PITYRIASIS LICHENOID AND ACUTE VARIOLIFORM AS A DIFFERENTIAL DIAGNOSIS OF SEVERE CUTANEOUS ACTIVITY IN SYSTEMIC LUPUS ERYTHEMATOSUS: A CASE REPORT

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BACKGROUND

Pityriasis lichenoides et varioliformis acuta (PLEVA) is a rare disease, characterized by a lymphoproliferative disorder of CD8+ T cells. Its main manifestation consists of disseminated erythematous to violaceous papules, mainly on the extremities and trunk. It can affect several systems (mucosal, gastrointestinal, pulmonary and neurological), with fatal complications such as bacteremia and sepsis – observed in the Mucha–Habermann variant (Chin 2021). It presents numerous differential diagnoses, including systemic lupus erythematosus (SLE). The clinical manifestations of SLE are variable, and may present more severe skin involvement, as in cases of vasculitis. In this report, we describe a case of severe SLE with significant cutaneous involvement, which initially did not meet the EULAR criteria (2019), and an extensive diagnostic investigation was carried out.

CASE REPORT

LRS, female, 20 years old, with 5 months of polyarthritis of small and large joints, alopecia, autoimmune hemolytic anemia, thrombocytopenia, cylindruria, proteinuria and hypocomplementemia, with 3 previous negative ANA HEp-2 tests. In 3 weeks of evolution, she presented papular lesions disseminated throughout the entire integument, with a varioliform aspect, in addition to necrotic ulcers, with erythematous infiltrated borders (Figure 1), the largest with serosanguineous exudate in the inframammary region, suggestive of Mucha–Habermann disease, as classified by clinical inspection by the dermatology team, also associated with other serious systemic manifestations: interstitial pneumonitis and tonic-clonic seizures (infectious etiologies were excluded after bronchoalveolar lavage, lumbar puncture and imaging exams were performed). Dermatological lesions then progressed to cutaneous abscesses and severe sepsis with a culture of wound secretion with ESBL-producing *Pseudomonas aeruginosa*, in addition to leukocytosis with neutrophilia and elevation of inflammatory proteins. Viral serologies were all negative. The therapeutic approach to control the lesions was methylprednisolone 1 mg/kg and human immunoglobulin 2 g/kg. The patient evolved with stagnation in the progression of lesions and healing of previous ulcers, as well as gradual clinical improvement. Subsequently received high titer ANA HEp-2 result.

Figure 1.

CONCLUSION

Despite its rarity, the Mucha–Habermann disease should be considered in patients with ulceronecrotic skin lesions and systemic manifestations, as it is potentially fatal. The most appropriate management is not well defined due to the small number of cases described and the prognosis is poor in adults, with a mortality rate of up to 33% according to the literature. Therefore, it is important to recognize this pathology, as it has the potential to mimic more frequent diseases, such as SLE.

KEYWORDS

Systemic lupus erythematosus, Acute cutaneous lupus, PLEVA, Mucha–Habermann disease.