AUTOIMMUNE BULLOUS DISEASES

COMPARISON OF CLINICAL OUTCOMES IN PATIENTS WITH MUCOUS MEMBRANE PEMPHIGOID TREATED WITH BIOLOGICS - CRITICAL REVIEW.

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Background: Mucous membrane pemphigoid (MMP) is an autoimmune mucocutaneous blistering disease with autoantibodies to basement membrane zone proteins. Conventional immunosuppressive therapy is not always successful and can produce catastrophic side effects, therefore newer biologic agents have been used in its treatment.

Objective: Comparing clinical outcomes in patients treated with Rituximab (RTX), Intravenous immunoglobulin (IVIg), and RTX+IVIg.

Materials and Methods: Data was obtained from case reports and case series published in English from 1999 to 2018. We identified 197 patients with direct immunofluorescence proven diagnosis of MMP. Rituximab was used according to the Lymphoma and Rheumatoid arthritis protocol. IVIg was used at a dose of 2 g/kg/cycle.

Results: Clinical remission was observed in 80 - 95 % of patients with MMP treated with RTX, IVIg and RTX+IVIg. Patients with ocular cicatricial pemphigoid showed a higher frequency of partial responses. Treatment with IVIg prevented disease progression, especially blindness, in many patients. Relapse rate of 45% was reported in patients treated with RTX only, requiring additional treatment with RTX, corticosteroids, or immunosuppressive agents. Long term sustained remissions were reported in patients treated with IVIg and RTX +IVIg. The incidence of serious adverse events and mortality was highest in the group treated with RTX only. A major limitation of this analysis is the variation in the information provided by the authors in the individual publications.

Conclusions: Rituximab alone produces a high incidence of clinical remission but in the majority of patients is associated with a relapse. IVIg produces the same frequency of remissions as RTX, but the remissions are longer in duration. The highest frequency of clinical remission was observed in the group treated with RTX and IVIg. Clinical response

was rapid in onset, without relapse, and was sustained for prolonged durations.