Miller Fisher syndrome in a Chinese octogenarian presenting with dizziness

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ABSTRACT
We report a case of Miller Fisher syndrome in an 80-year-old woman who presented with a 3-day history of worsening dizziness associated with vertigo. The patient had an episode of upper respiratory tract infection 1 week earlier. She later developed diplopia in bilateral lateral gaze and drooping of eyelids. She was treated conservatively without the use of intravenous immunoglobulin or plasmapheresis. Her symptoms improved gradually after 2 weeks of hospitalisation. At the 10-week follow-up, she had regained all the reflexes and full ocular movements with no residual ptosis.

Key words: Aged, 80 and over; Miller Fisher syndrome

INTRODUCTION
Miller Fisher syndrome is characterised by acute onset of oculomotor dysfunction, ataxia, and loss of deep tendon reflexes with relative sparing of strength in the extremities and trunk. The ataxia is produced by peripheral sensory nerve dysfunction and not cerebellar injury. Facial weakness and sensory loss may also occur. The process is mediated by autoantibodies directed against a component of myelin found in peripheral nerves.

CASE REPORT
In April 2012, an 80-year-old woman presented to the accident and emergency department with a 3-day history of worsening dizziness associated with vertigo. The patient had had an episode of upper respiratory tract infection 1 week earlier. She was diagnosed with vestibular neuronitis by her general practitioner and prescribed symptomatic treatment. However, her symptoms persisted, and she developed diplopia in bilateral lateral gaze and drooping of eyelids. Apart from mild distal upper limb numbness, she had no headache, limb weakness, or swallowing, breathing or sphincter disturbances.

She was transferred to a geriatric unit. On examination, bilateral partial ptosis and impaired bilateral eye movements (especially in elevation, abduction, and adduction) were noted (FIGURE). Pupils were unremarkable, and examinations for cranial nerves, limb power, and sensory were normal. There were no pyramidal signs and the plantar reflex was down going. Cerebellar test, head impulse test, and Hallpike manoeuvres were negative. However, the patient was areflexic throughout and had an ataxic gait with a positive Romberg sign.

Magnetic resonance imaging of the head did not reveal any abnormal signal or mass lesion in the brainstem. The cavernous sinus appeared intact with satisfactory enhancement. The superior ophthalmic veins were not dilated. There was no restricted diffusion and the ventricles were not dilated. Lumbar puncture and cerebral spinal fluid (CSF) analysis revealed a protein level of 1.17 g/L (elevated) and a glucose level of 4.0 mmol/L, and
no cells were detected. Bacterial, viral, and fungal cultures of the CSF were all negative, and the CSF test for syphilis was non-reactive. Nerve conduction study demonstrated reduced sensory nerve action potentials and absent H reflexes over the limbs, indicating generalised peripheral neuropathy and demyelination. Anti-acetylcholine receptor antibodies were within the normal range. Blood tests for anti-GQ1b antibodies, however, were strongly positive, which was indicative of Miller Fisher syndrome. Tumour markers and chest radiographs did not suggest any underlying occult malignancy.

The patient was treated conservatively without the use of intravenous immunoglobulin or plasmapheresis. Her symptoms improved gradually after 2 weeks of hospitalisation. Ptosis and extraocular movement improved, especially in the lower vertical gaze and left lateral gaze (Figure). At the 10-week follow-up, she had regained all the reflexes and full ocular movements with no residual ptosis.

**DISCUSSION**

Miller Fisher syndrome accounts for approximately 5% of patients with Guillain-Barré syndrome worldwide and up to 9% to 18% of those in East Asia. It is a disease of middle age, and the median age at presentation is about 41 years. It is rarely reported in patients older than 80 years and may be mistaken for stroke. In an 11-year hospital-based review of Miller Fisher syndrome in Taiwan, the oldest patient was aged 78 years. The syndrome affects more males than females, with a ratio of 2:1.

Ophthalmoplegia, ataxia, and areflexia are classic symptoms of the disease. The common initial symptom is diplopia owing to bilateral extraocular muscle weakness, with horizontal, followed by vertical, gaze palsies. In the Taiwan series, dizziness was the commonest presenting symptom, accounting for 54.5% of the initial symptoms. Other presenting symptoms include ptosis, blurred vision, nausea and vomiting, headache, paresthesias, as well as facial and limb weakness. Most patients had a recent history of infection, and the average time from infection to neurological manifestation was 7 days.

More than 80% of patients with Miller Fisher syndrome have in their serum anti-GQ1b antibodies. The anti-GQ1b antibody activities are associated with the disease severity. Dense concentration of GQ1b ganglioside in the oculomotor, trochlear, and abducens nerves may explain the association between anti-GQ1b antibodies and ophthalmoplegia. However, GQ1b antibodies may also be found in patients with Bickerstaff brainstem encephalitis, which is a closely related condition where alterations of consciousness or long tract signs are present in addition to ophthalmoplegia and ataxia. Both conditions form a continuous spectrum with variable central and peripheral nervous system involvement. In addition, Miller Fisher syndrome is associated with a normal neuroimaging study, albuminocytological dissociation, reduced sensory nerve action potentials, and absent H reflexes.

Although the use of immunoglobulin and plasmapheresis may hasten recovery, there is no...
difference in the outcome of patients who receive immunomodulatory treatment and those who do not.\textsuperscript{1,6} The prognosis of Miller Fisher syndrome is usually good, with recovery after a mean of 10 weeks.\textsuperscript{1,5}

REFERENCES