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A CASE REPORT OF MILLER FISHER VARIANT OF GUILLAIN BARRE SYNDROME

General Medicine

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ABSTRACT

Guillain-Barré Syndrome (GBS) is a rare autoimmune disorder characterized by acute onset, rapidly progressive symmetrical weakness and areflexia. Miller Fisher variant is a rare subtype of GBS presenting with ophthalmoplegia, ataxia, and areflexia, often associated with antibodies against gangliosides. This case report describes a patient presenting with sudden onset double vision, bilateral ptosis and ataxia, subsequently diagnosed as Miller Fisher variant of GBS. The patient was managed with intravenous immunoglobulin (IVIG) and steroids, showing clinical improvement over hospitalization. This case highlights the importance of early recognition and multidisciplinary management of GBS variants.

KEYWORDS

INTRODUCTION

Guillain-Barré Syndrome (GBS) is an acute immune-mediated polyradiculoneuropathy characterized by symmetrical weakness and areflexia. Miller Fisher syndrome (MFS) is a rare variant of GBS, presenting with ophthalmoplegia, ataxia, and areflexia, typically without significant limb weakness(1). It is associated with antibodies against gangliosides, particularly anti-GQ1b antibodies (2). Here, we report a case of MFS presenting with classical features and discuss its clinical course, investigations, and management.

Case Presentation

A 30-year-old male, X-ray technician by occupation previously asymptomatic, presented with sudden onset diplopia and drooping of both eyelids, and difficulty in eye movements since 2 days.Diplopia was sudden in onset, painless and not associated with diurnal variation. These symptoms were followed by involuntary movements of both upper limbs on reaching the objects which were associated with tingling sensations. Subsequently, he developed difficulty in getting up from the ground and noticed a change in voice with increased nasal twang. He reported a history of dry cough and sore throat for the past 8 days but denied fever or gastrointestinal symptoms. There were no associated complaints of headache, blurring of vision,loss of consciousness, involuntary passage of urine or stools, constipation, drooling of saliva or water from angle of mouth, difficulty in hearing, difficulty in chewing, involuntary tongue movements ,back pain, band like sensation, stiffness, spasms or cramps in limbs. There is no history of drug or toxin ingestion. Patient had no significant past medical or surgical history and no history of similar illness in the past.

On examination, the patient was afebrile and normotensive with no tachycardia and maintaining 98% saturation on room air without any respiratory distress. Neurological examination revealed bilateral ptosis, complete ophthalmoplegia in all directions, pupils bilaterally dilated and not reacting to light, loss of nasolabial folds and absence of forehead wrinkles. Motor examination showed normal tone and power graded as 4+ across all joints in both upper and lower limbs. There was evidence of limb ataxia with positive finger nose finger test and heel to shin test. Sensory examination revealed no abnormality and there were no involuntary movements at rest. Deep tendon and abdominal reflexes were absent and plantars were bilaterally mute . Cranial nerve examination showed absence of taste sensation over the whole tongue with absence of gag reflex.Hearing & olfaction was intact and nystagmus could not be assessed due to ophthalmoplegia. There were no abnormal movements of tongue or deviation to any side.

Routine blood investigations including complete blood count, renal and liver function tests, electrolytes, and blood glucose were within normal limits. Cerebrospinal fluid analysis showed total counts of 5 with lymphocyte predominance and albuminocytological dissociation. MRI brain was done which was found to be normal. Nerve conduction studies (NCV) revealed generalized, severe demyelinating with secondary axonal, pure sensory polyneuropathy affecting both upper limbs and severe, pure axonal, facial neuropathy bilaterally. Serum ganglioside antibodies panel (Antibodies against GM1, GM2, GM3,GD1a, GD1b, GT1b and GQ1b) was found to be negative.

TESTS	VALUES	Reference values
Hemoglobin (g/dL)	16.3	12 - 15
White blood cells (mm3)	7.68	4 - 10
Platelets (mm3)	277	150 - 410
Urea (mg/dL)	21.6	19.3 - 49.2
Creatinine (mg/dL)	0.77	0.5 - 1.3
Sodium (meq/L)	144	132 - 146
Potassium (meq/L)	4.37	3.5 - 5.5
SGPT (IU/L)	20	10 - 49
SGOT (IU/L)	21	0 - 34
Total bilirubin (mg/dL)	0.13	0.3 - 1.2
Total protein (g/dL)	7.1	5.7 - 8.2
Serum albumin (g/dL)	4.2	3.2 - 4.8
ESR (mm)	8	0 - 13
TSH (mcIU/mL)	1.86	0.35 - 5.5
Creatinine Phosphokinase (U/L)	75	22 - 198
Vitamin B12 (pg/mL)	470	200 - 1100
HIV (rapid test), HBsAg, anti-HCV antibodies	Non reactive	-
Urine routine examination	Normal	-

SGPT: serum glutamic-pyruvic transaminase
SGOT: serum glutamic-oxaloacetic transaminase
ESR : Erythrocyte sedimentation rate
TSH : Thyroid stimulating hormone

CSF EXAMINATION

Appearance - Clear
Total cells - 5 (Lymphocytes - 100%)
ADA - 1.7 (Normal <10)
Sugar - 68 mg/dL (40-80 mg/dL)
Protein- 19 mg/dL (15-60 mg/dL)
CSF culture - Negative

Based on clinical findings and investigations,diagnosis of Guillain-Barré Syndrome - Miller Fisher variant was made. The patient was managed with intravenous immunoglobulin (IVIG) for 5 days, followed by a course of injectable steroids for 3 days due to minimal improvement post immunoglobulin therapy.Treatment of complications like aspiration pneumonia was also given along with chest and limb physiotherapy. Over the course of hospitalization, patient showed gradual improvement in symptoms in the form of reduced intention tremors, return of abdominal reflexes, and subtle improvement in eyelid movements. Patients' SBC (Single Breath

Count) improved from 26 To 40 during the hospital stay. After 18 days of hospitalization he was discharged in vitally stable condition and was advised regular follow-up for monitoring of the symptoms and rehabilitation. Subsequent follow-up visits demonstrated continued improvement in neurological deficits, albeit with persistent mild residual symptoms.

DISCUSSION

Miller Fisher syndrome (MFS) is a rare variant of Guillain-Barré Syndrome (GBS) characterized by a distinctive clinical presentation and specific immunological features. Named after the neurologist Charles Miller Fisher who first described it in 1956, MFS constitutes approximately 5% of all GBS cases, making it a relatively uncommon but notable subset of this autoimmune neuropathy.The hallmark clinical triad of MFS includes: Ophthalmoplegia: Weakness or paralysis of the muscles responsible for eye movement, leading to diplopia. Ataxia: Loss of coordination of voluntary movements, affecting gait and balance and Areflexia: Absence of deep tendon reflexes, such as the knee-jerk reflex (3).

These symptoms typically develop rapidly over a period of days to weeks, often following a preceding infection, most commonly respiratory or gastrointestinal in nature (4). The onset of symptoms can be acute and progress to involve other cranial nerves, resulting in additional manifestations such as facial weakness or bulbar symptoms (difficulty swallowing and speaking).

Diagnosing MFS relies on a combination of clinical findings and supportive diagnostic tests. CSF analysis typically shows elevated protein levels without an increase in white blood cells, reflecting the albuminocytological dissociation commonly seen in GBS variants(5). Electrodiagnostic Studies like Nerve conduction studies and electromyography may reveal findings consistent with demyelination or axonal injury. Serological testing includes detection of anti-GQ1b antibodies in the serum which is present in 90% of patients with Miller Fisher syndrome and supports the diagnosis, although not all patients will test positive (6) (7).

When considering a differential diagnosis for Miller Fisher Syndrome, several conditions may present with similar features.Bickerstaff Brainstem Encephalitis is another variant of GBS that shares some clinical features with Miller Fisher Syndrome, including ophthalmoplegia and ataxia (8). It is distinguished by additional signs of brainstem involvement, such as altered consciousness and long tract signs. Acute disseminated encephalomyelitis (ADEM) is characterized by widespread inflammation in the brain and spinal cord, leading to neurological symptoms that can include ataxia and cranial nerve deficits. Unlike MFS, ADEM typically presents with multifocal neurological deficits and may have other associated symptoms (9).Botulism can present with cranial nerve palsies, including ophthalmoplegia, due to the neurotoxin's effect on neuromuscular transmission. However, botulism lacks the characteristic areflexia seen in Miller Fisher Syndrome and other features like ptosis and descending paralysis may be more prominent (10). Myasthenia Gravis is also an important differential of Miller Fisher syndrome, characterized by muscle weakness and fatigue, often involving the ocular muscles (ptosis and diplopia). While ophthalmoplegia can occur in both conditions, myasthenia gravis is associated with fluctuating muscle weakness rather than the acute onset seen in MFS (11).

The treatment of MFS is similar to that of GBS. Treatment modalities include plasmapheresis and intravenous immunoglobulin (IVIG). IVIG is often considered as the first-line treatment for Miller Fisher Syndrome. The usual dose is 2 g/kg given over 5 days.Plasma exchange is an alternative treatment option for MFS, particularly for patients who do not respond to or cannot tolerate IVIG (12). The role of corticosteroids in treating MFS is controversial. Some studies suggest they may not be effective or could potentially worsen outcomes. Therefore, corticosteroids are generally not recommended as a first-line treatment but may be considered in certain cases under specialist guidance (13). After the acute phase of Miller Fisher Syndrome resolves, rehabilitation therapy (physical therapy, occupational therapy) may be beneficial to aid in recovery of motor function and coordination. Most patients show a complete resolution of all symptoms but certain symptoms may persist in a few cases.

CONCLUSION

This case report illustrates a classic presentation of MFS in a previously healthy 30-year-old male who developed acute onset diplopia, bilateral ptosis, and ataxia following a mild upper respiratory tract infection. Diagnosis was confirmed on the basis of clinical findings, cerebrospinal fluid analysis, and nerve conduction studies consistent with demyelinating and axonal neuropathy however, our patient had absence of GQ1b antibodies in CSF. Management included prompt initiation of intravenous immunoglobulin (IVIG) and subsequent steroid therapy, resulting in gradual clinical improvement over the hospitalization period. This case underscores the significance of early recognition and appropriate management of MFS. Continued monitoring and rehabilitation are crucial for patients, given the potential for residual neurological deficits despite overall improvement. Further research and clinical experience are necessary to refine treatment strategies and enhance long-term prognosis in rare autoimmune neuropathies like MFS.

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