

Letter to the Editor

Subclinical dysphagia in task-specific mouth tremor triggered by drinking



Task-specific movement disorders mostly affect the hand and include musicians' dystonia, writer's cramp and primary writing tremor. Recently, a rare condition characterized by focal tremor selectively involving the oro-facial region, only visible during drinking, has been reported (O'Gorman et al., 2014; Macerollo et al., 2016). Although in these reports the involuntary activity was triggered exclusively by a specific behavior (e.g., drinking), none of the patients complained of dysphagia and no further diagnostic examination was performed.

Herewith we report a 74-year-old right-handed male with a 5-years history of isolated lip tremor, which occurred only during drinking. The involuntary movement, limited to the mouth, did not arise during any other activities such as eating or speaking. There was no family history of Parkinson's disease, essential tremor or other neurological disorders, and the medical history was unremarkable. The patient reported neither exacerbating or reducing factors, such as alcohol intake, nor sleep disturbances, headache or jaw pain suggestive of bruxism or oral trauma. He complained of slight non-progressive swallowing difficulties, only when drinking from a cup. He adapted by drinking with a straw and assuming liquids slowly in small amounts, and never lost his original weight or developed bronchopneumonia.

On neurological examination, no tremor of the jaw, lips, head, or limbs was visible at rest. Cranial nerves, motor, sensory and cerebellar examination were normal. There was neither bradykinesia, rigidity, nor gait disturbances. Conversely, drinking from a glass elicited a clear tremor of the mouth and lips (see Video 1). He could not voluntarily suppress the movement, but tended to take short sips of liquid to reduce the duration of the tremor.



Supplementary video 1. The tremor of the mouth and lips is visible as the patient drinks water from a glass. No tremor can be observed when the patient drinks from an empty glass, or when he drinks water from a straw.

Complete blood count, hepatic profile and thyroid function were normal. Inflammatory markers, serum manganese, urinary copper, and mercury were within the normal limits. Brain MRI and DAT scan were normal.

Multichannel surface electromyography (sEMG) was performed recording from upper and lower orbicularis oris, digastric, masseter, and temporalis muscles bilaterally. The study was approved by the Ethics Committees of IRCCS Neuromed in Pozzilli, Italy. The patient gave written informed consent to the study. No abnormal muscle activity was recorded at rest. Muscle activity upon drinking from a cup, as shown in Fig. 1A, was characterized by a very regular 7.5–8 Hz tremor, mostly involving the upper orbicularis oris and digastric muscles bilaterally. No abnormal muscle activity was observed or recorded with any position of the mouth, lips, or jaw, including when the patient pursed his lips, mimed drinking from an empty cup, held liquid in the mouth, drunk from a straw, held his mouth open, talked or laughed.

Electrophysiological study of swallowing (EPSS) was performed, recording EMG from both submental/suprahoid muscles (SMEMG) using two surface electrodes, and the laryngeal-pharyngeal mechanogram (LPM), using a piezoelectric transducer that detects laryngeal movements during swallowing. This procedure represents a useful tool to identify subclinical dysphagia in different neurological conditions through a number of neurophysiological parameters (Alfonsi et al., 2013). Only when drinking from a cup, a repetitive and patterned activity, characterized by 7.5–8 Hz oscillations, was simultaneously recorded from submental muscles and from the LPM (Fig. 1B). No oscillatory activity was detected when drinking from a straw (Fig. 1C).

We investigated the reproducibility of the swallowing act, during repetitive drinking from a straw, calculating the similarity index (SI) of the LPM, a novel marker of intra-individual variability of swallowing. A reduced SI could be a consequence of impaired voluntary or automatic control of swallowing, and constitutes a distinctive feature of neurological dysphagia (Cosentino et al., 2017). The patient's parameters were compared with those of 10 age-matched male healthy subjects (Table 1). For each EPSS measure, patient's values were standardized with respect to the mean and standard deviation of healthy controls obtaining the z-score. A marked reduction of the SI was observed, in particular, the patient's z score value was -2.69 and fell outside the 95% "control" confidence interval, indicating altered control of the oropharyngeal phase of swallowing. To better investigate subclinical swallowing abnormalities, the EPSS was performed during swallowing of small amounts (3 ml) of water administered with a syringe. We found significant increase of the oropharyngeal phase duration, shown by prolonged LPM. The duration of SMEMG activity and the interval between SMEMG onset and LPM onset were not altered. EPSS was then performed during increasing single-bolus amount of water. The dysphagia limit under EMG recording was altered, as

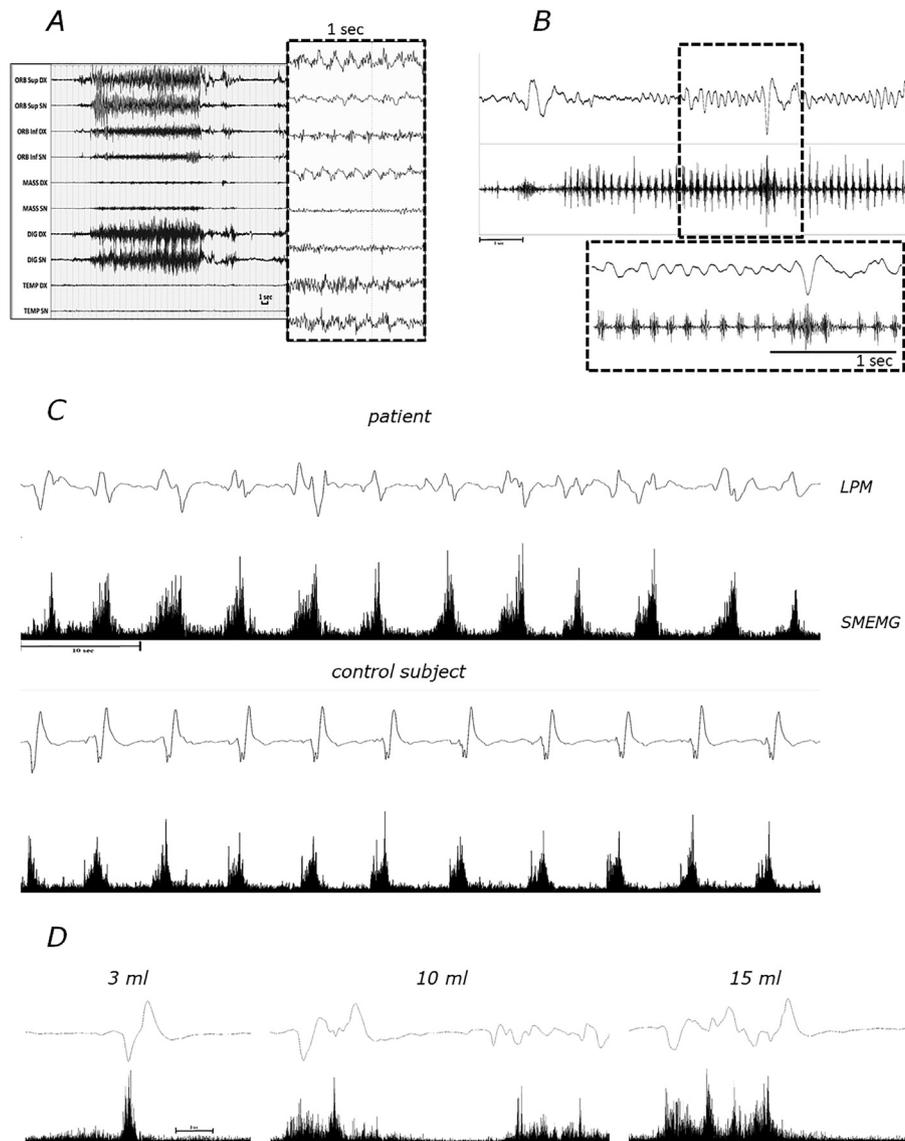


Fig. 1. EMG and electrophysiological study of swallowing. **Panel A:** Surface EMG recording from bilateral upper and lower orbicularis oris (ORB sup, inf), masseter (MASS), digastric muscles (DIG) and temporalis muscles (TEMP). Recording performed when the patient was taking a long sip of water from a cup and swallowing. Initial pursing of lips with activation of orbicularis oris is followed by onset of a regular 7.5–8 Hz tremor affecting orbicularis oris and digastric muscles, beginning with water inlet in the mouth. With cessation of drinking, the tremor stopped. **Panel B:** Electrophysiological study of swallowing (EPSS) during drinking water from a cup. A repetitive and patterned activity was recorded from submental muscles (SMEMG) characterized by a 7.5–8 Hz tremor (lower trace). A synchronous activity was also recorded in the laryngeal-pharyngeal mechanogram (LPM, upper trace). **Panel C:** EPSS during repetitive drinking from a straw. Tremor was not observed when the patient drank from a straw. The reproducibility of the LPM was significantly reduced in patient (upper trace) in comparison with controls (example recording in a control subject). **Panel D:** EPSS single bolus and dysphagia limit. Swallowing of increasing single bolus amounts (3, 10, 15 ml). SMEMG (lower trace), LPM (upper trace). Dysphagia limit is reduced, as piecemeal deglutition is observed already at 10 ml.

Table 1
Demographic/clinical characteristics and EMG parameters.

	Patient	Controls, mean (sd)	z-score
Age, years	74	72 (4.3)	
Similarity index	0.39	0.83 (0.16)	−2.69
LPM-D, ms	2659.1	2200.5 (108.75)	4.22
SMEMG-D, ms	1206	1065 (136.11)	1.04
SMEMG-LPM-I, ms	167	196 (28.77)	−1.02

z-score = (controls mean-patient's value)/controls sd. Abbreviations: similarity index (SI), laryngeal-pharyngeal mechanogram (LPM), standard deviation (SD), submental muscles EMG (SMEMG), interval between SMEMG and LPM (SMEMG-LPM-I).

piecemeal deglutition was consistently observed for 10–15 ml liquid boluses, confirming subclinical swallowing alterations (Fig. 1D). Notably, abnormal rhythmic EMG activity was not

recorded during the administration or the swallowing of the water bolus, or with increasing water volumes.

The activation of the ventral belly of the digastric muscles marks the beginning of the propulsive action of the tongue and is the main factor responsible for elevating the pharyngeal/laryngeal structures. In fact, in our patient the rhythmic EMG activity of the digastric muscles observed during drinking from a cup was synchronous with the abnormal oscillatory movements of the larynx shown by the LPM. Ertekin et al. (2005) had previously reported a patient with dysphagia associated with pre-swallowing abnormal rhythmic activity involving the submental muscles. Unlike our case, the pathological activity was observed during single bolus examination with increasing water volume. Our case highlights the role of EPSS in detecting subclinical alterations of the oropharyngeal phase of swallowing and shows the importance of investigating subclinical dysphagia in patients with orofacial

task-specific tremor. Our data show that involvement of the digastric muscles may interfere with swallowing even independently of the tremulous activity.

The patient was significantly inconvenienced by the tremor. He started propranolol (80 mg twice a day), with no clinical benefit and declined further treatments (botulinum toxin injections).

Declaration of Competing Interest

The authors declare no competing financial interests.

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