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Efficacy of rituximab in the treatment of epidermolysis bullosa acquisita

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Introduction & Objectives

Epidermolysis bullosa acquisita (EBA) is a subepidermal bullous disorder characterised by skin fragility and mucosal erosions, mediated by antibodies that bind collagen VII. The condition is difficult to treat and often requires combination immunosuppressant therapy. Rituximab is a monoclonal anti-CD20 biologic therapy that has been reported in small case series to be effective in the treatment of recalcitrant EBA. In this case series of we report the efficacy of rituximab in the treatment of 14 patients with severe EBA.**

Materials & Methods

Electronic records were used to identify all patients with a diagnosis of EBA who received rituximab in our centre from 2011-2021. EBA was defined as patients with skin fragility, positive direct immunofluorescence demonstrating linear IgG at the basement membrane with localisation to the base of salt split skin and a positive collagen VII ELISA. Clinic records for each patient's follow up visits over 600 days post treatment were then used to identify clinical response, collagen VII titre and prednisolone dosing. Complete remission (CR) was defined as the absence of new or established lesions on minimal therapy for 2 months, partial remission (PR) as transient lesions that heal within 1 week on minimal therapy and active disease (AD) as the development of new lesions. B cell quantification, immunoglobulin levels and reported adverse events related to rituximab were recorded for each follow up visit.

Results

14 patients were identified (male = 10 and female = 4) with a mean age of 58.1 years (age rage 35-91 years old) and a mean disease duration of 12.8 years at the time of rituximab administration. All patients were on combination corticosteroid and immunosuppressant therapy, consisting of one or more of mycophenolate mofetil, intravenous immunoglobulin, azathioprine or dapsone. 4 patients also had positive BP180/230 antibodies. Rituximab induced CR in 6 of 14 patients who remained on low dose prednisolone and mycophenolate mofetil. PR was seen in 4 patients with AD in the remaining 4. A reduction in collagen VII titre of 78% was observed over the follow up period. The mean reduction in prednisolone dosing was 40%. Rituximab also allowed withdrawal of intravenous immunoglobulin therapy in 9 of 14 patients. No significant adverse events were reported.

Conclusion

Rituximab was well tolerated and resulted in clinical improvement in 71% of our patient cohort and enabled dose reduction of prednisolone and other immunomodulatory therapies in the majority of patients. This study represents the largest reported cohort of EBA patients treated with rituximab and supports it's role in the management of this challenging disease.