GBS in Neuro ophthalmology : A jeopardize clinical scenario

Sheikh Mohammad Rashedul Haque[1]*, Md. Sibgatullah[2], Nazmun Nahar[3], Syeda Fatima Jinat[4], Sanjida Naharin[5], Salwa Khan[6], Utami Noor Syabaniyah[7]

Abstract

Purpose : *to report three cases of GBS with ophthalmic features.*

Method : *descriptive case reports*

Case report : *A 21 years young man presented with binocular diplopia following unsteady gait for 5 days. Then he developed bilateral variable ptosis with ophthalmoplegia , alteration of voice and nasal regurgitation of food during swallowing. Raised protein was found in CSF study. Ice pack test was positive. Miller Fisher Syndrome (MFS) was diagnosed on the basis of clinical features in addition to CSF study. So, clinically it was a case of MFS with Ocular Myasthenia gravis (OMG).*

A 33 years diabetic and hypertensive female, presented with sore throat for a week followed by binocular diplopia with ophthalmoplegia, bilateral ptosis and impaired speech for 3 weeks. The initial diagnosis was MFS on the basis of clinical presentation.

A young girl of 10 years old, came to us with sudden severe painful dimness of vision of both eyes. She was diagnosed as a case of GBS prior to this presentation. Hyperemic disc swelling found in fundus examination of both eyes. So Guillain- barre syndrome(GBS)with bilateral papillitis was an unique presentation.

Conclusion: *GBS along with its ophthalmic features may present to Neuro-ophthalmologist. So during neuro ocular work-up it should be kept in mind to confirm a clinical diagnosis.*

Keywords: *Guillain-barré syndrome (GBS), Miller fisher syndrome(MFS), Myasthenia gravis, Papillitis.*

Introduction

Guillain-Barré syndrome (GBS) is described as an acute inflammatory polyneuropathy characterized by rapidly evolving ascending weakness, mild sensory loss, and hypo- or areflexia. Besides the classic presentation of GBS, clinical variants are based on the types of nerve fibers involved (motor, sensory, sensory-motor, cranial or autonomic). Although the classic description of GBS is that of a demyelinating neuropathy with ascending weakness, yet many clinical variants have been reported in the medical literature, and variants involving the cranial nerves or pure motor involvement and axonal injury are not uncommon.[1,2]Miller Fisher syndrome (MFS), a clinical variant of GBS, occurs in 5% of cases in the USA, 25% of cases in Japan.[3]James Collier first recognized clinical triad of ataxia, areflexia, and ophthalmoplegia in 1932. It was subsequently described as a variant of Guillain-Barré Syndrome

1. Senior Medical Officer, Ispahai Islamia Eye Institute& Hospital, Bangladesh
2. Associate Professor Cum Senior Consultant, Ispahai Islamia Eye Institute& Hospital, Bangladesh
3. Professor Cum Senior Consultant, Ispahai Islamia Eye Institute& Hospital, Bangladesh
4. Assistant Professor Cum Consultant, Ispahai Islamia Eye Institute& Hospital, Bangladesh
5. Long term Fellow(Neuro-ophthalmology), Ispahai Islamia Eye Institute& Hospital, Bangladesh
6. Short term Fellow(Neuro-ophthalmology), Ispahai Islamia Eye Institute& Hospital, Bangladesh

Address of Correspondence: Sheikh Mohammad Rashedul Haque, Ispahai Islamia Eye Institute& Hospital, Farmgate, Dhaka 1205, Bangladesh, Email: huqrahat1986@gmail.com

(GBS) by Charles Miller Fisher in 1956. Patients with MFS whose clinical features mimic ocular myasthenia gravis have rarely been reported.[4]

Here, we will discuss 3 cases of GBS with ocular presentations.

Case report 1 :

A 21 years male came to Neuro-ophthalmology OPD with binocular variable diplopia for last 2 months. Prior to this presentation, he was hospitalized with complains of lower limbs weakness, generalized fatigability, instable gait followed by bilateral ptosis with ophthalmoplegia, alteration of voice and nasal regurgitation of food during swallowing. He had no history of fever or symptoms of respiratory tract infection and diarrhea prior to this event. His past medical history was not significant and he is non smoker, non alcoholic or non drug abuser. He had no history of exposure to neurotoxins. No family history of neurological disorder. He was evaluated by neurology team and routine investigations were done including lumber puncture for CSF study (raised protein found in CSF). Based on the above reports and clinical examination, it was diagnosed as GBS (Miller Fisher variant). He was feeling better gradually with conservative management but persistent diplopia was there . On examination we found diplopia in primary position and abduction defect on left side with bilateral moderate ptosis. Ice pack test was positive. Although test was normal, he responded with pyridostigmine . Follow up was suggested with Repetitive nerve stimulation (RNS) and other serological study to confirm OMG.

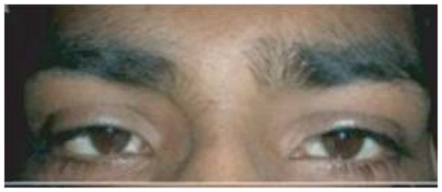

Fig 1a. Before Ice pack test

Fig 1b. After Ice pack test

Case report 2:

A 33 years diabetic and hypertensive female came to Neuro ophthalmology OPD with bilateral severe ptosis, ophthalmoplegia and diplopia for last 3 weeks following sore throat . She was admitted into ICU of a specialized hospital 2 weeks ago due to severe breathlessness. After thorough physical and neurological examination, she was diagnosed clinically as a case of GBS (Miller fisher variant). The patient was subsequently treated with intravenous immune globulin (IV IG) for five days. She was released from ICU after improving general condition but diplopia and ptosis persisted. We examined the patient and found severe ptosis (not variable), total ophthalmoplegia & negative Ice pack test. We advised to continue treatment as per advise of Neurologist with regular follow up and serological tests along RNS to exclude OMG.

Fig 2a. Severe bilateral ptosis

Fig 2b. Ophthalmoplegia in all gazes

Case report 3:

A 10 years young girl, came with sudden severe painful impaired vision of both eyes. She had sudden weakness of lower limbs which ascended up to lower trunk 2 months ago. Neurologist diagnosed GBS on basis of clinical presentation and observation was advised . She recovered gradually by conservative treatment. There was no history of fever, RTI , diarrhea or recent immunization. On ocular examination, Visual acuity- finger counting (FC) in both eyes, Color vision – 0/17 in both eyes by Ishihara chart, Pupillary light reflex – sluggish in both eyes with no RAPD and Fundus – hyperemic disc edema in both eyes. She was diagnosed as a case of bilateral papillitis. MRI of brain and orbit was normal. Neuromyelitis optica antibody (NMO) and Myelin oligodendrocyte glycoprotein antibody (MOG) test were negative. Intra venous methyl prednisolone and followed by oral prednisolone was given. After that gradual improvement of vision(Visual acuity -6/12 on Right and 6/9 on Left) was observed on both eyes.

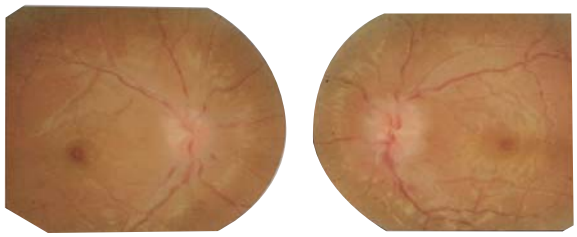

Fig 3a. Color fundus photograph (B/E) on initial presentation

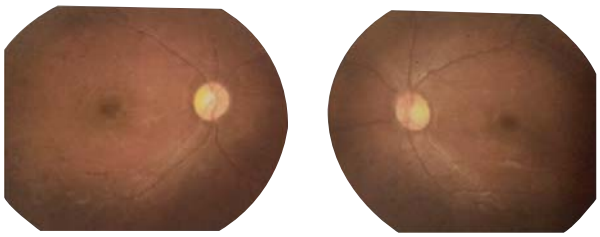

Fig 3b. Color fundus photograph (B/E) after treatment with IV Methyl Prednisolone and oral prednisolone

Discussion

GBS is an acute, immune-mediated polyneuropathies and incidence rate is 1–2 per 100 000 per annual in the world.[5] Miller Fisher Syndrome (MFS) is a geographically variable variant of GBS observed in about 1%- 5% of all GBS cases in Western countries, yet up to 19% and 25% in Taiwan and Japan, respectively.[6] There is an established male predominance at a ratio of 2:1 and a mean age of onset of 43.6 years, although cases of MFS have been reported in all age ranges.[7,8] As in GBS, an antecedent infectious illness can be identified in the majority of MFS cases. Campylobacter jejuni and Haemophilus influenza have been the most commonly implicated pathogens; however, multiple other are also associated, including Mycoplasma pneumonia, and cytomegalovirus. Upper respiratory infection is the most commonly described prodromic entity, followed by gastrointestinal illness.[6,7]

Neurological deficits follow a descending pattern in MFS , starting with diplopia due to external ophthalmoplegia, that is most common presenting symptom. But characteristic pattern of GBS is classical ascending weakness or paralysis.[7,9] Extremity weakness may develop in approximately one quarter of MFS patients like GBS. Acute ataxic neuropathy without ophthalmoplegia and acute ophthalmoplegia without ataxia are incomplete forms of MFS.[9,10]

Anti-GQ1b is an antiglanglioside antibody that is present in about 85–90% of all cases with MFS and this antibody is also found in patients with oculomotor nerve weakness and GBS.[11,12]GBS is diagnosed by clinical presentation initially. But raised protein is often found in CSF study while cell count remains normal in case of GBS and is initially present in about 50–66% of GBS patients following their first week of symptoms, and in 75% of patients in their third week.[9,13,14]

Disease modifying treatment options for MFS like GBS are intravenous immune globulin (IVIG) and plasmapheresis, although MFS is self-

limiting disease . Treatment options are helpful for early recovery of symptom and prevent development of complication. But Benefits of treatment are not clear in MFS.[15]Patients with MFS whose clinical features mimic ocular myasthenia gravis have rarely been reported.

Myasthenia gravis (MG) is an autoimmune disease affecting the neuro-muscular junction (NMJ) of the skeletal muscle causing variable weakness of skeletal muscles, which improves on resting. Ocular Myasthenia Gravis is a subtype of MG where the weakness is clinically isolated to the EOMs, levator, and orbicularis oculi.[16] Ptosis and diplopia are the initial signs of the disease in over 50% of MG patients.[17] The Ice test is a simple, but effective clinical test that can be used to confirm the diagnosis of MG.[18]

A case report, they found a patient with myasthenia gravis (MG) and Miller Fisher syndrome (MFS) overlap.[19]Another case report described MFS and OMG both auto immune diseases in same patient.[20]

Various subtypes of GBS have been described based on clinical, electro-diagnostic, and pathologic criteria. The most common underlying subtypes of the syndrome are acute inflammatory demyelinating polyradiculoneuropathy (AIDP). Another subtypes are acute motor axonal neuropathy (AMAN), and acute motor and sensory axonal neuropathy (AMSAN).[21] In GBS, optic nerve involvement is mostly seen in AIDP patients.[22,23] However, only one patient has been described with the axonal form and optic nerve involvement. Neuwirth et al. reported an adult patient with AMAN form GBS who presented with hyperreflexia and papillitis.[24]

Conclusion

It is important to emphasize on proper history taking and meticulous clinical examination. Patients may present with ptosis, ophthalmoplegia and sudden loss of vision because of optic neuritis to Neuro-ophthalmologist. We should take extra concern, as there is association of GBS with these ophthalmic features.

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