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Lingual Hyperkinesia as An Initial Manifestation of Wernicke's Encephalopathy: Evidence for Localization of Involuntary Hyperkinetic Movement of the Tongue

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Abstract

Background: Movement disorders caused by Wernicke's encephalopathy (WE) are very rarely reported, and involuntary hyperkinesia of the tongue as an initial manifestation of WE have not yet been reported. The study aimed to investigate the path mechanism and localization of involuntary lingual movement symptoms.

Methods: We present a patient with lingual hyperkinesia as an initial manifestation of WE, who had lesions of the periaqueductal area of the midbrain tectum and bilaterally medial thalami in Brain MRI. Patient's lingual symptoms were observed and analyzed sequentially for localization of involuntary hyperkinetic movement of the tongue.

Results: Lingual hyperkinesia symptoms of the patient diagnosed in WE, occurred in advance and were more earlier improved by thiamine treatment before other brainstem symptoms.

Conclusion: The clinical and neuro-radiological results discussed here may provide support for the localization of lingual hyperkinetic movement disorders. In addition to brainstem lesion, thalamic lesion should be considered in cases of acute or subacute-onset, involuntary hyperkinetic movement of the tongue.

Keywords: Lingual hyperkinesia; Wernicke`s encephalopathy; Localization; Thalamic lesions; Thiamine

Introduction

Wernicke's encephalopathy (WE) is an uncommon neurological complication of thiamine (vitamin B1) deficiency that is characterized by the acute onset of ocular motor signs, ataxia, and a confused mental state [1,2]. Typical neuro-radiological findings of magnetic resonance imaging (MRI) in WE include symmetric hyperintensity in the thalami, mammillary bodies, tectal plate, and periaqueductal area [3,4]. Movement disorders and complicated involuntary movement symptoms are very rarely reported as an initial manifestation of WE, and some case reports have described complications of WE presenting as dystonia, choreoathetosis, or dystonic tremor [5,6]. However, in the literature, involuntary hyperkinesia of the tongue as an initial manifestation of WE has not yet been reported.

Here, we report an atypical case in which typical initial symptoms were preceded by unusual involuntary movement of the tongue. The case developed with spontaneous upbeating nystagmus, mild ataxia, and confusion due to WE. After thiamine supplementation, not only the typical symptoms of WE but also the lingual hyperkinesia caused by symmetrical thalamic lesions were fully improved. Written informed consent was obtained from the patient to publish both video and brain imaging results for this article.

Patient and Method

Case presentation

A 41-year-old man, who had undergone a gastrectomy 6 months ago due to advanced gastric cancer, presented with subacute onset of dysarthria and involuntary movement of the tongue that commenced 3 days before the visit. Previously, he had been treated with some chemotherapy. However, he had not taken any anti-dopaminergic drugs such as gastrointestinal prokinetics, neuroleptics and antiemetic agents recently, and his latest concerning problem was the aggravation of poor oral intake. A neurological examination revealed not only involuntary hyperkinesia and motor impersistance of the tongue but also spontaneous upbeating nystagmus, mild limb ataxia, and confusion. The tongue movements were characterized by irregular slow, repetitive, involuntary protrusions and contractions involving the whole tongue. The movements were mainly back and forth but were combined with side-to-side tongue deviations similar to the involuntary tongue protruding and motor impersistance of choreic or dystonic patients. These tongue movements had a frequency of 1 Hz to 3 Hz similar to myorhythmic or slow myoclonic movements. Further, the tongue movements were mainly observed when the tongue was at rest, and the movements ceased during sleep. He also could not maintain the protruding position of the tongue due to the hyperkinetic involuntary tongue movement. For these reasons, he complained of moderate dysarthria, unsteadiness of the tongue and feeding difficulty without complaining of eye symptoms, which included spontaneous upbeating nystagmus. The soft palate was not involved (Video segment 1).

Routine laboratory tests were normal, except for a decreased vitamin B1 (thiamine) level (49.92 nmol/L; normal range, 66.5–200 nmol/L). Brain MRI showed high signal intensity in the medial thalamus bilaterally and periaqueductal area of the midbrain tectum, thus, he was diagnosed with WE (Figures 1A-1C). Also, ophthalmoplegia, opsoclonus, areflexia, headache and fever were not present, so we could exclude Miller Fisher syndrome, Bickerstaff`s encephalitis, and other encephalitis in differential diagnosis. Promptly after clinical diagnosis,

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Video segment 1: The patient showed irregular involuntary myorhythmic hyperkinesia and motor impersistance of the tongue in speaking serial Korean numbers from one to ten as well as protruding of the tongue. He also presented with simultaneous spontaneous upbeating nystagmus, without complaint of eye symptoms.

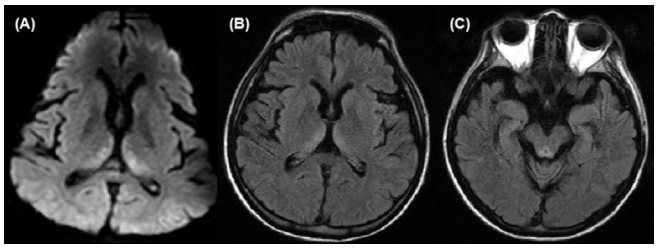

Figure 1: Axial brain imaging shows high signal intensity in the medial thalamus bilaterally and in the periaqueductal area of the midbrain tectum in diffusion MR imaging (A) and fluid-attenuated inversion recovery (FLAIR) MR imaging (B, C).

Video segment 2: After three days of treatment, the patient showed marked improvement of the dysarthria and lingual symptoms without involuntary hyperkinesia and motor impersistance of the tongue, although the mildly upbeating nystagmus could still be observed.

Video segment 3: After one month of treatment, the upbeating nystagmus and other neurological deficits almost fully resolved.

he underwent treatment with 100 mg of intravenous thiamine per day for the following seven days. The dysarthria and lingual symptoms improved markedly after three days of thiamine supplementation and supportive care, without a change in chemotherapy, although the mildly spontaneous upbeating nystagmus remained in the absence of other ocular motor signs including ophthalmoplegia and opsoclonus (Video segment 2). After one month, the spontaneous upbeating nystagmus and other neurological deficits almost fully resolved without any other complications or Korsakoff-like cognitive sequelae (Video segment 3). Although we recommended further pre-treatment laboratory evaluations, including electromyography (EMG) of involved facial muscles and analysis of erythrocyte trans ketolase activity or erythrocyte thiamine diphosphate levels for more accurate diagnosis, as well as follow-up checks of thiamine levels and neuro-radiological study, he declined to participate in further studies and evaluations after his rapid improvement.

Analysis on time-sequence of neurological symptoms

From onset of patient's lingual symptoms, I closely observed not only patient's lingual symptoms, but also other neurological symptoms by videotaping. And then, I analyzed the changes and improvements of each and all neurological symptoms sequentially for localization of involuntary hyperkinetic movement of the tongue.

Results

As a result of comparison between patient's lingual symptoms and other neurological symptoms, I found that the earlier improvement of involuntary lingual movement symptoms compared to other brainstem symptoms including spontaneous upbeating nystagmus, mild ataxia, and confusion.

Discussion

To date, few WE cases associated with movement disorders symptoms have been reported; however, to the best of my knowledge, this case is the first report of involuntary hyperkinesia of the tongue as an initial manifestation of WE in the English literature.

Involuntary movements of the tongue are rare and poorly understood. Several underlying conditions associated with isolated involuntary or abnormal tongue movements have been reported, including thalamic or pontine infarction, brainstem ischemia, radiosurgery for acoustic schwannoma, electrical injury, Arnol–Chiari malformation, Miller-Fisher syndrome, Bickerstaff's encephalitis, amyotrophic lateral sclerosis, drug-induced or drug-intoxication, chronic epilepsy and multiple sclerosis [7-20]. Among the above these reports, only some reports revealed the neuro-radiological localization of the involuntary hyperkinetic movements of the tongue (Table 1) [7,8,10,11,13,20]. The slower frequency and some of the phenomenological characteristics of the involuntary tongue movements observed in our case might be similar to the features of oculofacial or oculomasticatory myorhythmia, which is characterized by relatively rhythmic muscle contractions in ocular, facial, mastificatory, limb, and other muscles and is typically seen in brainstem or cerebellar disease and Whipple disease. However, this patient's involuntary hyperkinetic lingual movements and other neurological symptoms were somewhat different from typical oculofacial myorhythmia and previously reported episodic tongue hyperkinesia [9,10,21].

The brainstem, with involvement of the central tegmental tract at the pontine level or with of hypoglossal nuclei at the medullar level, was suspected to be the origin of the involuntary tongue movement [8,9,20]. WE possibly affect the brainstem, particularly the central

Types of IMS* of the tongue	Gender/Age [Reference]	Final diagnosis	Neuro-radiological localizations	Associated conditions	Onset of Symptoms	Time to improvement	Medication for IMS
Lingual dystonia	F/72 [7]	Thalamic infarction	Right posterolateral thalamus	Dysphagia	Acute onset (2 weeks ago)	4 months	No
Isolated continuous rhythmic tongue movements	F/82 [8]	Pontine infarction	Right ventrolateral portion of the mid and superior pons	No	6 months after infarction	NA†	Clonazepam
Episodic tongue hyperkinesias	M/74 [10]	Basilar artery ischemia	Bilateral thalamus, cerebellum, right temporo-occipital, left occipital areas	Dizziness, ataxia, dysarthria, dysphagia, quadriparesis	8 days after ischemia	7 months	Clonazepam
Isolated tongue tremor	F/70 [11]	Radiosurgery for acoustic schwannoma	Right pons and middle cerebellar peduncle	Audible click	1 month after radiosurgery	6 months	Propranolol
Isolated lingual myoclonus	M/61 [13]	Arnold-Chiari malformation	Herniation of cerebellar tonsils	No	15 days ago	1 week	Clonazepam
Isolated lingual dyskinesia	F/50s [20]	Multiple sclerosis	Brainstem, medulla, hypoglossal nuclei, pyramids	No	5 years ago	NA	No
Lingual hyperkinesia	M/41 [present case]	Wernicke's encephalopathy	Bilateral medial thalamus, periaqueductal area of the midbrain	Upbeating nystagmus, limb ataxia, confusion	Subacute onset (3 days ago)	3 days	Thiamine

IMS = involuntary movement symptoms; †NA = not available.

Table 1: Summary of clinical characteristics and neuro-radiological localizations in reported cases and present case of hyperkinetic involuntary movements of the tongue.

tegmental tract adjacent to the periaqueductal areas. Another possible mechanism for the involuntary lingual movement is a thalamic lesion, which would disinhibit the thalmocortical projection [7]. The excitatory thalamocortical projection is inhibited by the Globus pallidus pars internal/substantia nigra pars reticulata (GPi/SNr) complex, and blockade of these inhibitory fibers may result in hyperkinetic movement disorders by allowing excessive activation of corticospinal projections. A bilateral cortical projection to the lingual muscle might cause excessive contraction of both sets of styloglossus muscles, which results in retraction of lingual dystonia [22].

As previously reported, upbeating nystagmus may result from damage to the pathways mediating the upward vestibulo-ocular reflex or the neural integrators involved in vertical gaze holding [23]. Among these lesions, brainstem lesions including medulla, pons, midbrain, pontomedullary and Ponto mesencephalic junctions were mainly associated with upbeating nystagmus [23,24]. For these reasons, this report supports the hypothesis that abnormal tongue movements might be influenced more by bilateral thalamic lesions, rather than by brainstem lesions, due to the earlier improvement of involuntary lingual movement symptoms compared to other brainstem symptoms, including spontaneous up beating nystagmus, in our patient with WE. Considering the chronological aspect of neurological symptoms and neuro-radiological abnormalities, the reversible dysfunction of the thalamic lesions in WE may have a role in the pathogenesis of the lingual involuntary hyperkinetic movements.

In contrast to my case, rare clinical presentations of symptoms involving the tongue, including isolated glossoplegia, inability to protrude the tongue or complete tongue protrusion palsy, have been described in some reports. In these cases, lesions were primarily localized to the unilateral or bilateral precentral gyrus and were caused by capsular infarction or cortical stroke [25-27].

Based on the conflicting observations regarding the central localization of abnormal lingual movements, I hypothesize that hypokinetic movement or weakness of the tongue might be associated with brain cortical or capsular lesion and that involuntary hyperkinetic movement of the tongue might be associated with thalamic or brainstem lesion.

Conclusion

Here, I provide the first report of an unusual initial manifestation of WE, presenting as myorhythmic hyperkinesia and motor impersistance of the tongue. A diagnosis of WE should be considered if there are symptoms of abnormal involuntary hyperkinetic movement of the tongue that is accompanied with a confused mental status or ocular motor signs, such as upbeating nystagmus. Furthermore, in addition to brainstem lesion, thalamic lesion should be considered in cases of acute or subacute-onset, involuntary hyperkinetic movement of the tongue.

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