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Case report

A rare case of isolated myoclonus in an elderly male without a history of epilepsy

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ABSTRACT

Aim: Through this case report we attempt to highlight the presentation, initial investigation and management of lingual myoclonus as well as consolidate relevant literature.
Case: We present a unique case of a 72-year-old man who was admitted to the hospital for a sudden onset episodic speech arrest. Lingual myoclonus, an isolated movement disorder, manifested as an intermittent expressive aphasia secondary to the intrusion-protrusion movements of his tongue. During this time, the patient remained conscious and was able to continue to follow commands. Initial diagnostic evaluation with a CT scan, MRI and EEG failed to illicit a clear underlying etiology and the patient was empirically treated with valproic acid with complete resolution of his symptoms.
Discussion: This unusual presentation represents a rare disorder which is not well described in literature. Initial evaluation of which required excluding associated etiologies including strokes, seizures, medications/toxins or CNS infections. Without a clear etiology on initial diagnostic evaluation, the patient was empirically treated as no clear guidelines exist. This case presentation is an attempt to add to the current understanding of lingual myoclonus.

1. Introduction

We present a case of lingual myoclonus presenting as episodic speech arrest. This case report will review the presentation of this rare disorder, update the investigation and management of lingual myoclonus, and review relevant literature.

2. Case report

A 72-year-old Caucasian male with a relevant past medical history insulin dependent diabetes mellitus, dementia, essential tremors and a recent stroke presented with a sudden onset of intermittent expressive aphasia. The episodic speech arrest started 5 days ago. Since then, he has not had a single episode free day and has up to five episodes daily. Each episode preceded by "a numbness" on the left nasolabial fold, following which, the patient has an inability to speak. Each episode lasts less than five minutes and were followed by a quick and complete recovery. There were no precipitating or palliative factors. During the episodes, the patient could continue to sit, stand and walk and even reports being able to swallow a few sips of water.

His other medical issues included Diabetes mellitus, untreated Chronic lymphocytic leukemia, hypertension, essential tremor, mild dementia and a suspected brain stem stroke 5 months prior to his present admission. His recently underwent a right total knee replacement. He did not have a significant family history and only a remote history of tobacco use. The patient lives with his son and reports being high functioning at baseline.

On presentation, his vitals were within normal limits. He was a pleasant, well developed male who was comfortable at rest. His neurological examination was normal except for a chronic right sided pronator drift, a result of a prior stroke. His gait was slow due to recent total knee replacement. During the exam, the patient had a sudden onset of episode of speech arrest. Though he remained conscious, and able to follow commands, he was unable to speak. When asked to open his mouth, a spontaneous, uninterrupted intrusion-protrusion movement of the posterior tongue and adjoining hyoid was visualized. This rapid, jerking movement was not associated with any other facial movements. The patient was able to follow commands, open his mouth and even protrude his tongue further (Video 1). He returned to his baseline in 1–2 min.

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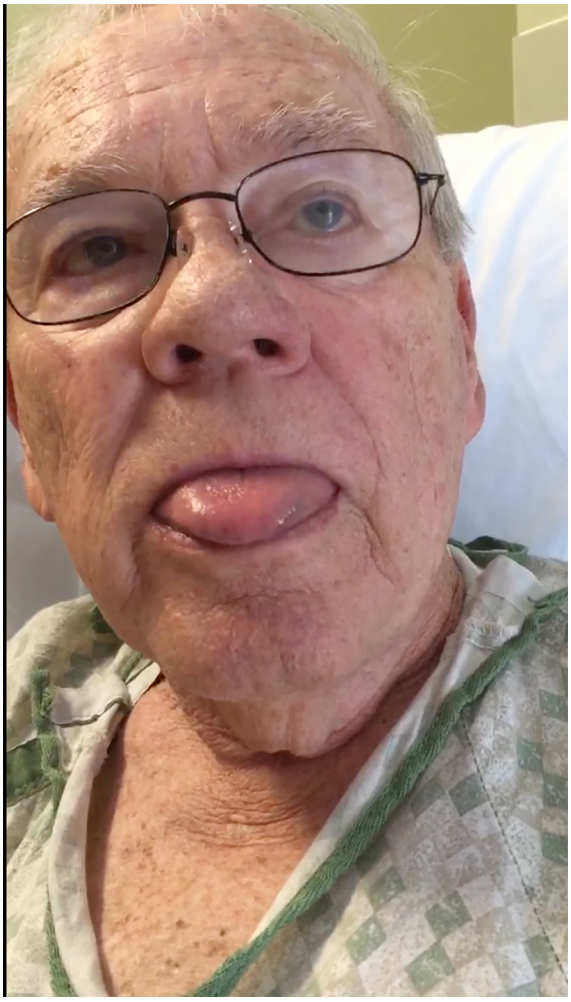

Video 1.

Initial laboratory data revealed significant lymphocytosis and smudge cells, consistent with his chronic lymphocytic leukemia. Other lab values were unrevealing except for mild hyperglycemia.

Based upon these findings, an initial differential diagnosis included lingual myoclonus, epilepsy, stroke, or a space occupying lesion. However, initial CT scan and MRI revealed old lacunar infarcts without acute ischemia (Fig. 1). The EEG was intensively discussed by several epileptologists, who concluded that the pattern was not consistent with myoclonic epilepsy, thus making the diagnosis of lingual myoclonus most likely. The patient was treated with valproic acid, resulting in a complete resolution of his symptoms.

3. Discussion

Isolated lingual myoclonus, as experienced by our patient, is an extremely rare phenomenon which is not well described in the literature. A myoclonus is described as a brief, shock like, involuntary movement (Marsden et al., 1982). Its incidence was studies in a population in Minnesota, which revealed an incidence of 1.3 cases in 100,000 person years with a lifetime prevalence of 8.6 per 100,000. (Caviness et al.,1999). The study further categorized myoclonus by etiology, The majority of known cases were labeled as secondary, followed by epileptic and finally essential myoclonus which made up 11%.

The differential diagnosis for myoclonus includes epileptic syndromes, focal central nervous (CNS) damage including ischemia, malignancy, trauma, infections, metabolic disorders and toxins. Usually, these are differentiated from one another on the basis of their clinical features, imaging and EEG. (Levy and Chen, 2016)

The lack of acute findings on imaging or EEG in our patient make this difficult. Based on our review of the literature, a focal, positive or excitatory myoclonus points towards a cortical, subcortical source, though, involuntary movements of the tongue have been reported in association with brain stem ischemia as keeping this in mind our initial differential diagnosis includes epileptic syndromes, focal central nervous (CNS) damage including strokes, malignancy, trauma, infections, metabolic disorders and toxins.

In our case, the EEG failed to show an epileptic focus. Epilepsy, however, is not completely excluded as a small epileptic focus, as in Epilepsia partialis continua, are not always evident (Niedermeyer et al., 1985). A new onset of seizures in an elderly male strongly suggests a prior stroke which fits with our patient's prior history. Though case reports of episodic tongue movements in children have been reported, epilepsy causing focal seizures in the tongue are not well described in adults (Emre, 1992; Jabbari and Coker, 1981).

Stroke is associated with several movement disorders (Bansil et al., 2012) and is the most common cause of symptomatic palatal tremor (SPT). (Deuschl et al., 1990). These brief rhythmic movements of the soft palate parallel lingual myoclonus. A prior study showed a 2–49 month delay from cerebral infarction to development of SPT. (Emre, 1992) Though a stroke-related movement disorder is a consideration, atypical features in our patient include intermittent symptoms, absence of symptoms during the night and a lack of focal deficits.

Though few case reports of lingual myoclonus exist, (Marsden et al., 1982; Jabbari and Coker, 1981; Deuschl et al., 1990; Jacobs et al., 1981) in our review, only once case of episodic, undulating

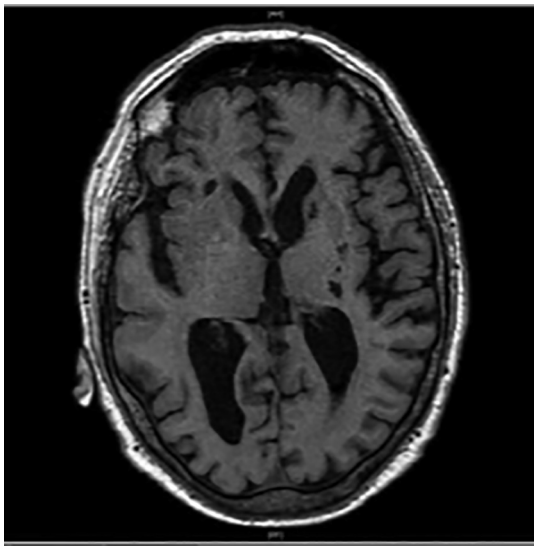

Fig. 1. Magnetic Resonance imaging of the patient which failed to show an underlying ischemic or structural pathology which could have been contributing to his symptoms.

tongue movements has been described. Similarities with our case extend to a negative EEG and MRI. However, in contrast to our case, this patient presented with continuous rhythmic contractions of both lingual edges which only improved after treatment with benzodiazepines. This ameliorated their patient's symptoms and there was no return of myoclonus even after the medications were discontinued.

As essential lingual myoclonus is such a rare phenomenon, guidelines dictating its treatment do not exist. In literature, benzodiazepines, antiepileptic drugs and botulinum toxin have been tried with variable results. Though surgical techniques for palatal myoclonus are available, no such intervention has been demonstrated in lingual myoclonus. In one case report, a novel device which was inserted into the mouth at night stopped an action oriented mixed lingual and myoclonus. However, again, a specific therapy for this disorder eludes us. (Mondria et al., 2007). Our patient was initially started on levetiracetam which has been shown to be of benefit in cortical subcortical and brainstem lesions(Levy and Chen, 2016), which unfortunately failed to ameliorate his symptoms. However, we were able to find a single case report suggesting the use of valproate (Gobernado et al., 1992), which when attempted in our patient at a dose of 250 mg BID ameliorated all further symptoms.

Conflict of interest

None.

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