Rare presentation of essential lingual myoclonus

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A 74-year-old woman presented to the Oral Medicine Department with a constant throbbing sensation affecting her entire tongue. She reported sudden onset of constant twitching of the tongue since January 2016. She described it as if “my heart is in my mouth.” She had visited both her dentist and her doctor, and was given a multitude of topical agents including Difflam, Nystatin, and Corsodyl, but none was effective.

The patient was unable to identify any aggravating or alleviating factors for her symptoms. She described the discomfort as 8/10 on the visual analogue pain scale, with considerable interruption of sleep and consequent fatigue and lowered mood.

The patient’s medical history included diagnoses of type 2 diabetes, hypertension, hypercholesterolemia, and gastric reflux. Her regular medications include metformin 500 mg OD, bendroflumethiazide 2.5 mg OD, lansoprazole 15 mg OD, and atorvastatin 80 mg OD. She was a nonsmoker and consumed alcohol very occasionally. The patient is retired, living at home with her husband.

The patient’s extraoral examination was unremarkable. Intraorally, on examination of the soft tissues, benign migratory glossitis was noted. This is a benign condition of the tongue; clinically the tongue appears to have red and white lesions and most patients are asymptomatic. More interestingly, the tongue appeared to be pulsating continuously in an involuntary rhythmic regular fashion as noted in the video, in isolation with no palatal involvement.

Ultrasound and MRI scan of the tongue did not reveal any abnormality; there was no degenerative change or space-occupying lesion, nor any sign of inflammatory disease that could explain the pulsating nature of the tongue. A further MRI as advised by the neurology department, with fine cuts through the medulla, ruled out a vascular event. MRI brain was reported as normal.

On subsequent review 2 months later, the tongue appeared to be pulsing in the constant rhythmic fashion noticed previously. Upon neurologic assessment, a provisional diagnosis of lingual myoclonus was made. The patient was started on 0.5 mg of clonazepam, which instantly improved her quality of sleep and she felt the throbbing was less “fierce.” She was advised to titrate and gradually increase the dose to between 1.5 and 3 mg in divided doses.

The patient reported a considerable improvement in her sleep; however, she was unable to tolerate more than 1 mg once a night due to hypersomnolence. On further review, she was referred to the movement disorders department for their expert opinion. She was started on a low dose of amitriptyline, which she felt helped her sleep with fewer side effects compared to clonazepam.

On subsequent examinations, the tongue appeared to be pulsing in the same fashion. A further EMG scan was arranged to investigate electrical activity within the lingual muscles. Bilateral

PRACTICAL IMPLICATIONS

Consider isolated lingual myoclonus as a differential when a patient who otherwise is fit and well presents with tongue tremor. Consider MRI and EMG readings to confirm diagnosis.

MORE ONLINE

▶ Video

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EMG of the genioglossus during relaxation revealed bursts of synchronous and rhythmic 1–2 Hz of burst of activity, each lasting approximately 200 ms. This fits well with limited literature available on cases of lingual myoclonus. There was no evidence of generalized neuropathy or signs of nerve degeneration.

Discussion

Lingual myoclonus is a spasmodic contraction of the lingual group of muscles resulting in rhythmic movement of the tongue. Conditions that can cause myoclonus include stroke, brain tumors, head or spinal cord injury, and infections. It can also be observed in chronic epilepsy.¹ Prolonged hypoxia depriving the brain from oxygen may result in posthypoxic myoclonus.² Myoclonus can occur in isolation; however, generally it is reported among a range of other symptoms relating to nervous system disorders. Our patient did not show any other neurologic signs or symptoms. Continuous rhythmic lingual movements may also present as part of myoclonus of the palatopharyngeal muscles, which is more commonly reported.

There are few reports of isolated cases of lingual myoclonus in the absence of neurologic disease. One case³ reported lingual myoclonus in a woman in whom no underlying pathology was found on extensive examination and tests. Three articles reported cases of isolated lingual myoclonus,^{4–6} the latest of which was more than 18 years ago. A recent review article⁷ acknowledged that tongue tremor occurring in isolation is a rare entity.

Rhythmic palatal myoclonus is well-documented, with theories suggesting either anatomical change or functional relations to

the inferior olive taken from animal experiments and neuro-anatomical positron tomography. Such theories are yet to be established regarding essential lingual myoclonus.

Author contributions

K. Patel: drafting/revising the manuscript, study concept or design, analysis or interpretation of data. A. Tappuni: drafting/revising the manuscript, study concept or design, analysis or interpretation of data, study supervision.

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