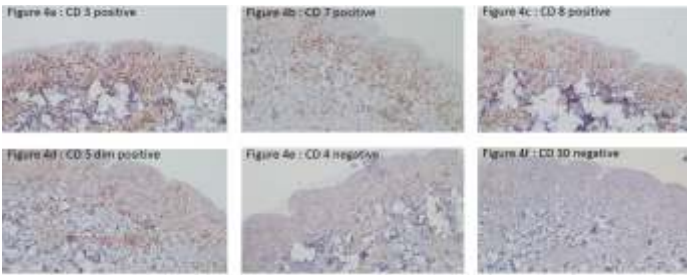


Figure 4(a -f) : IHC (200x) showed positivity for CD3+, CD7+, CD8+ and negative for CD 30-

evolve to form papules with a central punctum. The papule evolves to become vesicopustular, undergoes hemorrhagic necrosis and becomes ulcerated with overlying crusts. These lesions spontaneously heal to leave behind varioliform or smallpox-like scars and post-inflammatory hyperpigmentation and hypopigmentation. 1,2 Though the distribution of the lesions can be generalized, the trunk, flexors, and extremities are commonly involved.

The etiology of the disease still remains unknown though antibodies against *Toxoplasma gondii*, cytomegalovirus, parvovirus, adenovirus and Epstein Barr virus have been demonstrated in some studies. 1-3 T-cell dyscrasias and immune complex deposition have also been cited as probable causes in the literature.

The differential diagnosis for PLEVA includes lymphomatoid papulosis, arthropod bite reactions, varicella, Gianotti-Crosti syndrome, erythema multiforme, pityriasis rosea, guttate psoriasis, vasculitis, and secondary syphilis. Differentiation

between PLEVA and lymphomatoid papulosis is important as patients with lymphomatoid papulosis may develop systemic lymphoma. The presence of large CD30+ atypical lymphoid cells is the hallmark of lymphomatoid papulosis and clinically, the papules of lymphomatoid papulosis may develop into nodules, tumors, and large plaques.⁴

Clinical management of PLEVA is difficult due to exact pathology being unknown. Medications used till date include tetracycline, erythromycin, methotrexate, calciferol, chinoline and acridine derivatives, cyclosporine, intravenous gamma globulin, and retinoids.⁵ Systemic corticosteroids may have a role in severe cases of PLEVA. Topical corticosteroids and topical tacrolimus are helpful in symptomatic cases, but they do not alter the course of the disease. Despite a lack of randomized controlled trials, oral tetracycline and erythromycin have been prescribed most often in case series.

The existing literature outlines the mean duration of this rare disease to be 1.6 year with an episodic course, the frequency of which varies greatly from individual to individual.² The longest duration mentioned in literature is 8 years. Our case highlights a case PLEVA with a disease duration of 22 years with a subacute course which till now is the longest reported duration for the disease in literature.

We present this case to stress upon this rare presentation of PLEVA which can exist for years without any systemic manifestations and resolves spontaneously leaving behind post inflammatory hyperpigmentation. To give it a new nomenclature as "Subacute PLEVA" would not be wrong.

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