AUTOIMMUNE BULLOUS DISEASES

CLINICAL PROFILE AND PROGNOSIS OF PEMPHIGUS THROUGH A SERIES OF 76 CASES

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Introduction: Pemphigus is an autoimmune blistering disease involving the skin and mucosal areas. Clinical presentation is variable. Prognostic factors are rarely studied in the literature.

Objective: The aim of this study was to investigate the clinical profile and the prognostic factors of pemphigus in Tunisia.

Materials & Methods: We presented a retrospective study, including all patients with confirmed diagnosis of pemphigus between January 2002 and December 2016 (13 years).

Results: Seventy-six patients were enrolled in the study (53 women and 23 men; sex ratio W/M=2,3). The mean age was 52±35. There were 42 cases of pemphigus vulgaris (55%), 3 cases of pemphigus vegetans (4%), 21 seborrheic pemphigus (28%), 7 pemphigus foliaceus (9%) and 3 pemphigus herpetiformis (4%). Skin biopsy specimen revealed intraepidermal bullae and the diagnosis was confirmed with direct immunofluorescence in 59 cases (98,7%). Systemic corticosteroids (1mg/Kg/d) were used in 64 cases (84,21%). Immunosuppressive drugs were indicated in 9 cases. Disease control was obtained in an average of 18 days. The delay of the response was independent of the form of pemphigus, its initial severity or the patient's age at the first consultation. Most relapses (95%) occurred in the first 2 years of follow-up. There was no influence of age, sex, baseline severity, or clinical form on the long-term evolution of pemphigus. Only the presence of erosions of the oral mucosa was influencing the risk of recurrence (p = 0.006). Six deaths occurred in the first 4 months of follow-up.

Conclusions: Pemphigus is a rare life-threatening blistering disease. Complications are frequent and most of them occur within the first 2 years of follow-up. The presence of erosions of the oral mucosa at the time of diagnosis is identified as major prognostic factor.