Check for updates Page 1 of 1

AB024. A case of bullous systemic lupus erythematosus confined to the oral mucosa

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Abstract: Bullous systemic lupus erythematosus (BSLE) is a rare antibody-mediated blistering disease that presents in association with systemic lupus erythematosus (SLE). Features of BSLE can present as a diagnostic challenge as they often mimic other bullous skin diseases. We present a case of a 45-year-old female with history of celiac disease, Raynaud's syndrome, chilblains, and Sicca symptoms who presented with 10 months of recurrent vesicles and erosions of the gingiva. Review of symptoms was positive for arthralgias and recurrent photosensitive rashes. Physical examination was notable for multiple 4 mm vesicles with mild surrounding erythema confined to anterior upper gingiva without further mucosal or cutaneous involvement. Two biopsies obtained prior to presentation revealed a mixed inflammatory infiltrate with focal eosinophils, negative for viral cytopathic changes. A biopsy of the maxillary frenum for direct immunofluorescence (DIF) demonstrated a weak, fine linear deposition of IgG and IgA along the basement membrane zone initially suspicious for mucous membrane pemphigoid. Serum ELISA for antibodies against BP180, BP230, and collagen VII were

negative. Further lab workup was notable for an ANA of 1:160, anti-Ro60 774 (normal 0–19), anti-Sm 20.7 (normal 0–19), and C4 15.1 (normal 16.3–47.8). The patient was found to meet SLICC criteria (joint disease, chronic cutaneous lupus (chilblains), positive ANA, positive Anti-Sm, and low C4) for SLE diagnosis. The patient was started on dapsone 100 mg by mouth daily with improvement in symptoms. BSLE can affect the oral mucosa and has been described in the absence of anti-collagen VII positivity, but has not been described in the absence of cutaneous bullae. Although atypical, the DIF staining pattern in the context of SLE raises support for a diagnosis of oral BSLE. Teaching point: BSLE can manifest as an oral vesicular eruption and can be differentiated from other oral bullous diseases based on histologic and immunopathologic features

Keywords: Bullous systemic lupus erythematosus (BSLE); lupus; bullous disease

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