

## CASE REPORT

## Successful therapy with intravenous immunoglobulin in the management of polymyositis

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**Abstract:** Polymyositis is an inflammation of muscle tissue of unknown etiology. It is characterized by symmetric, mainly proximal muscle weakness, muscle fiber damage proved on biopsy, increased enzymes and myoglobin, and has corresponding electromyography findings. Other systems such as joints, lungs, heart, and gastrointestinal system are involved. Lung involvement is rather common. The most frequent symptom represents shortness of breath caused by muscle weakness.

We report a case of a 66 year old woman with primary idiopathic polymyositis. The clinical state of the patient was complicated by progressive muscle weakness, dysphagia, and respiratory failure. Due to the ineffectiveness of the treatment with corticosteroids and cyclophosphamide, treatment with high doses of immunoglobulins was started. A total of 100g of i.v. immunoglobulin therapy was administered beginning on the 13th day after hospital admission. The state of the patient progressively improved and after 7 weeks of treatment in a significantly improved state the patient was transferred to a Rehabilitation Unit.

We therefore conclude that IVIg therapy may be an effective therapeutic approach for the treatment of acute complications of polymyositis, especially in cases in whom other therapeutic strategies are ineffective or harmful (Ref. 10). Full Text (Free, PDF) [www.bmj.sk](http://www.bmj.sk).

Key words: polymyositis, intravenous immunoglobulin, cyclophosphamide.

Polymyositis is a chronic disease of unknown etiology characterized by inflammation of the muscle fibers. It starts when white blood cells, the immune cells of inflammation, spontaneously invade muscles. The muscles affected are typically those closest to the trunk or torso. This results in weakness that can be severe. Polymyositis is a chronic illness with periods of increased symptoms, called flares or relapses, and minimal or no symptoms, known as remissions. (1). Although myositis is the dominant clinical manifestation, internal organs may also be affected. It may be associated with other autoimmune diseases such as myasthenia gravis, Hashimoto's thyroiditis, systemic sclerosis and Waldenstrom's Macroglobulinaemia (4).

Intravenous immunoglobulins (IVIg) have been used for the treatment of various autoimmune disorders especially autoimmune thrombocytopenia and Kawasaki disease. Several series have also reported an overall beneficial effect of IVIg treatment in SLE, dermatomyositis, primary antiphospholipid syndrome and Churg Strauss syndrome (5, 6). We describe a patient with polymyositis who showed a prompt regression following a few doses of IVIg.

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### Case report

A 66-year-old Caucasian female was admitted because of fever, weakness, diffuse joint pain, muscle aches, and edema of the extremities. Severe weakness up to the extent of inability to walk or swallow was present. On admission, this patient was unable to rise from a seated position without help and to raise arms above her head.

Laboratory tests: Hematological examination revealed mild anemia (hemoglobin 11.2), leukocytosis (13.9) and thrombocytosis (657) with normal coagulation parameters (PT, APTT). Inflammatory parameters (FW, CRP, WBC) were increased and so was serum myoglobin indicating muscle tissue damage. Hepatic enzymes were also increased (AST 6.15; ALT 6.41; GMT 0.18) and hyperazotemia (urea 16.4; creatinine 104) with decrease glomerular filtration (GF) and mild proteinuria (0.3) were present.

Immunological examination detected auto-antibodies (ANA, ENA, anti JO-1) to be negative but anti-RO was slightly positive.

Electromyography detected acute myogenic lesions supporting the diagnosis. Neurological findings proved polymyositis.

Muscle tissue biopsy neither confirmed nor ruled-out the diagnosis of polymyositis.

Spirometry showed a restrictive ventilation disorder.

Contrast X-ray of esophagus during swallowing showed no evidence of organic disorder to cause the dysphagia.

Gastroscopic investigation proved pangastritis and *Candida* esophagitis.

Treatment was started on the 8th day of hospitalization with pulsating dose of 500 mg i.v. methylprednisolone. The next day progression of muscle weakness, hypotension, and respiratory failure were present. The patient was transferred to Intensive Care Unit where artificial ventilation and acute blood volume replacement were performed.

Meanwhile treatment with methylprednisolone 500 mg i.v. was continued (5 days). Prednisone 60 mg for 21 days in progressively decreasing doses to 45 mg was then administered. Pulsating treatment with cyclophosphamide was not successful. Because of the failure of all previous methods of treatment and worsening of blood biochemistry, treatment with IVIg 0.04 kg/day for 5 days was indicated. The clinical state of the patient started to improve with mild improvement of dysphagia and muscle strength. The patient was able to breathe independently 12 hours a day through a tracheostomy cannula without the aid of artificial ventilation. The patient was also administered antimicrobial and antimycotic treatment to prevent infection. Diarrhea was present during the course of hospitalization but microbiological investigation was negative. The suspected dysmicrobia was corrected with enteral diet which was administered by nasogastric tube. From the 7th day of hospitalization at Intensive Care Unit the patient began oral intake. After 5 weeks in the Intensive Care Unit the patient was transferred back to the Internal Clinic. The tracheostomy cannula was removed on the 12th day after the transfer. Chronic therapy continued with oral administration of Prednisone, LMWH and with rehabilitation. After the patient was in a significantly better state, she was transferred to Rehabilitation Unit.

## Discussion

In this case report we describe a patient with primary idiopathic polymyositis. The clinical state of the patient was complicated by progressive muscle weakness, dysphagia, and respiratory failure with a quick improvement after treatment with IVIg.

Polymyositis is a type of inflammatory myopathy, related to dermatomyositis and inclusion body myositis. Polymyositis tends to become evident in adulthood, presenting with bilateral proximal muscle weakness, often noted in the upper legs due to early fatigue while walking. Sometimes the weakness presents itself by the person being unable to rise from a seated position without help, or inability to raise their arms above their head.

The weakness is generally progressive, accompanied by lymphocytic inflammation (mainly cytotoxic T8 lymphocytes). The cause is unknown, but seems to be related to autoimmune factors, genetics, and perhaps viruses.

Treatment generally involves glucocorticoids, especially prednisone. At present, a number of studies are underway to determine whether patients diagnosed with polymyositis will benefit from newer drugs inhibiting the biologic effects of TNF alpha, such as Infliximab ("Remicade") (7).

There are few reports documenting a rapid improvement after IVIg administration in cases when corticosteroid treatment

failed. In our patient meanwhile treatment with methylprednisolone with a dose of 500 mg daily i.v. was continued (5 days). Prednisone 60mg for 21 days in progressively decreasing doses to 45 mg was then administered. Pulsating treatment with cyclophosphamide was not successful. Because of the failure of all previous methods of treatment and worsening of blood biochemistry, treatment with IVIg 0.04 kg/day for 5 days was indicated.

IVIg is a normal human polyclonal IgG obtained from pooled plasma from a large number of healthy blood donors (9). Initially used as replacement therapy for patients with primary and secondary immune deficiencies. IVIg is now widely used for the treatment of a large number of autoimmune and systemic diseases (9, 10).

The mechanisms of IVIg action include enhancement of suppressor activity, Fc receptor blockade, complement regulation, idiotypic network regulation etc. The presence of anti-idiotypes to autoantibodies within IVIg preparations provides an explanation of its beneficial effect (7). In the study of Cherin in patients with polymyositis the author documented a significant clinical improvement in 15 from 20 patients after IVIg treatment (8). In another study, authors described an adverse allergic manifestations as a complication of IVIg treatment in 3 out of 15 patients (3). There were no allergic manifestations associated with IVIg therapy in our patient.

In conclusion, the IVIg therapy was an effective therapeutic strategy in the treatment of polymyositis when other therapy (methylprednisone, cyclophosphamide) was not successful.

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