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Bullous Erythema Multiforme or Rowell Syndrome: A Case Report

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Abstract **Case Report**

Bullous erythema multiforme is characterized by the presence of target lesions with a bullous center, distributed bilaterally and relatively symmetrically on the extremities. Rowell's syndrome is a type of subacute cutaneous lupus erythematous that mimics erythema multiforme. These two diagnoses have a similar histopathology with interface dermatitis. To differentiate these two entities, we use the criteria of Rowell *et al*., Lee *et al*., Zeitouni *et al*., or Torchia *et al*., According to these criteria, the diagnosis of Rowell syndrome requires the presence of lupus lesions and antinuclear antibodies. In our case, the pathologist mistook erythema multiforme for subacute cutaneous lupus because of the presence of interface dermatitis. However, our patient did not meet any of the criteria for the diagnosis of Rowell's syndrome. The patient was treated as having erythema multiforme with a good outcome

Keywords: Erythema Multiforme, Rowell Syndrome, Histopathology, Criteria.

INTRODUCTION

Erythema multiforme can be confused with Rowell syndrome which is a type of subacute cutaneous lupus erythematous. Although histopathology is similar, certain criteria developed over the years have made it possible to distinguish between the two entities.

We report a case of bullous erythema multiforme that was initially mistaken for Rowell's syndrome on the basis of pathology

CASE REPORT

A 16-year-old female patient with approximately 5 recurrent episodes of orolabial herpes per year was admitted to the hospital for a bullous dermatosis occuring by recurrent outbreaks of bilateral and symmetrical acral target lesions unresponsive to acyclovir self-medication.

The rash for which she was hospitalized had not been preceded by orolabial herpes, the patient took no new medication and she had no pulmonary symptoms.

Skin examination revealed polycyclic target lesions with bilateral, roughly symmetrical acral distribution (figure 1).

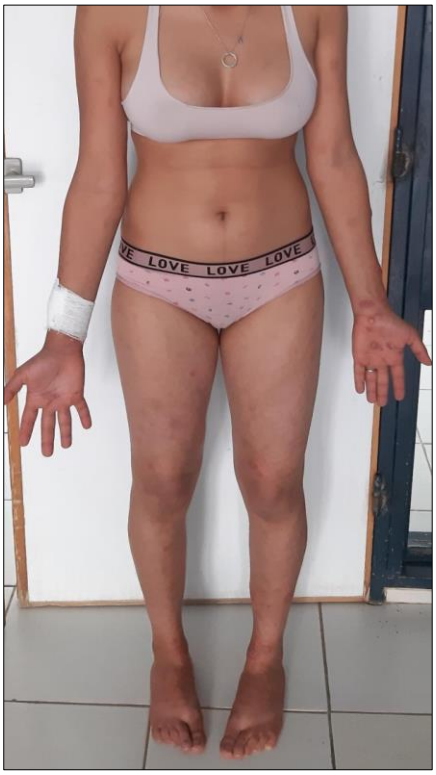

Figure 1: Polycyclic target lesions with bilateral, roughly symmetrical acral distribution

The target lesions have a papular, bullous or purpuric centre; a second embossed pale circle and a third erythematoviolaceous circle (figure 2).

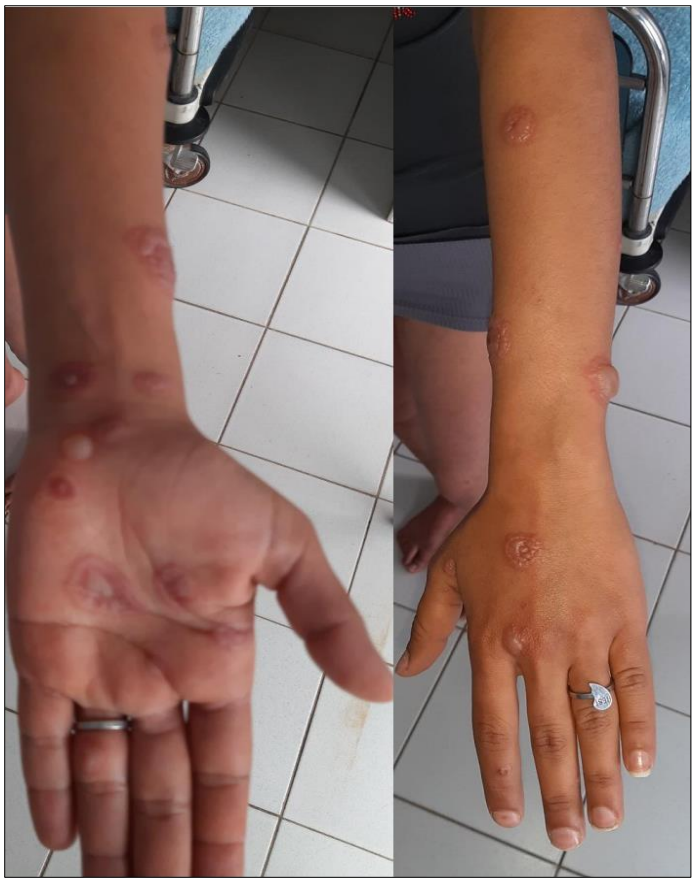

Figure 2: Clinical picture showing target lesions with a bullous center

The patient also had targetoid lesions and scars from previous outbreaks on the limbs. Mucosal examination revealed target lesions on the lip, polycyclic erosions on the hard palate, intact attached gingiva (figure 3) and unaffected genital mucosa. Examination of the skin appendages and other parts of the body revealed no abnormalities.

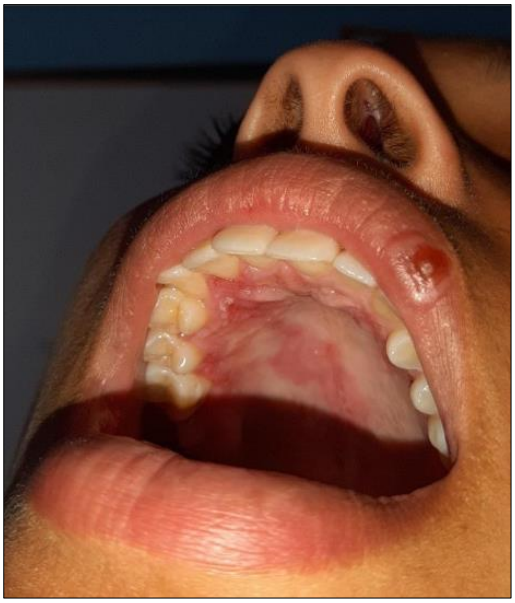

Figure 3: examination of the oral cavity showing target lesions on the lip and polycyclic erosions on the hard palate

We suggested the diagnosis of bullous erythema multiforme and ruled out Stevens-Johnson syndrome because of the absence of recent new medication intake and the absence of a diffuse cutaneous involvement and purpuric or slate-coloured macules; we also ruled out Rowell's syndrome because of the absence of skin involvement suggestive of lupus or abnormalities on examination of the other organs; we also ruled out pemphigus because the mucosal involvement was minimal and did not prevent the patient from eating and because of the presence of target lesions.

Biopsy was performed on a target lesion; we did not biopsy bullous lesions due to their location or size.

Results showed a thinned superficial epidermis covered by a thickened orthokeratotic stratum corneum. Dermal basal cells showed significant vacuolization. The superficial and mid-dermis harbored a band-like lymphocytic infiltrate that extended to perivascular and peripheral distribution (figure 4).

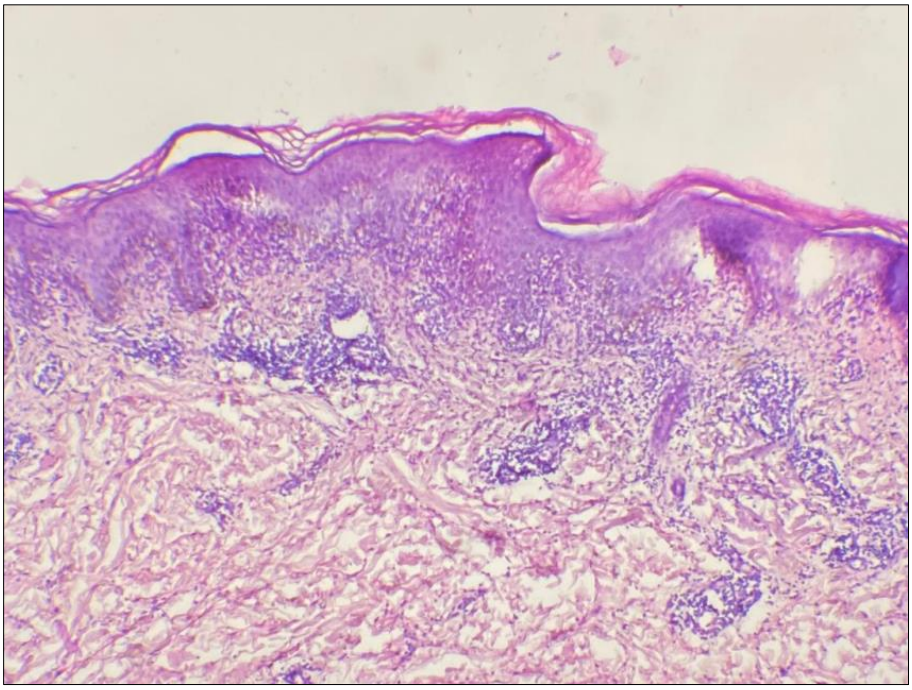

Figure 4: Histopathology showing a thinned superficial epidermis covered by a thickened orthokeratotic stratum corneum. Dermal basal cells showed significant vacuolization. The superficial and mid-dermis harbored a band-like lymphocytic infiltrate that extended to perivascular and peripheral distribution

The pathologist suggested that the skin biopsy was consistent with subacute lupus erythematous.

Immunologic tests were negative for antinuclear antibodies, Anti-DNA, Sm, SSA and SSB were also negative

Anti-EBV and anti-CMV virus serologies were positive.

We were puzzled by the clinical features strongly suggestive of bullous erythema multiforme and the pathology conclusion of subacute lupus erythematosus.

DISCUSSION

Subacute cutaneous lupus erythematosus sometimes simulates erythema multiforme in the diagnosis known as Rowell's syndrome.

Rowell et al's criteria for the diagnosis need the presence of lupus erythematous, erythema multiforme like lesion with the absence of precipitating factors, chilblains, speckled pattern of antinuclear antibodies, Anti-La/SSB antibody and a positive rheumatoid factor.

Lee *et al*'s criteria for the diagnosis need the presence of lupus erythematous, erythema multiforme like lesions with the absence of precipitating factor, speckled pattern of antinuclear antibodies, anti-La/SSB antibody and a positive rheumatoid factor.

Zeitouni *et al.*, define the diagnosis by the presence of 3 major criteria and 1 minor criterion. The major criteria are systemic lupus erythematous, discoid lupus erythematous or subacute cutaneous lupus erythematous; erythema multiforme like lesions and a speckled pattern of antinuclear antibodies. The minor criteria are chilblains, Anti-Ro/SSA or Anti-La/SSB antibodies and a positive rheumatoid factor.

Torchia *et al*., define the diagnosis by the presence of all major criteria and at least one minor criterion. The major criteria are the presence of chronic cutaneous lupus erythematous (discoid lupus erythematous and/or chilblains); the presence of erythema multiforme like lesions ; at least one positivity among speckled antinuclear antibodies, Anti-Ro/SSA, Anti-La/SSB ; and a negative direct immunofluorescence on lesional erythema multiforme like lesions. The minor criteria are the absence of infectious or pharmacologic triggers; the absence of typical erythema multiforme location and the presence of at least one additional ARA criterion for the diagnosis of systemic lupus erythematous besides discoid rash and antinuclear antibodies and excluding photosensitivity, malar rash and oral ulcers (Gallo L *et al*., 2020; Schissler C *et al*., 2017)

All these diagnosis set of criteria need the presence of both lupus lesions and antinuclear antibodies, which was not the case of our patient, so we reconsidered the pathological diagnosis of subacute cutaneous lupus erythematous. The most likely diagnosis was then bullous erythema multiforme.

In erythema multiforme, histopathology may show a vacuolar interface dermatitis with marked lymphocyte infiltration along the dermo-epidermal junction. One may also see dyskeratosis of basal keratinocytes and hydropic changes. In advanced lesions, one may see epidermal necrosis, subepidermal vesicles and blisters (Hafsi W *et al*, 2023; Samim *et al*., 2013)

So, our biopsy was consistent with the diagnosis of bullous erythema multiforme

CONCLUSION

If in doubt between Rowell's syndrome and bullous erythema multiforme, of which both histopathologies can show interface dermatitis, one can use the diagnosis criteria of Torchia *et al*., Zeitouni *et al*., Lee *et al*., or Rowell *et al*., When there are no lesions suggestive of lupus nor antinuclear antibodies, one can comfortably make the diagnosis of bullous erythema multiforme.

Acknowledgments: None

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