

Commentary

Difficult treatment – many options

Myasthenia gravis is an autoimmune disease that affects the neuromuscular junction. The disease is most often caused by nicotinic acetylcholine receptor (AChR) antibodies in the postsynaptic muscular part of the junction. In rare cases the disease is caused by antibodies against muscle-specific tyrosine kinase (MuSK) or against lipoprotein-related protein 4 (LRP4).

Myasthenia gravis is a relatively rare disease, with around 150 cases per million inhabitants and an incidence of around 10–15 new cases per million inhabitants per year. The illness manifests itself with ocular symptoms such as diplopia and ptosis, bulbar symptoms such as dysarthria and dysphagia, and generalised symptoms with paresis of the neck muscles and the proximal muscles of the extremities. Fluctuating symptoms on improvement after rest are typical.

The treatment of myasthenia gravis is symptomatic, by inhibiting acetylcholinesterase in the junction with pyridostigmine, and immunologically with corticosteroids or other immunosuppression (1). Thymectomy is indicated for patients with thymoma (approx. 10–15%) and for those with generalised myasthenia gravis. Maintenance treatment usually entails the use of pyridostigmine and low-dose prednisolone, possibly

with azathioprine as a steroid-saving drug. A number of other immunosuppressive drugs may be tried, such as mycophenolate mofetil, rituximab, methotrexate or cyclosporine (1, 2).

In this case a patient is described with known myasthenia gravis and myasthenic crisis. A number of factors can exacerbate myasthenia gravis, as can various drugs that affect the neuromuscular junction – and not least infections, as was the case here. The treatment of myasthenic crisis is intravenous immunoglobulin (IVIG) or plasma exchange. These treatments result in rapid improvement, and studies show that they are equally effective. High-dose corticosteroids can also result in improvement of myasthenic crisis if the aforementioned treatments have not had sufficient effect. These were administered in the course of treating the patient in question, in addition to antibiotics for pneumonia. Rituximab has also been shown to improve the course of illness and this drug should be tried if IVIG or corticosteroids have failed to give results (2, 3). The patient improved markedly following the latter treatment.

This case history illustrates some important points. Myasthenic crisis can be difficult to treat. Immunosuppressive treatment must often be given despite ongoing infection.

IVIG, corticosteroids and rituximab in that sequence are drugs of choice for myasthenic crisis.

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