Anti-CD20 monoclonal antibody (rituximab) in the treatment of autoimmune diseases. Successful result in refractory pemphigus vulgaris: report of a case

Haematologica 2007; 88(8):e113-e114

Pemphigus vulgaris is a potentially life-threatening autoimmune blistering disease of the skin and mucous membranes, characterized by flaccid bullae that rupture and leave erosions and scars.1 Its treatment is challenging. Although the use of systemic corticosteroids remains the cornerstone of effective therapeutic regimens for PV, their prolonged administration may lead to serious side effects. It is therefore necessary for many patients to add immunosuppressive agents or use chemotherapy to achieve remission but sometimes any treatment is unsuccessful.2 Rituximab is a human-mouse chimeric monoclonal antibody which reacts specifically with the CD 20 antigen, inducing marked and prolonged systemic B-cell depletion.

This effect leads to a strong reduction of antibody (and auto-antibody) production and justify its use in B-cell non-Hodgkin’s lymphoma3 and some auto-immune diseases.4,5,6 M. M. a 53-year old female first showed cutaneous manifestations of pemphigus vulgaris in 1992. After many years of steroid treatment and pulsed doses of methotrexate, the patient required 60 mg/day of methylprednisolone and 100 mg/day of cyclophosphamide to obtain an unsat-

Figure 1. Many flaccid blisters, erupted bullae, erosions and hemorrhagic crusting of the thoracic skin.

Figure 2. Hemorrhagic erosions on the thighs.

Figure 3. Complete healing of skin lesions three months after therapy.
satisfactory control of the disease (Figure 1 and Figure 2).

In June 2002 rituximab treatment at a scheduled dose of 375 mg/m²/week was started and completed in four weeks. The treatment was well tolerated and no side effects were observed. At the same time the previous therapy was progressively reduced. Three months after the end of the therapy we observed a complete healing of cutaneous lesions (Figure 3 and Figure 4). Cutaneous complete remission still persists ten months after treatment. Nobody has reported the duration of remission and we could expect a longer one than in clonal lymphoproliferative diseases such as NHL. This case confirms that rituximab is appropriate, effective and safe in the treatment of PV as recently reported. The considerable cost limits its utilization to such particular conditions as in the case described, when other immunosuppressive regimens have failed or the disease is life-threatening.

L. Virgolini, V. Marzocchi
Correspondence: Virgolini Luigi, Dipartimento di Medicina, U.O. Medicina 2, Azienda Ospedaliera Santa Maria degli Angeli, 33170 Pordenone, Italy. Telephone +39 0434 399236; Fax +39 0434 399700

References