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# Oral mucosal involvement in bullous lupus

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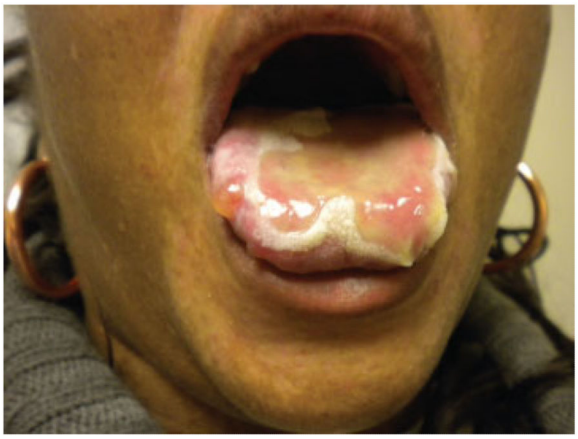

The patient, a 42-year-old woman with systemic lupus erythematosus (SLE), presented with malar rash, alopecia, arthralgia, photosensitivity, oral ulcers, and leukopenia 13 years ago, and she tested positive for antinuclear antibodies, anti-double-stranded DNA, IgM anticardiolipin antibodies, and anti-Sm antibodies. She had biopsy-proven bullous lupus that was refractory to treatment with dapsone, hydroxychloroquine, azathioprine, methotrexate, and prednisone at dosages of up to 120 mg per day. Except for one area on her breast, the bullous lupus lesions cleared after she received rituximab (2 infusions of 1,000 mg each given 2 weeks apart); for the last 2 years, she was maintained on a regimen that included 5 mg per day of prednisone, azathioprine, dapsone, and hydroxychloroquine. The disease recently relapsed with multiple tense bullae on the trunk and buccal mucosa and a 6-cm erosion of the tongue, prompting short-term, high-dose treatment with intravenous methylprednisolone and re-treatment with rituximab (2 infusions of 1,000 mg each). The disease again responded to treatment; 2 weeks after the patient received rituximab, the blistering process ceased, and existing lesions began to heal. Bullous lupus is an uncommon form of cutaneous lupus characterized by subepidermal autoantibody-mediated vesiculobullous skin lesions. The diagnosis is supported by a clinical history of active SLE, tense blisters on physical examination, subepidermal blisters with a dermal neutrophil-predominant infiltrate on histologic examination, deposition of immunoglobulins in the basement membrane on direct immunofluorescence analysis, and indirect immunofluorescence evidence of circulating antibodies to type VII collagen (1). Bullous lupus is usually responsive to dapsone, although patients with severe bullous lupus or patients with active SLE may require treatment with systemic steroids. Second-line therapies include azathioprine, mycophenolate mofetil, antimalarials, and cyclophosphamide (1). Although the use of rituximab for moderate-to-severe active SLE (as in the Exploratory

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Phase II/III SLE Evaluation of Rituximab study) and lupus nephritis (as in the Lupus Nephritis Assessment with Rituximab trial) has not been proven to be better than placebo (2), one other case in which rituximab was successfully used to treat bullous lupus has been reported (3).