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Case Report

Must we Treat Neuromyelitis Optica in Patients with Myasthenia Gravis More Aggressively? – A Case Report

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Abstract

Coexistence of neuromyelitisoptica (NMO) and myasthenia gravis (MG) has only rarely been documented. The outcome in these patients, despite all therapeutic interventions, is usually very poor.The aim of our study was to present a case and a short review of literature in order to encourage the neurologists to rethink about the treatment of these patients.We are reporting a case of 48-year old Caucasian woman with MG and the NMO that was treated with high doses of methylprednisolone and underwent plasmaferesis at her last in hospital stay but ended in spastic tetra paresis, bedridden, incontinent and blind. The course of NMO in patients with MG is quite severe with common relapses and poor outcome. Only by summing all our experiences we can bring out a good strategy for their treatment.

Keywords: Neuromyelitisoptica; Devic`s disease; Myasthenia gravis; Management

Abbreviations

NMO: Neuromyelitisoptica; MG: Myasthenia Gravis; AChR: Acetylcholine Receptor; ON: Optic Neuritis; Anti-nuclear, Anti SS-A, Anti SS-B, Anticardiolipin, Anticytoplasmatic MPO and PR3; MRI: Magnetic Resonance Imaging; C5 to C7 and from Th7 to Th9

Introduction

Neuromyelitisoptica (NMO, also known as Devic`s disease) is an idiopathic inflammatory syndrome of the central nervous system, predominantly affecting the spinal cord and optic nerves [1]. NMO is considered as an autoimmune disease and recent immune pathological evidences suggest that the target antigen is aquaporin-4, the dominant water channel in the central nervous system (CNS) [2]. NMO is occasionally associated with other autoimmune diseases, such as hypothyroidism, Sjögren's syndrome (SS), systemic lupus erythematosus (SLE) and pernicious anemia [3]. Neuronal and muscle antibodies commonly coexist with NMO and its marker, NMO-IgG [4].

Myasthenia gravis (MG) is also an autoimmune disorder caused by autoantibody that binds to the acetylcholine receptor (AChR) [5]. Muscle acetylcholine receptor antibodies were detected in around 12% of NMO patients [6,7]. Coexistence of MG and NMO has only rarely been documented; to the best of our knowledge only around forty patients with NMO and MG were reported and only half of them were Caucasians [8-12].

Though we are presenting only one case of coexistence of these two diseases, we believe that each case *per se* is important because of mostly very poor outcome in these patients. Only by summing all our experiences we can bring out a good strategy for their treatment.

Case Report

We can say that our 48-year old Caucasian woman fits the profile of previously reported patients [10,11]. Female with early onset of MG, mild course of MG and the NMO followed after more than a decade. More detailed; she was diagnosed MG at the age of twenty-four. She was treated with pyridostigmine; underwent thymectomy three years later (pathohistological findings unknown). At the age of forty four she was treated because of the optic neuritis (ON) for the first time. After ON the right eye she developed ON her left eye four years later and again one year later. Since then her vision was reduced dramatically. Visual evoked responses showed prolonged P100 latencies bilaterally. For each episode of ON she was treated with high doses of methylprednisolone. MG symptoms were no longer present and pyridostigmin was omitted from therapy. During her last episode of ON she was complaining on numbness of whole body, especially the legs. Few months later she developed tetra paresis. Hypothyreosis was diagnosed. Aquaporin 4 antibodies were positive. Other antibodies (anti-nuclear, anti SS-A, anti SS-B, anticardiolipin, anticytoplasmatic MPO and PR3) were negative. Brain magnetic resonance imaging (MRI) showed nonspecific punctiform demyelinating lesions in pons and bilaterally sub cortically. Spinal cord MRI discovered demyelinating lesions extending from C5 to C7 and from Th7 to Th9. Her cerebrospinal fluid showed intratecal synthesis of oligoclonal bands. She was administered high doses of corticosteroids (methylprednisolone) after which she underwent plasmaferesis but without benefit. At the end of our follow-up she was in spastic tetra paresis, bedridden, incontinent and blind on therapy with azathioprine.

Discussion

Though demyelinating diseases among patients with MG are rare, their incidence is much higher than expected in the general population [13,14]. The incidence of MG among patients with NMO spectrum disorder is around 2% [6,10]. In those patients MG usually remits or takes a mild course. But the course of NMO is quite severe with common relapses and unsatisfactory outcome. Most patients are treated with methylprednisolone [9,12] or prednisone [8] and azathioprine [8,11]. Though some authors reported favourable early

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outcome [9,12] it is usually associated with short disease duration and a long-term outcome is generally poor. More than 60% of patients have significant disability including bilateral blindness and/or para- or tetra paresis [10,11]. Unfortunately, our patient ended tetra paretic and blind. From this point of view we believe that she should be treated more aggressively, with all available sources. It is recommended to consider plasmaferesis in treatment of NMO if the therapy with methylprednisolon fails to bring improvement [7,15]. But maybe we should consider plasmaferesis as a first line therapy and even try double filtration or cryofiltration. As Kim et al. mentioned, in their review, early initiation of plasma exchange is tightly associated with neurological improvement [7]. Other options are intravenous immunoglobulin for the acute phase treatment and also as a "prophylactic" therapy in monthly infusions. Intermittent plasma exchange could also be used in prevention of relapses as well as rituximab [7].

Conclusion

It is important to bear in mind that patients with MG can have other associated autoimmune disorders. For those presenting with atypical motor or optic symptoms NMO should be considered. Common relapses and poor outcome are its characteristics. Quick diagnosis and aggressive treatment may make a difference.

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