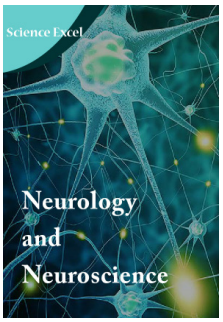
Neurology & Neuroscience

Case Report



A rare case of undulatory rhythmic lingual myoclonus in a patient with intracranial hemorrhage and diffuse pontine lesion

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- Received Date: 03 Mar 2022
- Accepted Date: 12 Mar 2022
- Publication Date: 04 Apr 2022

Abstract

We describe a 35-year-old man, who suffered a severe intraventricular hemorrhage and was admitted to the intensive care unit of our tertiary clinic. Two weeks after the incident, while in ICU, a neurological examination revealed persistent, undulating, rhythmic, myoclonic tongue movements. The diagnostic evaluation via CT scan, MRI and EEGs elicited a diffuse brainstem lesion as well as multiple corpus callosum lesions as a considerable underlying etiology.

This case presentation attempts to update the current investigation of the rare phenomenon of lingual myoclonus.

Introduction

Orofacial myoclonus in the form of lingual myoclonus represents a rare movement disorder of diverse and sometimes even unknown etiology, which in most cases is accompanied with other myoclonic movements such as palatal myoclonus. This report describes a case of persistent rhythmic wave-like lingual myoclonus with no involvement of other orofacial muscle in the setting of intraventricular hemorrhage and metabolic instability with required prolonged stay at the Intensive Care Unit (ICU).

Case report

A 35-year-old Caucasian male with a relevant medical history of alcohol abuse, smoking, a questionable episode of epileptic seizure several months ago, and blood hypertension, under medication with benzodiazepines and valproic acid in the context of a detox therapy, was admitted to the emergency room of our tertiary clinic due to a sudden-onset, severe impairment of consciousness level.

The Brain CT- Scan revealed an extended subarachnoid and intraventricular hemorrhage in the lateral ventriculi with hemorrhagic

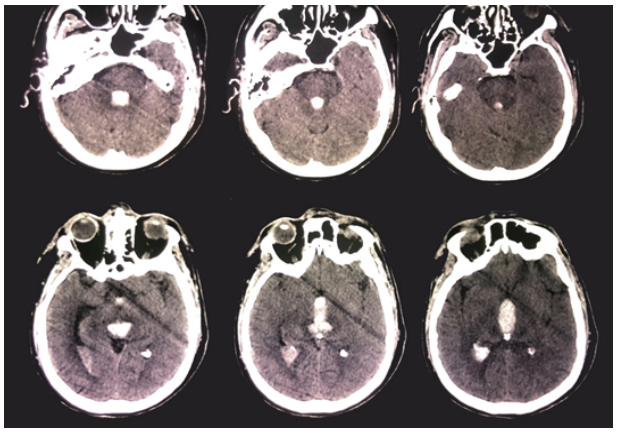

Figure 1. *Extended subarachnoid and intraventricular hemorrhage in the lateral ventriculi with hemorrhagic elements in the frontal horns bilaterally, mainly right, in the third ventricle, in the cerebral aqueduct, as well as in the fourth ventricle*

Citation: Zachariadi C, Alexandratou I, Katsaros V, Gatzonis S. A rare case of undulatory rhythmic lingual myoclonus in a patient with intracranial hemorrhage and diffuse pontine lesion . Neurol Neurosci. 2022; 3(1):1-3.

elements in the frontal horns bilaterally, mainly right, in the third ventricle, in the cerebral aqueduct, as well as in the fourth ventricle (Figure 1). An external ventricular drain with a CAMINO intracranial Pressure Device was placed and the patient was admitted to the ICU under general anesthesia with propofol. While in ICU the patient had some complications including bacteremia and a few peaks of intracranial pressure (ICP), which responded immediately to medical treatment. By his second week he had not managed to regain a sufficient level of consciousness (GCS<5) so a tracheostomy was considered necessary. At that point a new Brain CT-Scan showed a diffuse but marked brainstem (pons and midbrain) hypodensity of unknown etiology (low quality because of beam hardening artifacts). A cerebrospinal fluid analysis and culture excluded a central nervous system infection. The bed-side EEG was diffuse slow, without epileptiform, periodic or rhythmic discharges.

At that time, during the neurological evaluation, while opening the patient's mouth, we observed a series of persistent, rhythmic 4/s, undulatory/galloping tongue movements. No implication of other oral or facial muscles was noticed and the movements did not produce any kind of sound. A Magnetic Resonance Imaging (MRI) of the brain was performed (low quality because of motion artifacts), demonstrating multiple diffuse hyperintense lesions periventricular, scattered hyperintense lesions in the corpus callosum, as well as punctuate hyperintense and hypointense lesions in the posterior part of the pons and midbrain, bilaterally, mainly on the right, compatible with hemorrhagic transformation of ischemic lesions (Figures 2 and 3). The patient's status improved gradually. Almost two months after the episode, he was able to open his eyes spontaneously and obey simple commands. Vocalization was until that point not achieved. Through ought this period the lingual myoclonus was persistent. No symptomatic treatment was administrated, since it caused the patient no discomfort.

Discussion

A literature search revealed that lingual myoclonus is a rather rare movement disorder under the umbrella of myoclonus, one of the most frequent hyperkinisias. The case of our patient with orofacial myoclonus in form of persistent, rhythmic galloping movements strictly restricted to the tongue is unique. In most previous reported cases the tongue hyperkinisias were consisting of rhythmic predominantly bilateral symmetric jerking contractions or intrusion-protrusion movements [1,2] as part of myoclonus of the palatopharyngeal or orofacial muscles. Episodes of rhythmic undulating wave-form tongue movements as observed in our patient were described in only two patients after suffering head trauma [3] and in three children [4] accompanied by EEG pathology.

Palatal myoclonus is from the closest clinically speaking to tongue myoclonus, the best studied hyperkinesia. The underlying structural pathology is considered to implicate predominantly the olive with its input connections, as well as the central tegmental tract and the dentate nucleus (the Guillain-Mollaret triangle). The ipsilateral red nucleus projecting to the inferior olivary nucleus and the globus pallidi of the basal ganglia seem to be secondarily involved in the generation of the pattern [3-6] .

In our case a thorough investigation did not identify a certain cause for this phenomenon. Our patient was treated with a variety of antibiotics. An association with this pattern of extended orofacial hyperkinesia has not yet been reported. The phenomenon can extremely rarely occur in isolation without detection of an underlying pathology [1,5,7-10]. Ong et al. [11] reported a possible association based on one unique case between lingual myoclonus and neuropsychiatric lupus. Recently a case of isolated lingual myoclonus in an HIV-patient was reported [12]. It has also been sometimes described to have an epileptic origin [4,13,14]. The majority of the cases reported

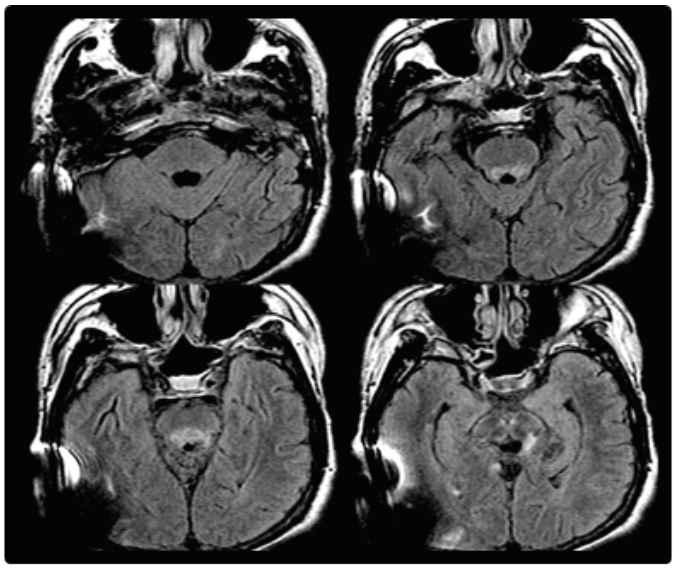

Figure 2. *Punctuate hyperintense and hypointense lesions in the posterior part of the pons and midbrain, bilaterally*

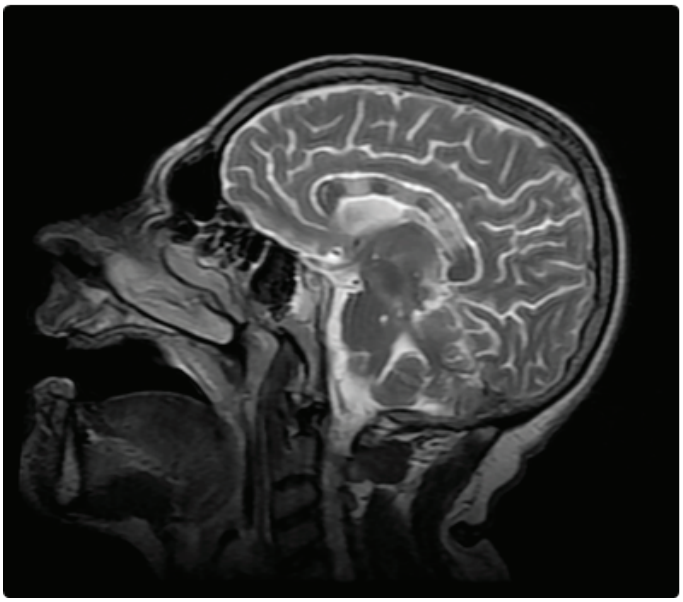

Figure 3. *Multiple scattered hyperintense lesions in the corpus callosum as well as diffuse hyperintense lesions periventricular*

were accompanied with an underlying structural lesion such as a medullary compression at the craniovertebral junction [2], an Arnold-Chiari malformation [15], a brainstem trauma [3,16,17] or ischemia [18] and even bilateral cortical ischemia [19]. Cartagena [6] reported two rare cases of involuntary craniofacial movements after septic encephalopathy and ICU admission, with the one presenting MRI findings representative of PRES and the other one showing an abnormal sign in the globus pallidi.

Our patient's MRI findings were consistent with a brainstem infarction, although not immediately affecting the Guillain-Mollaret triangle. A finding that we cannot ignore or underestimate is the extended multiple lesions in the corpus callosum. Although the disruption of transcallosal projections would rather lead to multifocal myoclonus [20,21] sensitive to stimuli, a rare pattern of focal cortical myoclonus is to be considered. No reports were found associating the origin of lingual myoclonus with corpus callosum injury.

Conclusion

According to our literature search, lingual myoclonus is a rare movement disorder under the umbrella of myoclonus, one of the most frequent hyperkinesias. Its origin is considered to be multifactorial. Ischemia, trauma, epileptic phenomena, infectious and autoimmune diseases are a few of the incriminating factors. Based on the MRI findings we consider the pons infarction to be the generator of the lingual myoclonus in the form of a "galloping" tongue at our patient. Nevertheless, the role of the extensive corpus callosum insult should not be confidently excluded. Future reports may offer some enlightenment respectively.

Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Funding

The authors received no financial support for the research and authorship of this article.

Availability of data

The data that support the findings of this study are available from the corresponding author, C.Z., upon reasonable request.

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