Control of Ocular Disease in Mucous Membrane Pemphigoid.

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ABSTRACT: Background: Mucous membrane pemphigoid is a rare immune-mediated disease. It is characterised by an abnormal binding of immunoglobulins to the basement membrane zone of mucous membranes and the skin. Conjunctival involvement in mucous membrane pemphigoid may lead to cicatrising conjunctivitis and eventually to corneal blindness. The factors that determine mild or progressive disease are not fully understood and need to be clarified. This study examines the features, progression and risk factors of patients with ocular involvement in mucous membrane pemphigoid. Methods: 36 eyes of 18 patients with the diagnosis of ocular disease associated with MMP were identified. Fornix depth and keratopathy were repeatedly assessed using a standardised protocol to identify progression. MMP was diagnosed based on the characteristic clinical and laboratory features. Endpoints of the study were the incidence of progressive disease and the development of keratopathy with and without systemic immunomodulatory therapy. Results: 12 eyes of 6 patients (33 %) showed progressive conjunctival cicatrisation. Obvious progression was observed in 2 patients who had refused systemic treatment at an early stage. 10 eyes showed progression while on systemic treatment. In these patients, however, systemic treatment was started at an advanced stage of the ocular disease. None of the patients receiving systemic treatment developed persistent keratopathy. Conclusion: Systemic treatment with diaminodiphenyl sulfone and/or cyclophosphamide allows one to control the further progression of cicatrising conjunctivitis. It prevents keratopathy. To be efficient, however, treatment has to be started at an early stage of the ocular disease.

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