Miller Fisher Syndrome presenting as unilateral abducens nerve palsy: A case report

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Abstract

Background : Miller Fisher Syndrome usually presents with complete ophthalmoplegia, areflexia and ataxia. We present an unusual case which presented with unilateral abducens nerve palsy. Case presentation : A 51 year old female patient presented with a history of difficulty in walking and double vision for 1 week which started 2 weeks following an episode of acute gastroenteritis. She didn’t have any bladder bowel incontinence or difficulty in breathing. On examination there was left side abducens nerve palsy and bilateral significant dysmetria. Upper limb and lower limb power was 4/5 with global areflexia. There was no fatigability or sensory deficit. Higher cortical functions were intact with Glasgow Coma Scale of 15/15. Brain stem cerebro vascular accident, alcohol, toxin or drug mediated disease, myasthenia gravis, Bickerstaff encephalitis and Miller-Fisher syndrome were considered as possible differential diagnosis. There was no history of alcohol consumption or any exposure to drugs or toxins. Her Non contrast brain and MRI brain were normal. Nerve conduction study showed asymmetrical sensory motor and F wave abnormalities consistent with a Guillen-Barre Sydrome variant. Cerebro Spinal Fluid analysis showed albumino-cytological dissociation. These findings suggested the diagnosis of Miller-Fisher Syndrome. She was started on plasmaphareis. Her vital parameters, vital capacity and neurological deficit were closely monitored. With 5 cycles of plasmapharesis she made a complete neurological recovery and she was discharged on 16th day of admission. Conclusion: Miller Fisher Syndrome can present as unilateral abducens nerve palsy and early diagnosis and treatment leads to excellent functional outcome.

Background

In 1956 Miller Fisher described an acute neurologic illness characterized by total external ophthalmoplegia, severe ataxia and loss of tendon reflexes1. In 1993 association between Miller Fisher Syndrome (MFS) and serum anti-GQ1b Ig G antibody was established2,3. Unilateral abducens nerve palsy is an uncommon presentation of Miller Fisher Syndrome4.

Case Presentation

A 51 year old lady presented to us with a history of difficulty in walking and double vision for 1 week duration. She had an acute gastroenteritis episode 2 weeks prior to her symptoms. Initially she noted imbalance of her body associated with difficulty in walking. On second day she also developed double

vision specially when looking to left side. She initially consulted her family doctor who referred to her to our ward for further investigation and management. There was no history of headache or falls. She didn’t complain of worsening of symptoms with activity. Also there was no bladder bowel incontinence or difficulty in breathing. Past medical and surgical history was not significant and she was not on any regular medication. Family history was non contributory. She was a housewife and mother of 2 children. There was no occupational exposure to neuro toxins. We couldn’t elicit a history of alcohol or substance abuse.

On examination she was not febrile, not pale and anicteric. There was no neck stiffness and Kernig’s sign was negative. She was conscious, rational and oriented in time, place and person. Mini mental state examination was normal. Her Glasgow coma score was 15/15. Pupils were symmetrical and equally reacted to light. There was left sided Abducens nerve palsy but there was no ptosis and rest of the cranial nerve examination was normal. Power of all four limbs was 4 but she had global areflexia. Plantar response was flexor. Fatigability of muscle power was not elicited. There was no objective sensory deficit. Romberg’s sign was negative. Unsteady gait was noted with impaired finger-nose test and heel-knee-shin test. Her vital parameters, single breath count and vital capacity were normal. Fundoscopy didn’t show papilloedema or any other changes.

Figure 1 : Timeline of patient history

Full blood count, renal function tests, liver function tests, serum electrolytes, thyroid stimulating hormone and inflammatory markers were normal. The urine toxicology screens were negative for common substances of abuse. The electrocardiogram was normal. The serum anti-cholinesterase antibody test results were negative, which eventually refuted myasthenia gravis as a diagnosis. The chest X-ray showed no significant abnormality. The carotid Doppler did not show any sign of carotid stenosis. Computed tomography (CT) and magnetic resonance imaging (MRI) of the brain demonstrated no mass effect or evidence of an acute infarct. The serum glucose was 102 mg/dl, and the hemoglobin A1c was normal. Monospot test was negative. A lumbar puncture was done, and the cerebrospinal fluid (CSF) analysis showed protein 135 mg/dl, glucose 106 mg/dl, white blood Cells 0/ul, red blood cells 0/ul and negative gram stain and cultures. Nerve conduction study showed

asymmetrical sensory, motor and F wave abnormalities consistent with a Guillain Barre Syndrome variant. The test for anti-GQ1b antibodies was not done due to financial constraints.

With above investigation results a diagnosis of Miller Fisher Syndrome was made and patient was started on plasmapharesis. Five cycles of plasmapharesis were done on every other day basis using a femoral venous line. She acquired an infection possibly through intra venous catheter which was successfully treated with antibiotics. Her vital parameters, vital capacity and neurological deficit were monitored throughout hospital stay.

With plasmapharesis her Abducens nerve palsy started improving. After 3rd cycle ataxia also improved and she started walking. Eventually she made a complete neurological recovery with minimal residual weakness. She was discharged on 16th day of admission. During the follow up visit 2 weeks after she didn’t have any significant symptoms.

Discussion And Conclusions

Miller Fisher Syndrome is a variant of Guillen Barre syndrome which represents 1-5% of cases. Median age of onset is 40 years with male preponderance5. Respiratory tract infection is considered the commonest preceding event (76%) 5,6. However gastroenteritis especially due to *Campylobacter jejuni* has also been reported as a possible triggering infection in MFS7. This probably was the case in this patient. Mean interval between infection and onset of symptoms had been 7 days with a range of 1-30 days5. Diplopia is considered the commonest presenting complaint8. Besides the characteristic clinical triad pupillary abnormalities, blepharoptosis and facial palsy are frequent in MFS, whereas sensory loss is unusual5.

However unilateral abducens nerve palsy found in this patient is a rare presentation of MFS. Kinno et al have reported a patient with unilateral Abducens nerve palsy due to mononeuropathy associated with anti-GQ1b antibody9. Tatsumoto et al have reported 100 patients who presented with isolated Abducens nerve palsy as a regional variant of Guillen Barre syndrome10.

Our patient presented with unilateral abducens nerve palsy and ataxia. Possible differential diagnoses were cerebro vascular accident, alcohol, toxin or drug mediated disease, myasthenia gravis, Bickerstaff encephalitis and Miller-Fisher syndrome. A cerebro vascular accident was excluded by

normal Computed Tomography (CT) scan and Magnetic Resonance Imaging (MRI) scan of brain. History and toxicology screening excluded alcohol, toxin or drug mediated disease. However ocular myasthenia gravis can closely resemble MFS11. Features of myasthenia gravis include ptosis, fatigability and positive acetyl choline esterase antibody12. All these were absent in our patient thus making myasthenia gravis an unlikely explanation for this presentation. Bickerstaff encephalitis is a disease entity which closely resembles MFS clinically and immunologically. They share clinical features such as ataxia and ophthalmoplegia and immunological features such as association with anti-GQ1b antibody. However altered level of consciousness and hyperreflexia are features of Bickerstaff encephalitis which help to differentiate it from MFS13. Clinical picture, nerve conduction study and cerebro spinal fluid analysis strengthened our diagnosis. However inability to perform anti-GQ1b antibody level due to financial constraints was the main limitation in our diagnostic approach. MFS has a good prognosis and patients usually recover with no residual weakness5. Since it is a variant of Guillen barre syndrome (GBS) and has the same patho-physiological mechanism it is managed in the same way as GBS. Plasmapharesis and intra venous immunoglobilins are the treatments with research proven benefit in GBS14,15. Therefore same treatment modalities are used in MFS. However there are case reports which have shown efficacy of plasmapharesis in MFS patients16,17. In this patient plasmapharesis was preferred as cost of intra venous immune globulins was much more and patient couldn’t afford it. This patient’s excellent recovery adds more evidence to the efficacy of plasmapharesis. However case control studies with large samples are required to establish plasmapharesis as the preferred treatment for MFS.

In conclusion unilateral Abducens nerve palsy associated with ataxia is a rare manifestation of Miller Fisher Syndrome which should be picked up early to institute timely treatment thus resulting in a more favourable prognosis.

List Of Abbreviations

MFS – Miller Fisher Syndrome

GBS – Guillain Barre Syndrome

Declarations

Ethics approval and consent to participate: Informed written consent was obtained from the patient to use her data for this case report. Ethics approval not applicable as this is a case report.

Consent for publication : Informed written consent was obtained from the patient to publish data regarding her disease.

Availability of data and materials : The data used for this case report are available from the corresponding author on reasonable request.

Competing interests: The authors declare that they have no competing interests.

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Authors’ contributions: SDNDS collected data and wrote the manuscript. SS contributed in literature survey and manuscript writing. CEDS provided advice and edited the manuscript. All authors read and approved the final manuscript.

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Figures

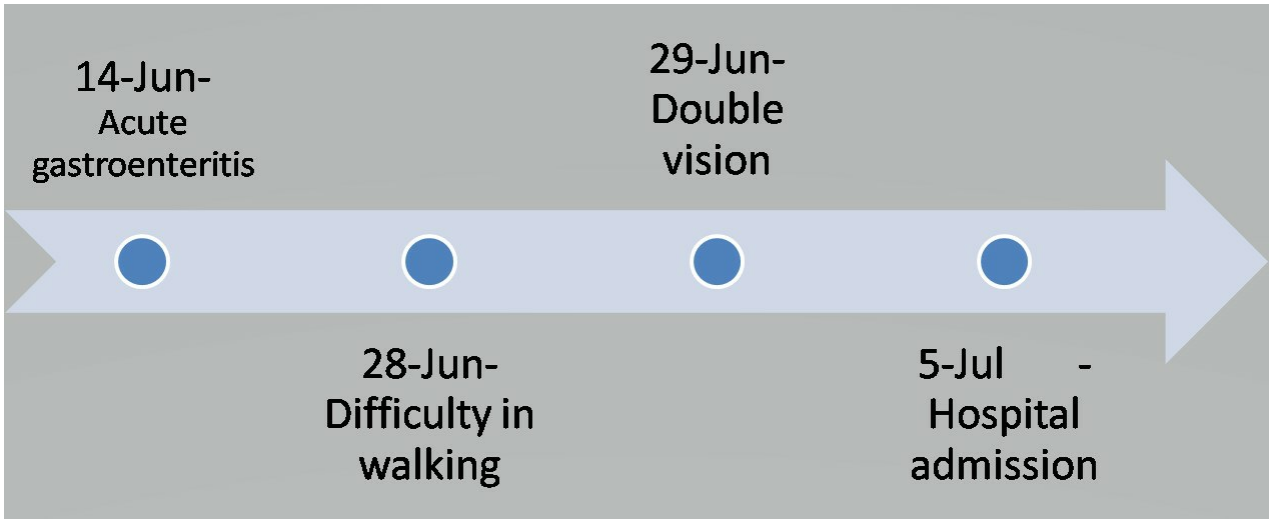

Figure 1

Timeline of patient history

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