AUTOIMMUNE BULLOUS DISEASES

THE TRANSITION OF PEMPHIGUS VULGARIS AND PEMPHIGUS FOLIACEUS IN IRANIAN PEMPHIGUS PATIENTS: A CASE SERIES

M Daneshpazhooh [1] - M Mohamadrezaee [1] - I Etesami [1] - K Balighi [1] - V Lajevardi [1] - H Mahmoudi [1]

Tehran University Of Medical Sciences, Dermatology, Tehran, Iran (islamic Republic Of) [1]

Introduction: Pemphigus vulgaris (PV) and pemphigus foliaceus (PF) are the two major subtypes of pemphigus that are distinct clinically and histologically and have different antigenic profiles. Transition between these two subtypes are well-documented, but rarely reported.

Objective: Our aim was to search for cases of transition of pemphigus subtypes.

Materials and methods: We reviewed the records of pemphigus patients presenting to the Autoimmune Bullous Diseases Clinics in a University Hospital in Tehran, Iran and searched for cases of transition of these two subtypes. Baseline histology, direct immunofluorescence, anti-desmoglein (anti-Dsg)3/1, clinical presentation and the changes during the disease course were extracted.

Results: Twenty-seven cases of transition of PV to PF and two cases of transition of PF to PV were identified. The majority of cases showing transition of PV to PF (74%) had mucocutaneous phenotype at the onset. The initial diagnosis of PV was based on histopathology. From these patients, 11 cases had anti-Dsg1/3 results before and after transition. All these 11 patients were positive for anti-Dsg3 and 10 out of 11 for anti-Dsg1 before transition. After the shift shown clinically, only anti-Dsg1 was positive. The shift occurred after treatment in all cases and the developing PF was not severe.
Only two patients had an initial diagnosis of PF clinically, histologically and serologically (negative anti-Dsg3 and positive anti-Dsg1) that shifted to PV with orocutaneous lesions and positive anti-Dsg3.

Conclusions: Transition from PV to PF is more common. Shifts are usually noticed after treatment and during relapses. The reason of the shift to PF is still unknown. B cells producing anti-Dsg3 may have become inactive and only those producing anti-Dsg1 may have escaped treatment. Transition to PV may be attributed to epitope spreading.