

## Remembering PLEVA: An Old Disease in Need of Renewed Attention

### Recordando a PLEVA: Uma Doença Antiga a Merecer Atenção Renovada

**Keywords:** Pityriasis Lichenoides/diagnosis  
**Palavras-chave:** Pitiríase Liquenoide/diagnóstico

We present the case of a 9-year-old boy with a history of chickenpox two years prior who presented to the emergency department with a two-week history of a rapidly progressive papular eruption. The lesions were erythematous, scaly, and intensely pruritic, initially developing on the dorsum and subsequently spreading to the thorax and genital areas, while sparing the lower limbs, scalp, and face. The dermatosis was suggestive of pityriasis lichenoides et varioliformis acuta (PLEVA). The mother declined skin biopsies, but laboratory workup including infectious serologies and TASO was performed, yielding normal results. The patient was treated with clarithromycin, with complete resolution of lesions within three weeks and no recurrence at two-month follow-up.

Pityriasis lichenoides is a rare self-limited autoimmune condition, with two main variants: pityriasis lichenoides chronica (PLC) and PLEVA. In rare cases, the disease progresses to febrile ulceronecrotic Mucha-Habermann disease (FUMHD), a severe variant, characterized by acute

symptoms, rapidly necrotic lesions, and high mortality, requiring urgent systemic treatment.<sup>1</sup>

The exact cause of PLEVA is unknown, but it is thought to represent an inflammatory or lymphoproliferative process triggered by infections, immune dysregulation, medications or vaccines. Its lymphoproliferative nature is suggested by the presence of clonal T-cell populations in some lesions, histological similarities to cutaneous T-cell disorders, and rare cases that evolve into lymphoma.<sup>2</sup>

Pytiriasis lichenoides et varioliformis acuta presents as successive crops of small erythematous macules and papules, predominantly on the trunk and flexural areas, often at different stages of evolution – an important diagnostic clue. Lesions may develop fine scale or central puncta, or progress to vesiculopustules with hemorrhagic necrosis and crusting, usually healing without scarring, although varioliform scars may occur. Systemic symptoms are uncommon. Dermoscopy usually displays three concentric zones of central brownish cloud, an intermediate ring of white scale, and a peripheral vascular ring.<sup>3</sup>

It typically affects children, posing a significant diagnostic challenge due to its resemblance to more common conditions such as chickenpox, lymphomatoid papulosis, Gianotti-Crosti syndrome and arthropod bites.<sup>4</sup> The absence of pustules, sparing of face and scalp, and



Figure 1 – Trunk showing discrete and confluent erythematous papules

characteristic necrotic papules favor PLEVA.

This condition may resolve spontaneously within weeks or months. Antibiotics like erythromycin or tetracycline are first-line options. Other options include topical steroids, phototherapy or systemic agents, used mainly for symptom control or refractory cases.<sup>5</sup>

The role of T-cell dyscrasia *versus* an inflammatory response to external antigens is still debated, with recent immunophenotyping studies showing a predominance of cytotoxic CD8+ T cells in PLEVA lesions, but without clear evidence of clonality. In the future, multiomics analysis will likely clarify the pathophysiology and etiology of this entity.<sup>6</sup>

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#### AUTHOR CONTRIBUTIONS

GAS: Study design, writing and critical review of the manuscript.

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All authors approved the final version to be published.

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#### PROTECTION OF HUMANS AND ANIMALS

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the Helsinki Declaration of the World Medical Association updated in October 2024.

#### DATA CONFIDENTIALITY

The authors declare having followed the protocols in use at their working center regarding patients' data publication.

#### PARENTAL CONSENT

Obtained.

#### CONFLICTS OF INTEREST

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