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CASE REPORT

Subcutaneous immunoglobulin administration: an alternative to intravenous infusion as adjuvant treatment for dermatomyositis?

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Abstract Monthly high-dose intravenous administration of human polyclonal immunoglobulins (IVIG) has been shown to be effective as an adjuvant treatment for dermatomyositis. We report a patient with dermatomyositis treated with high doses of immunoglobulins by subcutaneous infusion (SCIG). SCIG was used because of the lack of peripheral and central vein access to continue effective IVIG therapy. The treatment was administered at home, was well tolerated, and was associated to the stabilization of the disease after a 1-year follow-up. Thus, our report suggests that SCIG could be an alternative to IVIG in the treatment of dermatomyositis.

Keywords Dermatomyositis · Idiopathic inflammatory myositis · Intravenous immunoglobulins · Subcutaneous immunoglobulin

High doses of intravenous immunoglobulins (IVIG) have been shown to be effective in idiopathic inflammatory myopathies in patients resistant or with a partial response to conventional therapy [1, 2]. Doses are usually of 2 g/kg of body weight every 4 weeks during 6 months and require a good venous access. Immunoglobulins can also be administrated weekly subcutaneously (SCIG) in primary antibody deficiencies. SCIG is safe and associated with a reduced side effect in primary immunodeficiency but by using much lower doses (between 0.2 and 0.8 g/kg every 3 weeks) than those required for inflammatory myopathies [3]. Only a few cases report the use of SCIG for their immunomodulatory and anti-inflammatory properties in chronic inflammatory demyelinating polyneuropathy as an alternative to IVIG [4, 5]. We report here the 1-year follow-up of the use of SCIG in a patient with dermatomyositis at a monthly dose of 1.7 g/kg. The SCIG was used after the patient passed an informed consent because of the contraindication of an implanted central venous catheter and the lack of peripheral vein access for IVIG administration.

A 55-year-old woman was referred to our clinic for the treatment of dermatomyositis after the failure of a first-line treatment with corticosteroids, methotrexate, and ciclosporin A. She presented with typical skin rash, severe muscle weakness, lung fibrosis, and creatin phophokinase (CPK) level of 7,000 UI/L (N<145 UI/L). Dermatomyositis was associated to anti-JO1 antibodies, and the muscle biopsy showed typical perivascular inflammatory exudates with intracapillary deposits of the membrane attack complex. Monthly IVIG at 2 g/kg and cyclophospamide (600 mg/m^2) pulses associated to oral corticosteroid were initiated. At 6 months, the patient recovered normal muscle strength (CPK 113 UI/L), and skin rash disappeared. After 1 year, IVIG infusions were extended to every 2 months, associated to oral corticosteroids. Two months later, the patient relapsed (CPK 9,000 UI/L). Monthly IVIG infusions and corticosteroid pulses decreased CPK levels to 435 UI/L and improved muscle strength after 3 months. For a second time, an infection of an implantable central venous catheter was associated to a Gram-negative bacteriemia and

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was removed. Because of the several infectious episodes, the patient refused a new implantable central venous catether. Thus, IVIG was stopped because of the lack of a peripheral venous access. Oral corticosteroids were associated to azathioprine. Three months later, the patient status worsened, and azathioprine was switched for mycophenolate mofetil 2 g/day. One month later, her state was unchanged with persistence of proximal muscle weakness and CPK levels of 4,495 UI/L. The skin rash reappeared. We proposed to the patient to use SCIG as an alternative to IVIG. Her body weight was of 90 kg. We decided to use an intermediate dose of 1.7 g per month because of the volume limitation. The SCIG (Vivaglobin 160 mg/mL; CSL Behring) was administered by a programmable pump (CRONO super PID) with a syringe capacity of 30 mL. For 2 consecutive days a week, the patient received 4.8 g immunoglobulins on four different sites, administered by two pumps, at an initial infusion speed of 15 ml/h. Administration was done at home. Transient subcutaneous swelling was well tolerated without local or general side effects. The infusion speed could be progressively increased to 20 ml/h. The mycophenolate mofetil regimen was maintained unchanged, but the corticosteroid could be tapered. Skin rash disappeared in 1 month. Progressively, clinical symptoms of muscle weakness improved, and CPK levels decreased (2,890 UI/L at 3 months, 1,644 UI/L at 4 months, and 225 UI/L at 10 months). The SCIG is well tolerated with a 1-year follow-up, and the patient had no infectious complications.

In the present case, we report that SCIG, used at high doses for an anti-inflammatory effect in dermatomyositis, is well tolerated without side effects. We cannot demonstrate SCIG efficacy but our report suggests an anti-inflammatory effect in our patient in regard to the clinical stabilization and symptom improvement with a follow-up of 1 year. Two previous reports have suggested the efficacy of SCIG in chronic inflammatory demyelinating polyneuropathy [4, 5], another autoimmune disease. The subcutaneous way of administration was well tolerated, as in our case, but with the use of much lower doses. In these cases, it is also suggested that SCIG efficacy is comparable to IVIG [4, 5]. Thus, SCIG should be further evaluated as an alternative to IVIG administration in autoimmune diseases, as it has the advantage to allow with safety the administration of immunoglobulins at home.

Disclosures None.

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