Miller Fisher Syndrom in Intensive Care Unit: 2 Case Report and Review of Literature
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ABSTRACT
We report a case of Fisher/guillainbarré overlap in a 14 years old patient with antecedent of an infectious respiratory event one week before the installation of Miller fisher Syndrome. The symptomatology rapidly evolved into a case of guillainbarré with respiratory and deglutition disorders. The patient received 2 courses of IvIg, 5 days each, with a 2 weeks interval which resulted in a total decline of paralysis and motor recovery. This case shows the importance of meticulous surveillance of patients with Miller Fisher syndrome in order to act on first symptoms of a guillainbarré overlap.

Introduction
Guillain Barre Syndrome (GBS) is a rare but serious affection in which the body's immune system attacks part of the peripheral nervous system. It is described in literature that GBS can occur after a viral or bacterial infectious event, especially after a pulmonary infection. GBS can be described as a collection of clinical syndromes that manifest as an acute inflammatory polyradiculoneuropathy resulting in muscular weakness and diminished reflexes[1]. The weakness may progress over hours to days to involve the arms, truncal muscles, cranial nerves, and muscles of respiration.

Miller Fisher Syndrome (MFS) was first described by Dr. Charles Miller Fisher in 1956 as a limited variant of guillain Barre Syndrome. It is characterized by the clinical triad of ataxia, ophthalmoplegia, and areflexia.

We report here a rare case of 2 children with MFS syndrome that was complicated by tetraplegia, respiratory disorders and dysautonomia.

Case report
Case N1
A 14 years old girl with no medical history, presented to our emergency department, one week after a flow episode, with sudden weakness, ophthalmoplegia, double vision, ataxia, areflexia and respiratory disorder.

Upon her admission, evaluation of vital signs revealed pyrexia (T: 37°C), tachycardia with a pulse of 123 beats/min, blood pressure was high (150/70 mmHg), tachypnea of 30 breaths/minute, pulse oximetry was 75% at ambient air and 89% with high concentration mask. Neurological exam finds an agitated patient with altered mental status (Glasgow coma score GCS at 13/15), tetraplegia and ophthalmoplegia with ptosis, cranial nerve examination showed bilateral ptosis and total external and internal ophthalmoplegia, with total facial bilateral and symmetrical paralysis, which neither exaggerated on fixing of gaze upward nor improved with neostigmine. On motor system examination, strength in triceps, biceps, and supinator were 3/5; deltoids and pronator were 3/5; wrist extensors and flexors were 3/5 bilaterally; hip flexors, quadriceps, and hamstrings were 2/5 bilaterally; dorsiflexors and plantar flexors were 1/5 bilaterally. Her sensation was intact on upper extremities but diminished to touch and pin prick on lower extremities. Deep tendon reflex (DTR) 1+ and symmetrical on deltoid absent on biceps, brachioradialis and triceps; DTR was absent on knees and ankle.

At this point, the patient was put in mechanical ventilation due to respiratory distress.

Blood tests did not reveal any infectious process. Thyroid test were normal. We realized an MRI that showed no abnormalities. Her Cerebrospinal fluid analysis (CSF), showed no albuminoctycological dissociation.

As the patient showed clinical triad of MFS – ataxia, areflexia, ophthalmoplegia initially, then was aggravated by Bilateral symmetrical ascending motor weakness, with Respiratory and deglutition disorders, the patient was diagnosed to have Fisher/Guillain-Barré overlapping syndrome.

A 5-day course of IVIG (0.4 g/kg/day) was administered. After 2 days, the patient showed improvement of her limbs weakness. 2weeks later, the patient had significative improvement of limbs motility, the axial motility and tonus were also improved; after 3 weeks, we noticed regression of the ptosis and paresis of the left oral facial muscle. The forth week was marked by regression of the ophthalmoplegia and facial paralysis. Deglutition disorders diminished the beginning of the fifth week. After 45 days from admission in ICU, The patient was admitted in a general Neurology ward for additional support.

Case N2
A 12 years old boy, with no medical history, presented to our emergency department for acute diplopia, dysphonia, facial paralysis, and swallowing troubles. Evaluation of his vital signs revealed pyrexia (T: 37°C), heart rate 80 pulse/min, blood pressure at 110/70 mmHg, tachypnea at 39 breaths/min, pulse oximetry 60% on ambient air. Neurological exam finds a confused patient with altered mental status (Glasgow coma score GCS at 13/15), ophthalmoplegia with ptosis, cranial nerve examination showed bilateral ptosis and total external and internal ophthalmoplegia, with total facial bilateral and symmetrical...
paralysis. On motor system examination, strength in triceps, biceps, and supinator was 2/5; deltoids and pronator was 2/5; wrist extendors and flexors were 3/5 bilaterally; hip flexors, quadriceps, and hamstrings were 1/5 bilaterally; dorsiflexors and plantar flexors were 1/5 bilaterally. Her sensation was intact.

The patient was put in mechanical ventilation due to respiratory distress.

Blood tests did not reveal any infectious process. Thyroid test were normal. We realized an MRI that showed no abnormalities. Her Cerebrospinal fluid analysis (CSF) showed no albuminocytological dissociation.

As the patient showed Fisher/Guillain-Barréoverlapping syndrome.

After receiving 5-day course of IVIG (0.4 g/kg/day), the evolution was favorable and the patient was extubated and transferred to neurological department.

**Discussion:**

Guillain–Barré syndrome is an acute paralytic inflammatory demyelinating polyneuropathy. It represents the first cause of acute and extensive paralysis in the world with about 100000 people developing the disorder every year worldwide. Most studies that estimate incidence rates of Guillain Barré syndrome showed a similar range of 0.8–1.9 (median 1.1) cases per 100000 people per year [2,3]. Guillain–Barré syndrome is a typical post-infectious disorder. Two-thirds of adult patients report preceding symptoms of a respiratory or gastrointestinal tract infection within 4 weeks of onset of weakness [4]. Guillain barre can reach all ages but seems to increase with age and to touch more male than females [5,6]. The typical presentation of guillain barré syndrome is a rapidly progressive ascending bilateral weakness that usually starts in distal lower extremities.

During the progressive phase, 20–30% of patients develop respiratory failure and need artificial ventilation at an Intensive care unit as in our case [5]. GBS syndrome has different forms of presentation and variants. Miller Fisher syndrome is characterized by the triad of ophthalmoplegia, ataxia, and areflexia. In practice, Miller Fisher syndrome is frequently accompanied by other cranial nerve involvement and can progress to weakness of the limbs [6]. Guillain–Barré syndrome (GBS) overlaps with Miller Fisher syndrome (MFS) in about 5% of cases of MFS [5]. K Funakoshi et al had shown in their study that FS/GBS patients significantly needed mechanical ventilation more often. Such patients showing titubation and descending tetraparesis need to be carefully monitored as the illness progresses because those clinical features are helpful predictors of respiratory failure [6] (8). Our case of FS/GBS also needed ventilation in the first week of onset.

Guillain–Barré syndrome is a potentially life-threatening disease. Both general medical care and immunological treatment are essential. Metiliculous surveillance and supportive care are the main keys to prevent complications. A Respiratory and hemodynamic monitoring in an Intensive care unit might be needed. Among the other issues that need attention are prophylaxis for deep vein thrombosis, management of possible bladder and bowel dysfunction, early initiation of physiotherapy and rehabilitation, and psycho social support.

Immunotherapy and plasma exchange are now the main therapies that have proved effective in GBS. Whether the total IVlg dose (2 g/kg bodyweight) given in 2 days (1 g/kg per day) is more beneficial than when given in 5 days (0.4 g/kg per day) is not known.

Oral steroids and intravenous methylprednisolone are not beneficial in the disorder [10]. In our centre we used the 5 day course (0.4 g/kg per day) that we repeated after 2 weeks and showed significant improvement after each cure.

Although IVlg and plasma exchange have proved effective, 20% of patients with Guillain-Barré syndrome still develop severe weakness and have a long disease course, often with incomplete recovery, pain, and fatigue [11,12]. Mortality rates in Europe and North America vary between 3% and 7% [13,14]. A better early recognition system and more efficient treatments are therefore needed.

**Bibliography**