AUTOIMMUNE BULLOUS DISEASES

NEW INSIGHTS INTO BULLOUS PEMPHIGOID

Hideyuki Ujiie [(1)]

Hokkaido University Graduate School of Medicine, Department of Dermatology, Sapporo, Japan [(1)]

Bullous pemphigoid (BP) is the most common autoimmune blistering disorder, and it is characterized by itchy edematous erythema and tense blisters on the whole body. It is mainly caused by autoantibodies to a major hemidesmosomal component at the dermal-epidermal junction of the skin, BP180 and BP230. Recent studies demonstrated an increased risk of BP during dipeptidyl peptidase-4 inhibitors (DPP-4i) exposure in diabetic patients administered DPP-4i. Vildagliptin is reported to be associated with the highest risk among DPP-4i. We recently reported unique clinical and immunological features of DPP-4i-related BP. In particular, DPP-4i-related BP tends to show a noninflammatory (or less inflammatory) phenotype with few erythematous lesions, and the noninflammatory patients have lower titers of antibodies to the noncollagenous 16A (NC16A) domain of BP180. Furthermore, we recently reported a strong association between HLA-DQB1*03:01 and the noninflammatory DPP-4i-related BP in Japanese patients. It has also been reported that BP patients frequently have comorbidities associated with neurological disease, such as dementia, stroke, Parkinson's disease and multiple sclerosis. A recent study demonstrated a presence of anti-BP180 autoantibodies in stroke patients. These new clinical and experimental findings give us important clues to the mechanism of the breakdown of self-tolerance to BP antigens.