AUTOIMMUNE BULLOUS DISEASES

A CASE REPORT OF PEMPHIGUS FOLIACEUS CONVERTING INTO BULLOUS PEMPHIGOID WITH CLINICAL AND IMMUNOPATHOLOGICAL FEATURES

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Background: Pemphigus foliaceus (PF) and bullous pemphigoid (BP) are two different autoimmune blistering dermatosis with different clinical, histopathological and immunologic characteristics. The conversion between these two kinds of autoimmune diseases is rare, and the mechanisms underlying this disorder remain unknown. Here, we report a patient who developed BP 7 years after PF was diagnosed, which is the first reported in China.

Observation: A 58-year-old Chinese woman who developed BP 7 years after PF was diagnosed. On her first presentation in Feb 2010, the patient presented with erythema and erosion in her abdomen, histological examination (HE) demonstrated intraepidermal blistering and acantholysis in the upper stratum spinosum, indirect immunofluorescence revealed the binding of IgG antibodies against the intercellular spaces of epidermis, with the titer of 1:80. After prescripted with high dose of immunosuppressants, the lesions were gradually cleaning. In Apr 2017, ELISA detected the positive of antibody to desmogleins 1, negative of antibodies to desmogleins 3 and BP180, respectively. In Jun 2017, the patient abdomen appeared erythema, tense blister and erosion on the basis of normal skin, suddenly. HE demonstrated a subepidermal blister with eosinophils and lymphocytes, direct immunofluorescence revealed deposition of IgG and C3 along the basement membrane zone. ELISA showed the positive of antibody to BP180, negative of antibodies to Dsg1 and Dsg3. After given low dose glucocorticoid and immunomodulators, the lesions were quickly cleaning, and the anti- BP180 shifted to negative.

Key message: This rare case that pemphigus foliaceus converting into bullous pemphigoid with clinical and immunopathological features, is the first reported in China. The intermolecular epitope spreading mechanism may have involved with the pathological process. However, an exact explanation for the conversion of this two autoimmune diseases remains to be illuminated.