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CASE REPORT

MYASTHENIA GRAVIS WITH SUPERIMPOSED SPINAL CORD INJURY: A CASE REPORT

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Both myasthenia gravis and traumatic spinal cord injury are uncommon disorders and their concurrence is extremely rare. We report here the case of a man with stable myasthenia gravis with spinal cord injury due to a motor vehicle accident. His muscle strength and sensory function in all 4 limbs partially recovered during the initial hospitalization. However, after a later episode of pneumonia and urinary tract infection, muscle strength deteriorated and weakness of the ocular muscles occurred. A relapse of myasthenia gravis was confirmed by the elevated anti-acetylcholine receptor antibody titre. Muscle strength recovered rapidly after control of infection and treatment of myasthenia gravis exacerbation. In this case report, we show that spinal cord injury-related complications, including infection or emotional stress, could trigger a relapse of myasthenia gravis. This highlights the importance of recognizing an exacerbation of myasthenia gravis in spinal cord injury patients with a history of myasthenia gravis, since they are prone to urinary and pulmonary infection. Although exacerbation of myasthenia gravis might prolong hospitalization, the functional outcome of spinal cord injury might not be affected if the exacerbation is diagnosed and treated quickly.

Key words: spinal cord injuries, myasthenia gravis, rehabilitation.

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INTRODUCTION

Myasthenia gravis (MG) is an autoimmune disorder of neuromuscular transmission in which autoantibodies are formed against nicotinic acetylcholine receptors at the neuromuscular junction (NMJ). The clinical symptoms are progressive weakness of the ocular, bulbar, limb, and respiratory muscles following repeated contractions, and recovery of muscle strength after a period of rest (1–2). MG is a relatively uncommon disorder, with a reported annual incidence of 50–125 cases per million people (1, 3). The most common age of onset of MG is the second to third decades in women and the sixth to eighth decades in men. Traumatic spinal cord injury (SCI), which leads to sensorimotor deficits below the level of injury, is also an uncommon event, with an estimated annual incidence of 19 per million people in Taiwan (4) and 30–60 per million people in most industrialized countries (5–6). It is therefore very rare for a patient with MG to have superimposed SCI. We report here the case of a man with MG who suffered from traumatic SCI due to a motor vehicle accident.

CASE REPORT

A 45-year-old man with a 2 year history of MG presented with fluctuating ptosis and diplopia, but no weakness of the facial, neck, or limb muscles. The symptoms were worse after a period of daily activity, especially in the afternoon, and were relieved by rest. The diagnosis of MG was confirmed by a series of studies, including the edrophonium test, repetitive nerve stimulation test, single fibre electromyography, and serum antibodies test. He underwent thoracoscopic thymectomy and received oral pyridostigmine therapy. His symptoms of ocular motor dysfunction then subsided completely and no further neurological symptoms recurred on pyridostigmine treatment.

In May 2006, he suffered weakness of all 4 limbs immediately after a motor vehicle accident. He was sent to the emergency department, where neurological examination revealed markedly reduced muscle strength in the 4 limbs (MMT grade 0–1/5), absence of sensation below the C4 dermatome, absence of tendon reflexes, urine retention, and loose anal tone. These findings suggested a spinal cord lesion. Magnetic resonance imaging (MRI) of the cervical spine showed posterior bulging of the intervertebral disc at C3–4, C4–5 and C5–6, with compression of the dura and cervical cord. T2 weighted images showed a high signal intensity lesion at C3 and C4 in the spinal cord. No spine fracture or brain injury was noted. Based on the diagnosis of traumatic cervical cord injury with tetraplegia, he was immediately treated with high-dose parenteral methylprednisolone. He then underwent laminectomy plus plate fixation for cord decompression and stabilizing the spine. His neurological function improved gradually after the above treatment. Muscle strength in all 4 limbs improved to fair (MMT grade 3/5) and sensory function partially recovered after an 8-week inpatient rehabilitation programme. At discharge after the first hospitalization, he was able to walk with the aid of a walker

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for a short distance, but needed a wheelchair for long-distance locomotion. In terms of activities of daily living, he needed moderate assistance in feeding, grooming and dressing. Due to a large post-voiding residual volume (> 100 ml), a urethral indwelling catheter was used for bladder management.

Approximately one week after discharge, he had an acute onset of fever, malodorous urine, and cough with purulent sputum and was sent to the emergency department, where laboratory tests disclosed pyuria and leukocytosis, and chest radiograph showed increased infiltration over the right lower lung field. These findings suggested urinary and respiratory tract infection. The infection was treated with parenteral antibiotics and the fever subsided within a few days. However, one week after re-admission, the weakness of the ocular muscles and all 4 limbs worsened rapidly (from MMT grade 3/5 to 1–2/5). However, sensory function did not deteriorate. Cervical spine MRI showed no significant cervical cord signal changes compared with the previous images. Levels of serum electrolytes, creatine phosphokinase, and thyroid hormones were all within normal limits. No medication which could affect NMJ function (such as aminoglycoside) was used during the admission. Relapse of MG was therefore suspected and was confirmed by serological studies, which showed an elevated anti-acetylcholine receptor antibody titre. After treatment with plasmapheresis and oral corticosteroid, the ocular motor disturbance subsided and the muscle strength in all 4 limbs improved to grade 3/5. For suspected impaired swallowing function with aspiration, he underwent videofluoroscopic examination, which revealed mild weakness of the pharyngeal muscle and trace silent aspiration during swallowing, which was compatible with the presentation of dysphagia in patients with MG. He resumed adequate oral intake and the nasogastric tube was removed after swallowing training. For bladder training, he was instructed in timed voiding and clean intermittent catheterization techniques. After a 9-week inpatient rehabilitation programme, muscle strength in all 4 limbs improved significantly (MMT grade 4/5). At discharge, he could walk with a walker under supervision for indoor activity and could feed himself independently using a universal cuff once this had been set up and the table arranged by a caregiver and perform other activities of daily living with minimal to moderate assistance.

He continued to undergo outpatient-setting rehabilitation training after discharge. Neither infection nor MG exacerbation occurred during the 6-month follow-up. He developed only mild spasticity during this period, which did not affect his functional performance.

DISCUSSION

This paper describes the case of a man with traumatic SCI who had a history of stable chronic MG. The concurrence of traumatic SCI and MG in the same patient is extremely rare and, as far as we are aware, has not been reported previously. In the case described here, MG activity remained stable during the acute stage of cervical SCI, but muscle strength in all 4 limbs deteriorated rapidly after an episode of fever. MRI of the cervical spine showed no structural change in the cervical cord that could account for the weakness and there was no deterioration of sensory function. The worsening of muscle strength was therefore unlikely to be due to a new spinal cord lesion. In addition, metabolic factors (e.g. electrolytes and thyroid function) or medications that could affect motor function were excluded. Exacerbation of MG was therefore suspected as the cause of the decline in muscle strength and was confirmed by an increased serum anti-acetylcholine receptor antibody titre and rapid improvement after plasmapheresis.

A myasthenic relapse has been reported to be triggered by factors including systemic illness (especially infections), emotional stress, increase in body temperature, drugs affecting neuromuscular transmission, hypothyroidism or hyperthyroidism, pregnancy, or the menstrual cycle (7–10). It is important to recognize a relapse of MG in patients with acute SCI, as they are typically under enormous emotional stress and are prone to urinary and pulmonary infection, which could lead to an exacerbation of MG. In our case, the urinary and respiratory tract infections could account for the relapse. Emotional stress appeared to play a relative minor role in this case, since his MG activity remained stable during the acute stage of SCI. Respiratory and genitourinary complications are the most common reasons for re-hospitalization in the SCI population (11–13), as in our case. Thus, pulmonary rehabilitation and neurogenic bladder management are important in patients with SCI with MG because proper treatment may not only reduce SCI-related morbidity, but also prevent MG exacerbation. Medications for the treatment of spasticity are commonly used in persons with chronic SCI, but have been reported as potential causes of a myasthenic relapse (9, 14). Since no marked spasticity developed in the presented case, no antispastic medication was used. Nevertheless, medical personnel should be reminded that antispasticity agents could exacerbate MG activity.

Our patient exhibited rapid recovery of muscle strength after plasmapheresis and steroid therapy and regained functional performance after rehabilitation training. This case shows that, although an MG relapse may prolong hospitalization, with early diagnosis and proper treatment, it has little impact on the functional outcome of rehabilitation.

In conclusion, we describe here a case of MG with superimposed traumatic cervical SCI. Complications of SCI, such as infection or emotional stress, might contribute to the exacerbation of MG, which, in turn, might prolong hospitalization. Nevertheless, with early recognition and proper management, exacerbation of MG should have little effect on the rehabilitation outcome of the SCI.

REFERENCES

1. Drachman DB. Myasthenia gravis. N Engl J Med 1994; 330: 1797–1810.
2. Keesey JC. Clinical evaluation and management of myasthenia gravis. Muscle Nerve 2004; 29: 484–505.
3. Phillips LH. The epidemiology of myasthenia gravis. Semin Neurol 2004; 24: 17–20.
4. Liang HW, Wang YH, Lin YN, Wang JD, Jang Y. Impact of age

on the injury pattern and survival of people with cervical cord injuries. Spinal Cord 2001; 39: 375–380.
5. DeVivo MJ. Epidermiology of traumatic spinal cord injury. In: Kirshblum S, Campagnolo DI, DeLisa JA, editors. Spinal cord medicine. Philadelphia: Lippincott, Williams & Wilkins; 2002, p. 69–81.
6. Pickett GE, Campos-Benitez M, Keller JL, Duggal N. Epidemiology of traumatic spinal cord injury in Canada. Spine 2006; 31: 799–805.
7. Grob D, Arsura, EL, Brunner NG, Namba T. The course of myasthenia gravis and therapies affecting outcome. Ann NY Acad Sci 1987; 505: 472–499.
8. Simpson JF, Westerberg MR, Magee KR. Myasthenia gravis. An analysis of 295 cases. Acta Neurol Scand 1966: 42 Suppl 23: 1–27.
9. Wittbrodt ET. Drugs and myasthenia gravis: an update. Arch Int Med 1997; 157: 399–408.
10. Richman DP, Agius MA. Treatment of autoimmune myasthenia gravis. Neurology 2003; 61: 1652–1661.
11. Cardenas DD, Hoffman JM, Kirshblum S, McKinley W. Etiology and incidence of rehospitalization after traumatic spinal cord injury: a multicenter analysis. Arch Phys Med Rehabil 2004; 85: 1757–1763.
12. Haisma JA, van der Woude LH, Stam HJ, Bergen MP, Sluis TA, Post MW, Bussmann JB. Complications following spinal cord injury: occurrence and risk factors in a longitudinal study during and after inpatient rehabilitation. J Rehabil Med 2007; 39: 393–398.
13. Lidal IB, Snekkevik H, Aamodt G, Hjeltnes N, Biering-Sorensen F, Stanghelle JK, Mortality after spinal cord injury in Norway. J Rehabil Med 2007; 39: 145–151.
14. Adams SL, Mathews J, Grammer LC. Drugs that may exacerbate myasthenia gravis. Ann Emerg Med 1984; 13: 532–538.