Case Report

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A Rare & Unusual Case of "Miller-Fisher Variant of Gullain-Barre Syndrome"

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Abstract

Miller Fisher syndrome (MFS) is seen in about 1-5% of all cases of Gullain Barre syndrome. MFS is characterized by a classic triad of Ophthalmoplegia, Ataxia and Areflexia. Additional symptoms include bulbar palsy, motor weakness and sensory loss.

However, ophthalmoplegia is the most important and consistent finding in this syndrome. The incidence in males and females is 2:1 and the mean age of presentation is around 43 years. The anti-GQ1b IgG antibody is found in about 90% of MFS cases and it is a very specific and a sensitive marker. We are presenting a case of Miller Fisher syndrome which manifested with ophthalmoplegia, hyporeflexia and ataxia with proptosis and chemosis in a 50 yrs old male.

Keywords: Gullain Barre Syndrome; Miller Fisher Syndrome; Ophthalmoplegia; Ataxia; Areflexia; Proptosis; Chemosis; GQ1b Antibody; IV Immunoglobulin.

Introduction

Gullain Barre Syndrome (GBS) is a rare but serious peripheral nervous system disorder in which the peripheral nerves are affected by the immune system. There are many variants of GBS each of which has typical features and some of which have specific antibodies.

The variants of GBS are tabulated as follows:

GBS AIDP	MFS	AMAN	AMSAN	PCB variant
Gullain Barre syndrome Acute inflammatory demyelinating polyradiculo neuropathy.	Miller Fisher syndrome	Acute motor axonal neuropathy Chinese paralytic syndrome	Acute motor and sensory axonal neuropathy	Pharyngo cervico brachial variant
Post infection with URTI and GI infection C. jejuni is the major infectious cause, followed by H. influenza. Vaccines with influenza and rabies.	URTI GI infection.	C. jejuni Zika viirus	C. jejuni	
Ashbury criteria Ascending paralysis Areflexia Sensory loss B/L 7th n LMN palsy Bladder normal Symmetrical paralysis (Prox ms,	Triad: Ophthalmoplegia Ataxia Areflexia Others: Bulbar palsy Motor weakness	Muscle weakness Sensations normal CN normal Areflexia	Muscle weakness Sensory loss CN normal 7th n LMN palsy.	Face, Throat, Neck, Shoulder muscles weakness Areflexia in UL

then Distal ms). 0.4 – 4.0/1,00,000 GM1 Ab Rx IVIG or Plasmapheresis	Descending paralysis+/- Sensory loss 1-5% cases of GBS GQ1b Ab Rx IVIG or Plasmapheresis	GD1a Ab Rx IVIG or Plasmapheresis	5-10% cases of GBS Rx Immunotherapy not very useful	Rare GT1a Ab GM2 Ab

MFS was first recognized as a distinct clinical entity in 1956 and was described by Canadian neurologist Dr Charles Miller Fisher.

MFS is mostly an acute self-limiting condition. With immunoglobulin therapy, the improvement is hastened and recovery takes about 2-4 weeks and by 6 months most patients recover.

Studies show about 85% patients recover to normal, however about 10% patients have sequellae and 5% deaths.

Case History

A 50 years old Nepalese male patient, a known hypertensive and hypothyroid was brought to the emergency department (ED) with h/o upper respiratory infection 1 week back, followed by slurring of speech, double vision (diplopia), difficulty swallowing (dysphagia), drooping of eyelids (ptosis) and some weakness in upper limbs.

On examination, patient was conscious and oriented and was responding to verbal commands.

Pulse rate = 82/min, regular, BP = 140/90 mmHg, RR = 18/min without any distress, SpO2 = 94% at RA, T = 98.6 deg F, RBS = 121mg/dl.

ECG showed normal sinus rhythm.

Heent

(Figures 1 & 2)

B/L LMN facial palsy

B/L Absence of forehead frowning

B/L loss of angle of mouth

B/L Ptosis

B/L Proptosis

B/L Eyelid edema

B/L Chemosis

Left 12th CN paresis

Pupils: Rt – Normal size with sluggish reaction.

Lt – Normal size with normal reaction.

Fundoscopy: No abnormality detected bilaterally.

Throat: Pooling of secrections+ (swallowing reflex impaired).

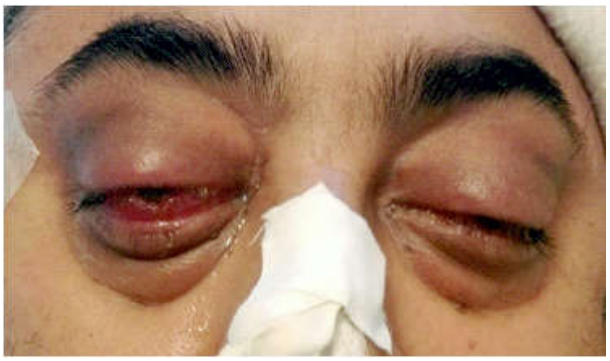

Fig. 1:

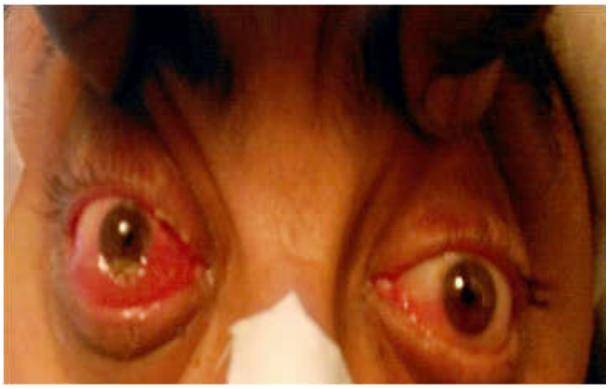

Fig. 2:

Neurological Examination

Conscious oriented

Power: 4+/5 at all joints except B/L upper limbs (3/5).

Sensations: within normal limits

DTR: Grade I at all joints (Hyporeflexia).

Plantars: B/L downgoing

Ataxia+ (LL>UL)

Respiratory System

Air entry B/L equal but minimal crepts on left basal area.

Cardiovascular System

S1 S2 normal; no murmur/bruit.

Per Abdomen

Soft, non tender, bowel sounds+, no organomegaly/guarding.

Extremities

Warm; no pedal edema/rash/hypotonia/tremors.

Bowel and bladder intact.

On Investigations

MRI brain showed foci of demyelination (Fig. 3 & 4).

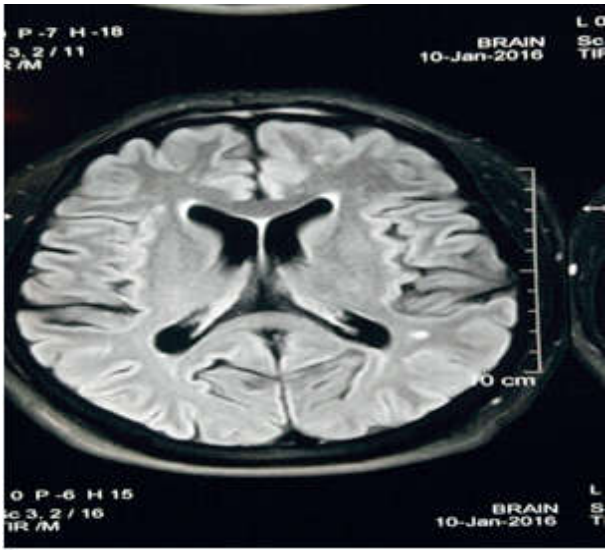

Fig. 3: MRI Brain

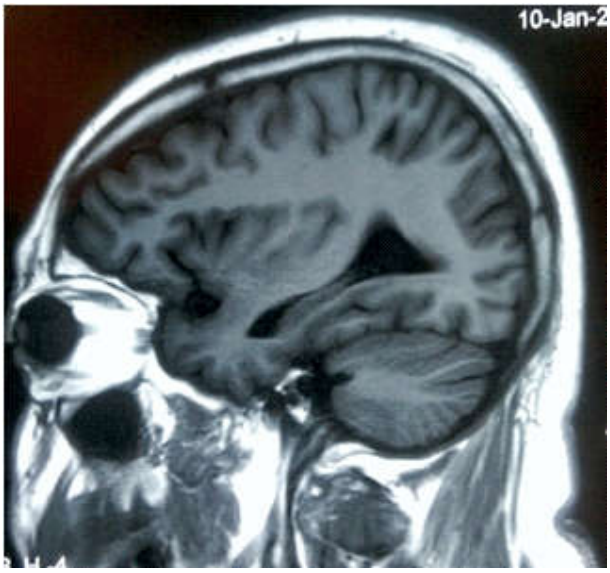

Fig. 4: MRI Brain

CSF study showed albumin-cytological dissociation with sugar 62mg/dl [Normal = 50-80mg/dl] and proteins 62.2mg/dl [20-40mg/dl].

Nerve conduction study of all limbs showed normal findings.

All other laboratory and radiological investigations showed normal findings.

However the Thyroid stimulating hormone (TSH) was raised which was 7.0 U/ml [0.3 – 5].

He was provisionally diagnosed as Polyneuritis cranialis ?GBS variant ?Miller Fisher syndrome.

He was started on IV Ig @0.4gm/kg [30gm] daily for 5 days and other conservative treatment such as Inj monocef 1gm IV BD, Inj Rabicip 20mg IV OD, Inj Emeset 8mg IV TDS, RT Feeds "150ml/2hrs, eye care with topical antibiotics and artificial tears, Facial physiotherapy with stimulation and physiotherapy of limb muscles.

Repeat MRI brain and CSF study after 7 days of presentation showed normal findings.

After a week's time, his speech improved, chemosis and ptosis improved, ataxia subsided and DTR improved.

He was discharged in stable condition after 2 weeks of hospitalization and at discharge, he had mild mild proptosis and ptosis, but other symptoms subsided and was able to take feeds orally.

Discussion

Our patient presented with ophthalmoplegia, ataxia and subnormal DTR after an episode of URI which strongly suggested Miller Fisher variant of GBS and dramatically responded to 5-days treatment with IV immunoglobulin and other conservative treatment.

The points favouring our diagnosis are URI followed by ophthalmoplegia, ataxia and hyporeflexia, LMN facial palsy, albuminocytological dissociation and dramatic improvement with IVIG therapy.

MFS is related to infectious, autoimmune and neoplastic disorders and in this case the patient developed symptoms after an URI which strongly suggested to be MFS.

The ganglioside GQ1b is found abundantly in cranial nerves (more in CN 3rd, 4th, 6th) and spinal roots and the anti-GQ1b antibodies are seen in about 90% patients of MFS. This antibody is associated with defective neuromuscular transmission. This is also

associated with GBS, PCB variant of GBS, and Bickerstaff Brainstem Encephalitis, all four of which come under the spectrum of 'Anti-GQ1b syndrome'.

We did not perform this test, firstly due to dramatic improvement of the patient and also the family members refused to go ahead with the test due to cost issues.

His TSH was raised and he was already on Thyroxine 50mcg OD and the proptosis caused by thyroid disorder would not have resolved even after thyroid treatment.

The treatment of choice for GBS or MFS is either IV Ig or plasmapheresis. Both have similar effects but the latter is more cumbersome and its availability is an issue in many centres. Many centres including ours prefer IVIG over plasmapheresis and gives good results.

IVIG is safer and easier to deliver.

Many studies show that steroids provide no benefit in treating GBS or MFS and infact one small study even showed delay in recovery after giving steroids.

Conclusion

Miller Fisher syndrome is a very rare variant of GBS and the classical triad of ophthalmoplegia, ataxia and areflexia may not be present in all cases. Patients may present only with ophthalmoplegia which is the most consistent finding.

Physicians must have a high suspicion to diagnose it clinically early, supported by investigations and to initiate appropriate therapy at the earliest.

If the treatment is delayed, patients may end up with sequellae and even death, though mortality is rare.

IVIG (or plasmapheresis) is the treatment of choice along with other supportive treatment and physiotherapy.

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