AUTOIMMUNE BULLOUS DISEASES

ATYPICAL PEMPHIGUS VULGARIS WITHOUT DETECTABLE SERUM AUTOANTIBODIES AGAINST DESMOGLEINS: A SERIES OF 10 PATIENTS WITH A CHALLENGING COURSE OF DISEASE

R Küçükoglu [(1)] - S Kiliç [(1)]

Istanbul University, Istanbul Faculty Of Medicine, Department Of Dermatology And Venereology, Istanbul, Turkey [(1)]

Background: Pemphigus vulgaris (PV) is a autoimmune blistering skin disease characterized by elevated levels of serum autoantibodies; anti-desmoglein (Dsg)3 and anti-Dsg1. Anti-Dsg3 and 1 are rarely undetectable by enzyme-linked immunosorbent assay (ELISA) in PV.

Objective: To assess the course of PV with an atypical serology and compare them with the literature

Materials/ Methods: This was a retrospective, single-center study conducted in our subspeciality policlinic of bullous diseases between 2005-2018. Among 422 diagnosed patients with PV, 10 patients with the absence of Dsg autoantibodies in the serum at the diagnosis and during the follow-up were included. All patients were diagnosed by both histopathological evaluation and DIF testing. Patients with elevated Dsg levels in ELISA once or more were excluded.

Results: The group consists of 10 patients(6 males and 4 females). Mean age at the diagnosis was 42.6 ±11.1 years. Mean duration from the first symptom to the diagnosis was 6.85±10.43 months. Oral lesions alone(n=5) and in combination with other areas(n=5) were affected. IIF titers were positive both at the diagnosis and persisted during the follow-up. Systemic corticosteroids with a (mean=80 mg/day) were iniated in all patients and at least one adjuvant therapy agent was started during the follow-up including;azathioprine(n=7), mycophenolate mofetil(n=5), methotrexate(n=1), rituximab(n=1) and IVIG(n=3). Full remission was achieved in only 4 patients, treatment was discontinued but restarted due to relapse. In 10 patients at least one (range:1-7) relapse was seen. Mean follow-up duration was 83.8±32.9 months.

Conclusion/ Discussion: Our patients with an atypical serology showed a chronic and

therapy-resistant course. Despite the general idea of the correlation between serum anti-Dsg levels and severity, recent publications showed that anti-Dsg3 and 1 may be absent or extremely low in the serum to be detectable occasionally but other autoantibodies like anti-desmocollins would be useful, is absent in our st