AUTOIMMUNE BULLOUS DISEASES

A CASE OF MUCOUS MEMBRANE PEMPHIGOID ASSOCIATED WITH ACQUIRED HEMOPHILIA A

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Background: Mucous membrane pemphigoid (MMP) is a rare autoimmune bullous disease caused by autoantibodies targeting various antigens including BP180, BP230, integrin α6/β4, and laminin-332. MMP predominantly affects mucous membranes including the oropharyngeal mucosa and occasionally involves the skin.

Observation: A 64-year-old man presented with many hemorrhagic bullae on the oropharyngeal mucosa and a few blisters on the upper trunk. Histopathological findings of the oral mucosal biopsy showed subepidermal blister filled with red blood cells. Direct immunofluorescence revealed linear IgG and C3 deposition along the basement membrane zone and indirect immunofluorescence using salt-split skin showed IgG deposition on the epidermal side. Isolated activated partial thromboplastin time was prolonged and serum coagulation factor VIII (FVIII) was decreased. Based on these results, we diagnosed him as MMP with acquired hemophilia A (AHA). He was treated with systemic steroid, intravenous immunoglobulin and recombinant activated factor VII. Remission of AHA was achieved after 2-month treatment and 7.5mg of prednisolone was used for the treatment of MMP.

Key message: AHA is a rare autoimmune disease caused by IgG autoantibodies against FVIII. There are several reports on autoimmune bullous diseases associated with AHA. Although it is unknown why both diseases concomitantly occur, cross-reactivity between two self-antigens is thought to be one of the possible mechanisms.