CASE REPORTS

# MILLER FISHER SYNDROME PRESENTING AS LINGUAL NEUROPATHY

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**Abstract**

Miller Fisher Syndrome typically presents as a triad of ophthalmoplegia, ataxia and areflexia. We report on a previously healthy 14 year-old female who presented with 3 -day history of lingual neuropathy followed by facial diplegia. Three days after hospital admission she had ataxia and areflexia. Cerebrospinal fluid (CSF) protein was high. Patient was diagnosed with Miller Fisher Syndrome even in the absence of ophthalmoplegia. Subsequently, she was treated with intravenous immunoglobulin. Complete recovery was seen in 6-month follow up. To our knowledge, this is the first case of Miller Fisher Syndrome presenting as lingual neuropathy.

**Keywords:** Lingual neuropathy, facial diplegia, Miller Fisher Syndrome.

## Introduction

Miller Fisher Syndrome typically presents as a triad of ophthalmoplegia, ataxia andareflexia. It was first described by Charles Miller Fisher in 1956 (1). In spite of this classical triad of presentation, Miller Fisher Syndrome may present with just one or two of them (2). It is considered as a variant of Guillain Barre Syndrome. In fact, many authors consider GuillainBarre Syndrome, Miller Fisher Syndrome and Bickerstaff's brainstem encephalitis as a spectrum of inflammatory polyneuropathy sharing manysimilarities (2).Isolated lingual neuropathy is unusual presentation of neurological disorders. It is mainly seen as a complication of dental procedures, mandibular blocking anesthesia and local maxillofacial pathology (3). It could also be a part of trigeminal neuralgia of different etiologies. (4) Isolated lingual neuropathy as a presenting feature of Miller Fisher Syndrome has not been reported in this context before.

## Case Report

A previously healthy 14 -year old girl presented with 3 day- history of tongue tingling with no taste impairment. This was followed by bilateral facial weakness manifesting as inability to smile, close her eyes tight or even hold water in mouth while brushing her teeth. She also had subjective fever with transient skin rash one week before presentation. There was no history of altered mental status, vomiting, visual changes or hearing abnormality. On initial examination, her vital signs were normal. Neurological examination revealed normal mini mental status. There was facial diplegia and bilaterally decreased pin prick and touch sensation in the anterior two thirds of the tongue. Taste sensation was intact. Pupillary light reflexes and extra ocular muscles were intact with no diplopia. Other cranial nerves, motor, sensory, deep tendon reflexes, cerebellar and gait exams were normal. Initial investigations showed normal blood count, blood glucose, electrolytes, calcium and magnesium. Urine toxic screen, erythrocyte sedimentation rate, thyroid stimulating hormone, thyroxin, glycosylated hemoglobin, creatinine phosphokinase and rheumatoid factor were normal. Blink reflex study showed absent response bilaterally. Brain MRI was unremarkable. Three days after admission, the patient's gait became significantly ataxic with areflexia. Muscle strength remained normal. Based on the clinical picture of ataxia and areflexia even in the absence of ophthalmoplegia, our patient was diagnosed with Miller Fisher Syndrome. Cerebrospinal fluid (CSF) analysis showed high protein of 204mg/dl (normal 20-40mg/dl) with normal cell count and glucose level. Bacterial culture was negative. Both CSF and blood lyme antibodies and angiotensin converting enzyme (ACE) were negative. CSF for Epstein-Barr virus panel, herpes zoster and cytomegalovirus were negative too. Anti-acetylcholine receptors antibodies were negative. CSF for antiGQ1b antibodies was not performed due to a technical problem. Nerve conduction velocity and electromyogram of the lower extremities were unremarkable. Negative inspiratory force was -41. Patient was treated with 2 gm/kg of intravenous immunoglobulin over 3 days. Mild clinical improvement was noticed after 1 month. However, complete recovery of her signs and symptoms was seen in 6 month- follow up.

## Discussion

The lingual nerve is a branch of the mandibular division of the trigeminal nerve that provides a general sensation to the anterior two thirds of the tongue. The chorda tympani, which is a branch of the facial nerve, joins the lingual nerve and carries the taste sensation from the anterior two thirds of the tongue to the nucleus solitarius. The preservation of taste and the restricted sensory deficit to the anterior two thirds of the tongue bilaterally localize the lesion to the lingual nerves in our patient. Lingual neuropathy is mainly seen as a complication of lower third molar surgical extraction, after anesthetic nerve block in dental procedures, entrapment of the nerve by lateral pterygoid muscle (5), temporomandibular joint disk displacement (6), compression from prone position during anesthesia (7), laryngoscopy and endotracheal intubation (8) and tumors of the oral cavity or after their excision (9). It could also be seen as a part of trigeminal neuropathy of different causes (4).While proceeding with the investigation toward excluding other possible causes of facial diplegia in our patient such as sarcoidosis, lyme disease, viral infection, infiltrative diseases and brain lesion mainly in the brainstem and at the cerebellopontine angle, the patient developed significant ataxia and areflexia suggestive of Miller Fischer Syndrome. Although missing in our patient, ophthalmoplegia is the most common presentation. About 65% present with diplopia as the ocular motor cranial nerves III, IV and VI have high concentration of GQ1b antigen (10). Abducens nerve palsy is the usual cause of diplopia and to a lesser degree oculomotor deficit. Ataxia is another important presenting symptom ranging from mild gait disturbance

to inability to walk independently. It might be due to proprioceptive impairment or cerebellar origin (11). Ataxia and ophthalmoplegia usually recover within 6 months. Areflexia is the third sign of the triad though it is a cardinal feature in Guillain Barre Syndrome. Some patients may have intact or even exaggerated deep tendon reflexes early in the disease course as in our patient. Other rare manifestation of Miller Fisher Syndrome could include dysarthria, hyper-nasalspeech (12), dysphagia, urinary retention, optic neuritis (13), headache (14) andautonomic dysfunction (15). By the second to the third week, most patients have significantly high protein concentration. AntiGQ1b antibodies are very helpful and are usually high in 80-90% of cases. GQ1b is a surface component that is concentrated in the ocular motor nerves which could account for the unique ophthalmic manifestations in Miller Fisher Syndrome. Though, it could be elevated in Guillain Barre Syndrome and Bickerstaff's brainstem encephalitis (16).

Treatment of Miller Fisher Syndrome depends on the severity of clinical course though spontaneous recovery is the rule. There are no randomized control trials regarding treatment of Miller Fisher Syndrome. However, most institutions apply the same protocols for Guillain Barre Syndrome which includes intravenous immunoglobulin 2g/kg in 2-5 days and or plasmapheresis (16). Many cases had been watched with no treatment (17). The long term outcome seems not to be altered with this treatment presumably due to the benign nature of this disease.

In conclusion, in spite of the classic clinical triad of presentation, Miller Fisher Syndrome should be considered in the presence of other atypical features like lingual neuropathy, as early diagnosis and treatment may halt the progression of the this inflammatory process.

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