CASE REPORT

The Differential Diagnosis of Acute Onset Truncal Ataxia: The Importance of Dysgeusia in Miller Fisher Syndrome

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Abstract:
Miller Fisher syndrome (MFS) can be difficult to diagnose, particularly in mild cases where some of the standard triad of symptoms (external ophthalmoplegia, ataxia, and loss of deep tendon reflex) are absent. We herein report a case of the incomplete form of MFS diagnosed in a 54-year-old Japanese man who presented only with ataxia symptoms and was positive for the anti-GQ1b antibody. However, the patient also suffered from dysgeusia, a significant impairment of taste perception. We propose that dysgeusia in acute-onset ataxia cases may constitute an important clinical feature to aid in the diagnosis of the incomplete form of MFS.

Key words: Dysgeusia, Taste impairment, Ataxia, Miller Fisher syndrome, GQ1b, Differential diagnosis


Introduction

Miller Fisher syndrome (MFS) is typically characterized by a triad of clinical symptoms (external ophthalmoplegia, ataxia, and loss of deep tendon reflex [DTR]) (1) and the presence of immunoglobulin G (IgG) antibodies against GQ1b gangliosides (2). The incomplete form of MFS, however, is characterized by acute ophthalmoparesis, acute ataxic neuropathy (AAN), acute ptosis, and acute mydriasis (2). Anti-GQ1b antibodies are also found in Bickerstaff brainstem encephalitis and in acute ophthalmoplegia without ataxia, which are collectively referred to as “anti-GQ1b antibody syndrome” (3). A growing body of evidence suggests that dysgeusia may represent an additional neurological symptom of MFS (1, 4-9). However, whether or not dysgeusia is also observed in cases of the incomplete form of MFS that present without all three triad symptoms is unclear.

We herein report a rare case of truncal ataxia with dysgeusia, due to AAN, that did not present with the other two triad symptoms. The present case suggests that dysgeusia may be useful for the differential diagnosis of MFS from other ataxia-related diseases. The patient provided his written informed consent for the publication of this report.

Case Report

A 54-year-old Japanese man was admitted to our department for the evaluation of gait disturbance, dysgeusia, and dysarthria, in addition to numbness in all four limbs and in the perioral area. Twelve days before the admission, he had nasal discharge and sore throat symptoms, indicating an upper respiratory infection. The patient also experienced gait unsteadiness seven days prior to admission and dysarthria accompanied by numbness in all four limbs and in the perioral area six days before admission. Furthermore, he could not perceive the salty taste of miso soup five days prior to admission. His medical history included postoperative mitral regurgitation and the insertion of a prosthetic right eye following a car accident. He did not drink alcohol and had no history of suspected toxic exposure to metals or solvents.

On admission, his vital signs were unremarkable. His Glasgow coma scale score was 15 (E4V5M6). A physical examination showed no lesions of the oral mucosa. A neurological examination revealed a wide-based gait, a tandem stance impairment, slightly slurred speech, and hypesthesia for cold sensation in his distal limbs. The left pupil was 3 mm in diameter with a prompt reactivity to light. The left extraocular movement was normal. There were no indica-
tions of facial nerve palsy. Both the DTR and deep sense of the limbs were normal, as was the jaw reflex. The bilateral plantar responses were flexor. There were no signs of limb weakness or limb ataxia. Although he could taste sweet and bitter foods normally, he found that oranges tasted bitter. Furthermore, he complained of a reduced ability to taste salty foods.

Laboratory evaluations showed normal levels of vitamin B1 (37 ng/ml), vitamin B12 (537 pg/ml), folate (7.2 ng/ml), total homocysteine (9.5 nmol/ml), blood glucose (107 mg/dl), hemoglobin A1c (5.7%), and Zn (90.0 pg/ml). Anti-SS-A/Ro and anti-SS-B/La antibody tests were all negative. A pharyngeal culture revealed *Haemophilus species*. A cerebrospinal fluid (CSF) analysis revealed normal cell counts (3 cells/μl), a total protein level of 24 mg/dl, and a glucose level of 62 mg/dl, with a concomitant blood glucose level of 107 mg/dl and a normal IgG index (0.63). Oligoclonal bands were negative. The Schirmer test, the Rose Bengal test, and gum test were all negative. Motor and sensory nerve conduction evaluations showed no abnormalities, including the minimal F-wave latency. Brain magnetic resonance imaging (MRI) findings was normal. Based on these results, we hypothesized that acute onset ataxia and peripheral neuropathy, resulting from AAN, were the likely causes of the clinical portrait of the patient.

After admission, we carefully monitored the patients’ neurological symptoms; however, no immunotherapy was prescribed due to his mild symptoms. On day three after admission, the dysarthria and the taste impairment disappeared. On day seven, both the truncal ataxia and the limb and perioral numbness were found to have improved. He was discharged on day 14 after admission. The patient was never treated with immunotherapy because follow-up assessments revealed that the ataxia never deteriorated, that there was no loss of DTR, and that eye movements were not restricted. On day 39 after the onset, enzyme-linked immunosorbent assays taken from a specimen at admission showed no IgG or IgM antibodies reacting with gangliosides GM1, GM2, GM3, GD1a, GD1b, GD3, GT1b, galactocerebroside, GalNAc-GD1a, or GD1a/GD1b. However, IgG reacting with GQ1b and GT1a were positive. We therefore finally diagnosed the patient with AAN.

**Discussion**

This case suggests that dysgeusia in acute-onset ataxia may constitute an important clinical feature to assist in the diagnosis of the AAN presenting without DTR abnormalities or external ophthalmoplegia. Midline cerebellar lesions are known to cause truncal ataxia (10). The differential diagnosis of sporadic ataxia cases that include truncal ataxia can be complicated, given the wide variety of diseases and conditions that present with similar symptoms: chronic alcohol use, exposure to toxic agents, immune-mediated inflammation (paraneoplastic cerebellar degeneration, anti-glutamic acid decarboxylase antibodies ataxia, Hashimoto encephalo-
important clinical feature to aid in the diagnosis of MFS, even when the diagnosis is challenging, such as in cases of AAN.

Dysgeusia in GBS is believed to result from a lesion of the chorda tympani branch of the facial nerve or from the involvement of an accessory taste pathway through the trigeminal nerve (20). In MFS, Uchibori et al. speculated that anti-GQ1b antibodies may affect the peripheral nerves associated with taste and taste buds (5). In the present case, dysgeusia improved quickly, likely because the taste receptors turnover in as little as 10 days (6). Alternatively, an accessory taste pathway through the trigeminal nerve may also be involved in dysgeusia, as our patient showed perioral numbness without facial nerve palsy.

In conclusion, dysgeusia in acute-onset ataxia may constitute an important clinical feature to aid in the diagnosis of AAN, an incomplete form of MFS. Physicians should investigate potential complaints of dysgeusia in patients with acute-onset ataxia.

The authors state that they have no Conflict of Interest (COI).

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References
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<th>Table: Clinical Features of Patients with Dysgeusia Due to Miller-Fisher Syndrome.</th>
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<td>Case</td>
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<td>Fisher M, 1956</td>
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<td>Uchibori A et al., 2006</td>
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<td>Dorr J et al., 2006</td>
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<td>Ohe Y et al., 2012</td>
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<td>Yagishita IV et al., 2015</td>
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<td>Yamamoto D et al., 2016</td>
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<td>F: female, M: male, EO: external ophthalmoplegia, DTR: deep tendon reflex, FNP: facial nerve palsy, URI: upper respiratory infection, IVg: intravenous immunoglobulin, NR: not reported</td>
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**Clinical Features of Patients with Dysgeusia Due to Miller-Fisher Syndrome.**

**Age**

**Sex**

**ED**

**Case**

**Antecedent infection**

**FNP**

**Anticardiolipin antibody**

**Neuropathy**

**Numbness**

**Facial nerve palsy**

**Limb and truncal ataxia**

**Truncal ataxia**

**Deep tendon reflexes**

**Taste impairment**

**Immunoglobulins**

**References**

11. Ogawa K, Suzuki Y, Oishi M, Kamei S. Clinical study of 46 pa-

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