AUTOIMMUNE BULLOUS DISEASES

PEMPHIGUS HERPETIFORMIS: IS DAPSONE ALONE EFFICIENT IN TUNISIAN PATIENTS?

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Introduction: Pemphigus herpetiformis (PH) is a rare subtype of intraepidermal autoimmune bullous disease. It combines the clinical features of dermatitis herpetiformis and the immunologic characteristics of pemphigus.

Objectives: We aimed to dertermine the epidemiological, clinical and immunological findings as well as the therapeutic management of PH in Tunisia.

Material and methods: A retrospective monocentric study was conducted in the Dermatology Department of Habib Thameur over a period of 33 years (1985-2018).

Results: We included 15 patients with a mean age of 44 years (24-71 years). Eleven of the patients were women (sex ratio M/F=0,36). The main clinical features were urticarial erythematous annular plaques (10 cases) and vesiculobullous eruption arranged in a herpetiform pattern (11 cases). Mucosal involvement was not observed in our patients. Histopathologic evaluation found eosinophilic spongiosis in 8 cases. Direct immunofluorescence microscopy revealed intercellular fluorescence of IgG and C3 in all cases. Reactivity targets identified when ELISA was performed (3 cases) were Desmoglein 1. First line treatment was systemic corticosteroids at the dosage of 1 mg/kg/day in 11 cases. Dapsone at the dosage of 100 mg was prescribed in 4 cases two of which did not respond to treatment. Relapses were observed in 6 cases.

Conclusions: In our country, PH was frequent in female patients with absence of mucosal involvement as a distinctive feature. Systemic steroids are the first line treatment since dapsone prescribed alone is often complicated with a high rate of therapeutic failure. Further research is needed to evaluate the efficacy of different treatment protocols in PH which seem to have a distinctive clinical course.