INTRODUCTION: Rituximab is a chimeric human-mouse monoclonal antibody, which binds to the CD20 antigen on B and pre-B lymphocytes and causes depletion of CD20+ cells in the mechanism of complement-dependent and independent cytolysis, antibody-dependent cell cytotoxicity and the mechanism of apoptosis. Rituximab is currently registered for the treatment of non-Hodgkin's lymphoma, chronic lymphocytic leukaemia and rheumatoid arthritis. Rituximab has also demonstrated efficacy in a number of other autoimmune diseases, including systemic lupus erythematosus and Sjögren's syndrome. In severe or recalcitrant cases of pemphigus vulgaris and pemphigus foliaceus, rituximab was applied as an adjuvant drug. OBJECTIVE: To present the efficacy of rituximab monotherapy in patients with pemphigus foliaceus, who had contraindications to classic immunosuppression, and a review of the literature. CASE REPORTS: We present 2 patients with pemphigus foliaceus, a 66-year old male and a 61-year old female, treated with rituximab in monotherapy as a first-line treatment. In the first patient serological negativisation and improvement were observed 4 months after therapy initiation. After 8 months further, significant clinical improvement, with sparse residual lesions, was observed. In the second case similar results were achieved. CONCLUSIONS: In patients with pemphigus foliaceus significant improvement was observed after rituximab monotherapy. Serological negativisation was achieved prior to clinical remission. This unusual phenomenon may be explained by rapid depletion of CD20+ cells and inhibition of antibody production, while in vivo bound pathogenic pemphigus antibodies remained active.