AUTOIMMUNE BULLOUS DISEASES

CLINICAL AND SEROLOGICAL CORRELATION IN 40 PATIENTS WITH PEMPHIGUS

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Introduction: pemphigus is an organ-specific autoimmune disease, characterized by autoantibodies against components of desmosomes. There are different types of pemphigus with specific clinical, histopathological and immunological characteristics.
In 2016, thanks to a research grant, it was possible the implementation of serology by ELISA, method unavailable in Argentina until that moment.

Objective: to correlate the clinical characteristics and serological autoantibodies profiles of patients with different pemphigus.

Material and methods: we analyze the clinical and serological profiles by ELISA of 40 patients with pemphigus evaluated in the "J.F.Muñiz" Hospital between November 2016 and August 2018.

Results: of the 40 patients, 18 had pemphigus vulgaris, 14 superficial pemphigus (PS), 4 pemphigus herpetiformis, 1 pemphigus vegetans, 1 pemphigus neonatal and 2 were suspected to have paraneoplastic autoimmune multi-organ syndrome(PAMS) that could not be confirmed.
Of the 18 patients with pemphigus vulgaris, 14 had mucocutaneous involvement and a positive ELISA for Desmoglein-1 and 3. In the remaining 4 patients, with exclusive mucosal involvement, the ELISA was positive only for Desmoglein-3.
In 11 of the 14 patients with PS the ELISA was positive for Desmoglein-1 and in the remaining 3 cases, which presented extended cutaneous involvement and therapeutical refractoriness, the ELISA was positive for both Desmoglein-1 and 3.
The 4 patients with herpetiformis pemphigus had ELISA positive for Desmoglein-1.
The patient with vegetating pemphigus and the one with neonatal pemphigus had positive Desmoglein-3.
Two patients with suspected PAMS were positive for Desmoglein-1, Desmoglein-3 and BP230 but were negative for envoplakine.

Conclusions: The serological findings, by the ELISA were concordant with the clinical presentation in most cases and with the international literature.
It has to be emphasized that the presence of Desmoglein-1 and Desmoglein-3 in PS is

correlated with greater severity.
In the patients with suspicion of PAMS, the presence of envoplakine could not be confirmed.