

Case Report

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Miller fisher syndrome: A case report and review

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Abstract

Guillain-Barre Syndrome (GBS) is a rapid-onset weakness caused by immune system damaging the peripheral nervous system. Miller Fisher Syndrome (MFS) is a variant of GBS characterized by weakness of the eye muscles, abnormalities in coordination and absent reflexes. The symptoms may develop over hours to a few weeks. Although rare, it has a tendency to recur, mostly affects younger age group and may have a long duration of course significantly affecting psychologically and economically. However, it remains a less reported and studied entity. There is a need for detailed study to provide recommendations for proper diagnosis and management to reduce the morbidity as well as adverse psychological, social and economic impact.

Keywords: Guillain Barre syndrome; Peripheral nervous system; Intravenous immunoglobulins.

Introduction

Guillain Barre syndrome is named after the French neurologist Georges Guillain and Jean Alexandra Barre who, together with French physician Andre Strohl described the condition in 1916. Two-thirds of people with GBS give a history of gastroenteritis or respiratory tract infection [1]. Approximately 30% of cases are provoked by *Campylobacter jejuni* bacteria, which cause diarrhoea. The first symptoms of GBS are numbness, tingling and pain, alone or in combination followed by weakness of the legs and arms that affects both sides equally and worsens over time [2,3]. Miller Fisher Syndrome (MFS), a clinical variant of GBS is characterized by a triad of weakness of the eye muscles, abnormalities in coordination and absent reflexes [3]. Although rare, it has a tendency to recur and affects young. However, it remains a less reported and studied entity. We present a case of MFS made evident by the presence of the characteristic triad of ataxia, ophthalmoplegia and areflexia.

Case report

A 22-year-old male presented with complaints of bilateral ptosis, numbness in both hands spreading to whole body and inability to walk since one day. The patient was asymptomatic

till one day prior when he felt inability to open eyes fully on waking up in morning. Two hours later, he started having numbness in distal part of both hands which spread to whole hands in three to four hours. The numbness spread to lower limbs also and he developed ataxia. He also gave history of rhinitis on and off since past eight months. He had also received three doses of covid vaccine including booster dose and three doses of hepatitis B vaccine, in which last dose was taken before one month.

On examination at presentation, he was conscious and oriented. Respiratory, cardiovascular and per abdominal examination were unremarkable. Nervous system examination revealed normal higher mental function, normal nutrition, tone and power (5/5) but bilateral absent deep tendon reflexes. Romberg's test was positive and incoordination was seen on tandem walking. Rest of the sensory system examination was unremarkable. On the basis of ophthalmoplegia, ataxia and areflexia, the patient was diagnosed as MFS and admitted in ICU. Routine investigations (Complete blood count, renal function tests, liver function tests, electrolytes etc.) were all normal. MRI brain and CSF were also within normal limits. Nerve conduction study showed features of demyelinating neuropathy (Table 1). Five cycles of plasma exchange were given to the patient along

with symptomatic and supportive treatment. With plasma exchange therapy still ongoing, his ophthalmoplegia disappeared and ataxia also started improving. After completion of plasma apheresis, at the time of discharge, there was no ophthalmoplegia and he was able to walk on his own with only mild ataxia.

Table 1: Lab reports.

Investigations	On day of admission
Hb (gm/dl)	15.2
WBC (/cu.mm)	13390
CRP (mg/L)	3.40
ESR(mm)	2
Platelet (lakh/cu.mm)	242000
PT(second)	13.6
INR	1.06
aPTT (second)	29.4
HbsAG	Negative
Anti HCV	Negative
Hba1c	5
Ca ²⁺ (mg/dl)	8.8
SGOT(IU/L)	29
SGPT(IU/L)	17
Na ⁺ (mEq/L)	144
K ⁺ (mEq/L)	4.6
Cl ⁻ (mEq/L)	109
Urea (mg/dl)	14
Creatinine (mg/dl)	0.6
Total protein (g/dl)	7.2
Albumin (g/dl)	4.6
Globulin (g/dl)	2.6
Total bilirubin(mg/dl)	0.7
Indirect bilirubin (mg/dl)	0.5
Direct bilirubin (mg/dl)	0.2
SGPT (IU/L)	35
Uric acid (mg/dl)	5.3
Nerve conduction study	Generalized sensory and median motor demyelinating neuropathy with imperistent f-wave in nerves of bilateral median, ulnar, peroneal nerve may be suggestive of early GBS in appropriate clinical setting.
MRI brain plain	No significant neuroparenchymal abnormality detected. Mucosal thickening in right maxillary, ethmoid and sphenoid sinuses.
X-ray chest AP view	Normal
ECG	Sinus rhythm
CSF routine micro	Total count – 03 cells/microliter Total RBC – 3-4/hpf Lymphocyte – 100% Protein – 11.7 mg/dl Sugar – 65 mg/dl

Investigations	27 th Nov 2022	28 th Nov 2022	29 th Nov 2022	30 th Nov 2022
Na ⁺ (mEq/L)	141	142	138	136
K ⁺ (mEq/L)	3.9	3.26	3.8	3.6
Cl ⁻ (mEq/L)	105	111	106	103
Hb (gm/dl)		13.4		
WBC (/cu.mm)		11060		
Platelet (lakh/cu.mm)		193000		

Discussion

GBS is an autoimmune disorder encompassing a heterogeneous group of acute, immune-mediated polyneuropathies [5,6]. It can affect all age groups and 20% increase in incidence has been reported every 10-years. It can present as paraesthesia (80%), dysautonomia (70%), back pain (66%) or severe respiratory weakness (50%). About 10% patients present with weakness beginning in either the arms or facial muscles [7-9]. MFS typically presents as ophthalmoplegia with areflexia and ataxia [10,11]. Respiratory symptoms precede neurological symptoms in about 76% of MFS cases (in contrast to GBS) while gastrointestinal involvement is reported in only 4% of MFS patients. The most common pathogen reported to trigger GBS is *C. Jejuni* which may be found in as many as 30% of GBS cases [12]. However, MFS has been found to be associated with *Haemophilus influenzae*, *Mycoplasma pneumoniae*, *Helicobacter pylori* as well as viral infection from Epstein-Barr virus, varicella-zoster virus, cytomegalovirus and HIV [12-15]. Although recurrent MFS is a rare disease, it may occur in genetically susceptible individuals [16]. Patients with younger age at presentation have been found to be more prone to recurrence [17].

Differential diagnosis of MFS includes GBS, Bickerstaff's brainstem encephalitis, brainstem stroke, botulism, myasthenia gravis, phenytoin and fluorouracil toxicity and Wernicke's encephalopathy. Rapid-onset ophthalmoplegia, lack of lateralisation of ataxia in MFS and associated features of history and clinical examination in other diseases helps in finalising the diagnosis. Plasmapheresis and Intravenous Immunoglobulins (IVIg) are the two main immunotherapy treatments recommended for GBS. The recovery is speeded up when plasmapheresis is used within four weeks of the onset of symptoms. If IVIg is started within two weeks of the onset of symptoms, it works as well as plasmapheresis and has fewer complications [4], but for MFS there is lack of randomised, double-blind, placebo-controlled trials. A retrospective analysis has reported that although IVIg may reduce ophthalmoplegia and ataxia to some extent, it doesn't affect the overall outcome significantly probably due to the frequency of natural recoveries [18].

In the studies published so far, complete resolution of ataxia by one month and resolution of ophthalmoplegia within three months are taken as outcome measures. However, no studies have compared the average duration of recovery in patients who received treatment with that in those who did not, although looking at the young age at presentation this might also be an important outcome measure as reducing the morbidity would have a significant impact. This case presented with rapid onset ophthalmoplegia and ataxia who responded very well to plasmapheresis and showed rapid recovery; with ophthalm-

moplegia completely disappearing during the treatment course itself. Also, he was able to walk on his own with remarkably reduced ataxia towards the last phase of plasmapheresis. Being at a young age of 28, this could mean returning fast to functional life with lesser adverse psychological, social and economical effect. Hence this case highlights the need to evaluate the treatment of this disease not only in terms of final mortality but also in terms of the psychological and economical impact.

Conclusion

MFS is a less studied entity although it affects young and may recur resulting in a long duration of functional, psychological as well as social affection. So it warrants detailed study to provide recommendations for prompt diagnosis and management to reduce the morbidity as well as adverse psychological, social and economic impact.

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