Accepted Manuscript

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PII: S1567-5688(17)30086-7

DOI: 10.1016/j.atherosclerosissup.2017.05.044

Reference: ATHSUP 336

To appear in: Atherosclerosis (Supplements) (Component)

Please cite this article as: Dietze J, Hohenstein B, Tselmin S, Julius U, Bornstein SR, Beissert S, Günther C, Successful and well-tolerated bi-weekly immunoadsorption regimen in pemphigus vulgaris, *Atherosclerosis (Supplements) (Component)* (2017), doi: 10.1016/j.atherosclerosissup.2017.05.044.

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ACCEPTED MANUSCRIPT

Successful and well-tolerated bi-weekly immunoadsorption regimen in pemphigus vulgaris

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All authors state no conflict of interest.

Key words: pemphigus, treatment interval, immunoadsorption, autoantibodies, steroid consumption

Abstract

Background

Pemphigus vulgaris is a chronic autoimmune disease characterized by blisters and erosions forming in the mucous membranes and the skin. Many patients are severely impaired by pain, weight loss and increased risk of infections. The disease is mediated by specific autoantibodies directed against desmogleins that contribute to connect keratinocytes in the epidermis. Autoantibody deposition in the skin causes inflammation and intraepidermal akantholysis. The concentration of autoantibodies in serum correlates with disease activity. Therefore, the removal of autoantibodies by immunoadsorption is a targeted therapeutic intervention for patients with pemphigus vulgaris.

Patients and methods

A total of 9 patients with pemphigus vulgaris resistant to the standard treatment regimen were treated by immunoadsorption using the TheraSorb[™]-Ig adsorber system and analyzed retrospectively. Patients received immunoadsorption on two or four consecutive days. Cycles were repeated every two or four weeks, respectively. Treatment was performed for a mean period of 17.5 months (range 6-26). Outcome was measured as improvement in clinical disease analysed by the investigators global assessment and the reduction of autoantibodies in serum measured by indirect immunofluorescence and ELISA. Tolerability of treatment by patients was evaluated using a visual analog scale.

Results

Retrospective analysis of 9 patients consecutively treated by immunoadsorption revealed an 80% reduction of the autoantibody concentration in serum after 6 months of treatment, led to a clinical improvement of disease in combination with classical immunosuppression. Steroidconsumption

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