AUTOIMMUNE BULLOUS DISEASES

RECALCITRANT PEMPHIGUS VULGARIS LOCALIZED ON THE SCALP AND NOSE

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Background: Pemphigus vulgaris (PV) is a rare autoimmune bullous disease (ABD). The principal lesions, blisters, are caused by circulating autoantibodies against desmogleins. Except for the skin, the oral mucosa is almost always involved. The typical clinical presentation is a widespread mucocutaneous bullous eruption, while the disease can have other, less recognizable forms.

Observation: A 59-year-old man presented with few erosions in the scalp and face, accompanied by itching, which began two months ago. He was applying local corticosteroids (CS), without improvement. Examination revealed scarcely dispersed tiny erosions and crusts on the nose and one in the right parietal part of the scalp, with no other skin or mucosal changes. According to history and presentation, we suspected ABD. DIF of the perilesional skin revealed intracellular deposits of IgG and C3 (++). Pathohistology confirmed PV. IIF (ICAb and BMZAb) and the first ELISA on anti-desmoglein 1/ 3 IgG were negative. In spite of introducing a combination of systemic CS and azathioprine, new scalp erosions appeared, while the facial lesions were especially stubborn. A year after, painful lesions on the buccal mucosa and larynx started to appear. During the most pronounced aggravation, ELISA detected positive anti-desmoglein 1 (53.1 RU/ ml) and anti-desmoglein 3 IgG (200.0 RU/ ml). Due to symptom´s persistence, we introduced rituximab, with consequent improvement.

Key message: Our patient had the atypical form of disease consisting of just a few tiny erosions on the nose and a scalp. Trunk or limbs lesions were not present at all, while the oral lesions appeared in the later course of illness. The disease was relentless, combined therapy was not successful, while the biologic finally achieved the amelioration. The PV is a serious ABD in which prompt diagnosis and disease control are mandatory, at the same time, the atypical presentations can be deceiving and misleading.