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Miller Fischer Syndrome – A Case Report

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ABSTRACT:

Miller-Fischer Syndrome(MFS) is a less common variety of Guillain Barre Syndrome(GBS) having different incidences and is reported in all the age groups. Ataxia, areflexia and ophthalmoplegia are the cardinal features based on which the diagnosis is made. Anti-GQ1b antibody titres help in establishing the serological diagnosis. This case report is of a female child aged 4 years who was brought with progressive generalised weakness, lethargy, vomiting and double vision following a minor upper respiratory illness. The characteristic features of ataxia, areflexia, and ophthalmoplegia characteristic to MFS were present. The provisional diagnosis of MFS was proved by serology which showed anti-GQ1b antibodies positive and the child was managed with Intravenous immuno globulin (IVIG) following which the child improved.

Keywords: Guillain Barre Syndrome, Miller-Fischer Syndrome, Neuropathy, Ataxia, Areflexia, Ophthalmoplegia

1. Introduction

James collier was the first to describe the the characteristic ataxia, areflexia, and ophthalmoplegia in MFS in1932 (1). Charles Miller Fisher in 1956, described this as a type of GBS and is named after him(1,2). It is now classified under GBS as heterogenous group of immune-mediated neuropathies. It has been reported across the world. Males have incidence twice that of females and the mean age of onset was reported to be 43 years. It is often predisposed by infectious diseases. The common causative organisms are Campylobacter jejuni and Haemophilus influenza; Mycoplasma pneumonia, cytomegalovirus and Zika virus are the other implicated organisms which are not very common. The prodromal symptoms are usually of upper respiratory infection or of gastrointestinal illness(2,3).

The disease is usually self limited in the course., but intravenous immune globulin (IVIG) and plasmapheresis can modify the progressive pattern of the disease just like in GBS. Benefits of the treatment include quicker resolution of the symptoms and decreasing the probability of developing serious complications (4).

Neurological symptoms or signs proceed in a descending fashion first causing external ophthalmoplegia (the most common feature being diplopia). GBS unlike MFS presents with ascending paralysis.(3) In a clinical series of 50 MFS cases reported from Japan, 77% began with diplopia, 48% presented with ataxia, and 33% with both the features . Dysesthesia of the legs, blepharoptosis, palsies of facial, bulbar, and pupillary nerves, grade 4 motor weakness are some of the features which can occasionally be seen. The complications associated with MFS are urinary incontinence or urinary retention.(3)

Case History:
A preschool female child aged 4 years presented with complaints of brief history of cough (non productive) and running nose with lethargy for 2days and history of 4 episodes of vomiting for 1 day which were non projectile and non bilious. On examination the child was found to be febrile with a temperature of 100F and pallor was present. Other systems showed no notable findings. She was initially diagnosed as acute febrile illness and was treated symptomatically. Blood investigations during admission revealed the following, Hb - 10 gm/dL, TLC of 13,000 cells/cu mm, differential count of 52% neutrophils, 45% leucocytes, 3% monocytes, platelets was found to be 120,500 cells/cu mm, and CRP - 22 mg/dL. Serum calcium, LFT and RFT were within the normal ranges. Chest X-ray did not reveal any abnormalities. Following admission and symptomatic treatment there were no fever spikes and complaints of vomiting subsided. The child was treated empirically with ceftriaxone in view of leucocytosis and positive CRP with pending blood culture reports.
Despite treatment, the child developed diplopia which was progressive and excessive fatigue with drowsiness (GCS was 14/15). On detailed examination the child had drooping of eyelids, right eye total ophthalmoplegia and restriction of adductor component of left eye.
Gait was unstable and there were also few cerebellar signs like past pointing on performing finger nose test. Fundus examination was normal with no evidence of papilledema. After obtaining the neurologist opinion, MRI brain with contrast was done suspecting post viral demyelination and was found to be normal. (Figure1-4)
She had progressive ophthalmoplegia, areflexia and hypotonia in all the 4 limbs over the next 2 to 3 days. The differential diagnosis were Myasthenia Gravis (MG) with an atypical presentation or Miller-Fischer Syndrome (12,13). Antibody testes were done, to come to a diagnosis, and the patient was also Pyridostigmine, which is a cholinesterase inhibitor, orally to treat for MG which was one among the possible diagnosis. As the child did not show any

clinical improvement after three doses of pyridostigmine, it was stopped. The patient was further managed with IVIG for 5 days considering MFS which is the other possible diagnosis. Serum antibody test result for GQ1b IgG was positive, hence proving the diagnosis of Miller-Fisher syndrome.

Following IVIG therapy the child had a dramatic improvement and regained normal power, tone and vision, and was discharged without any sensory or neurological deficits. The child is on periodic follow up and is doing well.

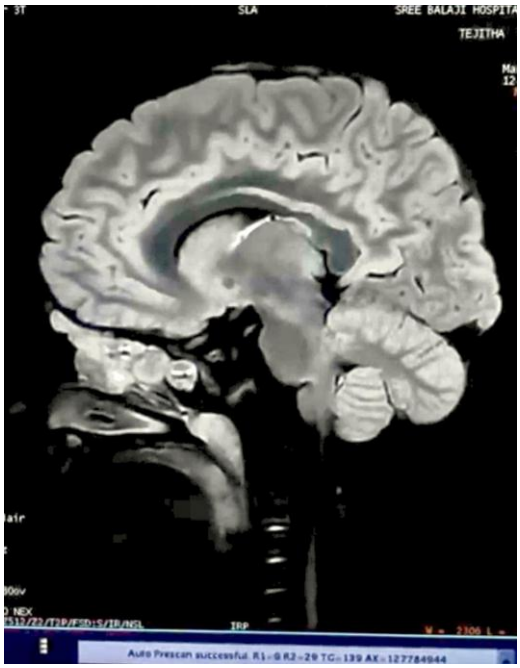

Figure: 1

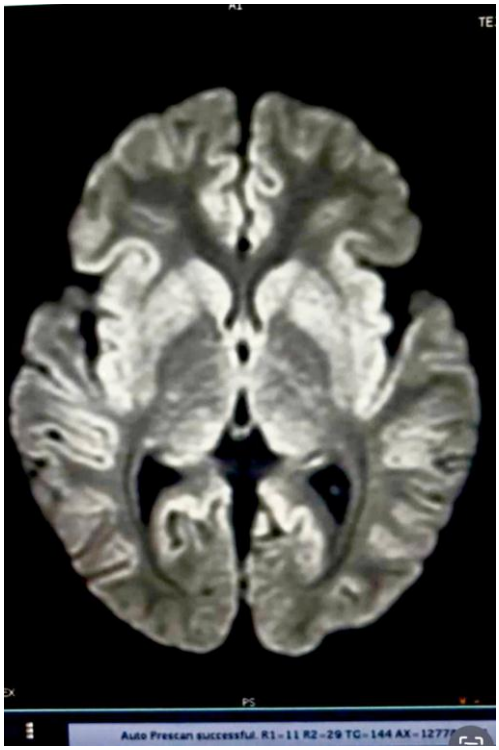
Figure: 2

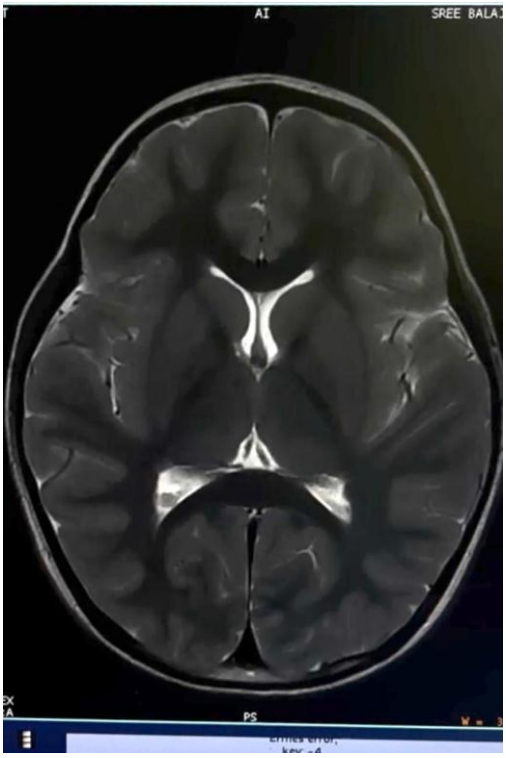

Figure: 3

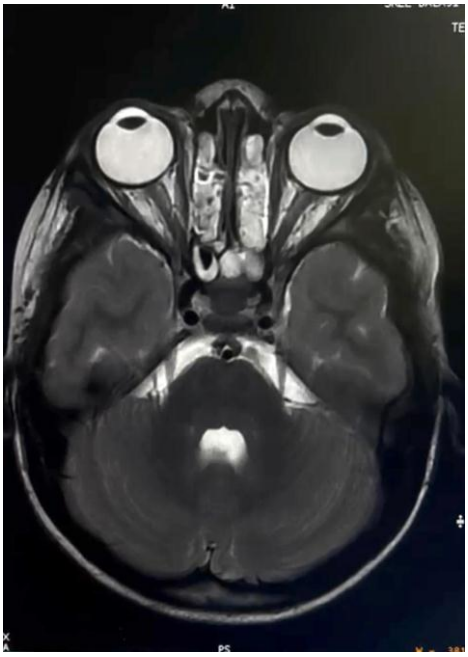

Figure: 4

2. Discussion

MFS is a benign, self-restraining condition, with prognosis being good. Most of the patients have a good improvement in the signs and symptoms within 6 months(10). Opthalmoplegia, ataxia, and areflexia without any other sensory deficit are the defining triad of Miller-Fisher Syndrome(11). It is a variety of Guillain Barre Syndrome. Acute idiopathic neuritis is an other term for it. Worldwide it is estimated that the incidence of GBS is found to be 0.6 per 1,00,000 ; of which about 5% were reported to be cases of MFS (7,8,9). There is growing data that

suggests that Miller-Fisher Syndrome and other types of GBS share a considerable number of neurological characteristics and that these neurological characteristics span a wide spectrum. Although evidence of central aspects has also been documented, the peripheral nervous system appears to be more significantly affected by Miller-Fischer Syndrome. It's noteworthy that during their sickness, our patient displayed a number of uncommon features.

As we can see from our case report, the onset of MFS is typically acute and usually follows an upper respiratory tract illness or gastrointestinal tract illness. The disease then progresses with neurological signs and then followed by the recovery period where clinical improvement of symptoms occur; although severe side effects including respiratory failure or arrhythmias of the heart, though reported to be rare can occur in child withMFS (5,6) Near complete recovery is anticipated within 6 months even without treatment. This recovery is dramatic with IVIG therapy (6).

According to numerous researches, MFS is specifically characterised by antibodies to the gangliosides, or IgG anti-GQ1b antibody. Ophthalmoparesis in MFS is hypothesised to be caused by anti-GQ1b antibodies which attack the neuromuscular junction(NMJ) at the cranial nerve innervation of eye muscles. Bickerstaff brainstem encephalitis (atypical MFS with central nervous system signs), post-infectious acute ophthalmoplegia (AO), incomplete MFS lacking ataxia, and Guillain-Barre syndrome with ophthalmoplegia (GBS-OP), all present with a positive GQ1b antibody. An enzyme-linked immunosorbent assay (ELISA) will yield positive results in roughly 70% to 90% of individuals. A small percentage of patients (up to 30%) are negative (GQ1b-seronegative), possibly because the presence of calcium-dependent ligands is necessary for the procedure to work., which is said in a study by Uchibori et al., 2016 in the Journal of Neuroimmunology.

The aforementioned case discusses targeted differential diagnoses, the anti-GQ1b antibody test, prognosis, and possible treatments in addition to highlighting important clinical characteristics of MFS. Knowing about this uncommon syndrome will help the doctor recognize MFS in patients who present with symptoms related to the eyes, ataxia, and areflexia.

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