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Case Report of Miller Fisher Syndrome a Variant of Guillain-Barre´ Syndrome

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Abstract: ***Miller Fisher syndrome (MFS), was first recognized by James Collier in 1932. MFS was named after Charles Miller Fisher who reported 3 cases of acute neurologic illnesses characterized by total external ophthalmoplegia, severe ataxia, and loss of tendon reflexes in 1956 as a limited variant of Guillain-Barré syndrome.MFS is mostly an acute, self-limiting condition. It is strongly associated with anti-GQ1b (antibody to ganglioside). MFS can be associated with infectious, autoimmune, and neoplastic disorders. Oculomotor dysfunction is partially explained by the tissue ganglioside concentration and distribution and the attraction of antibody-stimulating complement activation. Here, we present a case of a 25year old female patient who admitted with complaints of double vision that progressed to bilateral ophthalmoplegia, and ataxia with preceding gastro-intestinal infection. Subsequently she developed weakness of the all four extremities with proximal muscles affected more than distal muscles and areflexia. She was thoroughly investigated and is believed to have an axonal neuropathy in the form of Miller Fisher Syndrome (MFS) variant of GBS. She was initially treated with intravenous immunoglobulin, which significantly improved her symptoms.***

Keywords: Miller Fisher Syndrome, Guillain-Barré syndrome, ataxia, areflexia, ophthalmoplegia, antibody to ganglioside, intravenous immunoglobulins.

1. Introduction

Miller Fisher syndrome (MFS), was first recognized by James Collier in 1932. MFS was named after Charles Miller Fisher who reported 3 cases of acute neurologic illnesses characterized by total external ophthalmoplegia, severe ataxia, and loss of tendon reflexes in 1956 as a limited variant of Guillain-Barré syndrome [(1)]. MFS is mostly an acute, self-limiting condition. It is strongly associated with anti-GQ1b (antibody to ganglioside) [(2)].MFS can be associated with infectious, autoimmune, and neoplasticdisorders. Campylobacter jejuni is the most commonly identified agent causing antecedent infection in patients with acute MFS and those with GBS and related disorders, followed by Haemophilusinfluenzae. However, in most patients, no definite infectious association has been detected [(3)]. Several lines of evidence support molecular mimicry between ganglioside GQ1b and a *Campylobacterjejuni* lipopolysaccharide [(4)].

The anti-GQ1b test has been shown to be positive in approximately 85% of cases of MFS [(2)]. The rapid onset of ophthalmoplegia can help distinguish MFS from conditions that progress chronically, such as mitochondrial myopathies, oculopharyngeal dystrophy, myotonic dystrophy, thyroid eye disease, and some cases of ocular myasthenia gravis. The differential diagnosis of Miller Fisher syndrome includes other polyneuropathies, brainstem lesions, neuromuscular junction disorders, and cavernous sinus or orbital lesions [(5)]. The factors that determine which patients develop FS and which develop the related disorders remain unknown. According to Masahiro Mori et al study, ataxia and ophthalmoplegiadisappeared a median of 1 and 3 months after onset, and all patients had no or very little disability 6 months after onset. The natural course of MFS is characterized by good recovery [(6)].

2. Case Report

A 25 year old female patient teacher by profession brought to the hospital with two days history of sudden onset double vision for the near vision but not for far vision and her mother found that her eyes were fixed in the midline and that there was no absolute movement. Later in the day, while she was going to the bathroom, she felt slight transient dizziness, which gradually progressed in severity and she even felt dizziness in sitting position. Next day morning she was unable to stand from sitting position and complained pain in the bilateral shoulders and she was unable comb her hair. There is no history of fever, fall/ trauma but she recalled that she had 3 episodes vomiting and 5 episodes of loose stools one week back which resolved on its own over 2 days. No history thyroid disorder, congenital abnormalities, no previous history of similar complaints, no history of alcohol consumption or drug abuse.

Physical examination

On examination temperature was normal, all the peripheral pulses were felt and normal, blood pressure was 116/76 mm of Hg. There was complete ophthalmoplegia, eyes were fixed in the normal forward gaze, convergence and accommodation were absent, blinking was normal, bell's phenomenon was absent, pupils were equal and measured 4mm and both the pupils were sluggishly reacting to the light, corneal and conjunctival reflexes were present. Visual acuity and fundus examination were normal. No facial muscle asymmetry was noticed. Hearing was normal. Speaking and swallowing normal. Sternocleidomastoid muscle power was normal. Tongue protruded normally. Pain, temperature, and vibratory sensations were normal. Balance and co-ordination was not performed because patient was unable to stand. All the deep tendon reflexes

were absent, planters were flexors. Power in the proximal muscles was 2+, were in distal muscles it was 4+.

Investigations

On admission, routine blood tests, including complete blood count, biochemical and microbiological investigations were normal, ECG and chest radiograph, were normal. The urine toxicology screens were negative for common substances of abuse.The rapid plasma reagin for syphilis was negative. MRI scan of the brain and cervical spine were within normal limits. Electrophysiological study is suggestive of early, mixed (demyelinating and axonal) neuropathy. A lumbar puncture, performed 10 days after symptoms onset, which revealed cyto-albuminologic dissociation (protein 200 mg/dl, 4 lymphocytes per cubic millimetre), sugar 55mg/dl with negative gram stain and cultures. Acetylcholine receptor antibodies was negative and antiganglioside antibodies was reported positive after 2 weeks.

Treatment

Because of rapidly progressing symptoms she was initially admitted in the intensive care unit, then intravenous immunoglobulins were administered for 5 days for a total dose of 2g/kg body weight, despite 3 days of treatment, she developed severe global limb weakness. Significant clinical improvement was noticed after 8 days of treatment. Physical therapy specialists worked with the patient on a daily basis with daily range of motion exercises to prevent contractures. She was able to ambulate with some assistance on 12th day. She was discharged on the 18th day of her hospitalization to a subacute rehabilitation facility. Two months after her initial presentation, her power was 4 to 4+/5 in the upper limbs (4 on the left and 4+ on the right), with some similar pattern in the lower limbs but absent knee jerks and ankle jerks and flexor planters.

3. Discussion

Overview: This case had the clinical features of ataxia, areflexia and ophthalmoplegia with mixed neuropathy, cyto-albuminologic dissociation on CSF study and normal MRI, strongly suggesting Miller Fisher syndrome. In this case, the antiganglioside antibody testing including anti-GQ1b was positive.

The median age at the time of MFS onset was 40 years, predominating in spring [(6)]. Respiratory symptoms were observed predominantly within 1 month before MFS onset [(6)].MFS onset is typically acute, beginning with neurologic symptoms approximately 8–10 days (range of 1–30), following the antecedent illness[(6)].The disease then progresses until a clinical nadir is reached approximately a week (range of 2–21) after the initial neurologic symptoms. The most common presenting symptom of MFS is diplopia, which arises because of the acute onset of external ophthalmoplegia[(7)]. Pupillary abnormalities can include mydriasis, anisocoria, and a sluggish direct response to light [(7)]. Facial nerve involvement, which occurs in approximately 30% of patients, may result in orbicularis oculi weakness and, consequently, lagophthalmos[(6)]. Ptosis, if present, is often partial and can be unilateral or bilateral [(4)]. Other lid abnormalities reported include lid retraction, upper lid jerks, and lid nystagmus[(8)]. findings from previous study showed that the central components (e.g., supranuclear eye movement disorder) might be associated, but only rarely, indicative that the brainstem is not the main lesion site in MFS [(6)]. Gender, age, evidence of a prior infection, disability at the peak of illness, and latency to peak have no effect on the disease's outcome [(6)].

Although both MFS and ischemic event are acute, ataxic patients with cerebellar involvement show lateralization of ataxia while MSF patients typically lack lateralization of ataxia. It helps to differentiate MFS from most cerebellar lesions [(09)] . The rapid onset of eye symptoms in MFS can differentiate it from chronic diseases such as myasthenia gravis (MG), thyroid eye diseases (TEDs), and myotonic dystrophy (MD). Ptosis in myasthenia gravis worsen as the day progresses, where as MFS ophthalmoplegia progressively worsens until few days.In this patient there is no underlying thyroid disease or other clinical signs of TEDs. Features like myotonia, muscle wasting, testicular atrophy, cataracts, frontal balding, arrhythmia seen in myotonic dystrophy helps to differentiate from MFS.

MFS can also present with features that overlap Bickerstaff brainstem encephalitis (BBE), a syndrome characterized by ophthalmoplegia, ataxia, impaired consciousness, and hyperreflexia[(10)], and with features typical of GBS, in which limb weakness is the most prominent finding, sometimes accompanied by sensory loss and non-ocular motor cranial neuropathies [(11)]. These disorders can all develop after an antecedent infection and share a characteristic elevation of cerebrospinal fluid (CSF) protein.

Areflexia can be present in certain neurological conditions damaging lower motor neurons in the spinal cord or peripheral nerves. Diabetes and vitamin B12 deficiencies can cause peripheral neuropathies and subsequently can present as areflexia. Areflexia can also occur in the subacute phase of spinal shock. The anterior horn cell destruction viewed in amyotrophic lateral sclerosis (ALS) and polio can present as areflexia. Certain metabolic abnormalities like alcoholism hypomagnesemia, can also present with areflexia.

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