Miller-Fisher Syndrome: A Variant of Guillain-Barre Syndrome

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Guillain-Barre syndrome (GBS) is a rare neurological condition affecting all ages. It is rare in pediatric population; the disease is preceded by viral or bacterial infection. GBS present as a rapid onset ascending paralysis with pain and sensory impairment.

Miller Fisher syndrome is a rare but most recognized variant of GBS; it presents by the classical triad of symptoms: ataxic gait, absence of reflexes and ophthalmoplegia.

A healthy six-year-old boy, not known to have any medical illnesses, presented with the classical triad of symptoms after ten days’ history of gastroenteritis. Ganglioside GQ1B (IgG, IgM) antibodies were markedly elevated and the patient was started immediately on intravenous immunoglobulin (IVIG). After a few days, the patient showed marked improvement in gait with no further deterioration in clinical condition.

THE CASE

A previously healthy six-year-old boy presented with blurred vision and unsteady gait for two days.

The patient had a history of gastroenteritis for 10 days before his symptoms appeared. Physical examination showed a cooperative child with normal vital signs, restricted extraocular eye movement with reduced power over the lower extremities, absent reflexes in all limbs, and downgoing plantar response bilaterally. His gait was ataxic. Tone and other cranial nerves examination were normal.

Hematological and biochemistry blood test were normal. Stool culture showed Campylobacter. Lumbar puncture for CSF study showed normal hematological and biochemical variables. MRI brain and MRI spine were normal. Ganglioside GQ1B (IgG, IgM) antibodies revealed highly elevated (428%) AK-ratio (above 30 is positive) which is highly suggestive of Miller-Fisher syndrome.

Miller-Fisher syndrome, a variant of GBS was diagnosed and the patient was given two doses of IVIG.

The patient showed a positive response to medication without any further deterioration. His ataxic gait markedly improved; however, the patient fixed gaze and areflexia did not improve. The patient was discharged in a stable condition.

Three months after his initial treatment with IVIG, the patient showed full recovery of his extraocular eye movement; however, his reflexes were still absent.

The repeat ganglioside GQ1B (IgG, IgM) antibodies three-months later reduced from 103% AK-ratio to 428% AK-ratio.

DISCUSSION

Guillain-Barre syndrome (GBS) is the most common cause of acute ascending paralysis among the pediatric population. GBS is more common in males, and affect 1.1 to 1.8 per 100,000 persons per year. Most cases of GBS are preceded by a gastrointestinal infection which triggers acute immune system attacking peripheral nerves causing demyelinating polyneuropathy. GBS in children typically presents with symmetrical ascending weakness, areflexia and neuropathic pain along with sensory involvement.

Miller Fisher syndrome is a very rare variant of Guillain-Barre syndrome among the pediatric population. According to Snyder et al, the Miller-Fisher variant of GBS has classical features of ophthalmoplegia, areflexia and ataxia; similar findings were seen in our patient who presented with restricted eye movement, areflexia and unsteady gait.

Snyder et al found that the majority of patients (approximately 80%) who present with the Miller-Fisher variant of GBS showed positive serological test of anti GQ1B antibodies. Our patient showed very high levels of anti-GQ1B antibodies which confirmed the diagnosis of Miller-Fisher syndrome.

Our patient also showed positive stool culture of campylobacter jejune which is an important association in patients with GBS. According to Nyati et al, campylobacter jejune is a major infectious agent in the pathogenesis of GBS.

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Van Doorn et al found that IVIG is an effective therapeutic treatment in the management of GBS. Based on the clinical findings of our patient which was strongly suggestive of the Miller-Fisher variant of GBS, the patient was started on IVIG on the same day of his clinical presentation.

Our patient showed significant improvement in his clinical symptoms with a normal gait and no further deterioration after IVIG treatment. Follow-up after three months revealed full recovery of his eye movement.

According to Ryan, early recognition and treatment of pediatric GBS decrease the long-term morbidity and mortality.

**CONCLUSION**

Although Miller-Fisher is rare, it is important to diagnose it early and initiate the appropriate treatment with IVIG to prevent further deterioration and long-term disability.

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**REFERENCES**