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Brief Report

Bullous Sytemic Lupus Erythematosus

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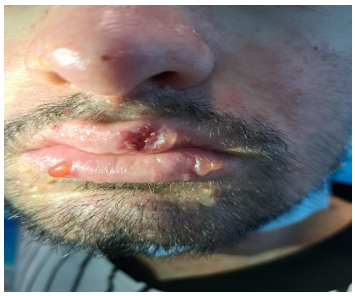
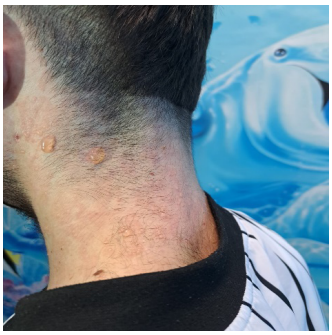
Case Report

A 19 years-old man presented to the hospital with blisters and bullae in the skin. Four months before presentation he had a diffuse pruritic erithematosus rash that resolved after 1 week of treatment with glucocorticoid. On physical examination several tense vesicles and bullae filled with clear or pink fluid were observed in the skin of the face, lips, neck chest, upper limbs and fingers; some irregular superficial erosions and scars were also observed in the same areas; a diffuse non-tender lymphadenomegaly and splenomegaly were also present. Laboratory studies showed a reduced white cell and platelets counts, low serum levels of complement C3 and C4, a positive antinuclear antibody with a titer of 1:640 and a speckled pattern, a positive anti-double-stranded DNA (379 UI/mL, reference range: < 16) and a positive antiribonucleoprotein autoantibody; additionally, autoantibodies against desmogleins 1-3 and BPAg1 and BPAg2 were all negative. Bacterial cultures of bullae aspirate yielded no growth. Direct immunofluorescence on involved skin biopsy showed granular continuous deposits of immunoglobulins (IgG, IgA and IgM), C3 and C1q localised at dermo-epidermal junction. A diagnosis of systemic lupus erithematosus with skin involvement was made; treatment with prednisone and dapsone was started; at the 4-weeks follow-up, the skin lesions had abated.

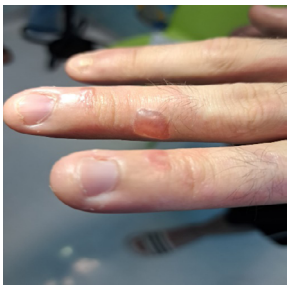
Bullous lesions are a relatively infrequent manifestation of SLE, and it predominantly affects women in the age range of second to fourth decade. This case underscores the importance for clinicians to be aware of this rare condition in male patients and the need of immunopathological examination of skin biopsy for the differential diagnosis with other blistering diseases (Panel A, B and C).



Panel A



Panel B



Panel C