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A Case Report on Miller Fisher Variant of Guillain Barre Syndrome in a Tertiary Care Hospital

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Abstract

Miller fisher syndrome is considered to be an uncommon cranial nerve variant of Guillain Barre Syndrome. It is characterized by the clinical triad of opthalmoplegia, ataxia and are flexia that can result from nerve damage caused by an aberrant autoimmune response to a bacterial or viral infection. The following report describes the case of a 7 year old female patient who presented with complaints of blurring vision and right eye drooping. Later she developed weakness of all limbs with respiratory failure that required ventilatory support and was also treated with immunoglobins, corticosteroids, antibiotics and other supportive medications. The patient gradually improved and was discharged in the recovery phase.

Keywords: Miller Fisher Syndrome • Guillain Barre Syndrome • Opthalmoplegia • Ataxia • Areflexia

Abbreviations: MFS: Miller Fisher Syndrome, GBS: Guillain Barre Syndrome, IV-IG: Intravenous Immunoglobin, PSV: Pressure Support Ventilation, SIMV: Synchronised Intermittent Mechanical Ventilation

Introduction

Guillain Barre Syndrome (GBS) is a type of neuromuscular paralysis that has several variants with distinct clinical and pathological features. The overall incidence rate of GBS increases with age and is reported to be 0.6 - 2.4 cases per 100, 000 a year where there is a slight predominance observed in males than in females [1]. Miller Fisher syndrome (MFS) is a clinical variant of GBS that is reportedly rare among children and is observed in only about 1% to 5% of all cases [2]. Etiology of the disease is incompletely understood but is believed to be due to autoimmune response to a preceding infection that is directed against peripheral nerve components because of molecular mimicry between the myelin found in peripheral nerves and the lipo-oligopolysaccharide of *Camphylobacter jejuni* which is the common cause of illness. Some of the other pathogens involved are *Haemophilus influenzae, Mycoplasma pneumoniae*, *Cytomegalovirus, Epstein-Barr Virus* and *Influenza* [3,4].

The onset of MFS is typically acute, beginning with neurologic symptoms approximately 8–10 days following the antecedent illness. The classic triad of symptoms include opthalmoplegia, ataxia and are flexia The CSF protein is elevated after the first week and the presence of anti-GQ1b antibodies are seen in 90% of patients with occulomotor (cranial nerve III), trochlear (cranial nerve IV) and abducens (cranial nerve VI) nerve having dense concentrations. This may explain the correlation between anti-GQ1b antibodies and external opthalmoplegia [2,5]. The most life-threatening complication of GBS is respiratory failure where one-third of patients are admitted to an intensive care unit (ICU), and many require mechanical ventilation. During this critical phase, they are at risk of systemic complications with the potential of substantial morbidity and consequent mortality. Thus, awareness of the risk factors of respiratory failure in children with GBS is important to prevent associated complications and even death [6]. Although MFS follows a self-limiting course, immunomodulatory therapies including intravenous immunoglobulins and plasmapheresis is of proven benefit and crucial, especially in patients with rapidly progressive weakness [3].

Case Report

A 7-year-old female patient came with a short history of right eye drooping, blurring and double vision. On arrival she was noted to have right lagopthalmos and bilateral lateral rectus nerve palsy. Her vitals signs were within the normal range and were fully alert and oriented. Next day she developed complete opthalmoplegia of right eye and lateral rectus palsy of the left along with progressive weakness of all four limbs starting from lower limbs and more on proximal muscles compared to distal muscles. On day 3 she had bilateral opthalmoplegia with both pupils briskly reactive. She was initiated on methyl prednisolone and IV-IG (2 g/kg over 2 days). NCV study showed neuropathy confirming the diagnosis of GBS with MF variant (Figure 1).

In view of unusual presentation of just eyes involved, myasthenia gravis was suspected and she underwent Neostigmine tests which produced a good response in the form of bilateral eye movements but no change in ptosis. Test for anti-Musk antibody and AchR antibody was negative. MRI of brain and spine was reported as normal. CSF study including Meningo-Encephalitis Panel (PCR) demonstrated no abnormality. She was haemodynamically stable with no clinical features of sepsis and normal renal and cardiovascular function. On 4th day, she developed erratic breathing pattern with absent gag reflex and drooling, suggestive of respiratory failure, hence she was intubated and started on ventilation. With regard to fast progress of her condition and nature of the illness, prolonged ventilation was anticipated. Tracheostomy was

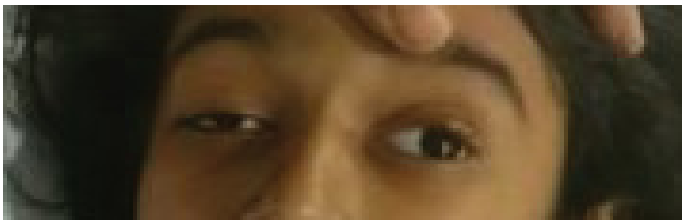

Figure 1. A 7-year-old female patient with a short history of right eye drooping, blurring and double vision.

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planned and done on 5th day. Subsequently she was ventilated with an aim to gradually wean ventilation once the recovery of respiratory muscle function and airway protective reflex achieves. She also received medications including analgesics, gabapentin, antibiotics and other supplements.

Later her enteral nutrition was restarted and gently established on oral feeds. Hypertension was then noticed which attributed to autonomic dysfunction as well as steroid therapy and was treated with amlodipine and carvedilol till the resolution of the issue. Gradually she had improvement of respiratory function with help of physiotherapy and hence weaned from ventilation by cycling with PSV and SIMV. Finally she was observed off ventilation support for 48 hrs, without having any signs of respiratory distress. She was stabilized by maintaining normal oxygen saturation on room air and limb muscle power 4/5 in all limbs with presence of cough and gag reflex. The patient was discharged in the recovery phase and the care giver has been advised about the course of the illness, long term requirement of rehabilitation and supportive care.

Discussion

MFS presents with a wide range of clinical features with most presenting symptom as bilateral opthalmoplegia. Our patient started with diplopia and blurring of vision which gradually progressed to opthalmoplegia of both eyes and then weakness of all limbs. A diagnosis of MFS can be made with clinical history, cardinal symptoms, CT or MRI findings, presence of albuminocytologic dissociation in the CSF and raised serum level of GQ1b ganglioside antibody [2]. However our patient was found with normal cell count and protein level in CSF with no significant MRI/CT findings though the results of nerve conduction studies supported the diagnosis of the syndrome.

MFS is fundamentally treated with adequate supportive care, management of pain, respiratory support and immunotherapy. IV-IG and plasma exchange are considered to be the disease modifying option for GBS and MFS with dysphagia and respiratory difficulties. Signs of respiratory muscle fatigue include tachypnea, tachycardia, abdomen movement and evident use of accessory muscles. Mechanical ventilation and ICU admission are required in 20-30 percent of patients along with endotracheal intubation and tracheostomy when needed [7]. A large retrospective study have shown that immunosuppressive treatment may hasten the resolution of symptoms but neither of them influence the patient's outcome because of self -limiting course of the disease [8]. Moreover Osulani et al. reported a case where the patient had a spontaneous and good recovery without immunomodulation therapies. On the other hand another case of severe MFS was reported by Ajena et al. in which patient experienced remarkable clinical improvement after the IV-IG cycles suggesting its effectiveness [9].

An optimal pain management is needed in the early course of the disease to accelerate recovery. A combination of medicines including anticonvulsants and corticosteroids are recommended but steroids be used only in the setting of neuropathic and radicular pain [10].

Conclusion

Physiotherapy is the mainstay of management in this case to recover function. The main goals of therapy is to achieve optimal muscle use as tolerated by pain and use supportive equipment to help patient regain functional activity as close to baseline as possible. The prognosis of MFS is usually good with mean recovery time range from 8-12 weeks. It can be shortened with appropriate treatment and complete resolution of symptoms is expected within few months. Therefore majority of people will recover completely and have no permanent muscle weakness.

Acknowledgment

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Conflict of Interest

The authors declare no conflict of interest.

References

1. Willison, HJ, Jacobs BC and Van Doorn PA. "Guillain-Barre syndrome" *The Lancet* 388 (2016): 717-727.
2. Bukhari, S and Taboada J. "A case of Miller fisher syndrome and literature review" *Cureus* 9 (2017).
3. Sudalugunta, Sreenivasa Rao, Mahesh Babu Sodalagunta, Mona Sepehrar and Hadi Khorram, et al. "Guillain-Barre syndrome clinical profile and management" *Medical Sci* 13 (2015).
4. Osalusi, BS and Ogun SA. "Miller fisher Syndome-A Case report" *Annals of Health Research* 3 (2017): 71-74.
5. Saha, R, Saha, SK, Islam MN and Kabir MR, et al. "Miller Fisher variant of Guillain Barre Syndrome" A Case Report. *Faridpur Med Coll J* 10 (2015): 44-45.
6. Hu Mei Hua, Chiung-Mei Chen, Kuang-Lin Lin and Huei-Shyong Wan, et al. "Risk factors of respiratory failure in children with Guillain-Barré syndrome" *Pediatr Neurol* 53 (2012): 295-299.
7. Cabrero, FR and Morrison EH. "Miller Fisher Syndrome" *In: Stat Pearls*.
8. Mori, M, Kuwabara S, Fukutake T and Yuki N, et al. "Clinical features and prognosis of Miller Fisher Syndrome" *Neurol* 56(2001): 1104-1106.
9. Ajena, D, Ferrari S, Romoto S and Zaglia F, et al. "A pediatric case of Miller Fisher syndrome with central involvement" *Neurol Sci* 34 (2013):1689-1690.
10. Liu, Jia, Lu Ning Wang and Ewan D McNicol. "Pharmacological treatment for pain in Guillain-Barré syndrome" *Cochrane Database Syst Rev* (2015): CD009950.

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