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# A Rare Presentation of Miller Fisher Syndrome, a Rare Variant of Guillain Barre Syndrome

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Letter to the Editor

# A Rare Presentation of Miller Fisher Syndrome, a Rare Variant of Guillain Barre Syndrome

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## Abstract

Guillain Barre syndrome presents as acute onset of gradually progressive ascending flaccid paralysis with hyporeflexia or areflexia. The variants are acute inflammatory demyelinating polyneuropathy, acute motor axonal neuropathy, acute motor sensory axonal neuropathy, and Miller Fisher syndrome. Miller Fisher syndrome includes symptoms that form a classical triad of ophthalmoplegia, ataxia, and hyporeflexia or areflexia. This case presents a case of Miller Fisher syndrome with atypical features wherein the patient had normal reflexes and his reflexes remained normal in follow-up.

**Key Words:** Areflexia, Guillain Barre syndrome, hyporeflexia, Miller Fisher syndrome, areflexia.

## Introduction

GBS presents as acute onset of gradually progressive ascending flaccid paralysis with hyporeflexia or areflexia, commonly without bowel or bladder involvement which occurs after an infectious disease, most commonly Campylobacter jejuni. The severity reaches its maximum over four weeks. Sensory symptoms if present start distally and asymmetrically. The clinical course, severity, and outcomes of patients with or without treatment (IvIg or plasma exchange (PLEX)) are highly variable [1].

The variants of GBS are acute inflammatory demyelinating polyneuropathy (AIDP), acute motor axonal neuropathy (AMAN), acute motor-sensory axonal neuropathy (AMSAN) and Miller Fisher syndrome [1].

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## Case Summary

This 43-year-old gentleman presented with complaints of (a) imbalance while walking, (b) numbness over the legs and face, (c) blurring of vision, and (d) difficulty in closing his eyes and drooling food from his mouth for 2two days. He had a history of fever with cough for a few days 1-2 weeks before presenting to the hospital. He was a known case of hypertension on irregular treatment and was detected to have diabetes (Table 1) in the hospital (had normal HbA1C six months back). He was conscious, oriented, and responsive to commands. He had facial weakness and had no closure of his eyes against resistance, and limb power was 5/5 in all limbs. He had a loss of joint position and vibration sense. His eye movements were restricted. His reflexes were normal in all limbs and both plantar reflexes were flexor (as demonstrated in the video). A possibility of Guillain Barre syndrome (GBS) was considered, with a strong possibility of variant Miller Fisher syndrome (MFS). The nerve conduction study (NCS) and blink reflex test (Table 1) both suggested a lower motor neuron pattern of weakness. The cerebrospinal fluid (CSF) test showed

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albumin-cytological dissociation (Table 1). Magnetic resonance imaging (MRI) of the brain and cervical spine showed no upper motor neuron lesion (Table 1, Figures 1, and 2). He was initiated on intravenous immunoglobulins (IvIg) therapy and improved on treatment and physiotherapy.

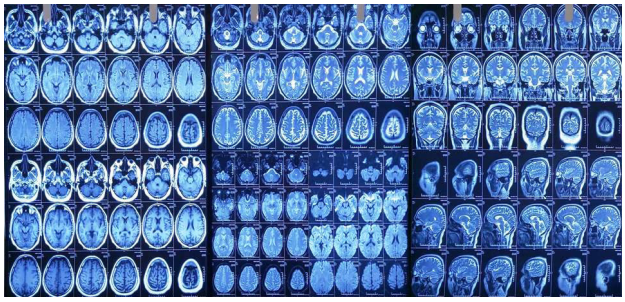

**Figure 1:** MRI of the brain was normal

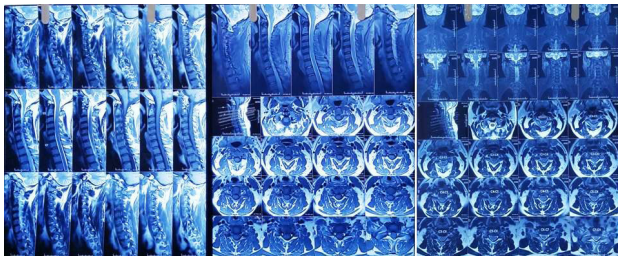

**Figure 2:** MRI of the cervical spine showed mild disc bulges at C4-C5 and C5-C6

## Discussion

The 1990 criteria of GBS need hyporeflexia or areflexia for the diagnosis. The initial presentation may have normal reflexes in a few patients. The reflexes may be normal more commonly in the upper limbs compared to the lower limbs. However, commonly over some time, the reflexes diminish in follow-up. Only a few patients may continue to have normal reflexes in upper limbs at follow-up. Some patients may however have normal or even exaggerated reflexes, due to unknown reasons, especially in the AMAN variant of GBS [1].

MFS includes symptoms that form a classical triad of ophthalmoplegia, ataxia, and hyporeflexia or areflexia. The syndrome may also have the involvement of cranial nerves, which include the facial nerve, glossopharyngeal nerve, vagus nerve, and hypoglossal nerve. MFS patients have dysfunction of the proprioceptive afferent system. The sensory ataxia may be due to the selective involvement of muscle spindle afferents [2, 3].

MFS on electrodiagnostic studies shows loss of sensory involvement too with axonal neuropathy more commonly compared to a demyelinating form of neuropathy [4].

In MFS, the decision on treatment options still needs a randomized control trial. Retrospective analysis, so far, has suggested a good outcome irrespective of the form of treatment- IvIg, PLEX, or no immunotherapy. However, early IvIg therapy showed an earlier response, especially in the more severe cases of MFS-GBS overlap syndrome [1].

Our patient had no weakness, which is an essential component of GBS, besides diminished reflexes. Ataxia on the other hand is not common in GBS. Our patient had ataxia, blurred vision, and restricted eye movements. As noted above, reflexes may sometimes be normal, at least initially, in a few variants of GBS. We, therefore, favoured the diagnosis of MFS.

## Conclusion

In this case, we represent a case of MFS who had normal reflexes. This is a rare presentation of MFS. Also, while most patients develop hyporeflexia during the illness, our patient had normal reflexes at the time of discharge and even during the first review at one month.

**TABLE 1: Relevant Investigations**

| TEST | RESULTS |
|---|---|
| Covid antigen | Negative |
| Dengue serology | Negative |
| Typhi DoT and Widal | Negative |

| | |
|---|---|
| Malarial antigen | Negative |
| Blink reflex test | Abnormal study with bilateral LMN facial weakness |
| NCS all four limbs | Sensorimotor axonal neuropathy |
| RNS study | Normal study |
| MRI brain | Normal Study |
| MRI cervical spine | Straightening of cervical spine curvature with mild disc bulge at C4-C5 and minimal disc bulge at the C5-C6 level |
| CSF study | |
| TLC | 0/cmm |
| DLC | 0 |
| Sugar | 121 mg/dL |
| Protein | 151 mg/dL |
| ADA | 14 U/L |
| Gram | No organism seen |
| Culture | No growth |
| AFB | No AFB seen |
| KOH | No fungal element seen |
| CPK | 10 |
| Echocardiography | Concentric LVH, EF 65% |
| HRCT chest | Normal study |
| NCCT head | Normal study |
| Blood culture | No growth |
| Urine R/M and culture | Normal study |
| Thyroid profile | |
| TSH | 4.07 mIU/mL |
| T3 | 1.32 nmol/L |
| T4 | 120.3 nmol/L |
| HbA1C | 6.10% |
| Vitamin B12 | 872 pg/mL |
| Vitamin D | 22.60 nmol/L |
| CBC, LFT, KFT | Normal |
| Lipid profile | |
| Cholesterol | 193 mg/dL |
| Triglycerides | 299 mg/dL |
| HDL | 43 mg/dL |
| LDL | 90.2 mg/dL |
| VLDL | 59.8 mg/dL |



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