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An Overlapping Case of Miller Fisher Syndrome and the Pharyngeal-Cervical-Brachial Variant of Guillain-Barré Syndrome

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ABSTRACT

A 55-year-old Caucasian male presented initially to the emergency room (ER) reporting myalgia, chills and fever. Physical examination and laboratory tests were unremarkable and he was discharged with symptomatic care. He returned to our ER 2 weeks later reporting dizziness, loss of balance, blurred vision, mild dysarthria and bilateral hand paraesthesia. On examination, he presented complete bilateral ophthalmoplegia, mild dysarthria, left finger-to-nose dysmetria, ataxia, areflexia and bilateral hand hypoaesthesia without fever. Blood tests and head computed tomography were normal. The patient was admitted to the Internal Medicine department. On the second day on the ward, the patient presented dysphagia. A head magnetic resonance angiogram showed no signs of ischaemia or vascular disease and a lumbar puncture was performed but no pleocytosis, albumin-cytological dissociation or hypoglycorrhachia was present. Despite the normal results we suspected a Guillain-Barré syndrome variant, and started treatment with intravenous immunoglobulin (IVIG) at a dose of 400 mg per kilogram which continued for 5 days with immediate neurological improvement. We present a rare overlapping case of Miller Fisher syndrome and the pharyngeal-cervical-brachial variant of Guillain-Barré syndrome.

LEARNING POINTS

- The overlap of Miller Fisher syndrome and pharyngeal-cervical-brachial variants of Guillain-Barré syndrome are rarely described in the literature but should be considered when multiple cranial nerves are involved with normal neuroimaging results, even with normal cerebrospinal fluid analysis.
- Due to similar clinical presentation, a misdiagnosis of cerebral ischaemia, botulism or ocular myasthenia gravis can delay treatment and put patients at risk.
- In its natural history, this syndrome evolves to respiratory arrest and death, but with accurate diagnosis and prompt treatment, the prognosis improves considerably.

KEYWORDS

Miller Fisher, Guillain-Barré syndrome, pharyngeal-cervical-brachial variant

CASE DESCRIPTION

We report a case of a 55-year-old Caucasian male, without a previous relevant medical record. He presented initially to the emergency room (ER) reporting myalgia, chills and fever (38.5°C) since 4 days previously. Physical examination and laboratory tests were unremarkable and he was discharged with a prescription of rest, ibuprofen and acetaminophen. He returned to our ER 2 weeks later reporting dizziness, loss of balance, blurred vision, mild dysarthria and bilateral hand paraesthesia in the previous 2 days. On examination, he presented complete bilateral ophthalmoplegia, mild dysarthria, left finger-to-nose dysmetria, ataxia, areflexia and bilateral hand hypoaesthesia without fever. Blood tests – including a complete blood count, sedimentation velocity, urea and creatinine, liver enzymes, folic acid, vitamin B12 and

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coagulation tests – were within normal ranges. Head computed tomography (CT) was reported as normal. The patient was admitted to the Internal Medicine department with a suspicion of a posterior cerebral stroke. On the second day, he presented with moderate dysphagia, only tolerating honey-thick liquids. A head magnetic resonance angiogram showed no signs of ischaemia or vascular disease. A lumbar puncture was performed but no pleocytosis, albumin-cytological dissociation or hypoglycorrhachia was present. Serology tests for human immunodeficiency virus, syphilis, cytomegalovirus and the Epstein-Barr virus were negative. Despite these normal results, we suspected a Guillain-Barré syndrome (GBS) variant, and started treatment with intravenous immunoglobulin (IVIG) at a dose of 400 mg per kilogram which continued for 5 days with immediate neurological improvement. On the third day of treatment, neurologic deficits were limited to maintaining difficulty in full abduction of both eyes. The patient started motor rehabilitation while on the ward and was discharged on day 10 to our Internal Medicine outpatient clinic. The patient was reevaluated in our outpatient clinic 2, 4 and 12 weeks after discharge. Currently, he has resumed his professional activity as a gardener and reports no deficits. The neurological examination was normalized.

DISCUSSION

GBS, Miller Fisher syndrome (MFS) and the pharyngeal-cervical-brachial (PCB) variant are thought to result from an aberrant acute autoimmune response to a preceding infection. Approximately two-thirds of cases have prodromic symptoms of upper respiratory tract infection or diarrhoea, and approximately 50% develop following an infection; in our case, we suspected a prior viral infection [1].

MFS is mostly associated with dysfunction of the third, fourth and sixth cranial nerves, but involvement of almost all other cranial nerves has been documented[2]. Patients with the PCB variant of GBS typically present with rapidly progressive oropharyngeal and cervicobrachial weakness associated with areflexia in the upper limbs[3]. In the case presented, only an overlap between these 2 entities would correlate with our patient's symptoms and signs.

Antibodies against the GQ1b ganglioside are a typical serological finding.

The worldwide incidence of GBS is approximately 1 to 2 in 100,000, with MFS representing a tiny subset of the cases (1 to 2 in 1,000,000). There are only rare descriptions of the PCB variant in the literature[4].

Albumin-cytological dissociation or a combination of a normal cell count and raised protein levels in the cerebrospinal fluid (CSF) are a strong feature in diagnosing GBS, MFS and PCB but up to 50% of patients show an unremarkable CSF.

As this case underlines, when multiple cranial nerves are involved with normal head CT and normal CSF analysis, the diagnosis can be challenging in the early stages of disease, but a detailed clinical examination narrows the differential diagnosis significantly. The gradual onset of GBS, MFS and PCB is a key factor in distinguishing these from acute brainstem stroke, and signs of ischaemia will appear in subsequent neuroimaging studies. Fluctuations of symptoms during the course of the day or fatigability of ocular muscles are more suggestive of ocular myasthenia gravis. The ice test can be performed – an ice pack is placed over a patient's closed eyelids for several minutes; if the ice pack relieves symptoms such as eyelid drooping or trouble with eye movement, it is an indication of ocular myasthenia gravis. Botulism is characterized by marked fatigue, weakness and vertigo, usually followed by blurred vision, dry mouth and difficulty in swallowing and speaking but can usually be excluded by the presence of sensory deficits or areflexia typically present in GBS, MFS and PCB[5].

The treatment is based on adequate supportive care with pain control and respiratory support. IVIG or plasmapheresis are both effective treatments. With correct treatment, the prognosis is usually good with case fatalities less than 5%. Although recurrence may occur, it is uncommon.

Our patient had a rarely described overlap syndrome, with the diagnostic work-up all within normal range, reinforcing anamnesis and physical examination as the eternal gold standard for all internists.

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