Case Study

Co-occurrence of Guillain-Barré syndrome and myasthenia gravis, the first report in Iran

Shaghayegh Rismanchi1, Mojtaba Rismanchi2* and Mehdi Karimian2

1Research Center Tower, Shiraz, Iran
2Clinical Neurology Research Center, Department of Neurology, Shiraz University of Medical Sciences, Shiraz, Iran

Abstract

Guillain-Barré syndrome and myasthenia gravis are two immune mediated neuromuscular diseases, both threaten respiratory function and may emerge as medical emergencies. Co-occurrence of the two entities though possible is a rare condition, and therefore poses special diagnostic difficulties. Herein, a report of co-presentation of Guillain-Barré syndrome and myasthenia gravis is discussed and the literature is studied regarding this rare combination.

Introduction

Myasthenia Gravis (MG) and Guillain-Barré Syndrome (GBS) are autoimmune disorders of neuromuscular junction and peripheral nerves respectively [1]. Both of these entities may produce similar presentations including bulbar and respiratory manifestations with rapid and excessive exacerbations leading to the need for Intensive Care Unit (ICU) care [1]. In fact it is highly probable to diagnose these disorders during ICU admission because of their rapid deterioration during their initial presentation.

Coincidence of these two disorders is extremely rare. The incidence of GBS is estimated to be 5 cases per million and that for MG to be 2 to 4 cases per million individuals [2]. It is also statistically estimated that the chance of coincidence of GBS and MG has to be approximately one of two billion people per year claiming that the coincidence is quite unlikely [2,3]. It is also noted that this combination of disorders are more prevalent between Chinese people which may in part point to the possible genetic background and ethnicity as predisposing factors for this rare combination [4,5]. In this report, we have presented a case of MG, newly diagnosed during her severe exacerbation, with clues for GBS particularly facial palsy that lead to the diagnosis of GBS after several days of ICU admission. To our knowledge this is the first co-occurrence of GBS and MG in Iran.

Case

A 32-year-old female with right hand paresthesia that progressed to quadriparesis in the next day was brought to Nemazee hospital affiliated to Shiraz University of Medical Sciences (SUMS). She had history of upper respiratory tract infection two weeks before the present weakness. She did not have aphthalmoplegia or sphincteric problems. In physical examination, the patient had right sided ptosis, symmetrical muscle power scale of 1 out of 5 in 4 limbs (quadriceps in lower and biceps and triceps in upper extremities) and absent deep tendon reflexes. No evidence of sensory level or upward plantar reflexes were found. The patient developed respiratory distress, was intubated and received mechanical ventilation. Based on history and significant ptosis in physical exam, Intravenous Immunoglobulin (IVIG) was started for her with the impression of MG which was then repeated after two weeks. During the treatment she further developed bilateral facial palsies that along with newly noticed symmetrical paresthesia
in distal extremities raised questions about diagnosis with suspicion of neuropathies like GBS. Lumbar puncture was done after 7 days of presentation showing 200mg/dl protein content and 5 lymphocytes. During the present admission her ptosis was also clinically approached in search for myasthenia. The patient had no past or present complaints involving bulbar muscles including diplopia, nasal speech or regurgitation. Anti acetylcholine receptor antibody was checked two times that were positive (3.33nmol/L and 8.33nmol/L). In the following days, Pyrdostigmine and prednisolone were started. They accompany restoration of muscle power and improvement of ptosis. She was discharged with oral prednisolone and pyrdostigmine.

Regarding laboratory investigations, Spiral chest computed tomography for Thymic mass or hyperplasia was negative. Electrodiagnostic study showed polynеuropathу (mainly axonal) with spontaneous muscle action potentials during Electromyography. Sural nerve sparing was also detected; all were in favor of GBS. Repetitive stimulation test was not performed due to possible interference of GBS with this modality (patient consent received).

Discussion

Co-occurrence of MG and GBS is extremely rare. However these two disorders share similar pathomechanism involving humoral immunity. There are several theories explaining the co-occurrence, among which one claims that there is cross reacting antibodies for these two disorders that cause both entities [1,6]. Another theory suggests that antibody titers for MG is low in an individual and this titer increases due to immunity response directed against infectious processes which themselves cause GBS [7]. Regardless of the cause of these disorders, both MG and GBS also share similar presentations including bulbar and respiratory manifestation [1] which may lead to the neglect of one entity in the presence of the other [4]. A relatively difficult diagnosis of both disorders in one patient may also explain the rare reports of MG and GBS co-occurrence [2]. Useful clinical features which can differentiate these disorders in cases of combined presentation include: prominent ptosis without ophthalmoplegia in MG, areflexia and sensory paresthesia in GBS, and distribution of weakness which is proximal and distal in case of GBS and proximal in MG [1]. Notably it is reported that one third of reported cases had the presentation of the second disease within the first month of diagnosing the first disorder. The co-occurrence is more pronounced in male individuals and all the cases are reported to have areflexia and ptosis and furthermore, almost all cases have positive results in anti acetylcholine receptor antibody assays [1]. In our case the co-occurrence was in a young woman and the co-presentations were almost simultaneous during first ICU admission.

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