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Successful management of a rare case of Miller Fisher variant of Guillain – Barre Syndrome in rural tertiary care

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Abstract

Guillain-Barre syndrome (GBS) is also known as acute idiopathic polyneuritis. It is a neuromuscular paralysis that occurs in all the age groups and has several variants. These include demyelinating polyneuropathy, acute axonal neuropathy, Bickerstaff brainstem encephalitis, pharyngo-cervical-brachial variant, Miller Fisher syndrome and polyneuritis cranialis. Miller Fisher syndrome (MFS) is a rare variant of GBS which is observed only about 1% to 5% of all cases. The incidence of the MFS is more common in males when compared to females by a ratio of 2:1. The lower and upper age groups reported with MFS are 13 and 78 years respectively. Here we report the successful management of rare form of GB syndrome.

Keywords: Guillian Barre syndrome, Immunoglobulins, Miller Fisher Variant, Paralysis

Introduction

Guillain-Barre syndrome (GBS) is also known as acute idiopathic polyneuritis or acute inflammatory demyelinating polyradiculoneuropathy (AIDP).[(1)] It is commonly characterized by rapidly evolving ascending weakness, mild sensory loss with hypo or areflexia. It occurs in both the sexes with a male preponderance and can occur at any age. Though in western countries, GBS is commonly reported in 5th decade but in India it occurs more commonly at a younger age.[(2,3)] Several variants are seen and these are demyelinating polyneuropathy, acute axonal neuropathy, Bickerstaff brainstem encephalitis, pharyngo-cervical-brachial variant, Miller Fisher syndrome and polyneuritis cranialis.[(4)] Miller Fisher syndrome (MFS) is a rare variant of GBS which is observed only about 1% to 5% of all cases.[(5-7)] Various clinical findings and laboratary tests help in the diagnosis of the GBS. The most commonly performed test is cerebrospinal fluid evaluation which demonstrates albuminocytologic dissociation in 90% of cases.[(8)]

Certain studies have shown slight preponderance of AIDP in Indian population. The Indian literature indicates that the incidence of GBS is more in June–July and Sept–October.[(9)] In Indian population the incidence of AIDP and acute motor axonal neuropathy (AMAN) are virtually equal.[(10)] Here we are reporting a case of successful management of Miller Fisher variant of GBS.

Case Report

A 17 yr old female patient presented with history of difficulty in breathing, cough with expectoration, fever and giddiness since 5 days. There was no history of nausea, vomiting or similar complaints in the past. The family and personal history were insignificant. All the symptoms were sudden in onset and gradually progressive in nature. At the time of admission only positive findings on examination of patient were bilateral basal crepitation in the lungs. A diagnosis of lower respiratory tract infection was made and patient was transferred to medical ICU for further management. After two days patient complained of difficulty in swallowing and irritation in the throat. The otolaryngologist did indirect laryngoscopy and found bilateral fixed paramedian position of the vocal cords. A differential diagnosis of bulbar palsy and bilateral vagal nerve palsy due to post viral infection was made. The magnetic resonance imaging study of brain was normal. In next 6 hours patient's oxygen saturation decreased and a decision of invasive mechanical ventilation was taken to assist the respiration. Patient later on developed ptosis and double vision (ophthalmoplegia), limb examination showed absence of deep tendon reflexes. Incoordination in right upperlimb was noted which was suggestive of ataxia. A differential diagnosis of Guillian Barre syndrome was made. A lumbar puncture was performed to analyze the cerebrospinal fluid (CSF) which was normal. By 2nd day of admission the muscle power in both the lower limbs was 0/5, right upper limb 2 /5 and left upper limb showed 1/5 grades. We were unable to do nerve conduction study bedside due to technical reasons. A decision was made to administer intravenous immunoglobulins (15 gms /day) for 5 days. Under closed observation I.V immunoglobulins were administered at 80 ml/hr. On second day, the patient's muscle power increased to 3/5 in lower limbs, 4/5 in right and 3/5 in left upper limbs. On 10th day of admission the CSF study was repeated, which showed the evidence of albuminocytological dissociation (Table 1). At the end of 5th dose, patient's muscle power improved in all the limbs but areflexia persisted. Weaning from the ventilator was carried out during the immunoglobulin therapy and patient met all the criteria of weaning at 2nd day of post immunoglobulin therapy. Extubation was successful and patient clinically improved. The patient was made to walk and we could notice the gait

abnormality after 24 hours of extubation which was subsequently improved after 48 hours. The patient was subjected for common peroneal nerve conduction study, which showed that decreased amplitude and reduction in conduction velocity with increased latency. All the deep tendon reflexes didn't reappear even after post immunoglobulin therapy. The patient was observed in the medical ICU for another 48 hours and then shifted to step down area. Patient was discharged on 23rd day of hospital admission.

Table 1: CSF Analysis

Features	On Admission	On 10th day of admission
Appearance	Clear & Colourless	Clear & Colourless
Biochemical	Glucose: 72mg/dl	Glucose: 48mg/dl
	Protein: 25mg/dl	Protein: 149mg/dl
	LDH: 192IU/L	LDH: 218IU/L
Microscopy	Total WBC- 2cells/cumm (Both lymphocytes) RBC & Malignant Cells- Absent	Total WBC- 4cells/cumm (Predominant lymphocytes) RBC & Malignant Cells- Absent

Discussion

GBS is a neurological disorder which is also known as acute idiopathic polyneuritis. It has a worldwide annual incidence of 1.3 cases/100,000 population. The classical presentation of GBS includes non-febrile, acute monophasic, postinfectious illness, ascending weakness and areflexia. Patient may also have sensory, autonomic, and brainstem abnormalities. Number of variants has been described till date. The clinical suspicion of GBS in our case was based on the presence of a ptosis, weakness, flaccidity and areflexia which was mainly in the lower limb and gradually progressed to both upper limbs. Differential diagnoses of poliomyelitis, occult snake bite and poisoning had to be considered. The main diagnostic tool for the diagnosis of GBS was albuminocytological dissociation in CSF which was absent initially but present on 10th day of presentation. We could not assess the anti-GQ1b IgG antibodies due to unavailability of such tests in our rural set up. Due to technical reasons the nerve conduction study could not be conducted initially but was conducted on 7th day of post admission. The bilateral common peritoneal nerve conduction study showed reduced amplitude and conduction velocity with increased latency suggestive of myopathy changes. In our patient at the time of admission patient did not complain of vision problem. With retrospective enquiry, our patient accepted the fact that she had vision disturbances like double vision which improved with due course of treatment. Rarely bilateral ptosis without ophthalmoplegia may be the initial presentation.[11]

CSF study shows albuminocytologic dissociation in 82–90% of the patients with GBS after 10–14 days from onset of the illness. A high CSF protein levels with normal cell counts and sugar levels were observed in the initial week of presentation. The normal CSF findings are seen during the first week of disease, albuminocytologic dissociation is seen in 82–90% by the end of second week of the illness. Proteins leak in to the CSF at the nerve root level due to demylination which is the cause of rise in the protein levels. The rises in inflammatory cells are not seen in CSF since there is no inflammation.[12] GBS has many variants, in that Miller Fisher syndrome is a rare variant which can be diagnosed by clinical triad i.e. areflexia, ataxia, and ophthalmoplegia. Our case patient had features suggestive of areflexia, ataxia and ophthalmoplegia.

According to literature the ataxia and opthalmoplegia may resolve within 1–3 months after onset and near complete recovery is expected within 6 months. In our patient the ataxia clinically recovered after 3 days of immunoglobulin therapy. The patient had areflexia persistently, but was not associated with functional disability which is again, reported in earlier cases.[6,13]

The present case could recover due to high index of suspicion of GBS as in rural setup all the facility may not be readily available. One should be aware of signs, symptoms consistent with MFS.

Conclusion

A high index of suspicion of GBS in patients with acute flaccid paralysis, irrespective of the age should be kept in mind. Nerve conduction study and CSF analysis are main diagnostic tests for GBS but clinical findings in rural set up play an important role. An initial CSF examination may not be always positive for the albuminocytological dissociation hence repeat investigation may confirm the diagnosis.

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