Sir,

Epidermolysis bullosa acquisita (EBA) is a rare acquired autoimmune disease. Antibodies reacting with collagen VII of anchoring fibrils lead to spontaneously arising or trauma-induced blisters (1). The course of EBA is usually protracted. Current immunosuppressive treatment is unsatisfactory and the disease can eventually result in irreversible extensive scarring.

CASE REPORT

A 55-year-old white man presented with a 2-month history of painful blisters. Tense bullae, erosions and haemorrhagic crusts were distributed on both arms, both legs and the trunk. Nikolsky's sign was negative. A biopsy specimen from perilesional skin showed haemorrhagic spongiosis of the epidermis, discontinuous subepidermal split formation and a superficial perivascular infiltrate composed predominantly of neutrophils. Direct immunofluorescence was positive for linear IgG, C3 and fibrinogen along the epidermal-dermal junction. Western immunoblotting of serum confirmed autoantibodies against a 290 kDa band corresponding to collagen VII.

Prednisolone 100 mg/day was given initially for 10 days but new blisters appeared. Combination therapy with prednisolone 100 mg/day and azathioprine 100 mg/day for 4 weeks followed by prednisolone 80 mg/day plus cyclosporin A 200 mg/day for a further 5 weeks did not reduce disease activity. Ten weeks after the initial treatment, the patient additionally received an intravenous infusion of 20 mg basiliximab (Simulect®; Novartis Pharma, Germany), which was well tolerated without any side effects. Cyclosporin A 200 mg/day was continued and prednisolone was subsequently tapered to 10 mg/day. Eruption of new blisters stopped within 2 weeks after the infusion of basiliximab and resolution of residual blisters, erosions and crusts was complete within 10 weeks, with partial atrophy and formation of milia.

DISCUSSION

Basiliximab is a chimeric anti-interleukin-2 (IL-2) receptor monoclonal antibody and is currently used to prevent acute rejection after cadaveric kidney transplantation (2). Prophylaxis with basiliximab reduces the incidence of acute rejection episodes significantly, with no clinically relevant safety or tolerability concerns having been reported so far in >1,000 patients (3). Basiliximab specifically binds to the z-subunit of the IL-2 receptor on activated T lymphocytes. The rationale for combining basiliximab with cyclosporin A in immunotherapy is that this combination both inhibits the IL-2 gene and blocks the IL-2 receptor as an additive synergistic modality. It allows one to considerably reduce steroid doses in a variety of (auto) immune diseases.

To our knowledge, this is the first published case of an autoimmune bullous disease treated successfully with basiliximab. We suggest that the combination of cyclosporin A and basiliximab may also be useful in the treatment of other potentially fatal autoimmune blistering disorders, such as bullous pemphigoid or pemphigus vulgaris. Although treatment was effective in the present case, spontaneous remission cannot be ruled out with certainty.

REFERENCES


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