Letters to the Editors

Sustained response to rituximab in a patient with Sjögren’s syndrome and severe refractory polyneuropathy

Sirs,

Rituximab has in some studies been reported to have efficacy in primary Sjögren’s syndrome (pSS) (1-3). Mekinian and associates have very recently investigated the efficacy of rituximab on the neurological symptoms of pSS (4-5). They found that rituximab was not effective in progressive MS-like manifestations of pSS patients with central nervous system involvement (4). However, they described improvement of peripheral nervous system symptoms evaluated 3 months after rituximab treatment in 65% of 17 pSS patients (5). We have a similar experience of favourable effects of rituximab therapy on peripheral nervous system symptoms in one patient with pSS. Moreover, her good response to the treatment has now been sustained for two and a half years. Her polyneuropathy is associated with systemic pSS, as previously noted in a majority of cases (6).

The patient is a 56-year-old female suffering from pSS since 2000, fulfilling the American-European consensus criteria for pSS (7). In her labial salivary gland biopsy more than two lymphocyte focuses/mm² were found. In addition to sicca symptoms, she has presented with fatigue, Raynaud’s phenomenon, gland swellings and occasional purpuric lesions on the lower extremities. In 2001 she developed muscle weakness, sensory or sensorymotor polyneuropathies. In 2001 the symptoms were consistent with a symmetric axonal sensorimotor polyneuropathy. In 2001, the patient was started. For the monoclonal gammapathy, related polyneuropathy symptoms andassociated polyneuropathy symptoms and generalised lymphadenopathy she received paracetamol and anti-histamine as pretreatment. In October 2009, serum IgM fell to 4 g/L, the RF titer decreased dramatically, and serum beta-2 microglobulin and C4 levels normalised. In December 2009, the findings in electrophysiology were comparable to those from 2004 and no more recent findings were observed. The lymphadenopathy disappeared, and up to January 2012 she has felt remarkably better, and her polyneuropathic symptoms have been alleviated. Her methylprednisolone dose has been tapered to 6 mg/day and serum IgM has remained normal up to January 2012. The laboratory and ESSDAI (9) values of our patient before rituximab therapy and at 3 and 30 months follow-up are given in Table I.

Our patient report is in accord with the results reported from the AIR registry (5) that rituximab was favourable in a pSS patient with a monoclonal gammapathy accompanied by an exceptionally severe polyneuropathy. The systemic activity index ESSDAI has been shown to correlate of patients with primary Sjögren’s syndrome: a new reappraisal. J Rheumatol 2011; 38: 1779-85.

References


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