ROWELL'S SYNDROME: ASSOCIATION OF LUPUS WITH ERYTHEMA MULTIFORME – CASE REPORT

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BACKGROUND

Systemic lupus erythematosus (SLE) is a heterogeneous autoimmune disease with a wide range of clinical and serologic manifestations that can affect virtually any organ. Most patients develop skin and mucous membrane lesions at some point during the course of their disease. The most common lesion is “the butterfly rash”. However, some forms of cutaneous manifestation of SLE are relatively rare, such as bullous eruptions, which represent less than 5% of the total. A rare form of bullous lesion in lupus patients is the so-called Rowell's syndrome.

CASE REPORT

Female, 42 years old, with SLE (serological profile: antinuclear antibodies – ANA thick nuclear reagent, anti-DNA/SM/RO/LA and rheumatoid factor positive) started after tanning a picture of annular and polycyclic lesions associated with peripheral vesicles in the trunk region with areas white in between. Histopathology showed superficial vacuolar interface dermatitis with marked necrosis of keratinocytes and epidermal detachment foci suggestive of erythema multiforme. Therapy based on temporary suspension of hydroxychloroquine and furosemide. The association of azathioprine and corticosteroid improved skin lesions.

CONCLUSION

The cutaneous SLE includes three subsets of skin diseases: acute cutaneous lupus erythematosus, subacute cutaneous lupus erythematosus and chronic cutaneous lupus erythematosus. Bullous lesions on the skin of patients with SLE require a careful differential diagnosis between the typical cutaneous lesions of the disease and those of another overlapping bullous disease. Rowell's syndrome is characterized by the association of systemic lupus erythematosus (SLE) and erythema multiforme-like lesions with the presence of immunological markers such as antinuclear antibodies with speckled pattern, anti-La antibodies and rheumatoid factor. Many factors, including infections, medications, malignancy, autoimmune disease, immunizations, radiation, sarcoidosis, and menstruation, have been linked to the development of erythema multiforme. It has been estimated that drugs induce the lesions in less than 10% of cases, including diuretics and antimalarials. Zeitouni et al. redefined the major and minor diagnostic criteria of Rowell's syndrome. Major criteria are lupus erythematosus, erythema multiforme-like lesions and speckled pattern of antinuclear antibody. Minor criteria include chilblains, positive anti-La (SS-B) or anti-Ro (SS-A) antibodies, and reactive RF. The diagnosis of this syndrome requires the presence of all the major criteria and at least one minor criterium. Rowell's syndrome describes a diagnostic challenge. Histopathologic and serologic findings can help differentiate this process from erythema multiforme.

KEYWORDS

Systemic lupus erythematosus, Rowell's syndrome, Erythema multiforme.