



AUTOIMMUNE BULLOUS DISEASES

SINGULAR SEROLOGICAL FINDINGS IN SUPERFICIAL PEMPHIGUS

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Background: superficial pemphigus is one of the classic pemphigus varieties. It typically affects the skin of seborrheic areas, although in some cases the skin involvement may be more extensive. Moreover the mucosa is usually preserved. It is produced by autoantibodies against desmoglein-1, which can be detected through serological techniques (ELISA). Subcorneal detachment is characteristically seen in the histopathology.

Observation: we present the cases of 3 patients with extended superficial pemphigus with striking serological findings, attended at the "F.J. Muñoz" Hospital in the period March 2017 – September 2018. It should be noted that this technique was incorporated for the first time in Argentina in November 2016, from the obtaining a research grant from our service.

Case 1: male, 61 years old. Flaccid blisters and erosions in the cephalic pole, trunk and extremities. 5 months of evolution.

Case 2: male, 75 years old. Erosions covered by scabs on scalp, face, trunk and upper limbs. 1 year of evolution.

Case 3: female, 55 years old. Dermatitis with outbreaks and remissions since 2014, characterized by generalized flaccid blisters and erosions.

In all 3 cases the mucous membranes were preserved. Moreover the skin disease involved a large extension of the body's surface, affecting the general health condition and the Nikolsky's sign was positive. In all of them, the histopathology revealed a subcorneal detachment and directed immunofluorescence informed IgG intercellular space staining pattern. ELISA was positive for desmoglein-1, as expected, and surprisingly also for desmoglein-3.

The 3 cases presented therapeutic relapse, with the need for high doses of steroids and biological behaviour similar to Pemphigus vulgaris.

Key message: despite the low casuistry, the concomitance of desmoglein-1 and 3 could be related to more severe forms of superficial pemphigus, with the possibility of turning to deep varieties at some point in their evolution.

