AUTOIMMUNE BULLOUS DISEASES

THE PRESENCE OF AUTOANTIBODIES PRIOR TO CLINICAL DIAGNOSIS IN PEMPHIGUS DISEASES

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Background: The pemphigus family of autoimmune blistering disease have been well characterized clinically and histopathologically, but due to the rarity of these diseases, the pathogenicity and timing of onset of the anti-desmoglein 1 and anti-desmoglein 3 antibodies has not been well explored. This is a unique study in that we are able to test serum samples from a pool of 7 million people banked by the Department of Defense Serum Repository, which has banked more than 30 million samples over 30 years.

Objective: We tested patients' banked sera for the presence of autoantibodies to desmoglein 1 or 3 prior to their respective dates of diagnosis to determine how long antibodies were present before clinical onset of symptoms.

Materials and Methods: This is a retrospective analysis of patients diagnosed with pemphigus vulgaris, pemphigus foliaceous, or paraneoplastic pemphigus by direct immunofluorescence between 2008-2017 at the Walter Reed National Military Medical Center Immunodermatology Lab. We reviewed patient charts to determine correlation with illness, medications, demographics or deployment history, and what common erroneous diagnoses were made prior to the correct clinical diagnosis or diagnostic biopsy. Patient sera were tested with a commercially available ELISA for anti-desmoglein 1 and anti-desmoglein 3 antibodies.

Results: 38% of patients with pemphigus had antibodies for as long as 27 months prior to clinical disease onset. 21.7% of patients had anti-desmoglein 3 antibodies, and 16.3% had anti-desmoglein 1 antibodies, with one patient with antibodies to both.

Conclusions: Titer elevations correlated with clinical onset of symptoms. However, with an average period of time between onset of symptoms to diagnostic DIF of 7 months (PF), and 4 months (PV), many patients experienced a delay in definitive diagnosis.