



Correspondence

Isolated lingual myoclonus in an HIV patient

ARTICLE INFO

Keywords:

Lingual myoclonus

HIV

Myoclonus

1. Introduction

Lingual myoclonus is an uncommon disorder, usually described in association with palatal myoclonus or other movement disorders involving oculofacial, diaphragmatic or shoulder muscles. Isolated lingual myoclonus, either episodic or continuous, is rare, with only a handful of cases reported in the literature [1,2]. Etiologies are diverse, ranging from secondary lesions of the brainstem region to focal cortical seizures [1,2]. We present a case of epileptic lingual myoclonus in the context of HIV infection. Written informed consent was obtained from the patient prior to video recording and publication.

2. Clinical case

A forty-one year old man with acute onset of speech difficulties attended the emergency room after 48 hours of persistent, uncontrollable tongue movement. He denied fever, presence of other neurological symptoms or medication use. Physical examination showed brief, continuous nonrhythmic isolated lingual movements, without associated palatal tremor or myoclonus (*video segment 1*). The remainder of the examination was normal.

Supplementary video related to this article can be found at <https://doi.org/10.1016/j.parkreldis.2018.10.020>.

Brain MRI with gadolinium was unremarkable. Initial investigations showed white blood cell count of 4870/mm³, mild increase of liver enzymes (SGOT 58 UI/l, SGPT 106 UI/l), erythrocyte sedimentation rate 7 mm/h, and CK 64 UI/l. Rheumatological (ANCA, FAN) and toxicological tests (cocaine, cannabis, benzodiazepines and barbiturates) were negative as well as VDRL testing. Lumbar puncture showed an opening pressure of 13 cmH₂O, and CSF biochemistry results were: 10 cells/ μ L (90% MNN), sugar 58 mg/dl (blood sugar 98 mg/dl), and 35.5 mg/dl protein level. PCR for: HSV types I and II, VZV, EBV and CMV in CSF were normal, as were microbiological CSF cultures (bacteria, fungi and mycobacteria). HIV (ELISA) test was performed in order to exclude secondary infectious diseases, as well as myoclonus related to HIV. The test yielded a positive result. Three days after lingual myoclonus started, the patient showed minor, involuntary and intermittent left facial involuntary movements followed by a secondarily generalized focal seizure. Treatment with 1000 mg of Levetiracetam was started, with complete myoclonus resolution and no seizure

recurrence. Both EEG and brain 3T MRI were normal.

After the HIV diagnosis, the patient was started on antiretroviral drugs (HAART). His anticonvulsant treatment was discontinued after a 2-year seizure-free period. To date, he remains neurologically normal, seizure and myoclonus-free. (*video segment 2*).

3. Discussion

The patient showed an arrhythmic, sharp (fast), lingual movement clinically interpreted as myoclonus of the tongue, not involving the palate. Symptoms were focal and continuous. The rapid onset, followed by sustained facial myoclonus and secondary generalization supports our belief that the lingual myoclonus was of epileptic origin. Although no EEG correlation was found, the fact that both episodes were separated by more than 24 hours (focal continuous seizure and focal seizure with secondary generalization), led us to identify the case as epilepsy under the ILAEs guidelines [3].

In addition to secondary forms, lingual myoclonus of epileptic origin has been previously described (Table 1), mainly associated with cortical frontal gliomas or cavernomas [1,2]. As in the present case, none of the previously reported cases demonstrated specific alterations in the EEG. In this case, however, secondary generalization and excellent response to anticonvulsant drugs strongly support an epileptic origin.

The association between myoclonus and HIV infection has already been addressed in previous reports [4]. Generalized and segmental myoclonus, specially spinal myoclonus were reported in HIV-patients [4]. Several mechanisms have been proposed for this association. Myoclonus has been suggested to be of subcortical origin in patients with HIV-associated dementia, whereas it has been proposed as a consequence of inflammatory CD4/CD8 dysregulation in patients with opsoclonus-myoclonus-ataxia syndrome; in which either seroconversion or immune reconstitution on HAART, results in brainstem-cerebellar circuit alteration. On the other hand, neurophysiological analysis of a single patient with HIV and myoclonic epilepsy suggested a cortical mechanism responsible for generating repetitive paroxysmal activity [5].

To the best of our knowledge, there are no prior reports of isolated lingual myoclonus in HIV, especially in uncomplicated infections. Even though main opportunistic infections were ruled out, we could not

<https://doi.org/10.1016/j.parkreldis.2018.10.020>

Received 26 June 2018; Received in revised form 30 September 2018; Accepted 15 October 2018

1353-8020/ © 2018 Elsevier Ltd. All rights reserved.

Table 1
Summary of clinical features of isolated lingual myoclonus (reported cases).

Case	Age	Sex	Primary cause	Latency	Characteristics	Characteristics II	Physical exam	Course	Treatment	Other findings	Authors
1	25 y.	M	Head trauma	8 days	Continuous (4–5Hz)	Bilateral contractions of the tongue	Mild dysarthria. Normal 24hs after trauma	Spontaneous recovery	DFH and DZP	EEG slowing/left temporal focus (theta waves)	Tropin and Kamm
2	56 y.	F	lupus	–	Continuous	Repetitive protrusion-intrusion movement of the tongue		Good response after 48 hours with VPA	VPA	EEG: normal MRI: diffuse cerebral atrophy and old infarct in left basal ganglia	Ong SW
3	21 y.	M	Unknown	–	Continuous	Rhythmic lingual movements		Refractory to drugs, good response with dental device.	Dental bit (CBZ, CNZ, GBP and VPA ineffective)	Endoscopy:lingual and palatal myoclonic movements MRI: normal	Mondria T.
4	53 y.	M	–	–	Continuous	The tongue is seen to be curled, and the left side raised, most of the tongue movements being directed towards the right side.	More or less rhythmic movements of his jaws and tongue, and also of his left foot.	?			Harris W
5	10 m.	M	Sub-acute encephalitis	2 days	Continuous (4Hz)		Normal			EEG slowing (temporal focus)	Sridharan (case 4)
6	55 y.	F	none	–	Continuous (2–4Hz)	Bilateral and symmetric jerking contractions that produced a narrowing of the anterior portion of the tongue with slight forward protrusion of the anterior portion, without lateral deviation. Apparently rhythmic, and variable in intensity.	Reduced facial expressiveness and low blinking frequency. Mild dysarthria.	Good response to VPA	VPA (CBZ ineffective)	MRI, EEG: normal EMG left genioglossus muscle: bilateral involuntary rhythmic discharges	Gobernado
7	61 y.	M	Arnold-Chiari	–	Continuous (3Hz)	Rhythmic low amplitude, symmetrical contractions of both lateral edges of the tongue affecting mainly its middle and posterior parts and causing a midline depression of the tongue. Repetitive movement with variable frequency.	Normal	Disappeared with CNZ (second week)	CNZ	MRI: Arnold-Chiari type I. EEG,BAEP: normal	Kulisevsky J.
8	41 y.	M	HIV	–	Continuous	Repetitive contractions of the entire tongue.	Minor intermittent left facial involuntary movements. Secondarily generalized focal seizure	Excellent response to LVT	LVT	LP: 10 cells. MRI and EEG normal.	present case
9	16 y.	F	Left frontal cavernoma	–	Episodic (5 Hz.)	Repetitive contractions of the entire tongue.	Involuntarily opening of her mouth.	Good response to TPM	TPM	MRI: cavernoma (left posterior frontal lobe) EEG: showed transient synchronized sharp waves over bilateral F (R > L)	Jie-Yuan Li.

(continued on next page)

Table 1 (continued)

Case	Age	Sex	Primary cause	Latency	Characteristics	Characteristics II	Physical exam	Course	Treatment	Other findings	Authors
10	59 y.	M	FP Oligodendroglioma	1 month after craniotomy	Episodic (1 Hz)	Rhythmic myoclonic jerks of the left part of the tongue and the lower lip.	Perioral numbness and slurred speech, which lasted about an hour. Lower lip involved.	Good response to VPA	VPA (no response to DFH)	MRI: right FP brain tumor. EEG: normal	Yilmaz R.
11	26 y.	F	Bilateral FT and insula stroke	3 weeks (first stroke) and at the same time of the second stroke.	?	Bilateral, nonrhythmic, asymmetric, and jerky movements involving the entire tongue.	Bilateral lower facial weakness and was severely dysarthric. Bursts of eyelid myoclonia.	One year after treatment (hydroxychloroquine and warfarin) the severity of lingual myoclonus was decreased. Eyelid myoclonia was not ameliorated by the treatment regimen and it persisted.	No specific treatment (just for the primary cause: hydroxychloroquine and warfarin)	EMG of genioglossus: non-rhythmic myoclonus. MRI: hypersignal intensity of bilateral FT and insular a reason T1, T2 and FLAIR.	Priya Jagota
12	2 m.	-	Epilepsy		Episodic		Hypotonia			CT: hypotrophy of right the hemisphere EEG: episodic desynchronization	Jabbari and Coker
13	6 m.	-	Epilepsy		Episodic		Cortical blindness			EEG: episodes corresponded with desynchronization	Jabbari and Coker
14	2 y.	-	Epilepsy		Episodic (50/hour)		normal			EEG: episodes corresponded with desynchronization	Jabbari and Coker
15	53 y.	M	Brainstem ischemia	14 days	Episodic 2/s undulation 4 per min		Facial paresis			MRI: normal Blinkreflex: brainstem lesion	Postert
16	26 y.	M	Head trauma	1 month	Episodic 3Hz		INO, facial paresis			Brainstem damage	Keane
17	19 y.	M	Head trauma	3 weeks	Episodic 3Hz		Abducens paresis	Spontaneous recovery	-	Brainstem damage	Keane

DFH, diphenylhydantoin; DZP, diazepam, EEG, electroencephalogram; VPA, valproic acid; CBZ, carbamazepine; CNZ, clonazepam; GBP gabapentin; BAEP, brainstem auditory evoked; LVT, levetiracetam. TPM, topiramate; MRI, Magnetic resonance imaging; EEG, electroencephalogram; FT, fronto-temporal; EMG, electromyography; VPA, valproic acid; CT, computed tomography; INO, internuclear ophthalmoplegia; potential.

determine whether this epileptic symptoms were consequence of direct viral action or not. Nevertheless, our case seems to stress the importance of considering and excluding an epileptic origin in such cases, in spite of scarce or unremarkable clinical and laboratory findings.

References

- [1] J.Y. Li, C.H. Lee, Episodic tongue hyperkinesias and alternating limb movements associated with basilar artery ischemia, *Mov. Disord.* 24 (8) (2009 Jun 15) 1249–12451. 1.
- [2] R. Yilmaz, B.S. Arica, A. Yigit, Lingual myoclonus associated with brain tumour: an epileptic origin? *Epileptic Disord.* 12 (4) (2010 Dec) 303–305 Epub 2010 Oct 25.
- [3] R.S. Fisher, J.H. Cross, C. D'Souza, J.A. French, S.R. Haut, N. Higurashi, E. Hirsch, F.E. Jansen, L. Lagae, S.L. Moshé, J. Peltola, E. Roulet Perez, I.E. Scheffer, A. Schulze-Bonhage, E. Somerville, M. Sperling, E.M. Yacubian, S.M. Zuberi, Instruction manual for the ILAE 2017 operational classification of seizure types, *Epilepsia* 58 (4) (2017) 531–542.
- [4] W. Tse, M.G. Cersosimo, J.M. Gracies, S. Morgello, C.W. Olanow, W. Koller, Movement disorders and AIDS: a review, *Park. Relat. Disord.* 10 (2004) 323–334.
- [5] R. Kapoor, G. Griffin, G. Barrett, C.J. Fowler, Myoclonic epilepsy in an HIV positive patient: neurophysiological findings, *Electroencephalogr. Clin. Neurophysiol.* 78 (1) (1991 Jan) 80–84.

Lucia Ameghino*, Mariano Marrodan
*Movement Disorders Section, Neuroscience Department, Raul Carrea
 Institute for Neurological Research (FLENI), Buenos Aires, Argentina*

Anthony E. Lang
*Edmond J. Safra Program in Parkinson's Disease and the Morton and Gloria
 Shulman Movement Disorders Clinic, Toronto Western Hospital, UHN,
 Division of Neurology, University of Toronto, Toronto, Ontario, Canada*

Marcelo Merello
*Movement Disorders Section, Neuroscience Department, Raul Carrea
 Institute for Neurological Research (FLENI), Buenos Aires, Argentina
 Argentine National Scientific and Technological Research Council
 (CONICET), Buenos Aires, Argentina*

E-mail addresses: lameghino@fleni.org.ar,
luameghino@gmail.com (L. Ameghino),
marianomarrodan90@gmail.com (M. Marrodan),
anthony.lang@uhnresearch.ca (A.E. Lang),
mmerello@fleni.org.ar (M. Merello)

* Corresponding author. Movement Disorders Section and Neuroscience Department, Raul Carrea Institute for Neurological Research (FLENI), Montañeses 2325 - Ciudad Autónoma de, Buenos Aires, 1428, Argentina.