Miller Fisher Syndrome Presenting with Idiopathic Central Serous Chorioretinopathy: A Rare Association

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ABSTRACT

Introduction: Miller Fisher syndrome is a rare autoimmune disorder characterized with acute onset of diplopia, bilateral complete ophthalmoplegia, ataxia and areflexia. It is a variant of Guillian Barre Syndrome and carries a risk of progression to GBS in rare instances. But unlike GBS, MFS is a self-limiting condition with annual incidence of 0.09/100,000 populations, and because of its similarity with GB syndrome, prompt diagnosis and follow-up is required. Typical MFS has never been reported previously with Idiopathic Central Serous chorioretinopathy [ICSC].

Case report: 40 years old male presented with sudden onset diminution of vision, diplopia and associated neurological features suggestive of MFS. B.P. at the time of presentation was 150/100mm of Hg with no prior history of hypertension. On further evaluation patient was diagnosed with ICSC right eye along with MFS. Patient was managed with supportive treatment and systemic steroid was avoided. Patient recovered completely by 4th week.

Conclusion: MFS is autoimmune neurological condition presenting as GBS. Because of its rarity, good clinical knowledge of this condition is essential for prompt diagnosis and management. MFS patients only require supportive treatment till complete neurological recovery. But rare severe cases may require immunosuppressive treatment. Association of ICSC with autoimmune conditions like Systemic Lupus Erythematous(SLE), Anti-phospholipid Ab syndrome is well known but association with MFS has never been reported earlier. In present case, association of ICSC with MFS and its resolution with resolution of systemic disease suggest some common pathophysiology, which needs further study.

Key words: Miller Fisher Syndrome, MFS, Idiopathic Central Serous Chorioretinopathy, ICSC, Central Serous Retinopathy, CSR, Guillian Barre Syndrome Variants

INTRODUCTION

Miller Fisher syndrome [MFS], also known as Fisher's syndrome, usually present with acute onset of three problems: 1) weak eye muscles, with double or blurred vision, often associated with drooping of eyelids and facial weakness; 2) poor balance and coordination with difficulty in walking and deglutition; and 3) on physical examination, loss of deep tendon reflexes, including the knee and ankle jerk.[1] Although the predominant ophthalmic feature of MFS is complete bilateral external ophthalmoplegia, it should be noted that the disease has variable associations with lid and pupillary dysfunction.[1]

MFS is considered as a limited variant of ascending paralysis, the Guillain-Barre syndrome [GBS]. Pure Fisher syndrome is quiet uncommon, with annual incidence of one in one million population and rarely the patients may go on to develop the prominent widespread weakness of GBS.[2] With diplopia being one of the most common presenting symptom of MFS, awareness of the disease among ophthalmologist is a needed for prompt diagnosis.

Association of ICSC with Miller fisher syndrome

Association of Idiopathic Central Serous Chorioretinopathy [ICSC] or Central serous chorioretinopathy [CSCR] is proven with autoimmune diseases like Systemic lupus erythematous [SLE], Anti-phospholipid antibody syndromes, gastroesophageal reflux diseases.[8] However there is no reported case of Miller fisher syndrome presenting with ICSC. We are reporting a case of MFS associated with U/L right eye Idiopathic central serous chorioretinopathy.

CASE REPORT

A 40-year-old male presented with sudden onset, progressive diminution of vision both eyes (right eye more than left eye) and double vision for last 14 days associated with history of cough and fever. Cough and fever resolved spontaneously within 1 week of onset. Difficulty in vision was associated with progressive difficulty in walking and doing fine work like taking the food up to the mouth. There was no history any loss of consciousness, seizures, any behavioral abnormality or any chronic illness like DM, Hypertension. The blood pressure at the time of presentation was 150/100mm of Hg.

On examination, the best-corrected visual acuity was 6/36 (Pin hole 6/18), N36 in the right eye and 6/6p, N36 left eye. Extraocular movements were absent in all cardinal gazes. But there was no associated eyeball deviation or ptosis. Corneal sensation was significantly diminished both sides. Slit lamp evaluation show bilateral dilated pupil with absence of light reflexes. On fundus evaluation under 90D lens, ring reflex with absent foveal reflex were noted in right eye. On OCT-FMT evaluation, macular edema with central macular thickness of 551 micron was noted in right eye. FFA of right eye showed typical smoke stacked hyper-fluorescence pattern suggestive of ICSC. Fundus, OCT FMT

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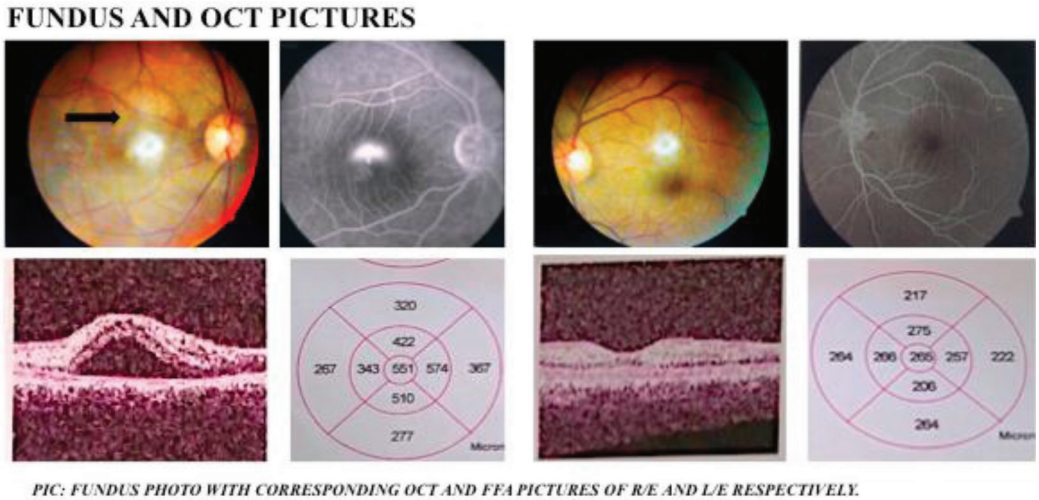

Figure-1: Fundus photo, FFA and OCT-FMT of both eye at presentation.

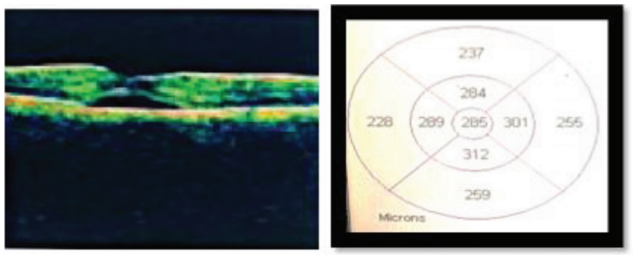

Figure-2: OCT of Right eye on 1st follow-up (At 2nd week)

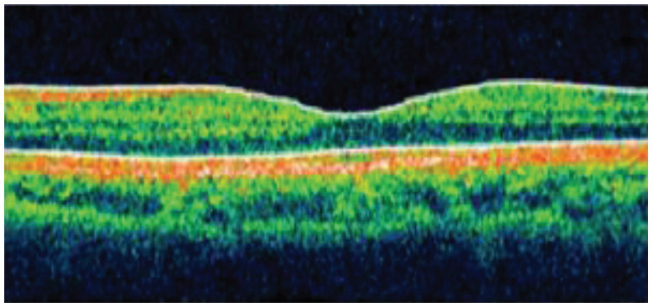

Figure-3: OCT of Right eye on 2nd follow-up (At 4th week)

and FFA evaluation were normal for left eye (Fig. 1).

On neurological examination, multiple cranial nerve palsy involving CN III with LPS sparing, IV, V, VI, VII (LMN type with right side more than left side) were noted along with complete areflexia (B/L absent deep tendon reflex with B/L plantar response upgoing) and cerebellar ataxia. CSF evaluation show raised CSF protein (113mg/dl). NCV (nerve conduction velocity) of upper and lower limbs show reduced latency and amplitude. VEP for optic nerve assessment was within normal limit. MRI brain and spinal cord was normal. Anti GQ1b IgM or IgG could not be done b/c of lack of availability of test.

Based on the clinical findings, a provisional diagnosis of Miller Fischer syndrome with Right eye ICSC with hypertension was made and patient was managed conservatively with weekly injections of Vit. B1, B6, and B12 and topical NSAID for 4 weeks along with tab amlodipine 5mg as antihypertensive mediation. Patient was followed closely.

On follow-up (after 2 week), the best-corrected visual acuity returned to 6/9p, N8 right eye and 6/6, N8 left eye and EOM recovered to full range with mid-dilated sluggishly reacting pupil in both eye. On repeat OCT evaluation the right sided macular edema reduced to 285-micron macular thickness (Fig.2). Ocular improvement was associated with improvement in systemic symptoms with complete recovery of normal gait and deep tendon reflexes.

On 2nd follow-up (4th week) there was complete recovery of vision (6/6,N6 both eye) along with resolution of ICSC in Right eye (Fig. 3) without any associated neurological deficit.

DISCUSSION

Miller Fisher syndrome is autoimmune neurological condition characterized by triad of: partial or complete ophthalmoplegia, ataxia, and areflexia. It is considered as a variant of GBS, accounting for 5 to 10% of all GBS cases, but unlike GBS it often presents with symptoms of diplopia.[2,3] The annual incidence of GBS is 1–2 cases/100,000 population, whereas MFS has a much lower annual incidence of 0.09 cases/100,000 population.[3] The incidence of MFS is more common in men and affects people of all ages, with the median age of onset being in the 4-5th decade (43.6yr).[4]

The clinical course of MFS is self-limiting and is similar to an acute phase primary immune response, in which a humoral response is initiated with subsequent nadir and then followed by spontaneous recovery.[1] The median time from the infection onset to development of neurological symptoms is approximately 8-10 days.[4] Miller fisher syndrome is autoimmune condition with antibody directed against neuronal myelin sheath especially against the GQ1b ganglioside. In more than 90% cases of miller fisher syndrome IgM or IgG anti GQ1b antibodies are found to be positive.[5] GQ1b ganglioside is highly enriched in CN III, IV and VI and ciliary ganglia. So, acute internal and external ophthalmoplegia is presenting feature of miller fisher syndrome. But association of ICSC as ophthalmic finding in miller fisher syndrome has not been reported earlier.

Although associations of ICSC with other autoimmune conditions like SLE, Anti-phospholipid syndrome is well established.[8] In our case ICSC in Rt. Eye may be the incidental finding associated with undiagnosed hypertension at time of presentation (B.P. 150/100mm Hg). However resolution of unilateral ICSC with resolution of systemic disease indicates some common pathophysiology, which needs further study to establish or rule out any positive association b/w ICSC and Miller fisher syndrome.

CONCLUSION

Because Miller Fischer syndrome most commonly present with acute onset of diplopia with bilateral variable degree of ophthalmoplegia and because of its rarity and similarity with Gullain Barre syndrome, familiarity of this condition among clinician and ophthalmologist is essential for prompt diagnosis and management.[2]

In present case report, association of Miller fischer syndrome with Idiopathic central serous chorioretinopathy and resolution of ICSC with resolution of disease activity suggest some common pathophysiology which needs further study in future.

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