



AUTOIMMUNE BULLOUS DISEASES

CHARACTERISTICS OF PEMPHIGUS HERPETIFORMIS IN SOUTH TUNISIA

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Background: Pemphigus herpetiformis (PH) is a rare subtype of pemphigus combining clinical features of dermatitis herpetiformis and the immunopathologic characteristics of pemphigus.

Objective : We aimed to analyze the epidemiological, clinical and immunological presentation and management of the disease in a cohort of south Tunisian patients with a long-term follow-up.

Methods: We included all patients with confirmed PH followed between 1987 and 2017 in the Dermatology department.

Results: We included 27 south Tunisian female patients with a mean age of 37 years [20–70], mainly from rural origin (74%). All patients had exclusive skin lesions. Clinical features combined erythematous, vesicular or bullous lesions, with circinate borders mainly on the trunk and limbs associated to a severe pruritus (88%). Peripheral hypereosinophilia was noted in 30% of cases. Histological examination demonstrated no intraepithelial cleavage (48%), suprabasal (30%) or subcorneal cleavage (22%). Eosinophilic spongiosis was seen in 33% of cases. On direct immunofluorescence, there was an intercellular staining in 92% of cases composed mainly of IgG and C3 (77%). Indirect immunofluorescence was positive in all cases. Reactivity was mainly directed against Dsg1. Dapsone alone was inefficient in 80% of cases (4/5). Systemic corticosteroids were used at first line or given due to the ineffectiveness of dapsone in 26 patients. Most of these patients (92%) relapsed. In two patients (8%), Azathioprine was associated with corticotherapy.

Conclusion: PH is characterized by the occurrence of exclusive skin lesions in young women with a good response to corticosteroids but not to Dapsone. Reactivity mainly against Dsg1 suggests that PH in south Tunisia is a particular variant of non-endemic pemphigus foliaceus.

