

Miller Fisher Syndrome Variant: The Incomplete Triad

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Abstract

Miller Fisher Syndrome (MFS) is a rare inflammatory peripheral neuropathy where the diagnosis is made based on the clinical triad of ophthalmoplegia, ataxia, and areflexia. It is considered a variant of Guillain-Barré Syndrome (GBS) and associated with antiGQ1b IgG positive serology. Reports on cerebellar ataxia and supranuclear gaze palsy in MFS suggested an additional involvement of the central nervous system, encompassing Bickerstaff's Brainstem Encephalitis (BBE) spectrum.

Keywords: Miller fisher syndrome • Bickerstaff brainstem encephalitis • AntiGQ1b antibodies

Introduction

We report a case of a healthy 39-year-old woman who presented with acute bilateral ophthalmoplegia, diplopia, severe headache, and ataxia following an upper respiratory tract infection a week prior. She also had hyperreflexia and other rare signs such as pupillary areflexia, nystagmus, absence of ocular somatic reflexes (vestibular-ocular reflex, optokinetic nystagmus and Bells reflex). Neuroimaging and blood results were unremarkable. A diagnosis of Miller Fisher variant, with a possible overlap of Bickerstaff Brainstem encephalitis, was made based on clinical findings and positive antiGQ1b IgG. Patient was managed conservatively and gradually improved in symptoms within one month.

Case Report

A previously well 39-year-old woman presented with complaint of double vision and wobbly gait. She had an upper respiratory tract infection a week before the symptoms started. She exhibited bilateral ophthalmoplegia, ataxia, hyperreflexia and finger dysmetria. Ophthalmic examinations revealed acuities of 6/6 for both eyes. Her pupil was fixed and dilated, not reactive to light and accommodation. She had variable ophthalmoplegia with vertical gaze restriction predominantly. Moreover, she demonstrated gaze evoked nystagmus and convergence retraction nystagmus downgaze. She also had gaze palsy on saccadic pursuit and absence of ocular somatic reflexes (vestibulo-ocular reflex, optokinetic nystagmus and Bell's reflex). Paresthesias, severe headache, weakness, and drowsiness rapidly developed within next 24 hours. Neuroimaging (CT scan and MRI) and blood results were unremarkable. CSF studies were not done as our patient refused to undergo lumbar puncture procedure. A diagnosis of Miller Fisher variant, with possible overlap of Bickerstaff Brainstem encephalitis was made based on clinical findings and positive serum antiGQ1b IgG. Patient was managed conservatively. No intravenous immunoglobulin was given but the patient gradually improved in symptoms. Her ataxia resolved but had residual ophthalmoplegia during two-month follow-up. At six months post illness she had full recovery without requiring further treatment.

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Discussion

MFS is primarily a clinical diagnosis based on the key presentation of ataxia, areflexia and ophthalmoplegia [1]. Although the clinical triad is the substantial diagnostic clue, multiple signs and symptoms beyond the triad have been reported. Our patient developed hyperreflexia which is contradictory to MFS and other rare signs such as papillary areflexia, nystagmus, absence of ocular somatic reflexes which were causing barriers to diagnosis [2,3]. Another case of atypical MFS was reported by Yeak et al. where his patient presented with acute ophthalmoparesis and hyporeflexic but without the ataxia [4]. Wang et al. reported a case with acute ophthalmoplegia with normal reflexes and without ataxia with positive antiGQ1b IgG [5]. A total of 194 cases reviewed by Odaka et al. studying the relation between MFS, Guillain-Barré Syndrome (GBS) with ophthalmoplegia, Bickerstaff's Brain Stem Encephalitis (BBE), and acute ophthalmoparesis without ataxia concluded that these three illnesses shows acute ophthalmoparesis together with positive antiGQ1b IgG are closely related therefore possibly forming a diversified findings. Jung et al. recruited 38 patients with MFS and evaluated the signs and symptoms beyond the clinical triad which resulted in more than 30% of patients can have atypical manifestations beyond the triad [6]. Chae et al. reported a case of overlapping MFS, GBS and BBE which shows involvement of both peripheral and central nervous system [7].

Conclusion

Our patient had evidence of both central (headache, hyperreflexia) as well as peripheral nerve involvement (ophthalmoplegia and absent ocular somatic reflexes) associated with positive antiGQ1b antibodies. This case represents an example of a patient who had features of both Miller Fisher Syndrome with overlap of Bickerstaff Brainstem Encephalitis suggesting that these two disorders form a continuous spectrum.

References

1. Fisher, Miller. "An Unusual Variant of Acute Idiopathic Polyneuritis (Syndrome of Ophthalmoplegia, Ataxia and Areflexia)." *New Eng J Med* 255 (1956): 57-65.
2. Hiroto Ito, Yuki Hatanaka, Yuki Fukami and Yumiko Harada, et al. "Anti-Ganglioside Complex Antibody Profiles in a Recurrent Complicated Case of GQ1b-Seronegative Miller Fisher Syndrome and Bickerstaff Brainstem Encephalitis: A Case Report." *BMC Neurol* 18 (2018): 72.
3. Odaka, Masaaki. "Anti-GQ1b IgG Antibody Syndrome: Clinical and Immunological Range." *J Neurol Neurosurg Psychiatr* 70 (2001): 50-55.
4. Justin Yeak, Mimiwati Zahari, Sujaya Singh and Nor Fadhilah Mohamad. "Co-Occurrence of Acute Ophthalmoplegia (without ataxia) and Idiopathic Intracranial Hypertension." *Euro J Ophthalmol* 29 (2019): 12.

5. Jeffrey W. Wang, Shira L. Robbins and Mark L. Moster. "One-Sided Triangle: A Case of Double Vision." *Survey Ophthalmol* 63 (2018): 880-883.
6. Jae Ho Jung, Eun Hye Oh, Jin-Hong Shin and Dae-Seong Kim, et al. "Atypical Clinical Manifestations of Miller Fisher Syndrome." *Neuro Sci* 40 (2019): 67-73.
7. Choong Sik Chae, Kyoung Min Kwon, Jung Soo Lee and Yeo Hyung Kim, et al. "A Case Report of Overlapping Miller Fisher Syndrome, Guillain-Barré Syndrome, and the Bickerstaff Brainstem Encephalitis." *Neuro* 23 (2018): 128-130.

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