A Case of Miller Fisher Syndrome with Unusual Features: Normal Muscle Stretch Reflexes and Facial Palsy and Dramatic Response to IVIG

Abstract
The Miller Fisher syndrome a variant of the Guillain-Barre syndrome characterized by the acute onset of oculomotor dysfunction, ataxia, and loss of deep tendon reflexes with relative sparing of strength in the extremities and trunk. Patient is presented here was a young female came with ataxia and ophthalmoplegia and unusual features of normal muscle stretch reflexes and had absent sural SNAP (Sensory Nerve Action Potential) in nerve conduction study. Patient was treated with IVIG and patient showed dramatic response to IVIG so she was symptom free in third day of treatment with IVIG.

Keywords: Guillain-Barre syndrome; Miller Fisher syndrome; IVIG

Introduction
A 34 years old female that was good till when developed with upper respiratory infection. Her problem was resolved at that time but two weeks later she developed double vision and “wobbly” gait with inability to walk and difficulty in standing. Few days later she was unable to keep open her eyes. Then she visited neurologist and received Dexamethason in outpatient setting with the diagnosis of cerebellitis after obtaining neuroimaging. But no improvement was seen in her condition after 2 weeks so she admitted in hospital Neurology ward for further evaluation. Patient had past medical history of Minor Thalassemia. Her family history for similar condition was negative. Her drug history was negative except the dexamethasone use for past 8 days. On physical examination, her blood pressure was 110/80 mm Hg, heart rate was 76/min, and respiratory rate was 14/min. Patient was alert, and oriented to time, place and person. On cranial nerve examination visual field and acuity were normal. Regarding the patient sever disability and inflammatory (Box 1). In the presented case involvement is among the most probable diagnoses. Common pathologies that involve brainstem are vascular, Neoplastic and inflammatory (Box 1). In the presented case involvement of brainstem ruled out by normal brain MRI scan. Wernick’s encephalopathy was another diagnosis that could have such presentation but our patient had no history of alcohol abuse or nutritional deficiency, so this diagnosis was not considered.

Discussion
In patient with ataxia and ophthalmoplegia brain stem involvement is among the most probable diagnoses. Common pathologies that involve brainstem are vascular, Neoplastic and inflammatory (Box 1). In the presented case involvement of brainstem ruled out by normal brain MRI scan. Wernick’s encephalopathy was another diagnosis that could have such presentation but our patient had no history of alcohol abuse or nutritional deficiency, so this diagnosis was not considered.
Acute painful ophthalmoplegia such as bilateral cavernous sinus thrombosis, Tolosa-Hunt syndrome and superior orbital fissure syndrome were not considered because they present with clearly distinctive features and MRI abnormalities. Bickerstaff brainstem encephalitis was considered because of the clinical presentation of multiple cranial nerve palsy and brisk reflexes, but it also seemed unlikely due to absence of disturbance in consciousness or limb weakness, and normal MRI and CSF study.

Miller Fisher syndrome is a type of GBS that has a classic triad of ataxia, ophthalmoplegia and areflexia [1]. Limb ataxia may resemble that seen with cerebellar disease. When above mentioned conditions ruled out Miller Fisher syndrome another condition that could present with such symptoms considered; but there was a problem with making the diagnoses of Miller Fisher syndrome which whether this syndrome can present without areflexia. In review of the published literature about Miller Fisher syndrome as in the study by Berlit et al that viewed 223 cases of this syndrome; prevalence of each symptom was investigated so that areflexia was present in 81.6% of cases and in 18.4% DTR was preserved [2]. In another study Yuki et al showed that DTRs could be normal or hyper-excitable during the entire clinical course in approximately 10% of GBS patients [3]. A case report by Yuzhu Tang et al was another example [4]. Cranial nerve involvement other than ocular motor nerves (III, IV and VI) as in our patient with facial palsy and 12th nerve involvement can be seen in association with Miller fisher syndrome. For example in the study of facial nerve in 45.7%, IX and X in 39.9%, and XII in 13% of patients were involved [2] and in study of Kuwabara et al facial palsy was present in 35% of Miller fisher syndrome cases. Normal CSF study was another feature of our case and in the same study of Kuwabara et al during the first week of illness, CSF albuminocytological dissociation occurred in 37% of cases and CSF pleocytosis was present in only 4% of the Miller fisher syndrome cases. In the second week CSF analysis showed albuminocytological dissociation in 76% of the cases. CSF pleocytosis was present in 5% of the Miller fisher syndrome cases [5]. Although CSF protein is mildly elevated, it is less so than in typical GBS.

In Miller Fisher syndrome Electrodiagnostic Studies show loss of sensory potentials, with milder axonal degeneration. Some studies have shown a demyelinating neuropathy, while others suggest purely an axonal process. But regarding past studies sensory nerves abnormality in NCS is more consistent than motor NCS [6,7], and our patient had bilateral absent sural SNAP. Mentioned studies indicate that neither normal DTR nor normal muscle power can rule out the diagnosis of Miller Fisher syndrome; even presence of sever muscle weakness in the setting of ophthalmoplegia and ataxia can make the diagnosis of Miller Fisher syndrome questionable [8].

![Box 1: Differential diagnoses for Miller Fisher syndrome, Bickerstaff’s brainstem encephalitis and pharyngeal-cervical-brachial weakness.](image)

**Conclusion**

MFS is characterized by ophthalmoplegia, ataxia and areflexia usually without limb weakness. Patients with additional involvement of the reticular formation, resulting in disturbance of consciousness, are said to have the central nervous system (CNS) subtype, known as Bickerstaff’s brainstem encephalitis [9]. Indeed some authors consider these two disorders as spectrums of unique condition. MFS can be diagnosed in patients with ophthalmoplegia, ataxia and areflexia. Patients with additional altered mental state have Bickerstaff’s brainstem encephalitis. although in some cases typical clinical picture of MFS may not be present in which ophthalmoplegia or ataxia is absent. Patients with MFS or Bickerstaff’s brainstem encephalitis who develop limb weakness can be diagnosed as having overlap with GBS. A problem that we encountered in confirming the diagnosis was that GQ1b immunoglobulin (Ig)G antibodies assay was not available in Iran [10]. But despite the above limitation we presented this patient as case of Miller Fisher syndrome based on clinical presentation and electrophysiologic findings whereas we lacking the Ab assay to confirm our diagnosis: but regardless of diagnosis dramatic response to IVIG maybe a therapeutic option for physician in similar condition in countries lacking that Ab assay for confirming diagnosis.

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