AUTOIMMUNE BULLOUS DISEASES

SERRATION PATTERN ANALYSIS REVEALED A CHALLENGING DIAGNOSIS: IMMUNOGLOBULIN A MEDIATED EPIDERMOLYSIS BULLOSA ACQUISITA

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Background: Epidermolysis bullosa acquisita (EBA) is a chronic sub-epidermal autoimmune blistering disease (AIBD) characterized by the presence of autoantibodies against collagen VII, essential component of the dermal-epidermal junction. In most of the cases, immunoglobulin (Ig) G are predominant, but in a smaller percentage of cases IgA can be found either exclusively or in combination with IgG.

Observation: A 69-year-old Caucasian man was referred to our clinic due to diffuse erythematous-violaceus urticarial plaques and tense blisters of the hands, associated with painful oral erosions. The eruption had a 2-month duration. The patient, whose medical history was relevant for cardiovascular diseases, renal adenocarcinoma and melanoma, was unsatisfactorily treated with high-dose prednisone elsewhere. Histopathological and immunopathological examinations were performed in order to achieve the diagnosis. Histopathology showed the presence of a subepidermal blister with neutrophilic infiltration, while indirect immunofluorescence (IF) on monkey esophagus and enzyme linked immunosorbent assay for IgG against epidermal and dermo-epidermal antigens were negative. Direct IF(DIF) revealed linear IgA deposition with a u-serrated pattern along the basement membrane zone (BMZ), and IgA bound to the dermal side of the BMZ when DIF was performed on 1 M NaCl lesional split skin (SSS). A diagnosis of IgA-mediated EBA was made. Treatment with dapsone 125mg/day resulted in complete and rapid healing of the skin lesions.

Key message: A correct differential diagnosis within the spectrum of AIBD is paramount for the correct management of the patients. In our case, the finding of a u-serrated pattern at DIF and the results of SSS-DIF, have proven to be the clue for the diagnosis of a very rare variant of EBA: IgA-EBA. This allowed to perform a more specific treatment such as dapsone, that, in this case, was more effective than systemic steroids and led to a complete resolution of the disease.