CASE REPORT

Ophthalmoplegia and Areflexia with Positive GQ1b Antibodies: A Limited Form of Miller Fisher Syndrome

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ABSTRACT

Miller Fisher syndrome is a variant of Guillain–Barré syndrome (GBS) which has a clinical triad of ophthalmoplegia, ataxia and areflexia. They are associated with anti GQ1b antibodies which are directed against the GQ1b epitope of cranial and peripheral nerves. This syndrome may present with ophthalmoplegia without ataxia and it may present only with ataxia. They are diagnosed by the presence of GQ1b antibodies in the serum. 95% of patients with Miller-Fisher syndrome possess anti-GQ1b antibodies.

We have a patient with bilateral ptosis and ophthalmoplegia and initially, we considered some common condition like thyroid disease, myasthenia gravis, cavernous sinus thrombosis, inflammatory conditions of eyes like inflammatory orbital myositis (IOM) and Tolosa-Hunt syndromes (THS). However, her initial investigations like MRI brain, nerve conduction studies, repetitive nerve stimulation and thyroid disease were normal. She did not respond to standard treatment of THS or IOM. Later on her GQ1b antibodies were present in the serum and she was discharged with a diagnosis of MFS. On follow-up visit after 1 week, she was much improved.

Keywords: Miller Fisher Syndrome, Anti GQ1b antibodies, Ophthalmoplegia

Introduction

Miller Fisher syndrome is an unusual variant of Guillain–Barré syndrome (GBS), which accounts for 3-5% of GBS cases.[1] It is characterized by ataxia, areflexia and opthalmoplegia.95% of patients with Miller-Fisher syndrome possess anti-GQ1b antibodies.[2] Miller Fisher syndrome may present with ataxia without ophthalmoplegia or it may present with ophthalmoplegia without ataxia.[3] Here we report a case of Miller Fisher syndrome that presented with ophthalmoplegia without ataxia and positive GQ1b antibodies.

Case Report

A 45 years old lady presented in the emergency department with diplopia and bilateral ptosis for the past 7days. She said that she developed difficulty in walking because of her double vision. She had normal speech, swallowing and she was not having weakness of limbs. Her eyesight, hearing, speech and comprehension were normal. She was a known case of hypertension on treatment and rest of systemic inquiry was unremarkable. There had no significant past medical history. She took Oral contraceptive pills once while going to Umrah. Family history included hypertension.

On examination, her pupils were equal, round and reactive. The fundoscopic examination was normal. She had complete ophthalmoplegia of both eyes with bilateral ptosis. Pupils were equal, round and reactive. Rest of the cranial nerves were normal. Motor examination revealed normal bulk, tone and power in all limbs but deep tendon reflexes were absent. Plantars were bilaterally down-

going. There was no ataxia. Her systemic examination was normal.

Her blood tests revealed WBC count was 19,100/cmm, Hb was 12.00 g/dl, platelets 355,000/cmm, INR 1.0 and normal LFTs, RFTs, serum electrolytes and lipid profile. Her ESR was raised (35 mm/1st hour) and CRP was borderline (5.61 mg/L, normal <5.0). Serum ANA was positive and C3, C4 levels were normal. Her CSF exam revealed WBCs <05/cmm, RBCs 20/cmm, Glucose 64 mg/dL and Protein 32.1 mg/dL.

Her MRI brain revealed T2 and FLAIR hyper-intense lesions suggestive of micro-vascular angiopathy, a large retention cyst with mucosal thickening obliterating the right sphenoid sinus antrum and mucosal thickening in the bilateral maxillary and ethmoid sinuses. There was no evidence of dural venous sinus thrombosis on MR Venography.

Nerve conduction studies and electromyography were normal. Her repetitive nerve stimulation was done for myasthenia gravis was also normal.

At this time our differentials included Tolosa-Hunt syndrome, ocular myasthenia gravis and Miller Fisher syndrome without ataxia. She was already taking pyridostigmine for one week for suspected myasthenia gravis but was not improving. Moreover, as her repetitive nerve stimulation and acetylcholine receptor antibodies were normal, it was stopped. As Tolosa-Hunt syndrome is an inflammatory disease of eye that responds well to steroids, she was given prednisolone 1 mg/kg but did not show improvement. Her serum anti GQ1b antibodies came out positive (titer 18 AU, normal < 5 AU).

She was discharged with a diagnosis of the limited form of Miller Fisher Syndrome. She was followed up after one week and her symptoms were found to be improving.

Discussion

Miller Fisher Syndrome (MFS) is one of the variants of GBS and comprises of a triad of ataxia, ophthalmoplegia, and areflexia. It can be post-infectious. There is a common lipopolysaccharide on the membranes of cranial and peripheral nerves at the GQ1b epitope and similar polysaccharide is present on the surface of bacteria and viruses.[4] Involvement of occulomotor nerve is common as it has a high concentration of GQ1b polysaccharides. CSF R/E is done to see albumin cytologic dissociation and to exclude other causes like infections and malignancies. Nerve conduction studies show absence or reduction of sensory nerve action potential with mild demyelinating neuropathy or axonal degeneration. [5, 6]

Most patients with MFS have a benign course and they start to improve after 2-4 weeks[7] with a mean recovery time of 10 weeks. Some patients develop swallowing difficulties and severe weakness and may require artificial ventilation.[8] Treatment is usually not required. Although immune modulatory treatment hastens recovery, no overall benefit has been reported.[9] Treatment is suggested when there is severe limb weakness or ataxia, swallowing difficulties or breathing problems.[10]

Antibodies against GQ1b (a component of nerve) are present in 95% of patients with MFS.[2] MFS may present with ataxia without ophthalmoplegia or it may present with ophthalmoplegia without ataxia.[3] These GQ1b positive antibodies syndromes may present with any combinations of signs and symptoms. Identification of these syndromes is important as they have similar etiology, investigations, and management.

Our case also is very similar to ocular myasthenia gravis which can have a similar clinical presentation and sometimes these cases are treated for myasthenia. However GQ1b antibodies are negative in myasthenia gravis.[11] Her MRI scan of the brain has ruled out any intracranial lesion like cavernous sinus thrombosis or retro-orbital tumors. Tolosa Hunt Syndrome and idiopathic orbital myositis may present with ophthalmoplegia and the latter condition is painful. They are inflammatory conditions and usually respond well to steroids. If a patient is having ophthalmoplegia with positive GQ1b antibodies, MFS must be strongly considered even if the patient is not having the complete triad of ophthalmoplegia, ataxia and areflexia.

Conclusion

Miller fischer syndrome is a triad of ataxia, areflexia and opthalmoplegia. However it may present only with ataxia or opthalmolplegia where it is difficult to diagnose as there are many differential diagnosis of these conditions. Anti GQ1b are very important in diagnosing isolated ataxia or opthalmoplegia as a limited form of Miller Fischer syndrome.

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