

Title: A case of Miller fisher's Syndrome presenting with dysphagia and nasal voice.

Leila Tamaoui¹, Mounia Rahmani¹, Hajar TOUATI¹, Leila Errguig², Maria Benabdeljlil¹, Saadia Aidi¹

1. Department of Neurology A and Neuropsychology
2. Department of Clinical Neurophysiology

Hôpital des Spécialités, IBN SINA University Hospital, Mohammed V University, RABAT, Morocco.

Correspondence: tamaouileila@gmail.com

Abstract: Miller Fisher Syndrome (MFS) is a rare variant of Guillain-Barré Syndrome (GBS). It is largely a clinical diagnosis based on the classical features of ataxia, areflexia, and ophthalmoplegia. Its clinical evolution is most often favorable. However, other neurological signs and symptoms may also be present. Supportive laboratory studies (positivity of antibodies, CSF albumin-cytological dissociation and nerve conduction studies) are useful especially in uncommon presentations. We report a case of a 74-year-old patient who exhibited dysphonia and difficulty to swallowing previously to the classic triad of ataxia, areflexia, and ophthalmoplegia, characteristic of MFS. CSF analysis demonstrates an albumin-cytological dissociation but anti-GQ1b antibody were negative. The patient has spontaneously and completely recovered after several weeks.

Key words: Miller Fisher Syndrome, Guillain Barré Syndrome variant, Dysphonia, Dysphagia.

1. Introduction:

Miller Fisher's syndrome (MFS) is a rare variant of Guillain Barré Syndrome (GBS) characterized by the acute onset of ataxia, ophthalmoparesis, and areflexia. The appearance of symptoms following an infection of the respiratory or digestive tract is a common ground for all GBS variants. The diagnosis of MFS is essentially clinical and it's mostly supported by serum positivity for anti-ganglioside antibodies [1]. Others supportives laboratories findings include CSF albumin-cytological dissociation and presence of sensitive abnormalities in nerve conduction studies. Currently, there are no consensus diagnostic criteria [2]. In addition to the classical form, the spectrum of MFS includes incomplete and extensive forms, with additional central clinical features such as the Bickerstaff rhombencephalitis phenotype or peripheral symptomatology suggesting overlap with classical GBS or its other variants [3]. The oculopharyngeal subtype of GBS, also known as polyneuritis cranialis is one of the subtypes of GBS which shares clinical findings with MFS [4]. Our patient illustrates an overlapping syndrome between this GBS variant and MFS given the involvement of bulbar cranial nerves responsible for difficulty of swallowing and dysphonia.

Case presentation:

A 74-year-old hypertensive and diabetic man presented to our emergency department for acute onset over 5 days of right ptosis, dysphonia with nasal voice, difficulty swallowing and horizontal diplopia. Few days later, he noted unsteady gait with dizziness sensation without nausea or vomiting. No evidence of previous infectious episode was found. His clinical examination found a complete bilateral ophthalmoplegia associated with a bilateral ptosis predominant on the right, a static ataxia not increased by the closure of the eyes, a diffuse osteotendinous areflexia, and vibration sense was decreased in the four extremities. There was involvement of the bulbar cranial nerves (IX & X) paresis of the soft palate, abolition of the nauseated reflex, dysphonia (with nasal voice) and dysphagia. The remainder of the examination was normal. Brain magnetic resonance imaging (MRI) revealed no abnormalities. Cerebrospinal fluid (CSF) analysis showed cytoalbuminologic dissociation with normal cells and elevated CSF protein of 1g/dl, and normal glucose levels. Electroneuromyographic examination was normal in the four limbs and in the facial nerve: sensory and motor nerve studies, F waves, H reflex,

and repetitive nerve stimulation. AntiGQ1B antibodies were negatives but anti GT1a were positives. The usual biological assessment (blood count-platelet count, blood crase, blood ionogram, renal and hepatic functions...), thyroid assessment returned normal. Viral serologies (syphilitis, hepatitis A, B, C, HIV) were negatives.

The evolution was marked by spontaneous rapidly favorable improvement, didn't require the use of polyvalent immunoglobulin, with complete resolution of symptoms in several weeks.

Discussion:

Our patient has presented with unusual initial symptoms of MFS, that were the nasal voice and dysphagia, previously to the classic triad. This situation may be challenging for the clinician and lead to additional investigations to narrow the differential diagnosis.

According to their clinical features, GBS and its variant MFS can be classified into different subtypes which together form a continuous spectrum of several overlapping syndromes. The involvement of cranial nerves other than the typical oculomotor ones, seems to be more frequent in the forms of MFS-GBS overlap: facial nerve in 30 to 50%, glossopharyngeal (IX), vagus (X), large hypoglossus (XII), responsible for disorders of swallowing and dysphonia, in 13% of cases [5,6].

The nasal voice related to palate paresis has been rarely described in MFS [7,8]. Also, dysphagia was found a less frequent sign (2%) in a clinical series of 50 Japanese patients [9].

The classic albumino-cytological dissociation of CSF is common, such as in our patient [1]. The electroneuromyographic studies have a more limited role in Miller Fisher's syndrome. They may be normal in true Miller Fisher's syndrome, similarly to our patient. However, mild abnormalities in sensory nerve conduction studies and in the blink reflex are often seen [2].

Antibodies against the GQ1b ganglioside are a typical serological finding but were absent in our patient. GQ1b-seronegative is reported in a minority of patients (approximately 10 to 17%), and may be related to an insufficient sensitivity method of detection or to the pathogenicity of antibodies against gangliosides other than GQ1b [7]. The anti GT1a antibodies often coexist with anti GQ1B, due to their cross-reactivity [1,10,11]. In our case, the positivity of anti GT1a may be of particular interest as they are commonly detected in pharyngeal-cervical-brachial variant of GBS patients and are associated with bulbar palsy in up to 70% of GBS patients [12, 13].

MFS is almost always a self-limited disorder with good prognosis and spontaneous recovery. Immunotherapy is not required unless there are features of an overlap syndrome with GBS variant. In patients with severe Miller Fisher syndrome who have swallowing and respiratory difficulties, IVIG should be considered despite lack of supporting evidence of benefit [1]. In our patient, the evolution was spontaneously and rapidly favorable.

Conclusion:

MFS, a variant of GBS, is a rare condition. It combines the typical clinical triad, acute ophthalmoplegia, ataxia and areflexia with an albumino-cytological dissociation of the CSF and an anti-GQ1b seropositivity. Rare extensive clinical forms with bulbar cranial nerves involvement are sometimes described. Our case presented particular symptoms with nasal voice and dysphagia, and suggests that an overlapping syndrome with polyneuritis cranialis variant of GBS might be considered.

Declaration of Conflicting Interests:

On behalf of all authors, the corresponding author states that there is no conflict of interest.

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