An Atypical Case of Miller Fisher Syndrome in a Young Man: A Neurological Dilemma

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Introduction

Guillain-Barré Syndrome (GBS) is an acute and fulminant symmetrical polyradiculoneuropathy with an autoimmune pathophysiology. Miller Fisher Syndrome (MFS), a variant of GBS, usually presents as a triad of ophthalmoplegia, ataxia and areflexia. 1 MFS is rare and only presents in 1-5% of patients with GBS. However, additional neurological signs or incomplete presentations mark a diagnostic challenge for clinicians and timely management is crucial.

Case Report

A 27-year-old gentleman presented with a two day history of paraesthesia in feet, diplopia, dysarthria, dysphagia and ataxia, preceded by a flu-like illness. He was diagnosed with Asperger’s syndrome in childhood. There was no history of similar episodes in the past, no significant family history, no history of substance abuse or recent vaccination.

Clinical Findings

There was bilateral ptosis with eyes deviated down & out, sluggish pupillary reflexes (Complex Ophthalmoplegia), absent deep-tendon reflexes and cerebellar signs in the form of nystagmus, intention tremor and incoordination of limbs. The gag reflex was depressed and slight impairment of fine touch was noted in the feet. Rest of the examination was within normal limits.

Investigations

Initial blood tests for renal and liver function were found to be normal. Furthermore, other blood tests indicative for electrolyte, infection and inflammatory markers were also normal. CT and MRI scans of the head were unremarkable. Albuminocytologic dissociation was present on CSF studies. Careful monitoring of respiratory function using spirometry was done. A diagnosis of Miller Fisher Syndrome was made. This was confirmed by elevated levels of the Anti-GQ1B antibody.

<table>
<thead>
<tr>
<th>Parameters</th>
<th>Levels</th>
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<tbody>
<tr>
<td>White Cell Count</td>
<td>2 per µl</td>
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<tr>
<td>Glucose</td>
<td>3.3mmol/L</td>
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<tr>
<td>Red Blood Cells</td>
<td>95 per µl</td>
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<tr>
<td>CSF Protein</td>
<td>0.63g/L</td>
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<tr>
<td>Culture</td>
<td>No growth of organisms</td>
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Management

The patient was started on IV immunoglobulins (IVig; 400mg/kg/day for 5days) with close monitoring of vital-capacity, cardiac & hemodynamic functions. He also underwent intensive spirometry with the help of the respiratory physiotherapy team. We also sought inputs from the speech and language therapy team regarding modifications to diet to minimise risk of aspiration. Patient was reviewed by the neurologist who agreed with the diagnosis and management plan. The patient recovered completely after three weeks and is currently doing well in follow-up clinic visits.

Discussion

GBS typically involves an ascending motor paralysis with or without sensory disturbances. 70% cases occur 1-3 weeks after an acute infectious process, usually respiratory or gastrointestinal. Diagnosis is based on clinical findings and CSF may reveal albuminocytologic dissociation.

In Miller Fisher variant (MFS), a triad of ophthalmoplegia, ataxia and areflexia is seen. 2 MFS is usually diagnosed clinically but is often challenging due to overlapping clinical signs. In our case, presence of bulbar weakness suggests overlap with pharyngeal-cervical-brachial weakness variant of GBS. 3 The presence of Anti-GQ1B antibody(85-90% of cases) is strongly associated with oculomotor nerve involvement.

Treatment should be initiated urgently with either IV immunoglobulins or plasmapheresis. Spirometry should be performed regularly to recognize need for respiratory support and transfer to the intensive care unit. Pain management usually involves neuropathic agents. Multidisciplinary team involvement with speech and language therapists and respiratory physiotherapists should be encouraged. 85% patients achieve a full functional recovery within several months to a year. 1 The physicians should be aware of atypical neurological presentations for better patient management.

References