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Case Report

A Rare Presentation of Miller Fisher Syndrome, A Rare Variant of Gb Syndrome

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Abstract

Guillain Barre syndrome presents as acute onset gradually progressive ascending flaccid paralysis with hyporeflexia or areflexia. The variants of GBS are Acute Inflammatory Demyelnating Polyneuropathy, Acute Motor Axonal Neuropathy, Acute Motor Sensory Axonal Neuropathy and Miller Fisher Syndrome. MFS 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 he had normal reflexes and his reflexes remained normal in follow up.

Keywords: *Guillain Barre Syndrome; Miller Fisher Syndrome; Hyporeflexia; Areflexia*

Case Summary

This 43 year old gentleman presented with complaints of (a) weakness of the limbs in which he had imbalance while walking, (b) numbness over the legs and face, and (c) difficulty in closing eyes and drooling of food from mouth for 2 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 detected to have diabetes (Table 1) in hospital (had a normal HbA1C 6 months back). He was conscious, oriented and responsive to commands. He had facial weakness and had no closure of eyes against resistance, and limb power was 5/5 in all limbs. His eye movements were normal. 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. NCS and Blink Reflex test (Table 1) both suggested a Lower Motor Neuron pattern of weakness. CSF test showed albumin-cytological dissociation (Table 1). MRI Brain and Cervical Spine showed no UMN lesion (Table 1 and Figure 1 and Figure 2). He was initiated on IvIg therapy and improved on treatment and physiotherapy.

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 4 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 C4-5 and minimal disc bulge at 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 micro IU/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

Table 1

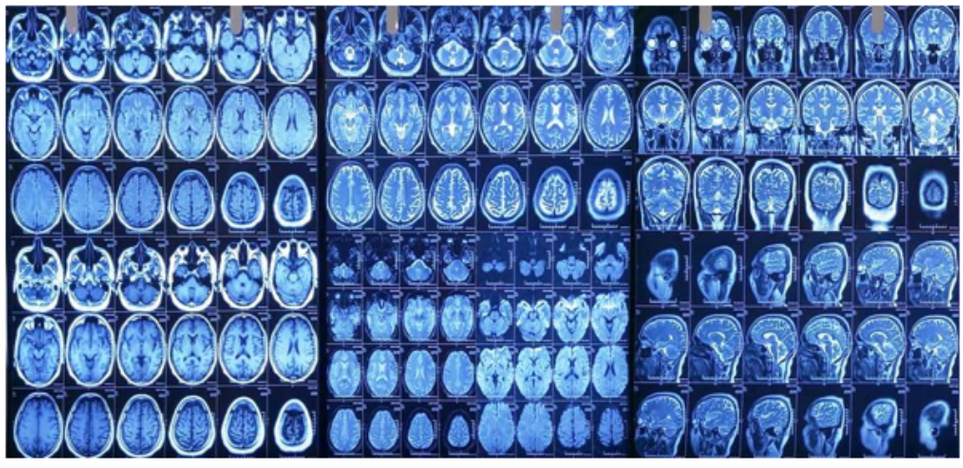

Figure 1: *MRI Brain was normal.*

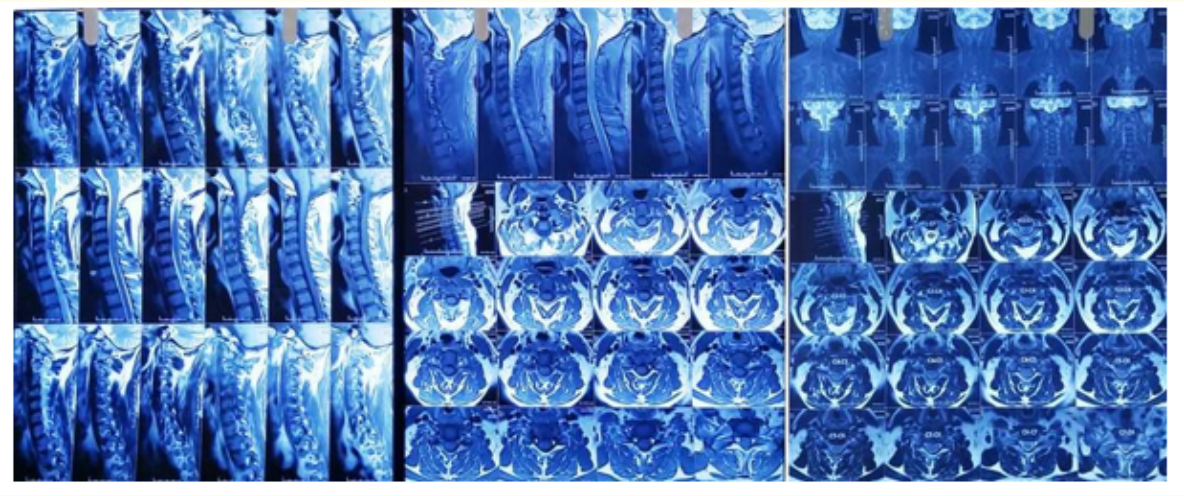

Figure 2: *MRI Cervical spine showed mild disc bulge C4-5, C5-6.*

Introduction

Guillain Barre syndrome (GBS) presents as acute onset 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 4 weeks. Sensory symptoms, if present start distally and asymmetrically. The clinical course, severity and outcomes of patients with or without treatment (Intravenous immunoglobulin (IvIg) or plasma exchange (PLEX)) are highly variable [1].

The variants of GBS are Acute Inflammatory Demyelnating Polyneuropathy (AIDP), Acute Motor Axonal Neuropathy (AMAN), Acute Motor Sensory Axonal Neuropathy (AMSAN) and Miller Fisher Syndrome (MFS) [1].

Discussion

The 1990 criteria of GBS needs 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 a period of 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 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 involvement of cranial nerves that include facial nerve, glossopharyngeal nerve, vagus nerve and hypoglossal nerve [2].

MFS on electrodiagnostic studies, shows loss of sensory involvement too with axonal neuropathy more commonly compared to demyelinating form of neuropathy [3].

In MFS, the decision on treatment options still needs a randomized control trial. Retrospectiva analysis so far have suggested a good outcome irrespective of 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].

Conclusion

In this case we represent a case of Miller Fisher Syndrome who had normal reflexes. This is a rare presentation for Miller Fisher Syndrome. Also, while most patients develop hyporeflexia during the course of the illness, our patient had normal reflexes at the time of discharge and even the first review at 1 month.

Acknowledgement

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Bibliography

1. Van Den Berg B., *et al.* "Guillain–Barré syndrome: pathogenesis, diagnosis, treatment and prognosis". *Nature Reviews Neurology* 10 (2014): 469-482.

2. Leila Tamaoui., *et al.* "Pharyngo-Cervico-Brachial Variant of Guillain-Barré or Miller Fisher Syndromes? When the Overlap Is Misleading". *Reports* 3 (2020): 34.

3. Talebi M., *et al.* "A Case of Miller Fisher Syndrome with Unusual Features: Normal Muscle Stretch Reflexes and Facial Palsy and Dramatic Response to IVIG". *Journal of Neurology and Stroke* 4.3 (2016): 00131.

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