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A Case of Miller Fischer variant of Guillain Barre Syndrome

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Abstract- We are presenting a case of 13 year old female , diagnosed as Miller Fischer variant of Guillain Barre syndrome who presented to us with sudden onset of bilateral facial weakness followed by limb weakness.

I. Case Report

A 13 year old female, 5th standard student presented to us with sudden onset of double vision followed by difficulty in walking for one day. Weakness developed within 1 day.

There was no complain of trauma, back pain , fever, diarrhoea , cough, convulsion, loss of consciousness, difficulty in swallowing. There was no sensory complains or bowel bladder involvement.

There was no past history of similar complain or family history of similar illness in parents or siblings.

On admission, patient's vitals were normal. On neurological examination, there was complete External Opthalmoplegia due to iii, iv and vi nerve palsy and mild ptosis in both eye. Both pupils were normal in size and reacting to light. There was also bilateral lower motor neuron type of facial weakness.

There was complete paraplegia and hypotonia in bilateral lower limb. There was bilateral hand grip weakness. Head flexion was weak.

All deep tendon reflexes were absent. Plantars were bilaterally absent. All sensations were preserved with intact bowel and bladder control.

On investigation, Electrolytes were normal, Urine for Porphyria was negative, S.CPK total was 59 mcg/L.

Nerve Conduction Study was suggestive of Demyelinating polyneuropathy suggesting Guillain Barre syndrome.

During stay in hospital, 5 cycles of plasmapharesis was done along with active physiotherapy.

Opthalmoplegia, ptosis and facial weakness improved. There was mild improvement in distal weakness in bilateral lower limb. Deep tendon reflexes were still absent.

II. Discussion

Guillain-Barré syndrome (GBS), also known as acute idiopathic polyneuritis, is a type of neuromuscular paralysis that has several variants.These variants all share similar patterns of symptoms and patient recovery. Miller Fisher syndrome (MFS) is a rare variant of GBS, observed in only about 1% to 5% of all cases of GBS in Western countries. Miller Fisher syndrome occurs in more men than women by a ratio of approximately 2:1.3The mean age of onset of MFS is 43.6 years, though onset has been documented in individuals between the ages of 13 and 78 years.

The main difference between MFS and more common variants of GBS is that the first nerve groups to be affected by paralysis in patients with MFS are those in the head, resulting in difficulty controlling eye muscles and balance. Paralysis in other forms of GBS typically begins in the legs. Moreover, MFS is characterized by a triad of conditions: areflexia, ataxia, and ophthalmoplegia.

The present report describes the case of a girl presented chiefly with complaints of diplopia and gait abnormality. Further evaluation revealed the triad of conditions characteristic of MFS. The examination found no signs of pupillary defects or nystagmus. Bilateral Ptosis was present with eye ball central in position. Bilateral hyporeflexia was detected throughout the patient's body. The patient showed no sensory deficits to light touch, pinprick, or proprioception.

Miller Fisher syndrome is an uncommon variant of GBS that can be diagnosed by tests for its characteristic triad of conditions: areflexia, ataxia, and ophthalmoplegia.

In cases of MFS, ataxia is primarily noted during the patient's gait, typically in the trunk and with lesser involvement of the limbs. A patient's motor strength characteristically is spared in MFS. Anti-GQ1b antibodies, activated by certain strains of *Campylobacter jejuni*, have a relatively high specificity and sensitivity for MFS. Dense concentrations of GQ1b ganglioside are found in the oculomotor nerve (cranial nerve III), trochlear nerve (cranial nerve IV), and abducens nerve (cranial nerve VI) of patients with MFS, which may explain the relationship between anti-GQ1b antibodies and ophthalmoplegia. Titers of anti-GQ1b antibodies in CSF that are greater than 1:40 are specific for MFS.

Onset of recovery from ataxia and ophthalmoplegia in patients ranges from approximately 2 weeks to 2 months, with a mean recovery onset at 10 weeks. Most patients will fully recover from ataxia and ophthalmoplegia, as well as areflexia, within 6 months. In some cases of MFS, post-gadolinium magnetic resonance imaging might show enhancement of cranial nerves.

III. Conclusion

The present case demonstrates that physicians need to be aware of symptoms and examination findings consistent with MFS so as not to confuse with other diagnostic possibilities.

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