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Variant of Guillain-Barré Syndrome

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Dear Editor,

A 13-year-old previously healthy boy experienced an upper respiratory tract infection one week prior to his presentation, returning from a school trip to Paris.

On admission, neuro-ophthalmological examination revealed symmetrically mydriatic pupils, unresponsive to light and near stimuli. Accommodation was impaired. Ocular movements of both eyes were limited in all directions. Funduscopic examination revealed normal findings. Systemic neurological examination showed that the strength of the facial muscles as well as the upper and the lower limbs muscles was normal. Ataxic gait was present. Deep tendon reflexes were absent with bilateral flexor plantar responses. There was no evidence on history or examination of involvement of sensory, respiratory, and autonomic systems of sphincter functions. Nasal dysarthria was noted on examination.

Motor and sensory nerve conduction studies were within normal limits. The blink response testing was normal and suggested intact bilateral afferent trigeminal nerve and efferent facial nerve. The results of routine blood chemistry tests, anti-HSV, as well as a panel of serology tests of autoimmune disorders were unremarkable. PCR screen of peripheral blood for Meningococcal DNA was negative. The cerebrospinal fluid (CSF) analysis revealed clear CSF, normal pressure, and no blood cells, normal glucose and protein levels. CSF culture and PCR for possible organisms, such as bacteria, mycobacterium tuberculosis, herpes viruses, yielded negative results. Electrocardiogram and chest X-ray were normal. CT brain and MRI brain and spine were normal. Stool analysis for *Campylobacter jejuni* was negative. CSF testing for anti-GQ1b and GM1 antibodies was negative.

Based on the clinical presentation Miller Fisher variant of Guillain-Barré Syndrome was the final diagnosis and intravenous immunoglobulin therapy was immediately initiated. The patient was treated with 0.4 g/kg/day IVIG over 5 days. The neurological symptoms dramatically resolved four days after the treatment. The patient was followed up in our outpatient clinic and one month after his hospitalisation he became symptom-free. During six months of observation no relapses were observed.

Miller Fisher syndrome is a rare, acquired nerve disease that is considered to be a variant of Guillain-Barré syndrome. It is characterized by abnormal muscle coordination, paralysis of the eye muscles, and absence of the tendon reflexes. Symptoms may be preceded by a viral illness.

A distinctive feature of our patient was the absence of anti-GQ1b antibody. Up to 95% of the patients have anti-GQ1b antibodies during the acute phase of MFS, owing to the high specificity and sensitivity, these antibodies are useful to confirm the diagnosis; however, a negative result does not exclude the diagnosis.
Because of no difference in the treatment response between the patients with anti-GQ1b antibody positive and negative, IVIG was initiated immediately with a favourable outcome. We believe that very immediate initiation of IVIG, when only mild immunological injury to the axon occurs, can yield a favourable outcome.

In conclusion, in the case of acute bilateral mydriasis, associated with ophthalmoplegia, diplopia, areflexia and ataxia, which poses a diagnostic challenge to the clinician, the diagnosis of Miller Fisher syndrome should be considered even in the absence of CSF/serum anti-GQ1b antibody.

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