Miller Fisher Syndrome – An Uncommon Clinical Presentation

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Abstract

Miller Fisher syndrome is an uncommon disease and it is a variant of Guillain-Barre syndrome. Miller Fisher syndrome also has rarer variants. Combined features of classic Guillain-Barre syndrome and Miller Fisher syndrome are uncommon. Here we are reporting a case of Miller Fisher variant with Guillain-Barre syndrome overlap in which ataxia, areflexia, oculomotor disturbance and limb weakness occurred within few days.

INTRODUCTION

Miller Fisher syndrome (MFS) is an uncommon variant of Guillain-Barre syndrome (GBS). MFS also has rarer variants. A case of Miller Fisher variant with GBS overlap having ataxia, areflexia, ophthalmoplegia and weakness is reported here.

CASE REPORT

A 50 years old male military person presented with predominant unsteadiness of gait, followed by development of partial weakness of lower limbs within five days. Initially he noticed imbalance and tendency to fall on either side during walking. He had tingling dysesthesia of all four limbs from the beginning. Later he noticed weakness in the lower limbs. Gradually the patient became bedridden at about 5th day of his illness. His imbalance was disproportionate to weakness. He had no history of fever, loose motion and upper respiratory tract infection in last one month. He had no recent history of vaccination or any surgery. The patient had no history of abnormal behaviour, headache, seizures, diplopa, dimness of vision, oscillopsia, aural symptoms (like tinnitus, deafness or ear discharge), vertigo, dysarthria and dysphagia. Bladder and bowel functions were normal. He had no history of drug abuse, alcohol addiction and extramarital sexual exposure. His family history was unremarkable.

On examination, the patient was conscious and co-operative. Pallor, cyanosis, jaundice, clubbing and lymphadenopathies were absent. His blood pressure was 126/78 mmHg with no orthostatic fall. His pulse was 82/minute and regular. Respiratory rate was 18/minute. On neurological examination, his higher mental function including speech and language was normal. He was oriented to time, place and person. Cranium and spine examinations were normal. On cranial nerve examination, eye movements were restricted in both lateral and upward direction. Pupillary response was sluggish bilaterally. Nystagmus was absent. Optic fundus was normal. Patient had mild bilateral lower motor type of facial weakness more on the left side. III, VI and VII cranial nerves were involved but other cranial nerves were normal. On motor system examination, tone of the muscles of the lower limbs was reduced. Power of lower limbs was 4/5 on both sides. Power of upper limbs was normal. Neither there was wasting of muscles nor any involuntary movement. Deep tendon jerks were absent in all four limbs. Plantar was bilaterally flexor. Abdominal reflex was preserved. On sensory examination, pain and temperature sensations were preserved. Joint position and vibration senses were impaired in lower limbs. Romberg's test was positive. He had wide based ataxic gait with tendency to fall on either side. Tandem gait was grossly impaired. His finger-nose test was mildly impaired but heel-shin test was grossly affected. There was no dysdiadochokinesia and rebound phenomenon. Examinations of other systems were normal.

His complete blood count was normal. Blood sugar, urea, creatinine, liver function tests and serum electrolytes revealed no abnormality. Hepatitis B surface antigen and HIV (ELISA) were negative. ANF was negative. CSF cell count was 10/cmm and protein 450 mg%. ECG and chest X-ray were normal. NCV study showed demyelinating motor sensory polyneuropathy. CT scan of brain was normal.

As the patient had ataxia, areflexia, mild ophthalmoparesis and weakness of lower limbs, clinically he was diagnosed to have Miller Fisher variant with GBS overlap. Patient was managed conservatively. No intravenous immunoglobulin was given but the patient gradually improved in symptoms including power, ataxia, ophthalmoparesis and
CNS involvement. The ataxia in MFS is due to peripheral spindles and kinesthetic information for joint receptors. MFS shows a higher level of sensory and proprioceptive input from muscle of MFS and GB. MFS shows a disorder akin to GB. Many studies showed the similarities in the pathogenesis of MFS and GB. MFS shows a higher level of sensory and CNS involvement. The ataxia in MFS is due to peripheral mismatch between proprioceptive input from muscle spindles and kinesthetic information for joint receptors.

Berlit et al. viewed 223 cases of MFS. The first symptom was diplopia (38.6%) or ataxia (20.6%). Areflexia was present in 81.6% of cases. The cranial nerves other than III were involved in 127 cases (56.9%); cranial nerves VII (45.7%), IX and X (39.9%), and XII (13%) were involved. In 53 cases, tetraparesis occurred. Elevated CSF protein was present in 134 patients (64.4%). CSF findings were normal in 56 patients, and 18 patients had mild pleocytosis. The prognosis of MFS was good. Recovery occurred after a mean time period of 10.1 weeks. Residual symptoms were present in 74 cases (33.2%), recurrence of MFS was reported in 7 patients, and 8 patients died.

Fross et al. reported in 10 patients with typical MFS electrophysiological abnormalities characteristic of axonal neuropathy or neuronopathy with predominant sensory nerve changes in the limbs and motor damage in the cranial nerves.

Uncini et al. reported an MFS patient with antibodies to GQ1b who developed limb weakness. Serial motor conduction velocities showed a marked reduction in the amplitudes of distal compound muscle action potential (CMAP), reaching low at 2 to 3 weeks, followed by a dramatic improvement in the 5th week. Motor conduction velocities were in the normal range, with the distal motor latencies changing only slightly without the conduction block between the root and the distal nerves. These data might suggest an axonal neuropathy or a distal demyelinating conduction block.

Variant forms of MFS—Different variants of MFS are present with a common tie of the GQ1b antibody. Some cases have only one or two symptoms out of the triad. Some patients have combined features of GBS and MFS, in which the oculomotor disturbance and limb weakness occur within a few days of one another (GBS overlap variant: ophthalmoplegia, weakness, areflexia and ataxia). Recurrent MFS is also reported rarely. There are also lower cranial nerve variants of GBS and atypical MFS. It was reported that the oculomotor nerves were involved early in 7 cases of the ophthalmoplegic variants of GBS, and the cranial nerves IX, X and XI were involved early in 9 cases of a lower cranial nerve variant of GBS.

Ataxic neuropathies - Kusunoki et al. found 5 patients with a variant form characterized by ataxia but no ophthalmoplegia in 149 patients who had the IgG anti-GQ1b antibody without profound weakness. These cases could fit with autoimmune ataxic neuropathy, having pathological lesions mainly in the dorsal root ganglion. GD1b antibody plays the key pathogenic role in autoimmune ataxic neuropathies. Because of the existence of cross-reactivity between GQ1b and GD1b, it is thought that the pathogenesis of this form is similar to that of MFS.

Mori et al. reviewed the clinical features and outcome of MFS for 50 consecutive patients including 28 patients who received no immunotherapy. Besides the characteristic clinical triad (ophthalmoplegia, ataxia, and areflexia), pupillary abnormalities, blepharoptosis, and facial palsy are frequent in MFS, whereas sensory loss is unusual despite the presence of profound ataxia. Patients with MFS usually had good recovery and no residual deficits.

In our case, ataxia and areflexia were predominant and initial signs. Eye signs were minimal with mild lateral and upward eye movement restriction due to involvement of
Ill and VI cranial nerves. The patient had sluggish pupillary responses. He also had mild and asymmetric (left>right) facial nerve involvement. The patient developed partial weakness of both lower limbs in course of time. Usually MFS is associated with axonopathy affecting predominantly the large diameter sensory fibers with only mild motor conduction abnormalities. F-wave latencies are usually normal in MFS. In contrast our case revealed demyelinating motor sensory polyneuropathy with normal F latencies. Anti- GQ1b antibody is one of the key factors in the pathogenesis of MFS, especially for ophthalmoplegia, and it is a useful marker in diagnosis of MFS but it was not done in our case because of unavailability of the test. Clinically our case is a Miller Fisher variant with GBS overlap with minimal ophthalmoparesis but with lower limbs motor weakness.

REFERENCES