# MILLER FISHER SYNDROME -A DIAGNOSIS TO LOOK FOR

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**ABSTRACT**

A 39 year old non-Diabetic, normotensive male presented to us with sudden onset diplopia with limitation of abduction in left eye that progressed to total external ophthalmoplegia in both eyes with ptosis and facial paresis within one week. There was no history of any dysarthria, dysphagia, bowel and bladder involvement, fever or upper respiratory infection. On neurological examination he was found to have ataxia and diminished reflexes with intact motor power. MRI brain was normal and nerve conduction velocity test suggested facial demyelination. He responded to IV immunoglobulin therapy with total recovery of ocular movements in 3 months. We present this case of Miller Fisher syndrome to emphasize the importance of a complete neurological evaluation in any case of acute progressive ophthalmoplegia and also to raise the level of awareness to the existence of this alarming neurological illness with a benign course

**Key-words:** Miller Fisher syndrome, acute progressive ophthalmoplegia, Guillain- Barre? syndrome

## INTRODUCTION

Miller Fisher syndrome (MFS) is a variant of Guillain-Barre? syndrome characterized by the triad of ophthalmoplegia, ataxia and areflexia, without significant motor or sensory deficit in the limbs.[1] The rather acute onsets of such ocular signs associated with cerebellar ataxia are apt to be alarming to the treating clinician. The aim of this report is to raise the level of awareness to the existence of this acute neurological illness with a benign course.

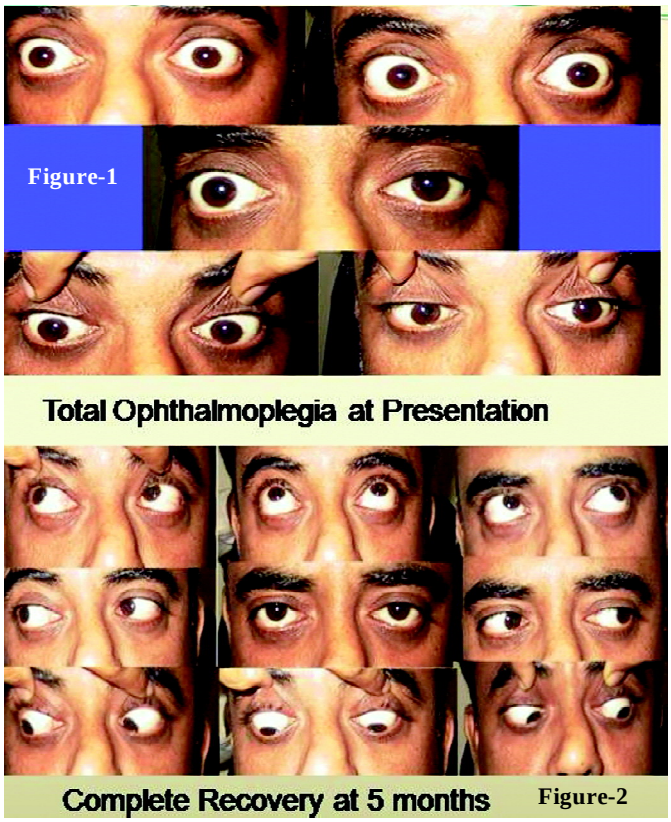
### Case History

A 39-year-old non-diabetic, normotensive male presented to us with sudden onset diplopia since one day. He also complained of paraesthesiae in his hands and mild weakness of lower limbs off and on for past 3-4 days. There was no history of any dysarthria, dysphagia, bowel / bladder involvement, fever or upper respiratory infection. On ophthalmic examination there was limitation of abduction of the left eye. The pupils were symmetrical and well reacting with no RAPD. MRI brain was unremarkable.

Three days later the patient developed ptosis of both eyes, more so in the left eye. There was gross limitation of all ocular movements (Figure 1). The pupils were slightly dilated but briskly reacting.

The patient was referred to a neuro- physician where he was found to have ataxia. The deep tendon reflexes were diminished with normal motor power in all limbs. A provisional diagnosis of Myasthenia Gravis was made and the patient was put on Tab. Pyridostigmine 60 mg QID for 5 days but no clinical or subjective improvement was noticed.

One week later, the patient noticed facial weakness on the left side with deviation of the angle of mouth. The patient was admitted for investigations. The routine hematological and blood biochemistry tests were within normal limits. Nerve conduction velocity test was suggestive of facial demyelination and repetitive nerve



stimulation was negative. MRI thorax and Acetyl cholinesterase antibodies were also negative. CSF analysis showed elevated proteins (65.6 mg/dl) with near normal WBCs (7 / $mm^3$)

A clinical diagnosis of Miller Fisher syndrome was made and the patient was put on intravenous immunoglobulin (IV IG) therapy for five days.

Three weeks after the IV IG therapy, the patient had recovered some vertical movements and was prescribed horizontal prisms for diplopia .At last follow up (5 months) the patient has full recovery of ocular movements with no diplopia (Figure 2).

## DISCUSSION

Miller Fisher syndrome (MFS) is one of a spectrum of acute demyelinating inflammatory polyneuropathies, which include Guillain – Barré syndrome, acute ophthalmoplegia and Bickerstaff's encephalitis. Originally described by Miller Fisher (1956), MFS is a triad of ataxia, areflexia, and ophthalmoplegia.

The male/female ratio is 2:1, with a mean age of 43.6 years at the onset of the disease. A viral infection which is seen preceding the neurological symptoms in 71.8% of the cases ,[2, 3] was not present in our case.

Diplopia is the presenting feature in 38.6% cases, with the remainder presenting with only ataxia (20.6%) or with both symptoms simultaneously.[1, 2]The disease progresses over five to ten days, sometimes up to three weeks. External ophthalmoplegia begins relatively symmetrically and most patients progress to have complete immobilization of the globes.

Additional features consistent with a peripheral neuropathy including paraesthesiae (50%), oropharyngeal weakness (26%), or bi-facial weakness (32%). Bladder dysfunction (16%) or more widespread dysautonomia is occasionally seen.[2, 3]

The diagnosis of MFS is still descriptive, depending on the presentation of the triad of MFS. Although, almost half of the cases are not the pure syndrome, it is important to emphasize that the diagnosis of MFS should be considered in any patient who develops an acute ophthalmoparesis with or without either ataxia or hyporeflexia, or even acute onset of either ataxia or areflexia without ophthalmoplegia.[3] Cranial nerves other than the oculomotor nerve may be involved in more than half of the cases (56 .9%), most commonly the facial nerve (45.7%) followed by IX and X (39.9%), and XII (13%). [4]

CSF examination may show an elevated protein value with a normal cell count. A tetrasyaloganglioside (GQ1b) antibody in patient serum may give clues in the diagnosis of MFS variants .Due to the linkage between this antibody and MFS, especially ophthalmoplegia; it may prove a highly sensitive and specific clinical marker for the diagnosis of typical and atypical MFS. It would also differentiate MFS ophthalmoplegia from ophthalmoplegia of other causes. GQ1b antibody is supportive but not essential for the clinical diagnosis of MFS.[3, 5]

Because of the relative scarcity of MFS, it is usually incorporated with GBS in the evaluation of therapy. The treatment options are steroids, plasma exchange (PE) and intravenous immunoglobulin (IVIG).[3]

Prognosis is good with recovery after a mean of 10 weeks, [2, 3] although as in the present case the total recovery may be delayed up to several months. Prisms may be prescribed in the meantime for the diplopia. Most patients show a complete remission without any residual symptoms.

## CONCLUSION

In summary, the importance of recognizing the significance of the triad of acute ophthalmoparesis, ataxia, and hyporeflexia, especially following either a gastroenteritis or upper respiratory infection, greatly limits the differential diagnosis. As discussed above, ophthalmoparesis without ataxia or areflexia does not eliminate a diagnosis of MFS, and therefore the diagnosis needs to be established with the combination of serologic studies, CSF, and electrophysiology. The authors present this case to highlight the existence of this acute neurological illness with a relatively benign course if recognized and managed appropriately.

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