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CASE REPORT

Rituximab as a first-line agent for the treatment of dermatomyositis

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Abstract B cells may play a pivotal role in the pathophysiology of DM, and reports have claimed that targeting B cells is a viable treatment option in patients with dermatomyositis. A 20-year-old girl presented in October 2007, with few weeks' history of proximal muscle weakness. Gottron's papules were noted on her knuckles. She had normal inflammatory markers and negative autoantibody screen. Her CPK was 7000 U/L (normal range of 0–170) with an LDH of 1300 U/L (normal range of 266–500). EMG and muscle biopsy was consistent with active myositis. She had normal pulmonary function tests. HRCT showed no interstitial lung disease. She was started with 60mg glucocorticoids (1mg/kg), with a good clinical response. However, any attempt to taper down the steroid dose led to recurrence of her symptoms. The options of available immunosuppressive therapies, including the experimental usage of rituximab, were discussed with her; averse to long-term systemic treatments, she opted to try a course of rituximab. She had rituximab 1,000 mg on days 0 and 14, and her glucocorticoids were tapered in next few weeks. Now, 24 months since her rituximab infusions, she remains in complete clinical and biochemical remission. She remains naïve to other immunosuppressive agents apart from glucocorticoids and rituximab. Depleting peripheral B cells with rituximab (one course) in our patient has led to not only complete resolution of muscle and skin disease (induction) rather she remains off all immunosuppressives including glucocorticoids.

Keywords Dermatomyositis · Rituximab · Corticosteroids

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The traditional treatment approach to dermatomyositis (DM) is immunosuppressive therapy, and targeted effective treatment for DM remains elusive. B cells may play a pivotal role in the pathophysiology of DM, and reports have claimed that targeting B cells is a viable treatment option in patients with dermatomyositis. Few case reports and two small open-label studies report the use of rituximab (an anti-CD20 monoclonal antibody) in patients with refractory disease [1–4], and the results of rituximab in myositis (RIM) study are anxiously awaited. However, little is known about the rituximab usage in immunosuppressive naive dermatomyositis. We would like to share our experience of using rituximab as the first-line agent for treating a patient with dermatomyositis (DM). To our knowledge, this is the first reported case of rituximab use in immunosuppressive naive dermatomyositis; moreover, in our patient, no maintenance immunosuppressives were used.

Case report

A 20-year-old girl (student of physical education) presented in October 2007, with few weeks' history of muscle weakness and generalized muscle aches and pains. Her physical examination showed symmetric proximal muscle weakness of upper and lower limbs, and manual muscle testing score of 4/5. Gottron's papules were noted on her knuckles. She was not taking any regular medications, and she had no family history of muscular disorders. On laboratory tests, she had normal complete blood count, normal liver function tests, normal kidney disease profile (including urea, creatinine, calcium, phosphate, albumin), normal inflammatory markers, and negative autoantibody screen (along with negative anti-Jo-1 antibodies). Her creatine phosphokinase was 7,000 U/L (normal range of 0–170) and

lactate dehydrogenase of 1,300 U/L (normal range of 266–500). EMG and muscle biopsy was consistent with active myositis, suggestive of dermatomyositis. She had normal pulmonary function tests. HRCT showed no interstitial lung disease, but showed changes consistent with a rare incidental diagnosis of lymphangioleiomyomatosis. According to these findings, she was diagnosed with DM and was started on 60 mg glucocorticoids (1 mg/kg), with a good clinical response. However, any attempt to taper down the steroid dose led to recurrence of her symptoms. The options of available immunosuppressive therapies including the experimental usage of rituximab were discussed with her; averse to long-term systemic treatments, she opted to try a course of rituximab. She received rituximab 1,000 mg on days 0 and 14, and her glucocorticoids were tapered in next 8 weeks. Now, 24 months since her rituximab treatment, she remains in complete clinical (with normal muscle strength) and biochemical remission regarding her dermatomyositis and has recently passed all her examinations relating to her physical education course. She remained naïve to other immunosuppressive agents apart from short courses of glucocorticoids and rituximab. About 6 months into her remission of dermatomyositis, she developed inflammatory oligoarthritis involving her bilateral knees and right elbow. This was associated with normal muscle enzymes, mildly raised inflammatory markers (CRP 15 mg/L), rheumatoid factor positive at a titer of 81 IU/ML, and the rest of her autoantibody status remained unchanged. Anti-CCP antibodies were not tested. Her inflammatory arthritis responded to one short course of oral glucocorticoids (3 weeks only), and she is asymptomatic since.

Discussion

Dermatomyositis is an idiopathic inflammatory myositis and is characterized by its classic skin and proximal muscle involvement. The main stay of treatment is corticosteroids and the use of different steroid-sparing agents (immunosuppressives). In refractory cases, there are mixed reports of experience with intravenous immunoglobulin (IVIG) [5, 6]; furthermore, a very recent trial has failed to show any efficacy of infliximab in these refractory cases [7]. In contrast to polymyositis and inclusion body myositis, humoral immune mechanisms appear to contribute to the pathogenesis of dermatomyositis; and the idea of targeting B cells seems intriguing. The mechanism of beneficial effects of rituximab in dermatomyositis is not precisely known but is thought to be related to modulation of pathogenic autoantibodies.

In summary, depleting peripheral B cells with rituximab (one course) in our patient has led to not only complete resolution of muscle and skin disease (induction) rather she remains off all immunosuppressives including glucocorticoids. For management of dermatomyositis, rituximab seems an important welcome addition to a limited armamentarium. Further long-term investigations are required to confirm these findings and to define the place of rituximab among the therapeutic options for this rare disease.

Conflict of interest statement None.

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