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Neurology & Neurotherapy Open Access Journal

A Case of Atypical Miller Fisher Syndrome

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Case Report

Volume 2 Issue 1

Received Date: February 21, 2017

Published Date: May 27, 2017

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Abstract

Miller Fisher syndrome (MFS) is considered to be a variant of Guillain Barre Syndrome (GBS). We describe an atypical presentation of MFS with retained reflexes, minimal ophthalmoplegia, no ataxia but positive GQ1b antibodies.

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

Case Report

A 72-year-old previously healthy male presented with asymmetrical combination of arm and leg proximal and distal weakness (L>R), impaired upward gaze with retained deep tendon reflexes (DTRs). There were no autonomic, sensory or cerebellar deficits. There was a preceding history of a non-specific upper respiratory tract infection. Two weeks after resolution of infection, patient developed difficulty holding his head up and raising arms above head. This was followed by leg weakness leading to fall. He denied diplopia or in coordination. Contrast MRI of the brain showed incidental colossal lipoma and partial agenesis of the splenium of the corpus callosum. Contrast MRI of cervical spine was unremarkable.

Acetylcholine receptor antibodies, muscle enzymes, inflammatory markers (ESR, CRP) and basic laboratory tests were normal. Lumbar puncture showed no pleocytosis orcytoalbuminological dissociation. Spinal fluid paraneoplastic antibodies, cytology and flow cytometry were negative. Electro diagnostic study revealed the presence of a sensorimotor demyelinating peripheral polyneuropathy. Ganglioside (GD1a) IGG/IGM was 405 (range 0-50IV), (GQIb) was 372 (range 0-50IV) confirming the diagnosis of MFS. Treatment with intravenous immunoglobulin led to gradual improvement of weakness.

Discussion

MFS was noted to be a Guillain-Barre Syndrome (GBS) variant due the shared finding of cytoalbuminological dissociation in spinal fluid and presence of a demyelinating peripheral polyneuropathy on electro diagnostic study [1,2]. Anti-GQ1b antibodies are a reliable marker of MFS. 90% of GQ1b antibody positive patients fulfill clinical criteria for MFS. False positive rates in healthy controls, patients with GBS or other auto-immune disorders are remarkably low. Antibody titers decrease with time and symptom improvement, suggesting a direct association between Anti-GQ1b antibody levels and neuronal dysfunction. Molecular mimicry between the GQ1b antibody and surface epitopes of *Campylobacter jejuni* suggests a link between preceding infection and cellular dysfunction [3].

Few patients with classic GBS have positive anti-GQ1b antibodies but those who do, present with significant ophthalmoplegia. Anti GQ1b antibodies are also seen in patients with Bickerstaff's brainstem encephalitis (BBE),

characterized by confusion, ophthalmoplegia, cerebellar ataxia and hyperreflexia. A significant correlation has been identified between GQ1b antibodies and the presence of ataxia and significant ophthalmoplegia in patient with MFS, GBS or BBE (no such correlation is seen with weakness, changes in mental status or are flexia) [3,4]. Our case is the first to demonstrate positive GQ1b antibodies in a MFS patient without ataxia, with intact reflexes, minimal ophthalmoplegia and asymmetric limb weakness. Other atypical cases of MFS reported include a 69-year-old woman with severe ophthalmoplegia, mild gait ataxia, preserved reflexes with positive GQ1b antibodies, a 59-year-old man with ophthalmoplegia, fluctuating pupil diameter without ataxia or are flexia and positive GQ1b antibodies and a 23-year-old woman with ophthalmoplegia, ataxia, psychosis, involuntary movements with positive GQ1b and glutamate receptor antibodies[5-7].

Conclusion

Our case emphasizes the importance of multimodal diagnostic testing in atypical presentations of GBS/MFS and the need for further research into the molecular mechanisms that underlie these presentations.

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